Congenital Heart Disease In Pediatric Patients: Recognizing The Undiagnosed And Managing Complications In The Emergency Department

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Abstract

Congenital heart disease is the most common form of all congenital malformations and, despite advances in prenatal and newborn screening, it may present undiagnosed to the emergency department. Signs and symptoms of congenital heart disease are variable and often nonspecific, making recognition and treatment challenging. Patient presentations can range from life-threatening shock or cyanosis in a neonate to respiratory distress or failure to thrive in infants. Advances in surgical techniques have improved short- and long-term survival of infants and children with congenital heart disease, but these children are at risk for a variety of complications related to the underlying or surgical anatomy and physiology. This review focuses on the recognition and initial management of patients with undiagnosed congenital heart disease presenting to the ED and touches on considerations for postoperative infants and children with complex congenital heart disease.

CME Objectives

Upon completion of this article, you should be able to:
1. Develop an approach to the history, physical examination, and basic investigations to elicit signs and symptoms suggestive of congenital heart disease.
2. Form a practical approach to the initial resuscitation and management of children presenting with shock, hypercyanotic spells, and acute congestive heart failure.
3. Recognize the special physiological considerations when therapies are initiated in a child with congenital heart disease.

Prior to beginning this activity, see "Physician CME Information" on the back page.

Case Presentations

An 8-day-old boy is brought to the ED by his mother for lethargy and "fast breathing." She states that he has not been feeding well for the past couple of days and his breathing has become faster and more labored over the past 24 hours. This morning he became lethargic and looked pale. She denies any fever, cough, vomiting, or diarrhea. The baby was born at term and delivered at home by a midwife, and there was little prenatal care. He has been exclusively breastfed, but feeds have become progressively shorter over the preceding 48 hours. At triage, the infant appeared ashen gray and limp, with the following vital signs: temperature, 36°C; heart rate, 194 beats/min; respiratory rate, 76 breaths/min; and initial oxygen saturation, 92% on room air. He was rushed back to the resuscitation room. As you enter the room to evaluate this critically ill neonate, you consider sepsis, metabolic disease, and congenital heart disease, and wonder how you can distinguish among these potential causes of critical illness in the first weeks of life. Given the clinical picture of this neonate, you administer broad-spectrum antibiotics, begin fluid resuscitation, and consider whether to initiate empiric prostaglandin, but you are not sure if this is necessary or safe without a clear diagnosis.

A 3-month-old girl is brought to the ED by her parents in January for difficulty breathing. Her mother has noticed a gradual increase in her work of breathing over the past few days, along with poor feeding. She has a slight runny nose but no fever or cough and no vomiting or diarrhea. Her 2-year-old sibling has had a cold for the past few days. The infant was born at 37 weeks after an uncomplicated pregnancy and spontaneous vaginal delivery, and discharged at 24 hours of life. Her pediatrician noted a heart murmur at her 2-month visit and referred her to a cardiologist for further evaluation, but the appointment is not until the next week. Upon further questioning, the mother says that she has been a difficult feeder, but that she seems to be getting worse, with shorter feeds and falling asleep at the breast, and she seems sweaty during feeds. She also noted that the infant is not gaining weight. At triage, her vital signs are: temperature, 37.6°C; heart rate, 180 beats/min; respiratory rate, 60 breaths/min; and oxygen saturation, 90% on room air. She is noted to have moderate respiratory distress. On examination, you note labored breathing with scattered rales, rhonchi, and mild wheezing, making it difficult to appreciate the heart sounds. You consider bronchiolitis, but decide to obtain a chest x-ray, given her history. The x-ray revealed a large heart, patchy perihilar opacities, and some fluid in the fissures. You suspect congestive heart failure and wonder if additional tests may be helpful and what medical therapies are indicated.

Introduction

Congenital heart disease (CHD) includes a spectrum of anatomic malformations of the heart and great vessels that occur during embryologic development of the fetus and can cause a wide range of physiologic perturbations and physical signs and symptoms. While many defects are identified prenatally through fetal ultrasound (including approximately 33% of all CHD and 57%-83% of critical lesions)[1] or diagnosed in the newborn period prior to discharge from the hospital, some CHD may go unrecognized and present without previous diagnosis to the emergency department (ED). The emergency clinician must maintain a high index of suspicion in these rare cases, as the clinical picture of undiagnosed CHD can be nonspecific, can mimic other common and benign childhood disease, or can present with a child in extremis. Infants and children with partially or fully corrected or palliated CHD may also present to the ED with complications related to the structural heart disease, the surgical repair, or as a result of concurrent illness in the setting of limited physiologic reserves. This review focuses primarily on the presentation, evaluation, and stabilization of undiagnosed CHD presenting to the ED, but will also touch on common emergencies in the patient with known heart defects.

Epidemiology

CHD is the most common major congenital anomaly, comprising one-third of all congenital malformations, and is the most common cause of mortality from birth defects in infants.[1,2] Differing definitions of CHD and methodologies make the exact determination of birth prevalence difficult; however, a 2011 systematic review and meta-analysis of 114 articles representing more than 24,000,000 births estimates a worldwide birth prevalence of 9.1/1000.[2] There is significant geographic variability, with the highest rates of CHD seen in Asia (9.3/1000), followed by Europe (8.2/1000), North America (6.9/1000), and the lowest rate noted in Africa (1.9/1000).[2] The birth prevalence of CHD appears to have increased worldwide over the past century, and leveled off since the late 1990s. Possible explanations for the increased prevalence include improvements in diagnosis (eg, fetal ultrasound and echocardiography), improved prenatal care with increased survival of preterm infants, or changing social and environmental determinants of disease (eg, delayed age of maternity, medication, or toxic exposures).

The spectrum of anatomic defects associated with CHD is broad, but 8 discrete lesions comprise more than three-quarters of all defects.[2] **(See Table 1, page 3.)** Complex and critical CHD, such as hypoplastic left heart syndrome (HLHS), total anomalous pulmonary venous return (TAPVR), and anomalous left coronary artery from the pulmonary artery (ALCAPA), are less common but important forms of CHD.

Presentations Of Congenital Heart Disease

While reviews and textbooks often categorize CHD based on the anatomy or physiology of structural lesions, it is more useful to the emergency clinician to consider the clinical presentations of CHD. Undiagnosed CHD can present in several ways, depending on the pathophysiology of the lesion(s), although individual variations may lead to overlapping features. Cardiovascular collapse/shock is typically seen in CHD and is characterized predominantly by left outflow tract obstruction. Cyanosis may be the presenting feature in lesions with limited pulmonary blood flow or right-to-left shunting of deoxygenated blood, or both. Respiratory distress from congestive heart failure (CHF) typically results from left-to-right shunting of blood, resulting in pulmonary overcirculation. **Table 2** summarizes the 3 main clinical presentations of CHD, including the symptoms, signs, and potential anatomic lesions associated with each.

Another potentially useful way to identify CHD is by the age of presentation. Lesions that depend on the ductus arteriosus for pulmonary or systemic circulation typically present with cyanosis or shock in the first week or weeks of life as the ductus closes. Lesions that result in pulmonary overcirculation leading to CHF more often develop gradually in the second or third month of life as falling pulmonary vascular resistance increases left-to-right shunting and results in pulmonary edema. **Figure 1 (page 4)** depicts the typical age of presentation for various types of CHD.

Two rare but important forms of CHD, ALCAPA and TAPVR, are particularly difficult to diagnose, as their presentations may vary considerably. ALCAPA can present early in the neonatal period with a shock-like state, as a result of myocardial infarction or it may present more insidiously, with recurrent periods of fussiness and gradual respiratory distress from CHF as a result of cardiac dysfunction from recurrent ischemia. Similarly, TAPVR may present in the neonate as cyanosis in cases with venous obstruction or later in infancy with CHF in cases without venous obstruction.

Table 1. Absolute And Relative Frequency Of The Most Common Cardiac Defects[2]

Cardiac Defect	Birth Prevalence Worldwide (#/1000)	Proportion of CHD
Ventricular septal defect	2.62	28.8%
Atrial septal defect	1.64	18%
Patent ductus arteriosus	0.87	9.6%
Pulmonic stenosis	0.5	5.5%
Tetralogy of Fallot	0.34	3.7%
Coarctation of the aorta	0.34	3.7%
Transposition of the great arteries	0.31	3.4%
Aortic stenosis	0.22	2.4%

Abbreviation: CHD, congenital heart disease.

Table 2. Clinical Presentations Of Congenital Heart Disease In Children

Clinical Presentation	Symptoms	Signs	Potential Congenital Cardiac Lesion
Shock	• Poor feeding • Fussiness • Progression to lethargy	• Extreme tachycardia • Pallor (often "ashen gray") or acral cyanosis • Weak peripheral pulses • Delayed capillary refill • Altered mental status • Hypotension with decreased BP in lower extremities vs right arm (in some lesions)	• Critical aortic stenosis • Coarctation of the aorta • HLHS • ALCAPA with myocardial infarction
Cyanosis	• Fussiness • Cyanosis	• Central cyanosis (mucus membranes/trunk) • Hypoxia not improved with oxygen administration (oxygen saturation typically < 80%-85%)	• Transposition of the great arteries • Tetralogy of Fallot • Tricuspid atresia • Truncus arteriosus • TAPVR with obstructed veins
Congestive heart failure	• Feeding difficulty • Sweating with feeds • Failure to thrive • Difficulty breathing • Fussiness	• Tachypnea with labored breathing • Rales • Hepatomegaly • Cyanosis if severe	• Ventricular septal defect • Patent ductus arteriosus • Atrioventricular canal • PAPVR • ALCAPA with recurrent ischemia and cardiac failure

Abbreviations: ALCAPA, anomalous left coronary artery from the pulmonary artery; BP, blood pressure; HLHS, hypoplastic left heart syndrome; PAPVR, partial anomalous pulmonary venous return; TAPVR, total anomalous pulmonary venous return.

Critical Appraisal Of The Literature

A literature search was performed in PubMed using combinations of the search terms *congenital heart disease or congenital heart defects and emergency department, epidemiology, etiology, embryology, genetics, congestive heart failure, shock, cardiogenic shock, cyanosis, prostaglandin, PGE$_1$,* and *vasopressors.* Only articles published in English whose subjects included children aged birth to 18 years were reviewed. Within CHD, only 25 clinical trials were available, none were conducted in the ED, and only 1 (on the use of prostaglandin E$_1$ [PGE$_1$]) was relevant to the acute management of infants and children with CHD. There was 1 practice guideline and evidence-based review of the management of pediatric heart failure that was not specific to CHD. The lack of high-quality evidence relevant to the ED management of infants and children with CHD is not surprising, given the rarity of ED presentation, the frequently critical nature of acute illness in these children, and the general difficulties related to clinical trials in the pediatric population. In the absence of evidence from clinical trials, the literature review was broadened to include review articles, systematic reviews, and case series related to pediatric CHD in the ED as well as the results of literature searches for specific therapies for pediatric cardiogenic shock, pediatric CHF, and complications of CHD and its surgical palliation and repair. In total, more than 70 peer-reviewed articles comprise the literature that informed this review.
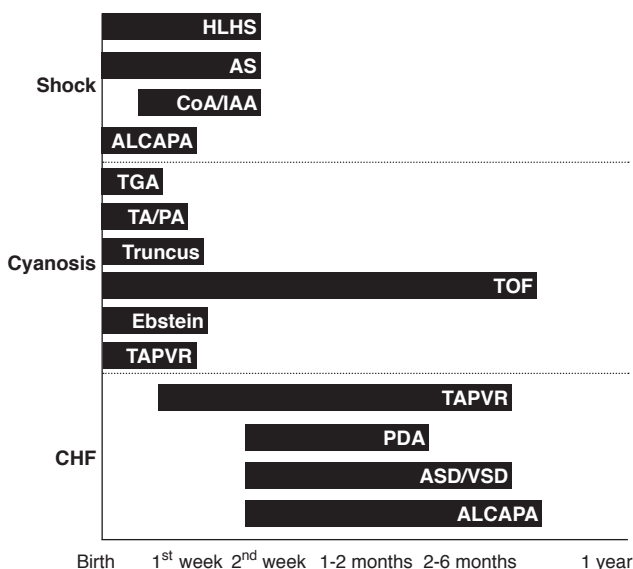
Etiology And Pathophysiology

The cause of CHD is often undetermined and is believed to be a multifactorial process with contributions from both genetic and environmental factors. Only approximately 15% of CHD has an identifiable etiology.[1] Genetics clearly play a role in some CHD, which is reflected in an increased risk of CHD in newborns with an affected sibling (2%-6%); the relative risk varies by the particular lesion of the first child, and can be as high as 20% to 30%. Specific chromosomal aneuploidies such as Down syndrome, Turner syndrome, trisomy 13, and trisomy 18 account for 8% to 10% of CHD.[1] Single-gene mutations are often associated with syndromes that include CHD such as DiGeorge syndrome (cardiac defects, abnormal facies, thymus aplasia, cleft palate, and hypocalcemia caused by deletion of the 22q11.2 region), Holt-Oram syndrome (CHD and upper limb malformations caused by mutations in the TBX5 and SALL4 genes), and Alagille syndrome (CHD and liver disease from JAG1 or NOTCH2 defects).[3,4] **Table 3, page 5,** summarizes some of the more common congenital syndromes and their associated CHD.

Fetal environmental factors can be identified in approximately 2% of CHD and include maternal factors, infections during pregnancy, and toxic exposures.[5] **(See Table 4, page 5.)**

Transition From Fetal To Neonatal Circulation

With the first breath at birth, the newborn's lungs expand with air, pulmonary blood flow and the partial pressure of oxygen in arterial blood (PaO$_2$) increase, and pulmonary vascular resistance falls. At the same time, the low-resistance placenta is removed from circulation and the ductus venosus closes, causing an increase in systemic vascular resistance. The lower pulmonary and higher systemic vascular resistance reverses the fetal right-to-left flow of blood through the ductus arteriosus, which becomes left-to-right instead. Over a period of hours to days, the ductus arteriosus constricts and gradually closes. Though the foramen ovale may remain anatomically patent, it becomes functionally closed by the increased pulmonary blood flow and higher left atrial pressures in comparison to the right atrium.

In some infants, the ductus arteriosus and foramen ovale may remain patent during the neonatal period. This failure of normal closure may be life-sustaining or may contribute to significant pathology in cases of CHD. For example, transposition

Figure 1. Timing And Presentation Of Congenital Heart Defects

Abbreviations: ALCAPA, anomalous left coronary artery from the pulmonary artery; AS, aortic stenosis; ASD, atrial septal defect; CoA, coarctation of the aorta; HLHS hypoplastic left heart syndrome; IAA, interrupted aortic arch; PA, pulmonary atresia; PDA, patent ductus arteriosus; TA, tricuspid atresia; TAPVR, total anomalous pulmonary venous return; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; truncus, truncus arteriosus; VSD, ventricular septal defect.

Table 3. Genetic Defects And Syndromes Associated With Congenital Heart Disease

Syndrome	Cardiac Lesion
Trisomy 21	AVSD, VSD, ASD
Trisomy 18	VSD, ASD, PDA, CoA, bicuspid aortic or pulmonary valve
Trisomy 13	VSD, ASD, PDA, CoA, bicuspid aortic or pulmonary valve
Turner (XO)	Bicuspid aortic valve, CoA
Fragile X	Mitral valve prolapse and aortic root dilatation
Alagille	Peripheral pulmonic stenosis, pulmonic stenosis, TOF
Holt-Oram	ASD, VSD, first-degree heart block
Crouzon	PDA, CoA
PHACE (Posterior brain fossa anomalies, facial hemangiomas, arterial anomalies, cardiac anomalies, CoA, eye anomalies)	VSD, PDA, CoA, arterial aneurysms
Noonan	Pulmonary stenosis, ASD, cardiomyopathy
CHARGE (coloboma, heart defects, atresia choanae, retardation, genital and ear anomalies)	VSD, ASD, PDA, TOF, AVSD
DiGeorge, CATCH22 (cardiac, abnormal facies, thymic aplasia, cleft palate, hypocalcemia, 22q microdeletion)	Aortic arch anomalies and conotruncal anomalies (truncus arteriosus, TGA, TOF, DORV, PA with VSD)
VATER (vertebral, anal, tracheoesophageal, radial, and renal anomalies)	VSD, TOF, ASD, PDA
Williams	Supravalvular aortic stenosis, peripheral pulmonic stenosis
Smith Lemli Opitz	VSD, PDA
TAR (thrombocytopenia and absent radii)	ASD, TOF
Scimitar	Hypoplasia of right lung with anomalous pulmonary venous return to IVC
Apert	VSD

Abbreviations: ASD, atrial septal defect; AVSD, atrioventricular septal defect; CoA, coarctation of the aorta; DORV, double outlet right ventricle; IVC, inferior vena cava; PA, pulmonary atresia; PDA, patent ductus arteriosus; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

Table 4. Risk Factors For Congenital Heart Disease[1,5-7]

Risk Category	Examples	Associated CHD	Relative Risk
Maternal Factors	Pregestational diabetes mellitus	VSD, d-TGA, PDA, HLHS, AVSD	3.1-18
	Phenylketonuria	CoA, HLHS	> 6
	Advanced maternal age	Conotruncal defects, TGA, CoA, VSD, ASD	1.3-2.1
	Febrile illness during pregnancy	Multiple	1.8-2.9
Infections	Influenza	Conotruncal defects, d-TGA, CoA, VSD, TA, obstructive lesions	2.1
	Rubella	PS, PDA, VSD	Unknown
Medications	NSAIDs	d-TGA, AVSD, VSD	1.86-2.5
	Sulfasalazine	Multiple	3.4
	Trimethoprim-sulfonamide	Multiple	2.1-4.8
	Lithium	Ebstein anomaly	Unknown
	Phenytoin	AS, PS	Unknown
	Thalidomide	Multiple	Unknown
Toxins	Marijuana	VSD, Ebstein anomaly	1.9-2.4
	Alcohol	VSD, ASD	Unknown
	Organic solvents	Multiple	2.0-5.6

Abbreviations: AS, aortic stenosis; ASD, atrial septal defect; AVSD, atrioventricular septal defect; CoA, coarctation of the aorta; d-TGA, dextro-transposition of the great arteries; HLHS hypoplastic left heart syndrome; NSAIDs, nonsteroidal anti-inflammatory drugs; PDA, patent ductus arteriosus; PS, pulmonic stenosis; TA, tricuspid atresia; TGA, transposition of the great arteries; VSD, ventricular septal defect.

of the great arteries creates 2 parallel circulations, which would be incompatible with life unless a shunt (such as a patent foramen ovale) is present; similarly, with severe coarctation of the aorta (CoA) or interrupted aortic arch, systemic circulation after birth depends on persistent patency of the ductus arteriosus. By contrast, a patent ductus arteriosus in the setting of prematurity or persistent pulmonary hypertension can lead to excessive left-to-right or right-to-left shunting of blood, which may lead to pulmonary edema or cyanosis, respectively. Medications can be used to manipulate the patency of the ductus arteriosus; PGE_1 can be used to maintain patency, and indomethacin can be used to promote closure.

Pathophysiology Of Clinical Presentations Of Shock

Regardless of its etiology, shock is a result of inadequate oxygen delivery to meet the metabolic demands of tissues. CHD presenting to the ED as shock typically arises from ductal-dependent left-sided obstructive lesions such as CoA, hypoplastic left heart syndrome, interrupted aortic arch, and critical aortic stenosis.[8,9] Neonates with ductal-dependent lesions may present in the first few weeks of life with signs of poor systemic perfusion and acidosis as the ductus arteriosus closes. In these patients, systemic blood flow is dependent on the patency of this anatomic shunt.

Pathophysiology Of Clinical Presentations Of Cyanosis

Cyanosis can be central (mucous membrane and lips) or peripheral (hands and feet). Peripheral cyanosis may be normal in neonates and young infants, related to cold temperature exposure and peripheral vasoconstriction. Central cyanosis is always pathologic. Because oxygen-carrying capacity is mainly determined by the hemoglobin concentration of blood and oxygen saturation, cyanosis may appear in a polycythemic child with adequate tissue oxygen delivery, while an anemic child without cyanosis may be unable to meet tissue oxygen needs.[10]

Most pulmonary causes of cyanosis improve with administration of supplemental oxygen, whereas cyanosis caused by cyanotic CHD typically does not respond to oxygen therapy. The 5 cyanotic CHD lesions traditionally described include (1) transposition of the great arteries (TGA), (2) tetralogy of Fallot (TOF), (3) tricuspid atresia (TA), (4) total anomalous pulmonary venous return (TAPVR), and (5) truncus arteriosus. In all of these lesions, there is mixing of oxygenated and deoxygenated blood and shunting of deoxygenated blood into systemic circulation, which manifests as cyanosis. While pulse oximetry is a critical piece of the assessment of all critically ill infants and children, its limitations must be kept in mind; readings may be falsely reduced by factors such as motion artifact, cool extremities, poor peripheral perfusion, ambient light, skin pigmentation, and probe position, and levels become inaccurate when oxygen is < 80% saturation.[10,11]

Pathophysiology Of Clinical Presentations Of Congestive Heart Failure

CHF is a clinical syndrome that is defined by the heart's inability to supply blood to the tissues to meet tissue metabolic demands.[12] Compensatory physiologic responses to decreased cardiac output include catecholamine release leading to increased heart rate, contractility, and peripheral vasoconstriction, all of which improve cardiac output. In addition, the renin-angiotensin-aldosterone system is activated, which leads to vasoconstriction, increased blood volume, and fluid retention. Eventually, there is myocardial apoptosis and areas of focal necrosis.

Although various forms of CHD can present early and initially with CHF (eg, left ventricular outflow tract obstruction with increased afterload), the classic presentation of CHD presenting with CHF is from large left-to-right shunts that cause increased preload, pulmonary overcirculation, and pulmonary vascular congestion and edema. The usual CHD lesions presenting in this way are ventricular septal defects (VSD) and atrioventricular canal defects. A patent ductus arteriosus can present similarly. In all of these conditions, pulmonary blood flow is initially restricted by high pulmonary vascular resistance at birth, but, as resistance falls during the first month of life, left-to-right shunting and pulmonary blood flow increase, and signs and symptoms of CHF typically develop between 4 and 12 weeks of life as systemic and pulmonary venous congestion progress.[8]

Differential Diagnosis

The differential diagnosis of undiagnosed CHD is broad and varies greatly by clinical presentation, which includes shock, cyanosis, or CHF. Shock is typically classified by pathophysiology into hypovolemic, distributive, obstructive, and cardiogenic. Cardiogenic shock can be related to structural problems (as with CHD) or to nonstructural causes such as dysrhythmias, myocarditis, and cardiomyopathy.

Because respiratory distress and feeding difficulties are the primary symptoms of CHD presenting with CHF, the differential diagnosis is broad and includes infection (sepsis, central nervous system concerns, pulmonary etiology), metabolic disease (hyperammonemia or metabolic acidosis), gastrointestinal emergencies (malrotation with volvulus), toxic ingestions, anemia, trauma (including nonaccidental), and nonstructural cardiac causes such as dysrhythmias, myocarditis, and cardiomyopathy.

The differential diagnosis for cyanotic CHD includes primarily infectious and pulmonary causes

such as pneumonia, bronchiolitis, and other respiratory tract infections, but also obstructive processes such as foreign body aspiration, severe asthma, and intrinsic or extrinsic lung disease. Acute respiratory distress syndrome from sepsis or neurogenic causes can also lead to cyanosis, as can CHF with pulmonary edema. Deoxygenated hemoglobin in the setting of polycythemia or toxins such as methemoglobin can also lead to cyanosis. Poorly perfusing cardiac dysrhythmias without structural abnormalities (eg, supraventricular tachycardia) and cardiac failure from acquired causes (eg, myocarditis or cardiomyopathy) can cause cyanosis, usually with respiratory distress and CHF. As previously mentioned, peripheral cyanosis may be a normal finding in neonates and young infants as a result of vasoconstriction in response to environmental cold stress.

Prehospital Care

Since definitive diagnosis and care of neonates, infants, and children with CHD often require advanced imaging, medication, or surgery, rapid transport of the pediatric patient to a tertiary children's hospital, once stabilized, is the primary goal. En route, support of airway, breathing, and circulation are paramount, with a few caveats. (See the "Controversies And Cutting Edge" section on page 20.) Patients presenting with signs of shock, cyanosis, or respiratory distress should receive supplemental oxygen, with continuous monitoring of pulse oximetry and heart rate. Vascular access should be obtained, but this is often difficult in this clinical setting and intraosseous access may be necessary. In the setting of poor perfusion or hypotension, a 10-mL/kg bolus of normal saline is indicated, and all obtunded or lethargic pediatric patients should have blood glucose checked and hypoglycemia treated with 5 mL/kg of 10% dextrose in water.[13] Infants and children with severe respiratory distress or respiratory failure may require support of airway, breathing, and ventilation using bag-valve mask positive-pressure ventilation. Controversy exists over the role of advanced airway techniques in the prehospital setting[14-17] and in the setting of CHD, as the complex pathophysiology may make these patients particularly vulnerable to decompensation during the process of rapid sequence intubation and positive-pressure ventilation. Therefore, endotracheal intubation is probably best avoided in this patient population if the diagnosis of CHD is strongly suspected.[12,18,19]

In cases of interhospital transfer, a number of studies suggest that outcomes among critically ill infants and children and patients with complex CHD are improved with the use of specialized pediatric critical care transport teams, and these should be utilized, if available.[20-23] In addition to critical care skills, these teams often carry medications such as PGE$_1$, which can be life-saving in those with ductal-dependent CHD.

Emergency Department Evaluation

Initial Evaluation And Stabilization

The evaluation of the pediatric patient with suspected or known CHD follows the usual ED approach of prioritizing airway, breathing, and circulation. Airway emergencies in this population may be related to associated anatomic abnormalities in children with certain syndromes (such as macroglossia in children with Down syndrome) and CHD. **(See Table 3, page 5.)**

Breathing is assessed by noting both the respiratory rate and work of breathing (subcostal, intercostal, suprasternal retractions, nasal flaring, grunting) as well as color, pulse oximetry, and mental status. Irritability may be a sign of hypoxemia, while lethargy may indicate hypercapnia or inadequate perfusion/shock. "Quiet tachypnea" (increased respiratory rate without significant work of breathing) may be noted in neonates with poor systemic or pulmonary perfusion (eg, CoA with a closing ductus arteriosus or TOF with significant pulmonary stenosis). Tachypnea with labored breathing suggests pulmonary pathophysiology, including pulmonary edema from CHF (eg, CHF from left-to-right shunting in large atrial septal defect [ASD] or VSD) or intrathoracic airway obstruction from anomalous large vessels associated with aortic arch abnormalities. Rales are often present with significant pulmonary edema from CHF, but "cardiogenic wheezing" may also be noted in cases of CHD presenting with CHF or vascular rings/slings and may mimic bronchiolitis or reactive airway disease.

Circulation is assessed by noting heart rate (tachycardia is the typical response to shock or initial hypoxia, but bradycardia may be a pre-arrest finding in critically ill neonates and infants with severe cardiogenic shock), the quality and difference between central and peripheral pulses, as well as preductal (right brachial or radial) and postductal (femoral or pedal) pulses, skin color and temperature, capillary refill, and mental status.

History

Obtain a thorough history starting with the chief complaint and the rate of progression of symptoms, which may be helpful in the context of the patient's age. **(See Figure 1, page 4.)** Relatively rapid progression of symptoms in a neonate suggests a ductal-dependent lesion. Ductal closure during the first or second week of life can cause pulmonary systemic hypoperfusion resulting in cyanosis or shock, respectively. In infants with corrected or palliated complex CHD, rapid onset of symptoms may suggest a thrombotic event such as clotting of a shunt or

Clinical Pathway For Management Of Congenital Heart Disease In The Neonate Presenting With Shock

Patient presents with signs and symptoms of shock
- Apply monitor
- Apply oxygen, if hypoxia present
- Obtain IV or IO access
- Draw bedside glucose, CBC, arterial blood gas, lactate, BUN, creatinine, and blood culture
- Obtain chest x-ray
- Consider broad-spectrum antibiotics

(Class II)

↓

Apnea or agonal respirations? — **YES** → **Utilize bag-valve mask and intubate expeditiously**

NO ↓

Abnormal cardiac examination?
- Murmur, gallop
- Hepatomegaly
- Unable to palpate femoral pulses
- Discrepant limb blood pressures

YES ↓   **NO** → **Treat other causes of shock:**
- Trauma
- Infection
- Metabolic disease
- Hypoglycemia
- Adrenal insufficiency

Patient aged < 1 month?

YES → **Start PGE$_1$ 0.05-0.1 mcg/kg/min infusion (Class III)**

NO ↓

Administer IV fluid bolus of 10 mL/kg normal saline

↓

- Obtain ECG
- Consult cardiology to obtain echocardiogram

Abbreviations: BUN, blood urea nitrogen; CBC, complete blood count; ECG, electrocardiogram; IO, intraosseous; IV, intravenous; PGE$_1$, prostaglandin E$_1$.

Class Of Evidence Definitions

Each action in the clinical pathways section of *Pediatric Emergency Medicine Practice* receives a score based on the following definitions.

Class I
- Always acceptable, safe
- Definitely useful
- Proven in both efficacy and effectiveness

Level of Evidence:
- One or more large prospective studies are present (with rare exceptions)
- High-quality meta-analyses
- Study results consistently positive and compelling

Class II
- Safe, acceptable
- Probably useful

Level of Evidence:
- Generally higher levels of evidence
- Nonrandomized or retrospective studies: historic, cohort, or case control studies
- Less robust randomized controlled trials
- Results consistently positive

Class III
- May be acceptable
- Possibly useful
- Considered optional or alternative treatments

Level of Evidence:
- Generally lower or intermediate levels of evidence
- Case series, animal studies, consensus panels
- Occasionally positive results

Indeterminate
- Continuing area of research
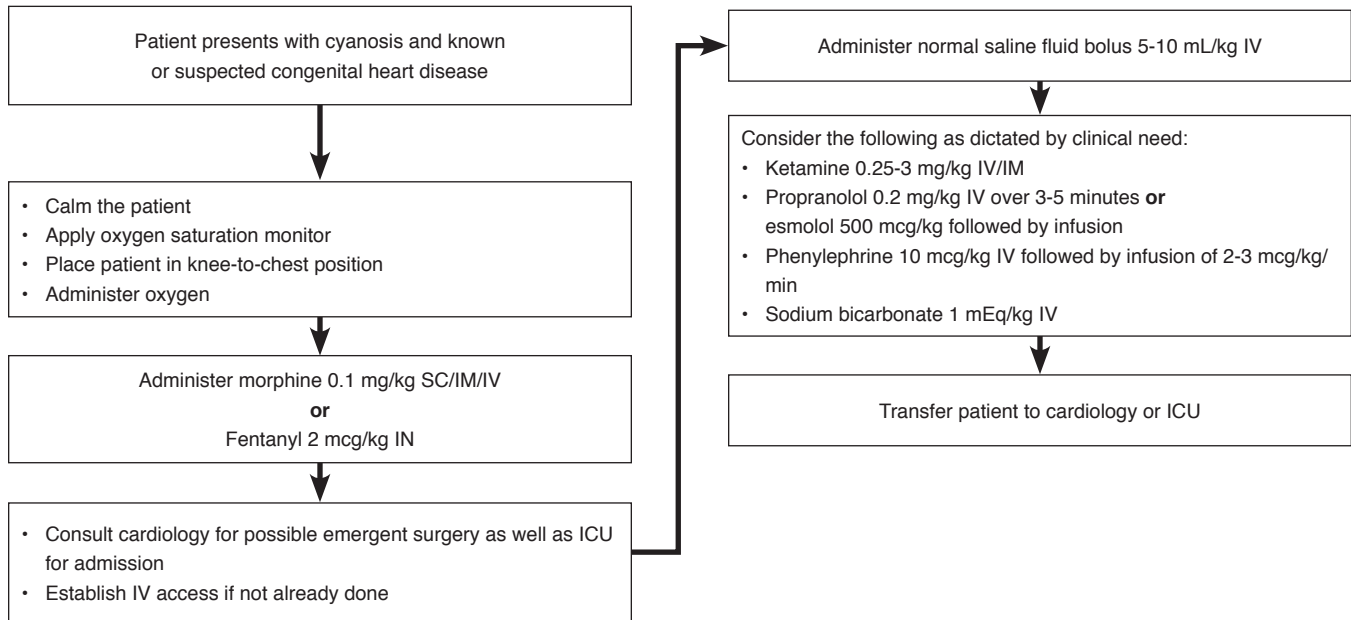- No recommendations until further research

Level of Evidence:
- Evidence not available
- Higher studies in progress
- Results inconsistent, contradictory
- Results not compelling

Clinical Pathway For Management Of Hypercyanotic Episode In Pediatric Patients With Tetralogy Of Fallot

Patient presents with cyanosis and known or suspected congenital heart disease

↓

- Calm the patient
- Apply oxygen saturation monitor
- Place patient in knee-to-chest position
- Administer oxygen

↓

Administer morphine 0.1 mg/kg SC/IM/IV
or
Fentanyl 2 mcg/kg IN

↓

- Consult cardiology for possible emergent surgery as well as ICU for admission
- Establish IV access if not already done

→

Administer normal saline fluid bolus 5-10 mL/kg IV

↓

Consider the following as dictated by clinical need:
- Ketamine 0.25-3 mg/kg IV/IM
- Propranolol 0.2 mg/kg IV over 3-5 minutes **or** esmolol 500 mcg/kg followed by infusion
- Phenylephrine 10 mcg/kg IV followed by infusion of 2-3 mcg/kg/min
- Sodium bicarbonate 1 mEq/kg IV

↓

Transfer patient to cardiology or ICU

Clinical Pathway For Management Of Congenital Heart Disease In The Pediatric Patient Presenting With Congestive Heart Failure

Patient presents with signs and symptoms of acute congestive heart failure:
- Gallop
- Hepatomegaly
- Poor pulses
- Tachypnea
- Jugular venous distention

→

Administer oxygen with caution. Patient protecting airway?

— **NO** →
- Intubate
- Notify ICU and cardiology for extracorporeal life support backup

YES ↓

Obtain chest x-ray. Cardiomegaly present?

— **NO** →

YES ↓

Administer furosemide 1 mg/kg IV
(Class II)

→

Consult cardiology to arrange echocardiography and to consider the following based on patient need:
- Spironolactone
- ACE inhibitors
- Beta blockers
- Digoxin
- Inotropes
(Class III)

Abbreviations: ACE, angiotensin-converting enzyme; ICU, intensive care unit; IM, intramuscular; IN, intranasal; IV, intravenous; SC, subcutaneous.

For Class of Evidence definitions, see page 8.

conduit. Sudden onset of cyanosis during episodes of crying may be seen in children with undiagnosed or uncorrected TOF (a "Tet spell"). More-insidious progression over days to weeks in slightly older infants is more characteristic of worsening pulmonary edema from CHF in lesions with pulmonary overcirculation from large left-to-right shunts.

Inquire about associated symptoms such as fever, rhinorrhea or nasal congestion, cough, vomiting, and diarrhea, as common viral illnesses (upper respiratory tract infection, bronchiolitis, or gastroenteritis) can often precipitate cardiac decompensation in infants with undiagnosed or palliated CHD.[24-26]

Elicit details of the feeding history, as this may yield clues for potential CHD. Episodes of fussiness, pallor, and sweating with feeds may be a sign of ischemia related to ALCAPA.[9] Prolonged feeds, sweating with feeds, and poor weight gain may suggest left-to-right shunting lesions such as ASD or VSD.

In patients without a known cardiac defect, ask about the pregnancy and prenatal care to identify potential risk factors for CHD such as maternal pregestational diabetes; febrile illness; influenza; toxoplasmosis, other (syphilis, varicella-zoster, parvovirus B19), rubella, cytomegalovirus, and herpes (TORCH) infections during pregnancy; maternal medications; and alcohol or drug use.[5] **(See Table 4, page 5.)** Do not rely on a history of normal prenatal ultrasound or a normal perinatal hospital course to exclude the possibility of CHD, as screening for CHD, while specific, is not 100% sensitive.[27,28] Ask about family members with CHD, as risk of recurrence is increased with ≥ 1 sibling or parent (especially the mother) with known CHD.

Physical Examination

The physical examination begins with general observation. Note dysmorphic features that may give clues to syndromes associated with CHD. **(See Table 3, page 5.)** Observe the patient's mental status for irritability or lethargy that could indicate hypoxemia, shock, or acidosis. Note the skin color, looking for pallor, mottling, or acral cyanosis (blue discoloration of the nails, hands, and feet that might indicate shock or may be a normal part of neonatal vascular instability), or central cyanosis (involving the mucus membranes and lips) that can be difficult to appreciate in infants and children with dark skin.

Examination of the cardiovascular system includes palpation of the precordium for thrills or heaves; assessment of central, peripheral, preductal (right brachial) and postductal (femoral) pulses; and auscultation for murmurs. **(See Table 5, page 11.)** Symmetric but weak right and left radial and pedal pulses may be seen in shock of any etiology, but a discrepancy between the strength of the right upper extremity and the lower extremity pulses suggests left outflow tract obstruction that is ductal-dependent, such as CoA or HLHS. Suspicion of pulse differential can be confirmed by measuring 4 extremity blood pressures in all patients with suspected CHD. Mean arterial pressure (MAP) should be equal in all extremities, and a 20-mm Hg gradient between the right arm (higher MAP) and either leg (lower MAP) should raise suspicion for CoA.[29]

Auscultate the heart, noting the timing, location, quality, and radiation of any murmurs. The absence of a murmur does not exclude CHD, as turbulent flow is required to produce the sound of a murmur. Paradoxically, large unrestricted shunts may not produce a murmur or the flow across a large defect may be restricted by elevated pulmonary pressures, diminishing the ability to detect the defect through auscultation. While benign murmurs are common throughout childhood, pathologic murmurs may be harsh or rumbling, and most diastolic murmurs are associated with pathology and warrant cardiology follow-up. In patients with surgically corrected or palliated CHD who have constructed shunts, a murmur is expected, and the absence of a shunt murmur may signal catastrophic thrombosis of the shunt.

Pay attention to the first and second heart sounds, noting the intensity and splitting of the second heart sound. Normal splitting of the second heart sound varies with respiration. A widely split S2 may be noted in TAPVR; a single S2 may be noted in truncus arteriosus or TA; and a fixed-split second heart sound can be heard in ASD. Finally, listen for the presence of a gallop rhythm (S3/S4), which can be heard in infants and children with CHF and may be the main clue in distinguishing CHF from bronchiolitis.

Complete the examination by carefully palpating the liver. Hepatomegaly, particularly in the setting of respiratory distress, a gallop rhythm, and rales suggests CHF. Unlike adults, CHF in infants rarely causes peripheral edema and assessment of neck veins for jugular venous distention in infants is difficult.

Diagnostic Studies

Laboratory Studies

The initial investigations in a child presenting with shock, respiratory distress, and/or cyanosis should include a complete blood count (CBC), electrolytes, blood urea nitrogen (BUN), creatinine, blood gas, and chest x-ray. An arterial blood gas is useful in the evaluation of any child presenting with shock, respiratory distress, or cyanosis. The pH provides an indication of acidosis, and helps distinguish respiratory and metabolic contributions in the critically ill child. The partial pressure of carbon dioxide (PCO_2) provides information on the ventilatory status of the patient in the context of the clinical examination findings. Also, the partial pressure of oxygen (PO_2) can be useful in distinguishing respiratory from cardiac pathology using the hyperoxia test. Provide 100%

fraction of inspired oxygen (FiO_2) to the hypoxic patient for 10 minutes. If the etiology is pulmonary, the pulse oximetry should increase by at least 10%[30] and the PaO_2 should generally rise to > 150 to 200 mm Hg, while cardiac etiologies rarely rise > 100 mm Hg.[31]

Obtain a blood culture for a source of infection in the setting of fever, hypothermia, or if an infectious etiology is suspected. Tailor additional investigations according to the differential diagnosis based on history, physical examination, initial investigations, and the patient's response to therapies initiated in the ED.

Biomarkers can be used to diagnose and stratify risk of CHF in children. The most commonly used and available is B-type natriuretic peptide (BNP). In a child with respiratory distress in whom you are questioning underlying CHD and CHF, BNP may help confirm or exclude a cardiac cause for the symptoms.[32,33] In children with known CHF, the elevation or trend of BNP can help predict disease progression.[34]

Electrocardiography

Normal electrocardiographic (ECG) findings in children differ significantly from adult ECGs. The neonatal heart demonstrates right ventricular dominance with a rightward axis deviation on ECG. This is manifested as an increased R-wave amplitude in leads V_1 and V_2, and decreased amplitude in V_5 and V_6. As the left ventricle becomes more dominant over the first months of life, the QRS axis shifts leftward.[35] Other differences include narrower QRS complexes, shorter PR intervals, and an age-related correction of QT intervals using the Bazzet formula. T waves in leads V_1 to V_3 are initially upright at birth, flip during the first week of life, and gradually revert to upright by adolescence. Upright T waves in early childhood may be an indicator of right ventricular hypertrophy.[35,36]

ECG abnormalities in CHD may not always be diagnostic (and the ECG can be normal in CHD), but they can provide clues to chamber enlargement or conduction abnormalities. Right ventricular

Table 5. Cardiovascular Examination Findings In Specific Congenital Heart Disease

Clinical Presentation	Heart Defect	Precordium and Heart Sounds	Type of Murmur	Comments
Cyanosis	TOF	Systolic thrill at left mid or lower sternal border, loud single S2, aortic ejection click	Loud SEM +/- continuous PDA murmur, possible VSD murmur	Cyanosis depends on degree of PS: greater obstruction leads to left-to-right shunting with cyanosis
	TGA	Loud single S2	None	Systolic murmur if VSD present
	TA	Single S2	Systolic regurgitant murmur at LLSB +/- continuous PDA murmur	N/A
	Truncus arteriosus	Single S2	Loud systolic regurgitant murmur at LSB with high-pitched diastolic decrescendo murmur or rumble	N/A
	TAPVR	RV heave, fixed-split S2 (if obstructed, loud and single S2 and gallop)	SEM at LUSB with diastolic rumble at LLSB (no murmur if obstructed)	Unobstructed TAPVR may present with CHF later in infancy
Shock	AS	Systolic thrill at RUSB, supra-sternal notch, carotid and ejection click	Systolic murmur at RUSB or LUSB radiating to the neck	May have a gallop if associated with congestive heart failure
	CoA	None	Possible SEM	May have a gallop if associated with congestive heart failure; differential upper and lower extremity pulses/BP
	HLHS	Single heart sound	SEM	Decreased peripheral pulses
Congestive heart failure	PDA	None	Loud continuous "machinery" or "to-and-fro" murmur at LUSB with diastolic rumble	Bounding pulses
	ASD	Wide and fixed-split S2	SEM at LSB	May have middiastolic rumble if significant shunting
	VSD	Possible precordial thrill, narrow-split S2	Loud, harsh, holosystolic murmur at LLSB	N/A

Abbreviations: AS, aortic stenosis; ASD, atrial septal defect; BP, blood pressure; CoA, coarctation of the aorta; HLHS hypoplastic left heart syndrome; LLSB, left lower sternal border; LSB, left sternal border; LUSB, left upper sternal border; N/A, not applicable; PDA, patent ductus arteriosus; PS, pulmonic stenosis; RUSB, right upper sternal border; RV, right ventricle; SEM, systolic ejection murmur; TA, tricuspid atresia; TAPVR, total anomalous pulmonary venous return; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

hypertrophy beyond the neonatal or early infant period can be seen in conditions such as HLHS. **(See Figure 2.)** Left ventricular hypertrophy may be noted with left ventricular outflow tract obstructions or large VSDs and atrioventricular canal defects.[35] **(See Figure 3.)** Right atrial enlargement may be noted with ASD, atrioventricular canal defects, or TA, and left atrial enlargement may be noted with mitral stenosis or left ventricular outflow tract obstruction.[37] The ECG findings should be evaluated in the context of other tests (such as chest x-ray), and echocardiogram is usually required for definitive diagnosis. For patients with known CHD, compare with previous ECGs, if available, to identify acute changes.

Chest Radiography

While the definitive diagnosis of most CHD fre-

Figure 2. Electrocardiogram Of 1-Week-Old Patient With Hypoplastic Left Heart Syndrome, Showing Right Ventricular Hypertrophy And Strain
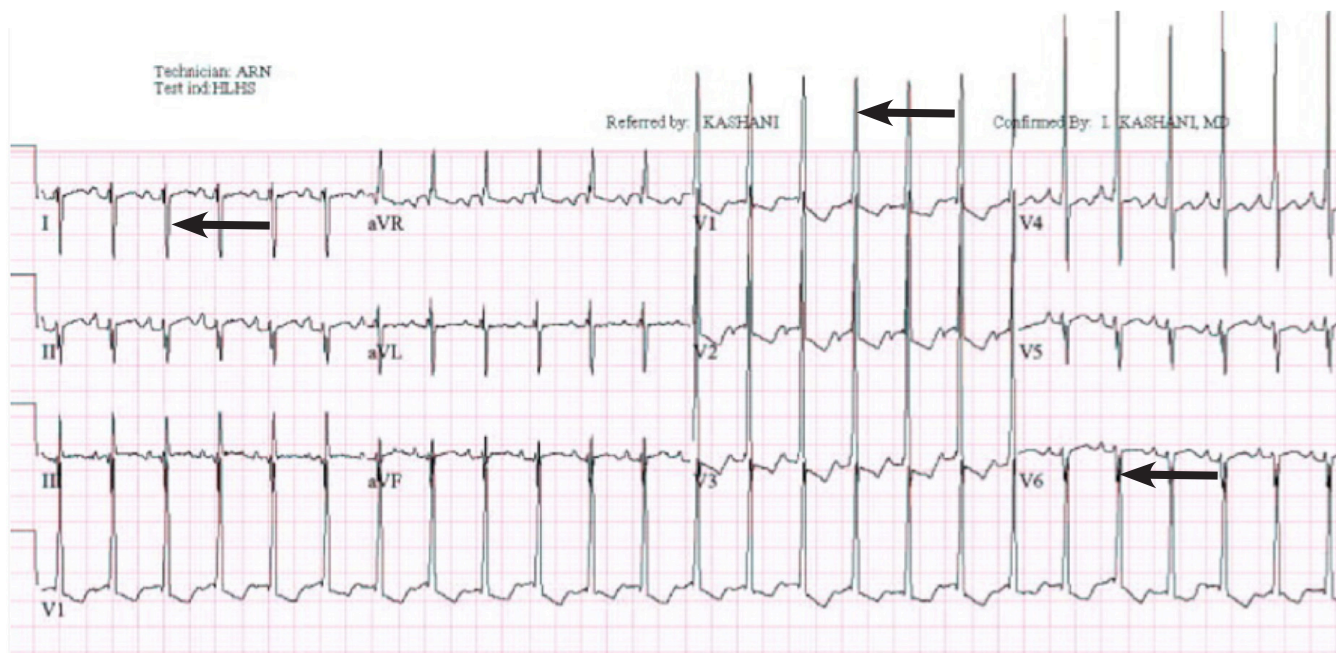
Right ventricular hypertrophy noted by dominant R wave in V_1 and S wave in V_6. Strain noted by rightward axis indicated by negative forces in lead I.
Reprinted from the *Journal of Emergency Medicine,* Volume 35, Issue 4, Theodore C. Chan, Ghazala Q. Sharieff, William J. Brady, Electrocardiographic Manifestations: Pediatric ECG, Pages 421-430, Copyright 2008, with permission from Elsevier.

Figure 3. Electrocardiogram Of 6-Week-Old Patient With Ventricular Septal Defect, Showing Left Ventricular Hypertrophy And Left Axis Deviation
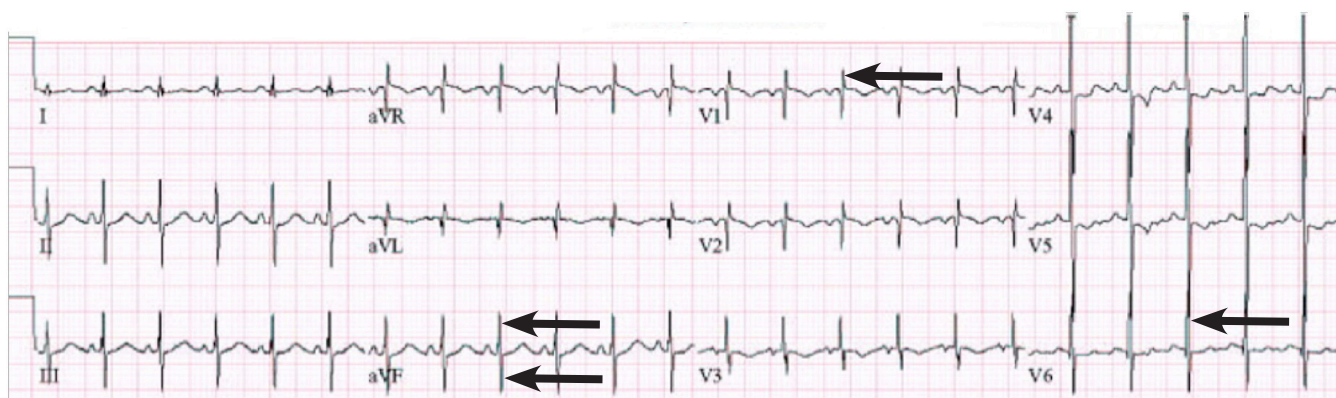
Left ventricular hypertrophy noted by increased S wave in V_1 and R wave in V_6. Left axis deviation noted by isoelectric or negative forces in aVF.
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quently depends on advanced imaging modalities (echocardiography, computed tomography, cardiac magnetic resonance imaging, magnetic resonance angiography, and diagnostic or interventional cardiac catheterization), the plain chest film can, in some circumstances, aid in the diagnosis and differential diagnosis of undiagnosed CHD and potential complications of corrected or palliated CHD.[38,39] Chest radiography may help to differentiate pulmonary or infectious causes of shock, cyanosis, or respiratory distress from CHD through careful evaluation of the airspace, cardiac silhouette, and pulmonary vascular markings. While focal airspace disease is typical of bacterial pneumonia, and diffuse airway inflammation may suggest viral lower respiratory tract disease, differentiating pulmonary edema from infection or inflammation can be difficult, and the conditions may coexist. **(See Figure 4.)**

Most significant CHD is associated with cardiomegaly (cardiothoracic ratio > 60%; **see Figure 5, page 14)**, and some forms of cyanotic CHD have the characteristic cardiac silhouette shapes, such as the boot shape of TOF or the egg-on-a-string shape of TGA, though these findings are not universal.[40] **(See Table 6, page 14.)** Assessing cardiac size can be challenging, particularly if the x-ray is limited to a single, portable anteroposterior view in which the heart appears artificially enlarged, or when the film is taken upon expiration, which can also increase the appearance of vascular markings.

Finally, for clinicians who view infant chest films infrequently, the appearance of the normal thymus may mimic cardiomegaly, pneumonia, atelectasis, or even a mediastinal mass, particularly if the posteroanterior or anteroposterior view is slightly rotated, obscuring the classic "sail sign" of the thymus. Obtaining a lateral view demonstrating the anterior position of the thymus can help distinguish these entities. **(See Figure 6, page 15.)**

Echocardiography

Echocardiography by an experienced clinician, such as a cardiologist, provides a more definitive diagnosis of the underlying cardiac lesion. The benefits of transthoracic echocardiography are that it can assess anatomy in the context of physiology and it is noninvasive. Doppler can be used to determine the direction of blood flow, estimate intracardiac pressures, visualize the function of the heart, and define the integrity of vessels. In addition, it gives information about possible vegetations, thrombi, and presence of pericardial fluid in the setting of postoperative CHD. Finally, ultrasound can be used to guide interventional procedures such as pericardiocentesis or emergent balloon atrial septostomy.[41]

Figure 4. Total Anomalous Pulmonary Venous Return Initially Complicated By Multilobar Pneumonia
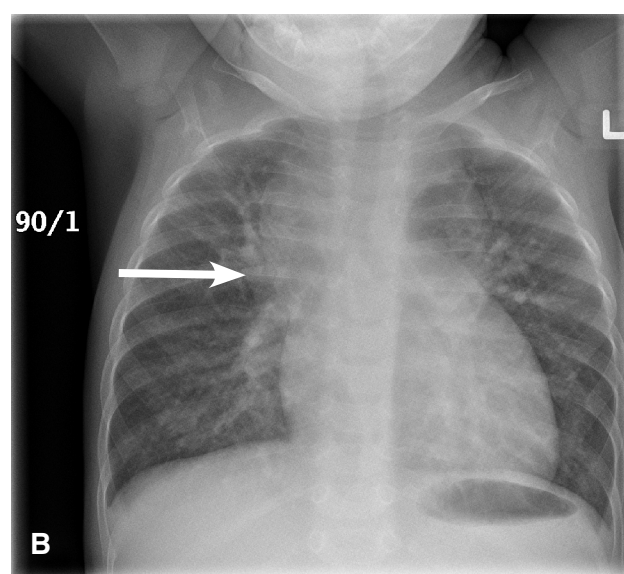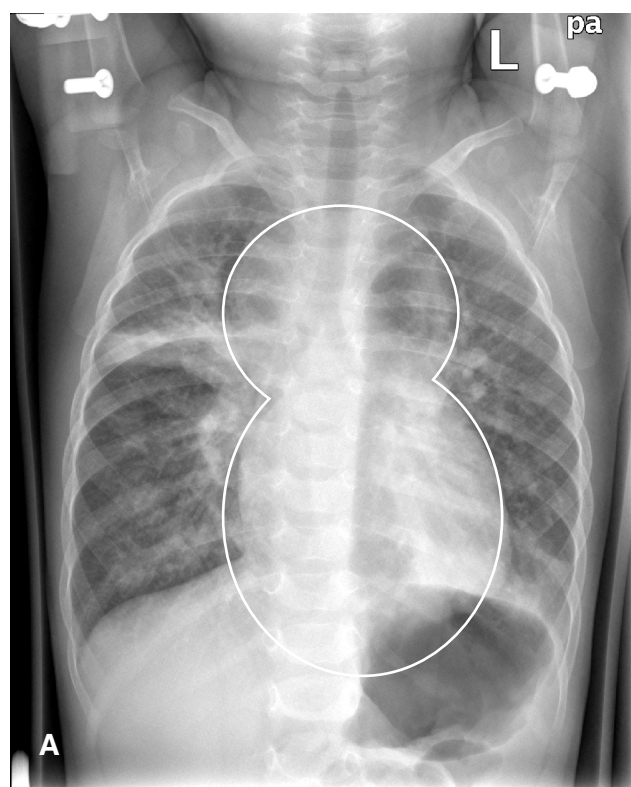
Subsequent resolution of pneumonia but persistent "snowman" configuration of upper mediastinum (View A) suggestive of obstructed pulmonary veins (View B).

Photos courtesy of Garth Meckler, MD.

Figure 5. Anteroposterior And Lateral Chest X-Ray Of An Infant With Anomalous Left Coronary Artery From The Pulmonary Artery
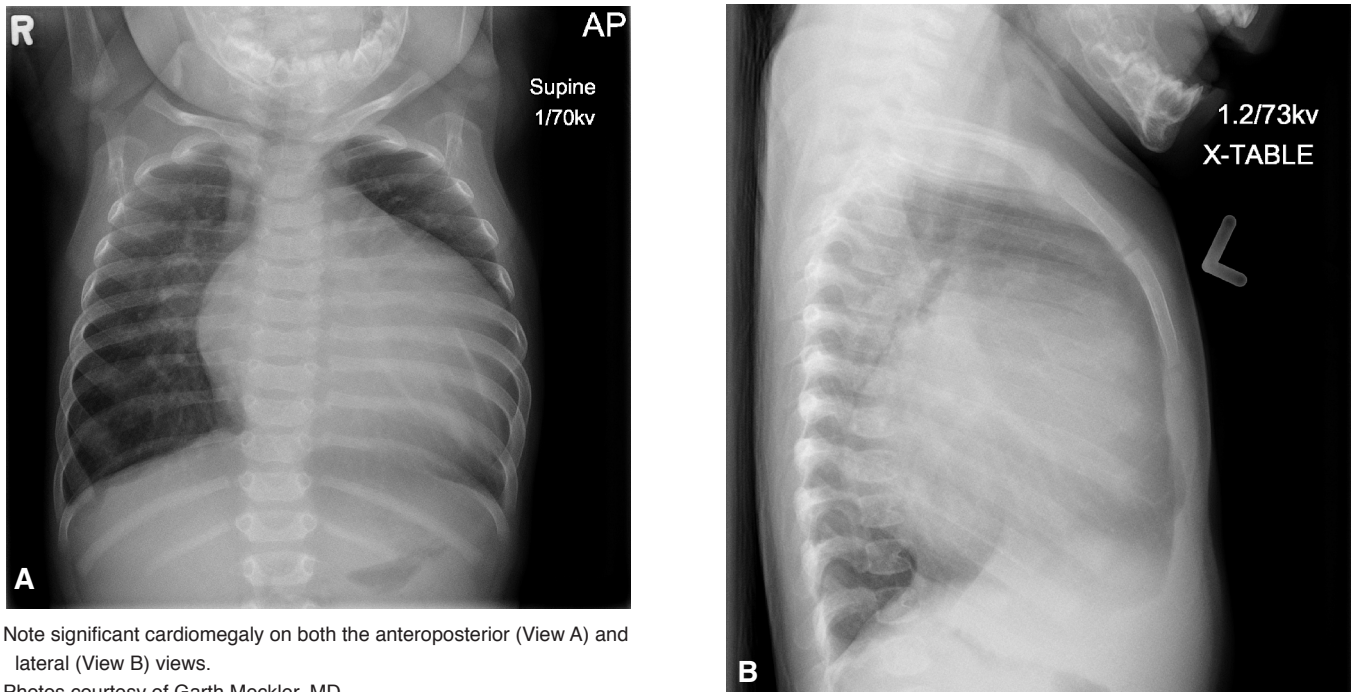
Note significant cardiomegaly on both the anteroposterior (View A) and lateral (View B) views.
Photos courtesy of Garth Meckler, MD.

Table 6. Possible Chest X-Ray And Electrocardiogram Findings In Congenital Heart Disease

Clinical Presentation	Heart Defect	Chest X-Ray Findings: Cardiac Silhouette	Chest X-Ray Findings: Pulmonary Vascular Markings	Electrocardiogram Findings
Cyanosis	TOF	Boot-shaped, normal size	Decreased	Right axis deviation, RVH
	TGA	"Egg-on-a-string" shaped (narrow mediastinum)	Increased	Right axis deviation, RVH
	TA	Normal to slight cardiomegaly	Decreased (increased with significant PDA)	Superior QRS axis with RAH, LAH, LVH
	Truncus arteriosus	Cardiomegaly of RV, absent thymus when associated with DiGeorge syndrome	Increased	Biventricular hypertrophy
	TAPVR	"Snowman sign," significant cardiomegaly	Increased	Right axis deviation, RVH, right atrial enlargement
Shock	AS	Cardiomegaly of LV	Normal or increased	LVH in severe cases
	CoA	Cardiomegaly	Normal or increased	RVH and RBBB (neonate), or LVH and rib-notching (child)
	HLHS	Small, normal, or enlarged	Increased	Right atrial enlargement, RVH, peaked P waves
	ALCAPA	Cardiomegaly	Increased	Pathologic Q waves in leads I, aVL, and V_4-V_6 with ST-segment elevations in V_4-V_6 (anterolateral infarct)
Congestive heart failure	PDA	Cardiomegaly of LA and LV	Increased	LVH, RVH with larger PDAs
	ASD	Cardiomegaly of RA and RV	Increased	Right axis deviation, RVH, RBBB
	VSD	Cardiomegaly of LA and LV	Increased	LAH, LVH, RVH with larger VSDs

Abbreviations: ALCAPA, anomalous left coronary artery from the pulmonary artery; AS, aortic stenosis; ASD, atrial septal defect; CoA, coarctation of the aorta; HLHS, hypoplastic left heart syndrome; LA, left atrium; LAH, left atrial hypertrophy; LV, left ventricle; LVH, left ventricular hypertrophy; PDA, patent ductus arteriosus; RA, right atrium; RAH, right atrial hypertrophy; RBBB, right bundle branch block; RV, right ventricle; RVH, right ventricular hypertrophy; TA, tricuspid atresia; TAPVR, total anomalous pulmonary venous return; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

Treatment

Though surgical correction or palliation represents definitive treatment of most CHD, emergent medical management is necessary to manage the patient until such options are available, and the ED approach is guided by the clinical presentation of the patient. In a critically ill child, early recognition and prompt, goal-directed therapy is necessary to avoid end-organ damage. The first steps are to optimize oxygenation and ventilation and establish vascular access, which may require insertion of an intraosseous line. If available, the intensive care team should be involved early and alerted to the possible need for extracorporeal life support.[42] After initial stabilization, specific medical therapies can be initiated in consultation with pediatric cardiology and cardiothoracic surgery, and arrangements can be made for rapid transfer to a tertiary pediatric hospital for definitive care. The ED management of patients with CHD presenting with shock, cyanosis, and CHF are detailed in the following sections and depicted in the Clinical Pathways on pages 8 and 9.

Shock And Congenital Heart Disease

Supplemental oxygen should be provided initially to any child presenting with shock, respiratory distress, or cyanosis. The clinical response must be carefully monitored, as neonates with ductal-dependent left ventricular outflow tract obstruction may decompensate with high FiO_2, and oxygen saturations < 100% may be acceptable (mid-70s are appropriate for single-ventricle physiology). Support the airway and breathing with positive-pressure ventilation as needed. Consider endotracheal intubation in cases of apnea or agonal respirations. Exercise caution, as the complex compensatory physiology and underlying pathophysiology may predispose the patient to decompensation with administration of rapid sequence intubation medications that can decrease preload. Mechanical ventilation may worsen metabolic acidosis that has been partially corrected by compensatory respiratory alkalosis.

Provide an initial intravenous or intraosseous fluid bolus of normal saline to any patient presenting with shock or with signs of poor perfusion. Fluid resuscitation will improve the clinical status of a child who has an etiology other than CHD, but it can worsen the status of neonates and children with cardiogenic shock, so it is prudent to begin with a 10-mL/kg bolus and reassess the clinical response before further fluid administration.

For neonates presenting with shock, consider empiric treatment with PGE_1, which may open the ductus arteriosus and provide life-sustaining systemic or pulmonary blood flow in cases of ductal-dependent CHD (such as CoA). Initiate PGE_1 as a continuous infusion, with a starting dose of 0.05 to 0.1 mcg/kg/min. PGE_1 can be started prior to echocardiography, based on clinical suspicion, and should result in clinical improvement (improved peripheral pulses and perfusion) within minutes in patients with ductal-dependent lesions.

There are few contraindications to PGE_1, and

Figure 6. Normal Infant Thymus May Be Mistaken For Cardiomegaly
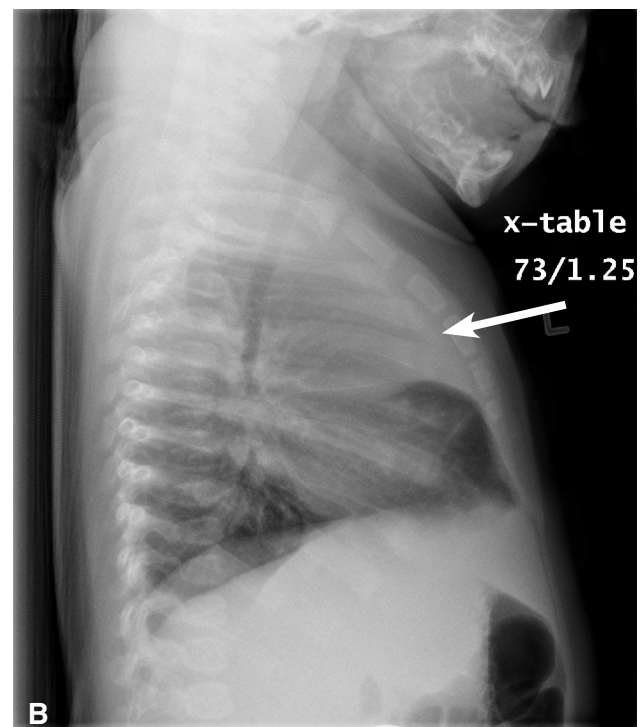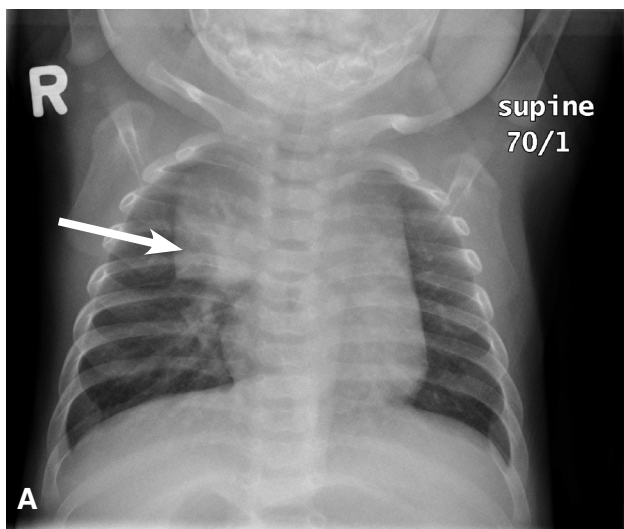
View A shows typical "sail sign" of thymus in the right upper chest on anteroposterior view (arrow).
View B shows interior position of thymus on lateral view (arrow).
Photos courtesy of Garth Meckler, MD.

side effects include local vasodilation (flushing), respiratory depression/apnea (12%),[43] fever, bradycardia, and jitteriness, but these are typically mild.[31] If starting at 0.1 mcg/kg/min, gradually titrate the rate of infusion to the lowest dose to maintain clinical response. Prophylactic intubation solely for the potential of respiratory depression/apnea or interfacility transport is not required.[44]

Given the broad differential diagnosis of the critically ill neonate presenting in shock, consider empiric treatment with broad-spectrum antibiotics for sepsis and administration of hydrocortisone for potential adrenal crisis from undiagnosed congenital adrenal hyperplasia. Treat hypoglycemia (from critical illness or undiagnosed metabolic disease) with 5 mL/kg 10% dextrose in normal saline peripherally, or 2 mL/kg 25% dextrose in normal saline if intraosseous or central access has been obtained.

Cyanosis And Congenital Heart Disease

The ED management of the pediatric patient presenting with cyanosis of unclear etiology is similar to the approach to the critically ill patient with shock. This includes cautious administration of oxygen, with close monitoring of the child's response to treatment (including the results of the hyperoxia test detailed previously), support of the airway and breathing, and provision of an initial intravenous fluid bolus. Respiratory etiologies of cyanosis should respond to these interventions with improvement in oxygenation. Failure to respond may be a clue to a cardiac etiology or methemoglobinemia and should prompt additional treatment. In the neonate with central cyanosis who is unresponsive to oxygen therapy, consider empiric administration of PGE_1 as discussed on page 15.

An exception to this general approach to cyanosis is the management of hypercyanotic episodes in neonates and children with TOF (Tet spells). TOF consists of 4 defects, including: (1) a misaligned VSD, (2) an overriding aorta, (3) right ventricular outflow tract obstruction, and (4) right ventricular hypertrophy. These patients are at risk for hypercyanotic episodes when they are agitated or crying, which triggers a catecholamine surge that can lead to infundibular spasm and decreased pulmonary blood flow.[45] Pulmonary outflow obstruction leads to increased right-to-left shunting through the VSD, further hypoxia and pulmonary vascular constriction, and additional release of catecholamines, which worsen the process and precipitate a vicious cycle of worsening hypoxemia. Severe Tet spells can continue to the point of syncope, seizures, or death.

The primary goal of treatment is to increase pulmonary blood flow through a combination of lowering the catecholamine effect on the infundibulum, increasing systemic vascular resistance (to reverse the right-to-left shunting across the VSD),

and decreasing hypoxemic pulmonary vasoconstriction. This is achieved through a progressive sequence of maneuvers, starting with calming the patient to decrease catecholamine release, providing a knee-to-chest position to increase systemic vascular resistance, and applying supplemental oxygen to promote pulmonary vasodilation. Additional interventions for refractory spells include intravenous fluids to increase preload, morphine (0.05-0.1 mg/kg) to decrease infundibular spasm, or phenylephrine to raise systemic vascular resistance.[46] Propranolol or esmolol can also be used to decrease right ventricular outflow spasm.[47] Sodium bicarbonate may help by increasing pulmonary vasodilation and decreasing pulmonary vascular resistance. Ketamine has been suggested and may have benefits related to both sedative effects and increased systemic vascular resistance.[26,48] One case report demonstrated efficacy of intranasal fentanyl as an alternative to morphine.[49]

Congestive Heart Failure And Congenital Heart Disease

CHF as an etiology for respiratory distress in the pediatric population requires a high index of suspicion in the presence of physical findings (gallop rhythm, hepatomegaly) and x-ray appearance (pulmonary vascular congestion and pulmonary edema with cardiomegaly). Severe pulmonary edema in CHF may be associated with hypoxia, and supplemental oxygen should be applied with the same caveats as described for shock, since oxygen is a potent pulmonary vasodilator and, in CHD, with overcirculation of the pulmonary circuit, oxygen may exacerbate symptoms. Similarly, exercise caution with fluid administration after carefully evaluating fluid status.

Medications
Medications include diuretics, angiotensin-converting enzyme (ACE) inhibitors, beta blockers, digoxin, and inotropes. Other than diuretics, these medications are best initiated after consultation with a pediatric cardiologist, as individual management preferences vary considerably.

Diuretics are the mainstay of acute management of CHF in the ED. A Cochrane systematic review of diuretics in CHF suggests that they reduce the risk of death and progression of CHF and improve exercise capacity in comparison to placebo.[50] Furosemide is the most commonly used diuretic, although there is no strong evidence to suggest its superiority over other agents.[8] The Canadian Cardiovascular Society Guidelines strongly recommend using furosemide in children with CHF if there is evidence of fluid overload, with a starting dose of 0.5 to 1 mg/kg intravenously or orally every 6 to 12 hours.[51] Though bumetanide is also used at times, there is little evidence for its use in pediatric CHF.[52]

There is no clear evidence for the use and benefit of inotropic infusions in the setting of acute CHF in children;[53] however, they should be considered as a temporizing measure when there is evidence of low cardiac output and shock.[51] Initial considerations include milrinone, dobutamine, and epinephrine.[51] Milrinone is an inotrope that has been shown to be effective in preventing low cardiac output syndrome following surgical repair of CHD in children.[54,55] Epinephrine improves cardiac contractility but increases myocardial oxygen demand, heart rate, and systemic vascular resistance, so it can have untoward effects, depending on the underlying physiology and compensatory status of a child with CHF.

Chronic management options for children with CHF include aldosterone inhibitors, ACE inhibitors, beta-adrenergic blockers, and digoxin; however, these agents play no role in the ED management of acute CHF.[8,56-62]

Special Populations

Complications Of Surgically Repaired/ Palliated Congenital Heart Disease

Though this issue focuses primarily on the emergency care of neonates and infants with undiagnosed CHD, reviews suggest that the majority of children presenting to the ED for emergency care have known CHD and are in various stages of surgical repair or palliation.[24,26] Most patients with previously diagnosed CHD present to the ED with common childhood illnesses such as upper respiratory tract infections or gastroenteritis; however, their underlying physiology often makes them susceptible to decompensation in the setting of increased pulmonary vascular resistance in respiratory tract infection, hypovolemia from vomiting or diarrhea, or fever.

Even without intercurrent illness, children with known CHD are at risk for certain complications related to their underlying anatomy and surgical history. Stenosis of vessels, valves, and surgical anastomoses may lead to decreased pulmonary or systemic blood flow, depending on the type of CHD and repair, resulting in increased cyanosis, CHF, or shock. Thrombosis of conduits can similarly precipitate acute decompensation. Dysrhythmias are commonly associated with some forms of CHD and certain surgical procedures. Infants and children with some forms of CHD (particularly left outflow tract obstruction) and heterografts are at increased risk for bacterial endocarditis. Children with persistent shunting are at increased risk for thromboembolic events, both pulmonary (pulmonary embolus) and systemic (eg, stroke).[19,63-65] Finally, children who take medications for their CHD are at risk for complications related to their medications, particularly in the setting of intercurrent viral illness. Though a complete review of the surgical approach to CHD is beyond the scope of this article, **Table 7, page 18,** provides an overview of some of the more common surgical procedures and their potential complications in complex CHD.

Emergency Management Of Surgically Repaired/ Palliated Congenital Heart Disease Presenting With Acute Decompensation

The ED approach to the acutely ill infant or child with known and surgically corrected or palliated CHD depends on the surgical anatomy and physiology of the patient and the nature of the acute insult. Upper or lower respiratory tract infection can precipitate hypoxia through pulmonary hypertension/vasoconstriction and should be treated initially with increased supplemental oxygen. Target oxygen therapy to baseline saturations, if known. For example, patients with mixing lesions, such as a single-ventricle patient at the Glenn stage, will have baseline saturations in the low 80s. Poor oral intake or gastro-intestinal illness causing dehydration can lead to poor systemic or pulmonary perfusion, depending on the anatomy, and should be addressed with intravenous fluid boluses of 10 mL/kg, with careful reassessment. A sudden and catastrophic decompensation in perfusion or oxygenation in a child with a surgical conduit (eg, Blalock-Taussig shunt) may represent conduit occlusion or thrombosis. In these cases, the expected shunt murmur may be absent and the patient may be in extremis. An initial intravenous fluid bolus should be initiated, and cardiology, the intensive care unit, and cardiac surgery should be consulted, as definitive management is often surgical, though thrombolysis may be considered as a temporizing measure.

Anomalous Origin Of The Left Coronary Artery From The Pulmonary Artery

ALCAPA results in poor blood supply to the left ventricle and can present during the neonatal or infant period as shock or CHF, or later in childhood with symptoms of exercise-induced angina, myocardial ischemia or infarct, or progressive CHF. After birth, as the pressure in the pulmonary artery falls, the perfusion pressure to the left coronary artery drops, resulting in myocardial ischemia and eventual infarction. At this stage, blood from the left coronary artery begins to flow into the pulmonary artery and myocardial steal syndrome occurs. In some cases, interarterial collateral anastomoses develop between the coronary arteries, and this may impact the age at which a patient presents.[9]

Neonates can present with symptoms of CHF in the context of a respiratory illness. Recurrent episodes of angina in preverbal infants may manifest as irritability (sometimes described as a high-pitched cry, often during feeding or episodes of fussiness), diaphoresis, pallor, and respiratory distress. Older children may present with more typical angina (chest pain) during exercise, and there may be a continuous murmur and gradual progression of CHF.

The continuous murmur represents a left-to-right shunt, where blood flows from the aorta to the right coronary artery, to the left coronary artery, and then to the pulmonary artery.

An infant in CHF presenting with respiratory distress or shock will have cardiomegaly on chest x-ray.

(See Figure 5, page 14.) The ECG may demonstrate a classic anterolateral wall infarction pattern, with a QR pattern with inverted T waves in leads I and aVL, and deep Q waves, elevated ST segments, and inverted T waves in leads V_5 and V_6.[66,67] (See Figure 7, page 19.) Emergent surgical correction is indicated.

Table 7. Common Surgical Procedures And Their Complications In Complex Congenital Heart Disease

Type of CHD	Surgical Procedure (Timing)	Repair	Postsurgical Complications	Comments
TGA	Arterial switch (at birth)	Switch of the aortic and pulmonary trunks, leaving original valve; coronary arteries reimplanted	Myocardial ischemia	N/A
	Rastelli (at birth)	RV to PA conduit and VSD closed with slight LV protrusion into RV	Supravalvular stenosis, atrial dysrhythmias	Typically performed in patients with LVOT obstruction
	Mustard/Senning (at birth)	Atrial switch with prosthetic (mustard) or native (Senning) intra-atrial baffle	Atrial dysrhythmias, ventricular failure	Rarely used, RV becomes systemic ventricle, LV becomes pulmonary ventricle
HLHS or tricuspid atresia	Staged repair: 1. Norwood (HLHS only) **+** BTS **or** Sano modification (at birth)	Norwood: Neo-aorta created from aorta and part of PA with ligation of main PA BTS: right subclavian artery to right PA conduit Sano: RV to PA conduit	Atrial arrhythmias, sudden death, thromboembolism (pulmonary > arterial), thrombosis of shunt, elevated CVP (SVC syndrome, ascites, hepatomegaly), pericardial or pleural effusion, protein-losing enteropathy	Single-ventricle physiology with mixing of blood through ASD and POX 75%-85% expected
	2. Glenn (age 6 months)	BTS or Sano taken down, SVC anastomosed to right PA or main PA (bidirectional Glenn)		POX 75%-85% expected
	3. Fontan (age 1.5 years)	IVC connected to PA		At this stage, passive blood flow to lungs, single ventricle pumps to body, mixing no longer occurs, POX normal
CoA, AS	Balloon dilation or surgical repair (timing variable)	Can include end-to-end anastomosis, patch repair, balloon dilation, or aortoplasty	Re-stenosis at original site or surgical anastomosis	N/A
Truncus arteriosus	Primary repair (around age 2 weeks)	Pulmonary arteries removed from common trunk and attached via conduit to RV and closure of VSD	Valvular insufficiency, dysrhythmias (RBBB and heart block), outgrown conduit	Pulmonary edema and hypertension from valvular insufficiency, hypoxia if conduit is outgrown due to decreased pulmonary blood flow
TOF	Complete repair +/- BTS (around age 6 months)	Augmentation of PA and valve, closure of VSD +/- right subclavian to right PA conduit (BTS)	Residual VSD, RV failure, conduction defects, valvular insufficiency	N/A

Abbreviations: AS, aortic stenosis; ASD, atrial septal defect; BTS, Blalock-Taussig shunt; CHD, congenital heart disease; CoA, coarctation of the aorta; CVP, central venous pressure; HLHS, hypoplastic left heart syndrome; IVC, inferior vena cava; LV, left ventricle; LVOT, left ventricular outflow tract; N/A, not applicable; PA, pulmonary artery; POX, pulse oximetry; RBBB, right bundle branch block; RV, right ventricle; SVC, superior vena cava; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

Heterotaxy Syndromes

Situs solitus describes the normal anatomical alignment of the lungs and abdominal viscera. With situs inversus, abdominal organs and lung lobation are reversed, with the right atrium positioned on the left and the left atrium on the right. This can result in heterotaxy, with either right or left isomerism. Right isomerism is characterized by a central liver, absent spleen (asplenia), and 2 morphological right lungs. Left isomerism is described by absence of the intrahepatic portion of the inferior vena cava, multiple small spleens (polysplenia), and 2 morphological left lungs.[68]

These heterotaxy syndromes are usually associated with complex CHD. The most important consideration for the emergency clinician is to recognize the risk of serious infection as a result of poor splenic function, and consider early administration of antimicrobial therapy in the setting of fever. It is also important to recognize the association between heterotaxy syndromes and complex CHD.

Aortic Arch Abnormalities And Pulmonary Slings

Vascular rings and pulmonary slings represent approximately 1% to 1.6% of all CHD.[69] Vascular rings are anatomical abnormalities wherein the trachea and esophagus are completely surrounded by an abnormal vascular structure originating from the aortic arch. **(See Figure 8, page 20.)** Examples of vascular rings include double aortic arch, right aortic arch with a left ligamentum arteriosum, aberrant left subclavian artery, and aberrant innominate artery. A pulmonary sling occurs when the left pulmonary artery arises from the right pulmonary artery and passes leftward between the trachea and the esophagus, potentially compressing the trachea and right mainstem bronchus.

Both vascular rings and pulmonary slings may present with subtle symptoms of airway obstruction (stridor, wheeze, difficulty feeding, respiratory distress), often in the context of or exacerbated by upper or lower respiratory tract infection. Consider these diagnoses in infants who present with recurrent symptoms that do not respond to usual treatment. Chest x-ray may demonstrate a right-sided aortic arch, but definitive diagnosis often requires advanced imaging such as magnetic resonance imaging, computed tomography, esophagram, or angiography.[38] Treatment of symptomatic vascular rings or slings is surgical.[69] Advanced imaging and surgical repair occur outside the ED.

Figure 7. Electrocardiogram Of 5-Month-Old Patient With Anomalous Origin Of The Left Coronary Artery From The Pulmonary Artery
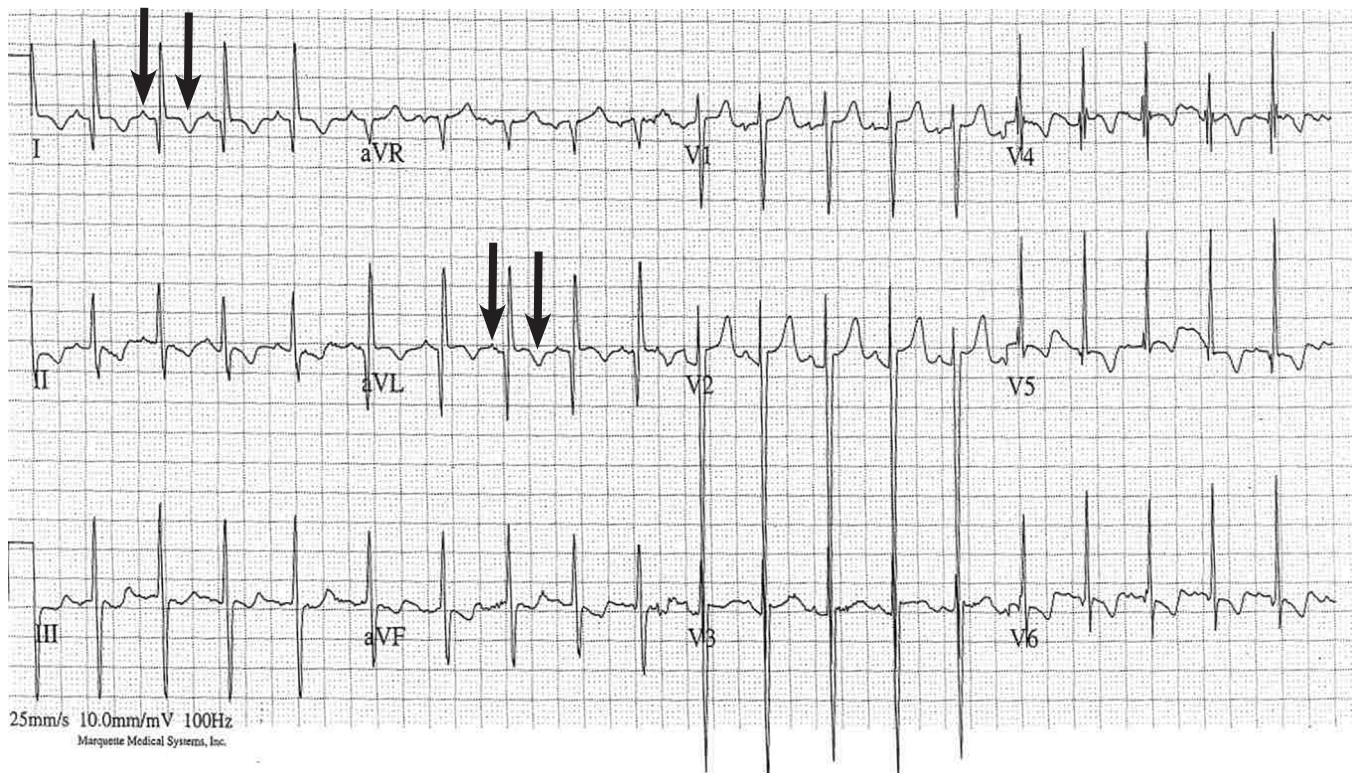
Q waves and T-wave inversion can be seen in the lateral leads, I and aVL. Though not seen in this patient, other classic findings in ALCAPA are abnormal Q waves in precordial leads V_3-V_6.

Used with permission from Emergency Presentation Of Congenital Heart Disease In Children, *Pediatric Emergency Medicine Practice*, Christopher W. Mastropietro, Susan P. Tourner, Ashok P. Sarnaik, May 2008.

Pulmonary Hypertension

Patients with either corrected or uncorrected CHD can develop pulmonary hypertension with increased pulmonary vascular resistance that predisposes them to pulmonary hypertensive crises during conditions of hypercarbia, hypoxia, and/or acidosis. The goal in managing pulmonary hypertension is to reduce right ventricular afterload by ensuring adequate ventilation, providing supplemental oxygen, correcting acidosis, and keeping patients calm.[69] While some forms of pulmonary hypertension are progressive and without cure, others may be reversible with nitric oxide or medications such as calcium-channel blockers, prostacyclin infusions, and oral pulmonary vasodilators. Oral pulmonary vasodilators include bosentan (an endothelin receptor antagonist) and sildenafil (a phosphodiesterase type 5 inhibitor). Occasionally, patients with pulmonary hypertension and a history of thromboemboli may require anticoagulation. All of these medical therapies are temporizing, and definitive therapy typical requires heart-lung or lung transplant.[70]

Dysrhythmias

Children with corrected or unrepaired CHD may present to the ED with dysrhythmias, and those with CHF or previous myocardial surgery are at higher risk.[67] In addition, prolonged dysrhythmias can lead to CHF. Dysrhythmias can be classified as narrow complex (supraventricular dysrhythmias) or wide complex (ventricular dysrhythmias). In a stable patient without evidence of end-organ dysfunction, supraventricular tachycardia is initially managed with vagal maneuvers, or with a rapid push of intravenous adenosine (0.1 mg/kg/dose, maximum of 6 mg; can be increased to 0.2 mg/kg/dose, maximum of 12 mg). If the patient shows signs of end-organ dysfunction (hypoxia, poor perfusion, altered mental status, or respiratory distress), treatment is with prompt synchronized cardioversion (0.5 to 1 J/kg). For ventricular tachycardia, adenosine can be considered for stable patients, but consultation with cardiology is advised for alternative treatments, including amiodarone or procainamide. Unstable ventricular tachycardia is treated with synchronized cardioversion at 0.5 to 1 J/kg (can increase to 2 J/kg).[71]

Controversies And Cutting Edge

Oxygen In Congenital Heart Disease

Most emergency clinicians instinctively provide supplemental oxygen in the setting of hypoxia or critical illness, and it is often an important adjunct to improve oxygen delivery to critical organs and tissues. In the setting of CHD, however, it is important to be aware of the fact that oxygen is a medication with powerful vasodilatory effects on the pulmonary vasculature. In complex CHD, undiagnosed or palliated, the balance between pulmonary and systemic vascular circulation through anomalous or surgically created shunts or conduits depends on the relative pulmonary and systemic vascular resistance.[19,65,72] In

Figure 8. Right-Sided Aortic Arch And Vascular Ring
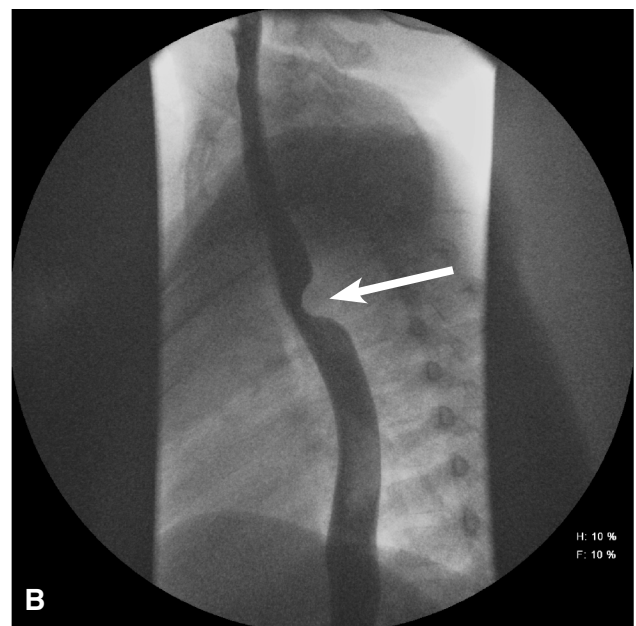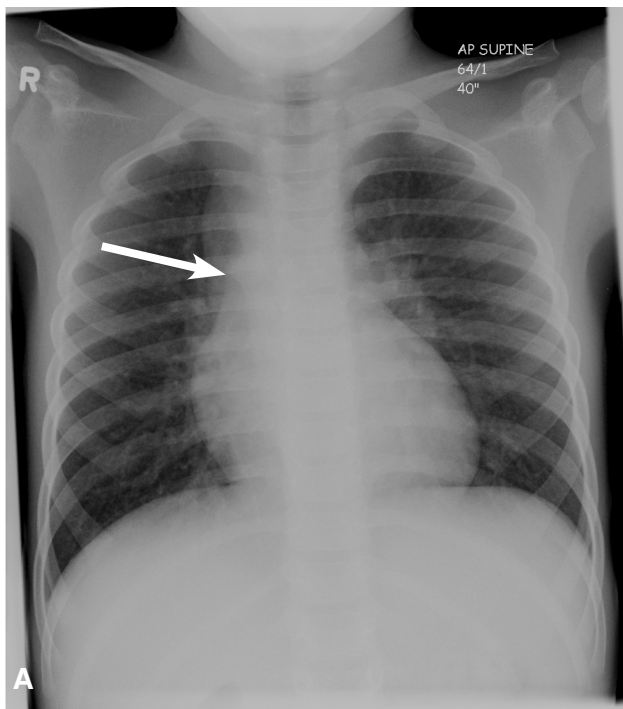
View A shows right-sided aortic arch.
View B shows vascular ring causing compression of the esophagus (arrow) demonstrated during fluoroscopic feeding study.
Photos courtesy of Garth Meckler, MD.

some circumstances, such as pulmonary valve or artery stenosis, Tet spells, or patients with a completed Fontan procedure and passive pulmonary perfusion experiencing hypoxemia from decreased pulmonary blood flow, oxygen therapy is indicated. In other situations, such as in infants with pulmonary vascular overcirculation and CHF from large left-to-right shunts, or infants with ductal-dependent systemic circulation (such as HLHS or critical CoA), supplemental oxygen may worsen symptoms through its vasodilatory effects on the pulmonary vasculature, increasing pulmonary edema in the former, and causing "pulmonary steal" with diminished systemic perfusion in the latter. The goal should be to target baseline oxygen saturations in patients with known and partially repaired CHD (eg, 75%-85% in those with mixing lesions, Blalock-Taussig shunts, or Glenn shunts), and target 90% to 95% in neonates and young infants without a clear diagnosis.

Intubation And Positive-Pressure Ventilation

Endotracheal intubation should be approached cautiously and with a well-thought-out management plan in a child with CHD in consultation with cardiology and the intensive care unit. Anatomical anomalies associated with CHD or genetic syndromes can make bag-mask ventilation and endotracheal intubation difficult. The sedation used and the vagal effects of endotracheal intubation can be very hazardous in such a decompensated child. Adult literature suggests that high FiO_2 bag-mask ventilation followed by noninvasive positive-pressure ventilation is well tolerated in patients with acute CHF.[73,74] However, children with CHD have higher pulmonary vascular resistance and are more predisposed to pulmonary hypertensive crisis. Once the airway is secure, positive-pressure ventilation can alter hemodynamics by decreasing venous return or preload to the heart, increasing pulmonary vascular resistance or right ventricular afterload, decreasing left ventricle afterload, and decreasing oxygen demand by supporting ventilation.

Prostaglandin E1

As discussed previously, PGE_1 can be life-saving for neonates with ductal-dependent pulmonary or systemic circulation in the setting of CHD and should be used empirically when the diagnosis is strongly suspected in a critically ill neonate. Controversy exists over the ideal dosing, and while the original studies started with 0.1 mcg/kg/min, there is increasing evidence that initiating therapy at lower doses, such as 0.02 to 0.05 mcg/kg/min, may be equally effective. Though there is no clear correlation between infusion dose and the risk of side effects (including apnea), lower doses may be associated with less risk.[75,76] Finally, many textbooks and review articles recommend prophylactic intubation of all neonates receiving PGE_1 prior to transport, out of fear for potential apnea, but a retrospective study of interfacility transports of children receiving PGE_1 found an increased risk for adverse events among those who were intubated prophylactically compared to those transported without intubation.[44] Given the risks associated with rapid sequence intubation medications and endotracheal intubation in these fragile patients, intubation solely for transport is likely not necessary and may be harmful.

Subacute Bacterial Endocarditis And Antibiotic Prophylaxis

While some infants and children with CHD are at increased risk for subacute bacterial endocarditis (SBE), this is not true for all congenital or repaired lesions. A large population-based analysis of SBE in children with CHD found an overall cumulative incidence between birth and 18 years of age of 6.1/1000 children. Lesions at highest risk included cyanotic CHD, endocardial cushion defects, and left-sided lesions. Cardiac surgery within the previous 6 months and age < 3 years also conferred risk among children with CHD. By contrast, ASD, VSD, PDA, and right-sided lesions were at low risk.[77]

Table 8 summarizes the antibiotic prophylaxis recommendations for patients with CHD.[78] Invasive dental or respiratory procedures and urinary catheterization or skin procedures in the setting of infection should receive prophylaxis. Obtaining peripheral intravenous access, administering subcutaneous or intramuscular injections, placing an intraosseous line, or utilizing urinary catheterization in patients without infection does not require prophylaxis.[65,78,79]

Table 8. Indications And Antibiotics For Endocarditis Prophylaxis In Congenital Heart Disease[78]

Indications	Antibiotic Prophylaxis*
• Unrepaired cyanotic CHD • CHD with palliative shunts and conduits (eg, BTS, Glenn, Fontan) • Completely repaired CHD with synthetic material or device (both surgical or catheter-inserted) for the first 6 months following the repair • Repaired CHD with residual defects at or adjacent to the site of prosthetic patch or device • Prosthetic valves • Cardiac transplant	Oral: • Amoxicillin 50 mg/kg **or** • Cephalexin 50 mg/kg **or** • Clindamycin 20 mg/kg **or** • Azithromycin 15 mg/kg Parenteral (IV/IM): • Ampicillin 50 mg/kg **or** • Cefazolin or ceftriaxone 50 mg/kg **or** • Clindamycin 20 mg/kg

*Administer single dose, 30-60 min prior to procedure.
Abbreviations: BTS, Blalock-Taussig shunt; CHD, congenital heart disease; IM, intramuscular; IV, intravenous.

Disposition

All children with suspected or ED-diagnosed CHD needing resuscitation require admission to the hospital, typically to an intensive care unit. This may entail transfer to a pediatric tertiary care hospital, ideally by a specialized pediatric transport team. Patients with mild CHF or increased cyanosis who are hemodynamically stable may be candidates for admission to a pediatric hospital ward, preferably a unit capable of continuous cardiac monitoring. Patients who are hemodynamically stable and have normal or baseline oxygen saturations (if surgically repaired or palliated) can be considered for discharge and outpatient cardiology follow-up. The disposition of patients with complex CHD should be discussed with pediatric cardiology, as there is great variability in individual anatomy and physiology across the spectrum of CHD and individual considerations are often best known by the treating cardiologist.

Summary

CHD includes a spectrum of anatomic malformations of the heart and great vessels, and while many defects are identified prenatally through fetal ultrasound or diagnosed in the newborn period prior to discharge from the hospital, some CHD may not manifest with signs and symptoms until the infant or child is older. The emergency clinician must maintain a high index of suspicion in these rare cases, as the clinical picture of undiagnosed CHD can be nonspecific, can mimic other common and be-

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Risk Management Pitfalls In Pediatric Congenital Heart Disease
(Continued on page 23)

1. **"This neonate had normal prenatal care, including a prenatal ultrasound, so CHD has been ruled out. There must be another cause for his shock."**
   While prenatal ultrasound has advanced significantly over recent decades, only about one-third of all CHD and 57% to 85% of critical CHD are detected before birth. Normal prenatal care and screening ultrasound do not exclude the possibility of significant CHD.

2. **"I don't hear a murmur or a gallop, so this isn't CHD."**
   The absence of abnormal heart sounds does not preclude underlying structural disease. A murmur requires turbulent blood flow across a defect, usually from a significant pressure gradient. In the first days of life, high pulmonary vascular resistance can minimize left-to-right shunting across a large ASD or VSD, and a murmur may not be detected prior to discharge from the nursery.

3. **"Although this 1-week-old is in shock, we can't get an echocardiogram, and I'm not sure what is going on, so I don't want to start PGE_1 until we have more information. I'll just fluid resuscitate…."**
   In the critically ill neonate presenting with shock, PGE_1 can be life-saving and should be empirically initiated if there is no response to an initial 10-mL/kg bolus of intravenous fluids. Careful monitoring of clinical response is all that is needed and the infusion can be stopped if the clinical condition worsens.

4. **"All children with significant hypoxia require 100% FiO_2 to normalize oxygenation."**
   While oxygen can be beneficial and is first-line therapy for many conditions associated with hypoxia or poor perfusion, its potent pulmonary vasodilatory effects must be considered in the context of CHD with significant shunting lesions, as decreased pulmonary vascular resistance can lead to worsening pulmonary edema or decreased systemic perfusion as a result of exacerbation of left-to-right shunting. Baseline oxygen saturations should be targeted in patients with complex CHD, if the baseline is known, and oxygen saturation of 90% to 95% should be targeted if the baseline is unknown. Wean oxygen if clinical deterioration is observed after initiation of therapy.

5. **"Although he is breathing on his own, this child with a Fontan procedure and gastroenteritis is hypoxic and tachypneic, so I should intubate. I don't expect a difficult airway."**
   While intubation may be required for infants and children with apnea or agonal respirations, the switch to positive-pressure ventilation and the vascular and cardiac effects of preintubation medications must be carefully considered in patients with complex CHD who may be dependent on preload. In addition, airway anomalies may be associated with some CHD. Consultation with anesthesia or cardiology is recommended in all but the most emergent cases in which intubation is considered.

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nign childhood disease, or can present in a child in extremis. Undiagnosed CHD may present with signs of shock in neonates with ductal-dependent cardiac malformations in the first weeks of life. This catastrophic presentation may be difficult to distinguish from other neonatal critical illness, such as sepsis or metabolic disease, but requires unique resuscitation priorities, including administration of PGE_1 to maintain ductal patency. Cyanosis may be the presenting symptom of undiagnosed CHD with restricted pulmonary blood flow or right-to-left shunting, or may be the result of decreased pulmonary perfusion in patients with surgically palliated complex CHD in the setting of concurrent illness. Treatment focuses on support of the airway and breathing and provision of an initial intravenous fluid bolus. CHD presenting with CHF can mimic common viral lower respiratory tract infection, such as bronchiolitis or

pneumonia, and requires a high index of suspicion for diagnosis. Treatment involves diuretics and optimization of cardiac output in consultation with a pediatric cardiologist.

Case Conclusions

Although sepsis, metabolic disease, and CHD can all cause shock and present in a neonate in extremis, your examination of the 8-day-old revealed a significant difference between the strength of the right brachial pulses and the femoral pulses, with some femoral delay. There was also a difference between upper and lower extremity blood pressures: right arm, 80/45 mm Hg; left leg, 40/20 mm Hg. Given the critical nature of the presentation, you decided to administer broad-spectrum antibiotics empirically, but your suspicion for a ductal-dependent cardiac defect was

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Risk Management Pitfalls In Pediatric Congenital Heart Disease
(Continued from page 22)

6. **"This is the fourth bad case of bronchiolitis I've had this shift! She's getting worse despite intravenous fluids, so I'll just admit her and try nebulized epinephrine."**
   CHD presenting with CHF can mimic common viral illness such as bronchiolitis, and, during epidemics, it is easy to overlook heart disease as a cause of respiratory distress in an infant. Worsening of clinical condition with usual treatment (such as intravenous fluids for presumed dehydration in bronchiolitis) should alert you to the possibility of CHF, for which diuretics are first-line therapy. A BNP and chest x-ray may help in these circumstances.

7. **"I'm going to start PGE_1 on this neonate with suspected CoA and transfer him to a children's hospital. I'd better intubate prior to transport in case he develops apnea, even though he is breathing well on his own now."**
   Although often recommended in textbooks, prophylactic intubation is likely not necessary in the absence of observed apnea or agonal respirations prior to transport. One study found a higher rate of adverse events among neonates on PGE_1 who were prophylactically intubated compared to those who were not intubated for transport.[44]

8. **"The nurse got an ECG at triage for this 6-year-old with chest pain and it looks like ischemia! That's impossible in a child with no past medical history, so it must be a technical error."**
   Although rare, children can develop myocardial ischemia or infarct from ALCAPA. Though ALCAPA typically presents in early infancy, it can escape detection and present later in life with acute myocardial infarction or progressive CHF from recurrent ischemia.

9. **"I need to refer this 2-year-old with TOF to a pediatric dentist for outpatient extraction of multiple carious teeth. Her last surgery was > 6 months ago, so I don't think she needs antibiotic prophylaxis prior to oral surgery."**
   The 2010 American Heart Association guidelines on antibiotic prophylaxis for bacterial endocarditis eliminated many of the indications for prophylaxis, but children with cyanotic CHD and allografts are at higher risk for SBE and should receive preprocedural antibiotic prophylaxis, with a single dose of oral or parenteral antibiotics 30 to 60 minutes prior to the procedure.

10. **"This 5-month-old has a systolic ejection murmur and a slight diastolic rumble, but I think it is an innocent murmur. She doesn't require cardiology referral."**
    A systolic ejection murmur can be benign and a common finding in many children, but a diastolic murmur is usually pathologic and should be referred to a cardiologist for further evaluation.

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sufficiently high from your physical examination that you started a PGE_1 infusion at 0.05 mcg/kg/min. Within 10 minutes, you noted improved peripheral circulation. You consulted the cardiologist, who performed a bedside echocardiogram and identified a critical coarctation of the aorta. The patient was admitted to the pediatric ICU and subsequently underwent surgical correction of his CHD.

For the 3-month-old, you suspected CHF as a cause for the respiratory symptoms, despite the prevalence of bronchiolitis this time of year, given the cardiomegaly and apparent pulmonary edema on chest x-ray and the patient's feeding history and failure to thrive. You ordered a BNP that returned abnormally elevated, further supporting your diagnosis. You obtained intravenous access and administered 1 mg/kg IV furosemide and consulted cardiology. Within 30 minutes, the patient had significant urine output and the respiratory rate decreased from 60 to 40 breaths/min with a slight improvement of oxygen saturations to 94% on room air. An echocardiogram revealed a large VSD with left-to-right shunting. Cardiology thanked you for your clinical acumen and admitted the patient to the hospital ward for further management and digitalization.

Time- And Cost-Effective Strategies

- Consider intranasal fentanyl or intramuscular or subcutaneous morphine for a hypercyanotic Tet spell, if intravenous access will cause significant delay. These routes of administration are rapid and available without the need for intravenous access, which can be difficult in the critically ill infant or child. Moreover, obtaining intravenous access can exacerbate a Tet spell by further upsetting the child.
- Proceed to early placement of an intraosseous needle in the critically ill neonate, infant, or child requiring resuscitation or medication when intravenous access is difficult or time consuming. Intraosseous access is rapid, safe, and can be used to administer intravenous fluids and all vasoactive medications.

Abbreviations

ALCAPA: Anomalous left coronary artery from the pulmonary artery
ASD: Atrial septal defect
AVSD: Atrioventricular septal defect
CHD: Congenital heart disease
CHF: Congestive heart failure
CoA: Coarctation of the aorta
HLHS: Hypoplastic left heart syndrome
PA: Pulmonary atresia
PDA: Patent ductus arteriosus
PGE[1]: Prostaglandin E_1
TA: Tricuspid atresia
TAPVR: Total anomalous pulmonary venous return
TGA: Transposition of the great arteries
TOF: Tetralogy of Fallot
VSD: Ventricular septal defect

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 are included in bold type following the references, where available. The most informative references cited in this paper, as determined by the authors, are noted by an asterisk (*) next to the number of the reference.

1. van der Bom T, Zomer AC, Zwinderman AH, et al. The changing epidemiology of congenital heart disease. *Nat Rev Cardiol*. 2011;8(1):50-60. **(Review article)**
2. van der Linde D, Konings EE, Slager MA, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. *J Am Coll Cardiol*. 2011;58(21):2241-2247. **(Systematic review and meta-analysis; 114 studies)**
3. Fahed AC, Roberts AE, Mital S, et al. Heart failure in congenital heart disease: a confluence of acquired and congenital. *Heart Fail Clin*. 2014;10(1):219-227. **(Review article)**
4. Bajolle F, Zaffran S, Bonnet D. Genetics and embryological mechanisms of congenital heart diseases. *Arch Cardiovasc Dis*. 2009;102(1):59-63. **(Review article)**
5.* Jenkins KJ, Correa A, Feinstein JA, et al. Noninherited risk factors and congenital cardiovascular defects: current knowledge: a scientific statement from the American Heart Association Council on Cardiovascular Disease in the Young: endorsed by the American Academy of Pediatrics. *Circulation*. 2007;115(23):2995-3014. **(Scientific statement)**
6. Miller A, Riehle-Colarusso T, Siffel C, et al. Maternal age and prevalence of isolated congenital heart defects in an urban area of the United States. *Am J Med Genet A*. 2011;155a(9):2137-2145. **(Cohort study; 5289 infants and 1,301,143 live births)**
7. Fung A, Manlhiot C, Naik S, et al. Impact of prenatal risk fac-

tors on congenital heart disease in the current era. *J Am Heart Assoc.* 2013;2(3):e000064. **(Case control study; 2339 cases and 199 controls)**

8. Dedieu N, Burch M. Understanding and treating heart failure in children. *Paediatr Child Health.*23(2):47-52. **(Review article)**

9.* Brown K. The infant with undiagnosed cardiac disease in the emergency department. *Clin Pediatr Emerg Med.* 2005;6(4):200-206. **(Review article)**

10. Sinha IP, Mayell SJ, Halfhide C. Pulse oximetry in children. *Arch Dis Child Educ Pract Ed*. 2014;99(3):117-118. **(Review article)**

11. Fouzas S, Priftis KN, Anthracopoulos MB. Pulse oximetry in pediatric practice. *Pediatrics.* 2011;128(4):740-752. **(Review article)**

12. Yee L. Cardiac emergencies in the first year of life. *Emerg Med Clin North Am.* 2007;25(4):981-1008. **(Review article)**

13. Kleinman ME, Chameides L, Schexnayder SM, et al. Pediatric advanced life support: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care. *Pediatrics*. 2010;126(5):e1361-e1399. **(Guidelines)**

14. Gerritse BM, Draaisma JM, Schalkwijk A, et al. Should EMS-paramedics perform paediatric tracheal intubation in the field? *Resuscitation*. 2008;79(2):225-229. **(Prospective observational study of pediatric intubations by EMS vs specialized air transport team)**

15. Wang HE, Lave JR, Sirio CA, et al. Paramedic intubation errors: isolated events or symptoms of larger problems? *Health Aff (Millwood)*. 2006;25(2):501-509. **(Retrospective study of pediatric EMS intubations)**

16. Gausche M, Lewis RJ, Stratton SJ, et al. Effect of out-of-hospital pediatric endotracheal intubation on survival and neurological outcome: a controlled clinical trial. *JAMA*. 2000;283(6):783-790. **(Controlled prospective trial of intubation vs BVM; 830 consecutive pediatric patients treated by EMS)**

17. Young KD, Gausche-Hill M, McClung CD, et al. A prospective, population-based study of the epidemiology and outcome of out-of-hospital pediatric cardiopulmonary arrest. *Pediatrics*. 2004;114(1):157-164. **(Secondary analysis of data; 599 patients from above prospective interventional trial of pediatric EMS airway management)**

18. Lee C, Mason LJ. Pediatric cardiac emergencies. *Anesthesiol Clin North America*. 2001;19(2):287-308. **(Review article)**

19.* Szlam S, Dejanovich B, Ramirez R, et al. Congenital heart disease: complications before and after surgical repair. *Clin Pediatr Emerg Med.* 2012;13(2):65-80. **(Review article)**

20. Orr RA, Felmet KA, Han Y, et al. Pediatric specialized transport teams are associated with improved outcomes. *Pediatrics*. 2009;124(1):40-48. **(Prospective single-center cohort study; 1085 pediatric transports)**

21. Edge WE, Kanter RK, Weigle CG, et al. Reduction of morbidity in interhospital transport by specialized pediatric staff. *Crit Care Med*. 1994;22(7):1186-1191. **(Concurrent prospective comparison of 2 centers with different transport teams and 141 total transports)**

22. Singh JM, MacDonald RD, Ahghari M. Critical events during land-based interfacility transport. *Ann Emerg Med*. 2014;64(1):9-15. **(Retrospective cohort study; 5144 transports)**

23. Lim MT, Ratnavel N. A prospective review of adverse events during interhospital transfers of neonates by a dedicated neonatal transfer service. *Pediatr Crit Care Med*. 2008;9(3):289-293. **(Prospective observational study; 346 pediatric transports)**

24.* Lee YS, Baek JS, Kwon BS, et al. Pediatric emergency room presentation of congenital heart disease. *Korean Circ J.* 2010;40(1):36-41. **(Retrospective study; 368 pediatric patients)**

25. Tibbles CD, Bouton M, Lucas JM, et al. Emergency department management of pediatric patients with cyanotic heart disease and fever. *J Emerg Med*. 2013;44(3):599-604. **(Retrospective review; 809 pediatric patients)**

26.* Savitsky E, Alejos J, Votey S. Emergency department presentations of pediatric congenital heart disease. *J Emerg Med*. 2003;24(3):239-245. **(Retrospective review; 77 pediatric patients)**

27.* Wren C, Richmond S, Donaldson L. Presentation of congenital heart disease in infancy: implications for routine examination. *Arch Dis Child Fetal Neonatal Ed*. 1999;80(1):F49-F53. **(Retrospective review; 1590 infants)**

28. Thangaratinam S, Brown K, Zamora J, et al. Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies: a systematic review and meta-analysis. *Lancet*. 2012;379(9835):2459-2464. **(Systematic review)**

29. Crossland DS, Furness JC, Abu-Harb M, et al. Variability of four limb blood pressure in normal neonates. *Arch Dis Child Fetal Neonatal Ed*. 2004;89(4):F325-F327. **(Prospective study; 40 neonates)**

30. Sharieff GQ, Wylie TW. Pediatric cardiac disorders. *J Emerg Med*. 2004;26(1):65-79. **(Review article)**

31. Colletti JE, Homme JL, Woodridge DP. Unsuspected neonatal killers in emergency medicine. *Emerg Med Clin North Am*. 2004;22(4):929-960. **(Review article)**

32. Koulouri S, Acherman RJ, Wong PC, et al. Utility of B-type natriuretic peptide in differentiating congestive heart failure from lung disease in pediatric patients with respiratory distress. *Pediatr Cardiol*. 2004;25(4):341-346. **(Prospective study; 49 infants and children)**

33. Maher KO, Reed H, Cuadrado A, et al. B-type natriuretic peptide in the emergency diagnosis of critical heart disease in children. *Pediatrics*. 2008;121(6):e1484-e1488. **(Prospective case control study; 33 patients with CHD and 70 controls)**

34. Rossano JW, Shaddy RE. Heart failure in children: etiology and treatment. *J Pediatr*. 2014;165(2):228-233. **(Review article)**

35.* Chan TC, Sharieff GQ, Brady WJ. Electrocardiographic manifestations: pediatric ECG. *J Emerg Med*. 2008;35(4):421-430. **(Review article)**

36. O'Connor M, McDaniel N, Brady WJ. The pediatric electrocardiogram. Part I: age-related interpretation. *Am J Emerg Med*. 2008;26(2):221-228. **(Review article)**

37. O'Connor M, McDaniel N, Brady WJ. The pediatric electrocardiogram. Part III: congenital heart disease and other cardiac syndromes. *Am J Emerg Med.* 2008;26(4):497-503. **(Review article)**

38. Tonkin IL. Imaging of pediatric congenital heart disease. *J Thorac Imaging*. 2000;15(4):274-279. **(Review article)**

39. Russell J, Justino H, Dipchand A, et al. Noninvasive imaging in congenital heart disease. *Curr Opin Cardiol*. 2000;15(4):224-237. **(Review article)**

40. Ferguson EC, Krishnamurthy R, Oldham SA. Classic imaging signs of congenital cardiovascular abnormalities. *Radiographics*. 2007;27(5):1323-1334. **(Review)**

41. Bernstein D. Laboratory evaluation. In: Kliegman RM, Stanton BF, St. Geme JW, et al, eds. *Nelson Textbook of Pediatrics.* 19th ed. Philadelphia, PA: Elsevier Saunders; 2011. **(Textbook)**

42.* Gazit AZ, Oren PP. Pharmaceutical management of decompensated heart failure syndrome in children: current state of the art and a new approach. *Curr Treat Options Cardiovasc Med*. 2009;11(5):403-409. **(Review article)**

43. Lewis AB, Freed MD, Heymann MA, et al. Side effects of therapy with prostaglandin E1 in infants with critical congenital heart disease. *Circulation*. 1981;64(5):893-898. **(Case report review; 492 infants)**

44.* Meckler GD, Lowe C. To intubate or not to intubate? Transporting infants on prostaglandin E1. *Pediatrics*. 2009;123(1):e25-e30. **(Retrospective review; 202 neonatal transports receiving PGE)**

45. Kothari SS. Mechanism of cyanotic spells in Tetralogy of Fallot--the missing link? *Int J Cardiol*. 1992;37(1):1-5. **(Review)**

46. BMJ Best Practice. Tetralogy of Fallot (management of hypercyanotic (tet) spells). Available at: http://bestpractice.bmj.com/best-practice/monograph/701/treatment/step-by-step.html. Accessed January 15, 2016. **(Guidelines)**

47. Apitz C, Webb GD, Redington AN. Tetralogy of Fallot. *Lancet*. 2009;374(9699):1462-1471. **(Review article)**

48. Barata IA. Cardiac emergencies. *Emerg Med Clin North Am*. 2013;31(3):677-704. **(Review)**

49. Tsze DS, Vitberg YM, Berezow J, et al. Treatment of Tetralogy of Fallot hypoxic spell with intranasal fentanyl. *Pediatrics*. 2014;134(1):e266-e269. **(Case report)**

50.* Faris R, Flather MD, Purcell H, et al. Diuretics for heart failure. *Cochrane Database Syst Rev*. 2006(1):CD003838. **(Systematic review)**

51. Kantor PF, Lougheed J, Dancea A, et al. Presentation, diagnosis, and medical management of heart failure in children: Canadian Cardiovascular Society guidelines. *Can J Cardiol*. 2013;29(12):1535-1552. **(Professional society practice guidelines)**

52. Beggs S, Thompson A, Nash R, et al. Cardiac failure in children. 17th Expert Committee on the Selection and Use of Essential Medicines. Geneva, Switzerland: World Health Organization; 2009. **(Expert committee systematic review and recommendations)**

53. Thackray S, Easthaugh J, Freemantle N, et al. The effectiveness and relative effectiveness of intravenous inotropic drugs acting through the adrenergic pathway in patients with heart failure-a meta-regression analysis. *Eur J Heart Fail*. 2002;4(4):515-529. **(Systematic review and meta-analysis)**

54. Cuffe MS, Califf RM, Adams KF, Jr., et al. Short-term intravenous milrinone for acute exacerbation of chronic heart failure: a randomized controlled trial. *JAMA*. 2002;287(12):1541-1547. **(Prospective randomized double-blind placebo-controlled multicenter study; 951 patients)**

55. Hoffman TM, Wernovsky G, Atz AM, et al. Efficacy and safety of milrinone in preventing low cardiac output syndrome in infants and children after corrective surgery for congenital heart disease. *Circulation*. 2003;107(7):996-1002. **(Prospective double-blind placebo-controlled parallel group trial; 238 patients)**

56. Buck ML. Clinical experience with spironolactone in pediatrics. *Ann Pharmacother*. 2005;39(5):823-828. **(Prospective observational study; 100 consecutive pediatric patients)**

57. Hsu DT, Pearson GD. Heart failure in children: part II: diagnosis, treatment, and future directions. *Circ Heart Fail*. 2009;2(5):490-498. **(Review article)**

58. Bruns LA, Chrisant MK, Lamour JM, et al. Carvedilol as therapy in pediatric heart failure: an initial multicenter experience. *J Pediatr*. 2001;138(4):505-511. **(Retrospective multicenter study)**

59. Shaddy RE, Boucek MM, Hsu DT, et al. Carvedilol for children and adolescents with heart failure: a randomized controlled trial. *JAMA*. 2007;298(10):1171-1179. **(Prospective randomized, double-blind, placebo-controlled multicenter study; 161 pediatric patients)**

60. Frobel AK, Hulpke-Wette M, Schmidt KG, et al. Beta-block-

61. Hood WB Jr, Dans AL, Guyatt GH, et al. Digitalis for treatment of congestive heart failure in patients in sinus rhythm. *Cochrane Database Syst Rev*. 2004(2):CD002901. **(Systematic review)**

62. Gaca AM, Jaggers JJ, Dudley LT, et al. Repair of congenital heart disease: a primer-part 2. *Radiology*. 2008;248(1):44-60. **(Review article)**

63. Gaca AM, Jaggers JJ, Dudley LT, et al. Repair of congenital heart disease: a primer-part 1. *Radiology*. 2008;247(3):617-631. **(Review article)**

64.* Woods WA, McCulloch MA. Cardiovascular emergencies in the pediatric patient. *Emerg Med Clin North Am*. 2005;23(4):1233-1249. **(Review article)**

65. Bernstein D. Anomalous origin of the coronary arteries. In: Kliegman RM, Stanton BF, St. Geme JW, et al, eds. *Nelson Textbook of Pediatrics*. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011. **(Textbook)**

66. Frazier A, Hunt EA, Holmes K. Pediatric cardiac emergencies: children are not small adults. *J Emerg Trauma Shock*. 2011;4(1):89-96. **(Review article)**

67. Bernstein D. Abnormal positions of the heart and heterotaxy syndromes. In: Kliegman RM, Stanton BF, St. Geme JW, et al., eds. *Nelson Textbook of Pediatrics*. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011. **(Textbook)**

68. Healy F, Hanna BD, Zinman R. Pulmonary complications of congenital heart disease. *Paediatr Respir Rev*. 2012;13(1):10-15. **(Review article)**

69. Bernstein D. Pulmonary hypertension. In: Kliegman RM, Stanton BF, St. Geme JW, et al., eds. *Nelson Textbook of Pediatrics*. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011. **(Textbook)**

70. Kleinman ME, Chameides L, Schexnayder SM, et al. Part 14: pediatric advanced life support: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care. *Circulation*. 2010;122(18 Suppl 3):S876-S908. **(Professional society practice guidelines)**

71. Yun SW. Congenital heart disease in the newborn requiring early intervention. *Korean J Pediatr*. 2011;54(5):183-191. **(Review article)**

72. Tallman TA, Peacock WF, Emerman CL, et al. Noninvasive ventilation outcomes in 2,430 acute decompensated heart failure patients: an ADHERE Registry Analysis. *Acad Emerg Med*. 2008;15(4):355-362. **(Retrospective study; 2430 patients)**

73.* Kantor PF, Mertens LL. Clinical practice: heart failure in children. Part I: clinical evaluation, diagnostic testing, and initial medical management. *Eur J Pediatr*. 2010;169(3):269-279. **(Review article)**

74. Huang FK, Lin CC, Huang TC, et al. Reappraisal of the prostaglandin E1 dose for early newborns with patent ductus arteriosus-dependent pulmonary circulation. *Pediatr Neonatol*. 2013;54(2):102-106. **(Retrospective review; 33 patients)**

75. Sharma M, Sasikumar M, Karloopia S, et al. Prostaglandins in congenital heart disease. *Med J Armed Forces India*. 2001;57(2):134-138. **(Review article)**

76.* Rushani D, Kaufman JS, Ionescu-Ittu R, et al. Infective endocarditis in children with congenital heart disease: cumulative incidence and predictors. *Circulation*. 2013;128(13):1412-1419. **(Population based study; 47,518 children in Quebec)**

77. Wilson W, Taubert KA, Gewitz M, et al. Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young,

and the Council on Clinical Cardiology, Council on Cardio-vascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. *Circulation*. 2007;116(15):1736-1754. **(Professional society practice guidelines)**

78. Allen UD, Canadian Paediatric Society. Infective endocarditis: updated guidelines *Paediatr Child Health*. 2010 (reaffirmed 2014);15(4):205-208. **(Professional society guidelines)**

CME Questions

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1. **Which of the following lesions is not expected to present with shock in a 7-day-old neonate?**
   a. Hypoplastic left heart syndrome
   b. Coarctation of the aorta
   c. Critical aortic stenosis
   d. Tetralogy of Fallot

2. **Regarding the initiation of PGE$_1$ for a neonate with suspected CHD, which of the following is TRUE?**
   a. Initiate PGE$_1$ only after the diagnosis of CHD is confirmed by echocardiography.
   b. Initiate PGE$_1$ empirically in a neonate presenting with shock, as he has a ductal-dependent lesion.
   c. Intubate all neonates prior to initiating PGE$_1$, due to risk of apnea.
   d. While on a PGE$_1$ infusion, the neonate should have a definitive airway established prior to any transport.

3. **Which of the following is NOT a side effect of PGE$_1$?**
   a. Hypotension
   b. Hypothermia
   c. Jitteriness
   d. Apnea

4. **Which of the following physiologic changes leads to the presentation of CHF around 2 months of age in infants with a large ventricular septal defect?**
   a. Decreased pulmonary blood flow
   b. Decreased pulmonary vascular resistance
   c. Decreased left-to-right shunting
   d. Decreased fetal hemoglobin

5. **Which is the most useful initial investigation in a child presenting to the ED with acute CHF?**
   a. Complete blood count
   b. B-type natriuretic peptide level
   c. 12-lead ECG
   d. Chest x-ray

6. **A child with tetralogy of Fallot presents with a hypercyanotic spell. You have tried knee-to-chest position, oxygen, morphine, and a fluid bolus, without response. What is the best next step?**
   a. Intubate the patient using ketamine and rocuronium.
   b. Call cardiothoracic surgery to prepare for emergent surgery.
   c. Wait for the effects of the morphine and the fluid bolus.
   d. Give a dose of propranolol or esmolol.

7. **Which medication has the most evidence to support its efficacy as the initial treatment for a child presenting to the ED in respiratory distress due to acute CHF?**
   a. ACE inhibitors
   b. Digoxin
   c. Diuretics
   d. Beta adrenergic blockers

8. **Which of the following cyanotic CHD lesions can present outside of the neonatal period as recurrent respiratory distress and CHF?**
   a. Transposition of the great arteries
   b. Truncus arteriosus
   c. Total anomalous pulmonary venous return
   d. Tricuspid atresia

9. **With regard to ventilating an intubated child with CHD, which of the following statements is FALSE?**
    a. Positive-pressure ventilation decreases venous return and preload.
    b. Positive-pressure ventilation increases pulmonary vascular resistance and right ventricle afterload.
    c. Positive-pressure ventilation decreases left ventricle afterload.
    d. Positive-pressure ventilation and sedation increase tissue oxygen demand.

10. **When giving supplemental oxygen to a neonate suspected of having CHD, which of the following is TRUE?**
    a. The target for oxygen saturation on pulse oximetry is 100%.
    b. High FiO_2 should be provided in all cases of CHD.
    c. Giving oxygen will decrease pulmonary blood flow and decrease pulmonary edema.
    d. Oxygen administration leads to pulmonary vasodilation and increased pulmonary blood flow, and can worsen pulmonary edema or systemic perfusion in some lesions.

Physician CME Information