Cardiac Emergencies in Children

A Practical Approach to Diagnosis and Management

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Foreword 1

You are a pediatrician, a family practice physician, an emergency department physician who sees children, a pediatric hospitalist, a trainee, or a nurse practitioner caring for children and you encounter a cardiac emergency—it's somewhat uncommon and scary. Other than the little experience you receive during training, there are limited resources to help you to navigate the approach to care for such a patient, including no textbooks that focus on this problem, until now.

In this textbook, *Cardiac Emergencies in Children: A Practical Approach to Diagnosis and Management*, the editors and authors cover the pathophysiology of, diagnostic approaches to, and therapeutic rationale for children presenting with life-threatening conditions from a diverse group of congenital and acquired heart lesions, as well as the failing heart. They discuss topics ranging from the emergency presentations and recognition of cardiovascular disease in childhood in countries with resources and those with limited resources, to issues occurring in the child recovering from a surgical repair of congenital heart disease, to the use of mechanical circulatory support and problems seen in children after heart transplantation. In addition, emergencies in children with pulmonary hypertension, inflammatory heart disease, cyanotic heart disease, shunts, and arrhythmias are all detailed, and the practical approaches to their care are described. In delivering the needed information, the editors have utilized the talents of renowned leaders in the field of congenital and acquired heart disease, heart failure, and transplantation, including highly respected pediatric cardiologists and cardiac surgeons, pediatric and cardiac intensivists, and pediatric radiologists. The textbook they have produced will educate a wide variety of clinicians and trainees for many years to come and will become a "go-to" book for the current and future generations of clinicians planning to take care of children at risk for cardiovascular emergencies. Pediatric medicine owes its gratitude to the editors, Drs. Sarnaik, Ross, Lipshultz, and Walters III, as well as the authors of the excellent topical chapters, for their visions, insights, and recommendations which will undoubtedly help to care for children exposed to cardiac emergencies.

Memphis, TN, USA — Jeffrey A. Towbin, M.D.

Foreword 2

Ashok Sarnaik, Robert Ross, Steven Lipshultz, and Henry Walters III have set out to edit a unique book tailored to physicians who might encounter cardiac emergencies in children, but to whom such encounters are not their daily preoccupation. Some note of who these editors are helps to clarify their purpose.

Ashok Sarnaik is a senior pediatric intensivist whose preoccupations include the care of children who suffer cardiac emergencies. In the course of a day's work, he commonly receives transfers from physicians who have provided initial management. He recognizes the challenges they face and the limits to their expertise in this highly specialized clinical area. Robert Ross is a senior pediatric cardiologist with a grand overview and broad expertise in the pathophysiology of pediatric cardiac disease. Steven Lipshultz is a pediatric department chair who is also a senior pediatric cardiologist. As a pediatric department chair, Dr. Lipshultz has a broad overview of the skill sets of non-cardiologist pediatricians and others who see sporadic examples of pediatric cardiac emergencies. Henry Walters III is a pediatric heart surgeon with a broad understanding of the medical and surgical options for children who suffer or are prone to cardiac emergencies.

Together, the editors have undertaken to highlight the background needed to understand the nature of these emergencies, their basis in physiology, and their diagnostic elements and therapeutic options. The topics presented are tailored to the needs of the practitioners who face these challenging crises sporadically: adult and pediatric emergency physicians, hospitalists, primary care physicians, midlevel providers, and nurses.

This text is unique in its targeted readership, scope of content, and level of detail. It is both archival and educational. It can be used both to launch further literature inquiry and to quickly inform in a crisis. It is both problem oriented and diagnostically categorized.

It teaches both what to do and what not to do.

El Paso, TX, USA — Bradley Fuhrman

Preface

"There is no disease more conducive to clinical humility than aneurysm of the aorta."

Sir William Osler

Facing a critically ill child with a life-threatening cardiac emergency is extremely challenging for first responders. Children with either known or suspected heart disease most often present to an emergency department, including departments that predominantly serve adults, and some may present in a primary care provider's office. Many of these children have undiagnosed heart disease with congenital or acquired origins. The emergency manifestations of heart disease in a critically ill child can often mimic other common pediatric illnesses such as bronchiolitis, asthma, pneumonia, dehydration, shock, and sepsis, resulting in misdiagnosis and delays in beginning appropriate therapies, delays that can have catastrophic consequences. Also, commonly used therapies such as bronchodilators, fluid expansion, and supplemental oxygen can adversely affect the outcome in some situations.

With advances in surgery techniques, many children with complex congenital heart diseases are surviving with improved life expectancy and quality of life. Some of these children present with emergencies between several stages of cardiac surgical palliation. These patients have unique pathophysiological considerations and require individualized management approaches. Certain therapies that are life-saving in one situation can be paradoxically life-threatening in another. One such example is the potential danger of using supplemental oxygen in an infant who has recently undergone a first-stage Norwood procedure versus its potential benefit when used in that same infant, later in life, after undergoing a completion Fontan procedure. First responders without familiarity in managing such emergencies need a practical guide on how to quickly evaluate, stabilize, and initiate treatment on these patients and to know when to get immediate cardiology consultation.

Historically, pediatric cardiovascular surgery has often been limited in the developing world where children with heart disease received only supportive care. This is no longer the case; patients with heart defects such as septal defects, valvar stenoses, and vascular anomalies are now routinely operated on in developing countries. Even cardiac transplantation is now available with great success in many parts of the developing world. Thus, a normal life expectancy is an attainable goal for many

such children. At the same time, with increasing globalization and international travel, technologically advanced countries have much to learn from countries with limited resources about cardiac involvement in conditions that are uncommon to them, such as vitamin D deficiency cardiomyopathy and dengue myocarditis. The authors of the chapter "Cardiac Emergencies in Countries with Limited Resources" have addressed some of the special challenges they routinely face in their practice.

In this book on pediatric cardiac emergencies, we attempt to bridge the gap between cardiac critical care subspecialists and the emergency care providers in various settings. The goal is to provide the pathophysiological basis of cardiac dysfunction, improve clinical reasoning to facilitate the diagnosis, and explain the rationale for stabilizing and managing cardiac emergencies in children. We emphasize the importance of history and physical examination and interpreting ECG and chest radiographs and describe situations where there is a need to consult subspecialists. We do so by presenting real-life case scenarios and practical guidelines.

This book is aimed primarily at pediatric and adult emergency physicians who do a yeoman's work in diagnosing and stabilizing patients. Primary care physicians and pediatric hospitalists who encounter cardiac emergencies in their practice will also find the information valuable. The book will also be beneficial for nurses and physicians at all levels of training including students, residents, fellows, and other health-care professionals.

Finally, we must acknowledge that this book is a result of true team effort. Experts from various institutions and specialties have willingly and selflessly provided their knowledge and expertise with the sole purpose of improving outcomes of children with heart disease throughout the world. We thank them from the cockles of our hearts!

Detroit, MI, USA — Ashok P. Sarnaik
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Emergency Presentation of Heart Disease

1

Syana Sarnaik, Katherine Cashen, and Ashok P. Sarnaik

Introduction

Congenital heart defects (CHD) are the most common congenital malformations with an estimated prevalence of 6–8 per 1000 live births [1]. Advances in fetal ultrasound and echocardiography have improved the prenatal detection of CHD. In addition, screening for critical congenital heart disease (CCHD) was added to the US Recommended Uniform Screening Panel in 2011. Since that time, CCHD screening with pulse oximetry has become nearly universal for newborns born in the United States [2]. Approximately 25% of neonates with CCHD will not be detected by pulse oximetry alone, and many children will not be diagnosed until after discharge from the hospital. Reasons for undiagnosed CHD at birth include the lack of prenatal diagnosis due to no or poor prenatal care, standard ultrasonography missing CHD, postnatal detection failure through pulse oximetry, or home birth. Most cardiac emergencies due to CHD in undiagnosed children most often present in neonates and infants less than 1 year of age. When previously undiagnosed children present with life-threatening cardiac emergencies, it is often the physicians in the ED (including those who predominantly care for adults) or the primary care providers in an office setting who will first see the patient. Also, with improved surgical outcomes of complex CHD, there are more adults than children with CHD in developed countries. Emergency presentations of CHD often masquerade as other common entities delaying diagnosis and prompt institution of lifesaving therapy.

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Pediatric cardiac emergencies may result from anatomic abnormalities (congenital or acquired) or electrophysiological dysfunction. The latter is discussed in Chap. 8.

Symptomatology of Cardiac Emergencies in Children

There are three major symptom complexes that children with previously undiagnosed heart disease present with life-threatening emergencies. These are (1) respiratory distress, (2) shock, and (3) cyanosis. The clinical manifestations and pathogenesis of these symptom complexes and responsible underlying pathophysiologic states are outlined in Table 1.1. Of these symptom complexes, respiratory distress and shock are fraught with many diagnostic pitfalls and may be mistaken for other commonly encountered entities in children such as asthma, bronchiolitis, pneumonia (respiratory distress), dehydration, and sepsis (shock) (Table 1.2). The physician must look for diagnostic clues whenever presented with such scenarios.

Chronology of Presentation of Heart Disease in Children

Neonatal and infantile cardiopulmonary adaptation has a profound influence on the likelihood of a specific congenital heart lesion presenting at a given age. Before birth, the fetal circulation is in parallel circuits. The airless lungs have

Table 1.1 Emergency presentation of heart disease in children

Symptom	Clinical manifestations	Pathophysiology	Underlying state
Respiratory distress	• Tachypnea • Retractions • Wheezing • Grunting	• ↓ Lung compliance • Metabolic acidosis • Respiratory alkalosis	• Pulmonary edema • Airway obstruction • Anxiety • Baroreceptor stimulation
Shock	• Tachycardia • Poor perfusion • Hypotension	Decreased cardiac output	• Obstructive lesions • Decreased contractility • Increased afterload
Cyanosis	Central cyanosis	Hypoxemia	Right-to-left shunt

Table 1.2 Cardiogenic shock: diagnostic pitfalls

• Most present with respiratory distress (metabolic acidosis, stimulation of baroreceptors): misdiagnosed as asthma, bronchiolitis, pneumonia
• Early manifestations of pulmonary edema: wheezing, hyperinflation
• Treatment with bronchodilators worsens clinical state
• Apparent dehydration leads to inappropriate fluid expansion
• Lack of obvious findings such as jugular venous distension (JVD) or hepatomegaly
• Shock state is often blamed on sepsis especially in neonates
• Metabolic acidosis is blamed on hypovolemic state or sepsis
• Lack of cardiomegaly on chest X-ray in early stages of myocarditis

*Avoid low index of suspicion

suprasystemic vascular resistance accepting only 10% of cardiac output. A large portion of venous return to the right side of the heart is shunted across the foramen ovale (FO) and ductus arteriosus (DA) to the left side of the heart. The systemic circulation including the placenta accepts 90% of total cardiac output. The fetus tolerates complex congenital heart lesions as the FO and DA allow the right side of the heart to take over the function of the left side of the heart and vice versa.

Early Neonatal Period (1–7 Days)

Soon after birth, with initiation of air-breathing and oxygen-induced pulmonary vasodilation, the pulmonary vascular resistance (PVR) falls to 50% of systemic vascular resistance (SVR) which rises as placental circulation ceases to exist. Also, the FO and DA initially close functionally and then subsequently close anatomically. The circulation changes to being in series as the right ventricular stroke volume approaches that of the left ventricle with very little if any blood being shunted across the FO or DA. In this phase of early neonatal hemodynamic adaptation, several groups of congenital lesions are prone to manifest. These are:

Systemic arteriovenous fistula: The increasing systemic venous return flowing into the lung allows greater shunting across arteriovenous (AV) fistulae resulting in high-output congestive heart failure. The two places to look for AV fistula in a young infant with high-output failure are a) aneurysm of the vein of Galen and b) liver. It is important to auscultate for a bruit on the head and on the liver when dealing with such situations.

Left-sided obstructive lesions: Because of the functional closure of DA, the right ventricle cannot push blood into the descending aorta to maintain systemic perfusion. Lesions such as hypoplastic left heart syndrome, critical aortic stenosis, and interrupted aortic arch are likely to present in 1–7 days of life with systemic hypoperfusion, cardiogenic shock, and lactic acidosis. This hypoperfusion predominately manifests in the child as a gray and listless appearance rather than overt central cyanosis.

Right-sided obstructive heart lesions: Pulmonary blood flow can be maintained at a sufficient level for adequate oxygenation as long as the left ventricle can pump enough blood across the DA into the pulmonary artery. As the DA begins to close, an increasing amount of systemic venous return is shunted into the systemic arterial circulation. Lesions such as tricuspid atresia, hypoplastic right heart syndrome, and pulmonary atresia present in the early neonatal period with central cyanosis as the DA begins to close. Some cyanotic heart diseases such as tetralogy of Fallot may not manifest until a few weeks after birth as long as the right ventricular outflow tract (RVOT) is not too severely obstructed and allows reasonable amount of pulmonary blood flow. These patients are sometimes missed during the initial neonatal pulse oximetry screening. Cyanosis develops when RVOT obstruction worsens resulting in increased shunting across the ventricular septal defect.

Independent circuits: Transposition of the great vessels (TGV), where the aorta and pulmonary artery are transposed, presents soon after birth when functional closure of FO and DA prevent mixing of blood in the two circuits resulting in cyanosis.

Pulmonary venous obstruction: As pulmonary arterial flow increases after birth so does the pulmonary venous flow back to the left atrium. Lesions associated with obstruction to pulmonary venous flow such as total anomalous pulmonary venous return (TAPVR) and cor triatriatum will present in the early neonatal period with increasing pulmonary venous hydrostatic pressure and pulmonary edema resulting in respiratory distress and cyanosis. If the pulmonary venous return is less severely obstructed, such lesions may not manifest until later in life.

Early Infancy (6 Weeks–3 Months)

In the first 6 weeks to 3 months of life, the pulmonary vascular resistance continues to fall as the pulmonary arterial tunica media muscle continues to involute. By 3 months of age, the PVR declines to about 15% of SVR, a relationship that is maintained into adulthood. Since the tunica media muscle is mainly deposited in the last trimester of pregnancy, prematurely born infants have a more rapid fall in PVR compared to infants born after full term. The following lesions are more likely to become symptomatic at the age of 6 weeks to 3 months:

Left-to-right shunt: Ventricular septal defect (VSD), atrial septal defect (ASD), and patent ductus arteriosus (PDA) allow increasing pulmonary blood flow with decreasing pulmonary vascular resistance. The result is an increase in pulmonary blood flow (Qp) compared to systemic blood flow (Qs), hyperdynamic circulation, and congestive heart failure. Clinical manifestations include respiratory distress, tachycardia, bounding pulses, and eventually cardiac decompensation. If left untreated, left-to-right shunts result in hypertrophy of the medial musculature and intimal changes which lead to a rise in PVR and pulmonary hypertension limiting the amount of shunt. Without surgical intervention such children are at risk of developing suprasystemic PVR and Eisenmenger complex and right-to-left shunting.

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA): While children are born with ALCAPA, the lesion does not manifest itself at birth. Even though the left coronary artery is carrying deoxygenated blood, myocardial oxygen consumption is not compromised as long as the pulmonary artery pressure (PAP) remains sufficiently elevated. The myocardial oxygen consumption is more dependent on coronary perfusion pressure compared to coronary oxygen content since the heart is one of the most efficient extractors of oxygen. It is when PVR, and therefore the PAP, falls below a critical level, the anomalous left coronary artery fails to perfuse the myocardium with sufficient blood flow to maintain its oxygen demands. The manifestations are those of episodic screaming (angina), feeding difficulty (effort intolerance), and overt cardiogenic shock (myocardial infarction).

Any Age

Certain conditions are not dependent on postnatal cardiovascular adaptation. They can present at any age. Infection (myocarditis, pericarditis, sepsis) and cardiomyopathies should be considered at any age when cardiac dysfunction is suspected (Table 1.3).

Table 1.3 Age-specific lesions and most common ED presentations

Age	Lesion	Presentation
Newborn (less than 1 month)	Ductal-dependent pulmonary blood flow lesions • Tricuspid atresia • Pulmonary atresia • Tetralogy of Fallot • Critical pulmonary stenosis • Ebstein's anomaly • D-transposition of the great arteries • Double outlet right ventricle with severe pulmonary stenosis	Cyanosis, gray appearance, eventually shock
Newborn (less than 1 month)	Ductal-dependent systemic blood flow lesions • Critical coarctation of the aorta • Critical aortic stenosis • Aortic atresia • Interrupted aortic arch • Double outlet right ventricle with aortic stenosis	Hypoperfusion, shock, and pulmonary edema
Young infant (2–6 months)	Lesions dependent on pulmonary vascular resistance • Ventricular septal defects • Atrioventricular canal defects • Large patent DA • Unobstructed TAPVR	Volume overload, congestive heart failure • Tachypnea • Tachycardia • Poor feeding • Hepatomegaly • Pulmonary edema • Facial edema • Increased work of breathing, diaphoresis
Young infant (2–6 months)	Lesion dependent on pulmonary arterial pressure • Anomalous left coronary artery from the pulmonary artery (ALCAPA)	Shock
Any age	Acquired heart disease • Myocarditis • Cardiomyopathy • Pericarditis • Cardiac tumor Arrhythmias • Supraventricular tachycardia • Heart block	May present with mild, vague symptoms, older children may present with dyspnea, exercise intolerance Younger children may present with poor feeding, irritability, or shock

Table 1.4 Risks of commonly employed treatment strategies

• Oxygen—pulmonary vasodilator: worsening pulmonary edema, pulmonary circulation steal from systemic circulation—peripheral vasoconstrictor, increased afterload
• Fluid bolus—worsening pulmonary edema—decreased coronary perfusion (CPP = MAP–CVP)
• Albuterol—increased myocardial oxygen consumption—worsening tachypnea and metabolic acidosis
• Furosemide—worsened relative hypovolemia
• Treatment with bronchodilators worsens clinical state
• Apparent dehydration leads to inappropriate fluid expansion

Practical Considerations for Initial Care Providers

Ductal-dependent lesions should be a consideration in all neonates presenting in shock and/or cyanosis in the neonatal period. Alprostadil (PGE1) infusion may be lifesaving if instituted even prior to an established diagnosis. FO-dependent lesions (TGV, obstructed TAPVR) may need balloon atrial septostomy. All patients with suspected CCHD should be promptly transferred to tertiary care centers.

Oxygen administration, which is a common intervention for life-threatening pediatric emergencies, may have deleterious consequences in certain cardiac lesions. Elevated PaO_2 results in constriction of DA and pulmonary vasodilation. In left-sided obstructive lesions of the heart, constriction of DA would result in worsening of systemic hypoperfusion, shock, and lactic acidosis. Pulmonary vasodilation in single ventricle physiology (e.g., Norwood surgery for hypoplastic left heart syndrome) may result in a greater proportion of cardiac output going to the lungs (Qp) at the expense of the rest of the body (Qs). Also, in left-to-right shunts, pulmonary overcirculation resulting from vasodilation may worsen pulmonary edema. Oxygen administration in such situations must be carefully monitored to maintain SaO_2 around 75–80% in single ventricle physiology and around 90% in left-to-right shunts.

Administration of albuterol in a child whose respiratory symptoms are secondary to myocardial dysfunction will lead to increased oxygen demands on an already compromised heart. Excessive fluid resuscitation for decreased cardiac output and apparent dehydration in a situation with a failing heart will have similar adverse consequences (Table 1.4). Early identification of cardiac etiology as the underlying mechanism of life-threatening manifestations is of utmost importance.

Important Historical Findings in Infants with Heart Disease

Unlike older children and adults, infants and young children are unable to articulate their symptoms. The clinician needs to seek from the caretaker certain historical findings that may act as surrogates for symptoms that older children may complain about (Table 1.5). Tachypnea, rapid or congested breathing, and wheezing are suggestive of pulmonary edema from congestive heart failure. Feeding difficulty and

Table 1.5 Important historical findings of heart disease in infants

Symptoms/signs	Cardiac relevance	Condition
Fast breathing	Decreased lung compliance Pulmonary edema	Congestive heart failure
Wheezing/coughing/congested breathing	Pulmonary edema Compression of airways Vascular compression	Congestive heart failure Chamber enlargement Vascular ring/sling
Turning blue spells	Right-to-left shunting	Pink tetralogy Cyanotic heart disease
Chest pounding	Hyperdynamic circulation	Left-to-right shunt Cardiomegaly
Excessive sweating	Increased sympathetic activity	Heart failure
Episodes of screaming	Angina	Coronary ischemia
Feeding difficulty Failure to thrive	Effort intolerance Increased work of breathing	Congestive heart failure

failure to thrive could be manifestations of effort intolerance and increased work of breathing. Excessive sweating may result from increased sympathetic activity and obvious chest "pounding" could be a result of hyperdynamic circulation. Episodes of screaming may signify angina from coronary ischemia.

Case Presentation 1

A 4-day-old, full-term neonate is brought to the ED by his parents for poor feeding and lethargy. His vital signs are as follows: temperature 36.2 °C, heart rate (HR) 192 beats per minute, respiratory rate (RR) 63 breaths per minute, and blood pressure (BP) 62/37 mmHg with oxygen saturation (SpO2) of 92% on room air. On examination, the baby appears lethargic with cool extremities and capillary refill time of 4 s. He has nasal flaring with intercostal retractions. Liver edge is palpable 3 cm below the right costal margin. The saturations improve to 97% on 100% oxygen. Repeat vital signs after initiation of oxygen administration show a HR of 212 beats per minute, RR of 75 breaths per minute, and a right upper extremity BP of 54/23 mmHg. A chest radiograph demonstrates pulmonary edema and cardiomegaly (Fig. 1.1). An initial arterial blood gas (ABG) demonstrates pH 7.15, PCO_2 28 mmHg, PaO_2 94 mmHg, bicarbonate 10 mEq/L, and lactate 12 mmol/L.

The neonate in this vignette has signs of shock, severe metabolic acidosis, and marked hyperlactatemia. The etiology of shock in an infant needs to be determined and lifesaving measures employed rapidly. Cardiogenic shock should be suspected in all such infants in addition to hypovolemia and sepsis. In this infant presenting with shock in the first week of life, there are no historical findings to account for hypovolemia, hypotension, and poor perfusion. Therefore treatment for an underlying ductal-dependent cardiac disease is imperative. Obstructive lesions of the left side of the heart such as HLHS, critical aortic stenosis, coarctation of the aorta, and interrupted aortic arch are at the top of the list of differential diagnosis. The infant may appear relatively normal at birth as long as the DA remains open, and the

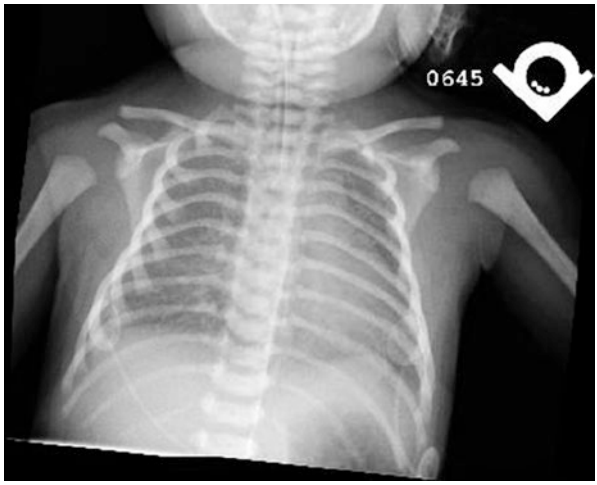

Fig. 1.1 Chest radiograph showing cardiomegaly and early pulmonary edema

postobstructive systemic circulation is maintained by the right ventricle through the pulmonary artery and DA. Symptoms appear when closure of the DA leads to systemic hypoperfusion. These children are not cyanotic but rather appear ashen gray because of hypoperfusion and increased peripheral oxygen extraction. In order to maintain ductal patency and establish systemic blood flow, alprostadil infusion must be started. Since alprostadil is a systemic vasodilator, judicious intravenous fluid expansion may be necessary to maintain normal blood pressure for age.

The use of oxygen in a child with shock due to a left-sided obstructive lesion can be of particular harm as oxygen itself is a potent DA constrictor which may further limit blood flow through a compromised DA. Ductal patency is a major factor that determines the difference between survival and death in a child dependent on the DA for systemic blood flow.

Unlike the infant in the vignette, a child with right-sided obstructive lesions (pulmonary atresia, tricuspid atresia,) or independent circuits (TGA) will present with central cyanosis and often has PaO_2 ~40 and oxygen saturations ~75%. In cyanotic infants, the hyperoxia test may be utilized as a clinical tool to differentiate between pulmonary and cardiac disease. The test is based on the principle that 100% oxygen will increase alveolar PO_2, leading to an increase in systemic arterial PO_2 in the absence of a fixed cardiac shunt. In cyanotic congenital heart disease, little or no rise in PaO_2 would be expected after breathing 100% O_2. An arterial blood gas analysis done both before and after the administration of 100% O_2 demonstrating an increase in PaO_2 to more than 100 mmHg would suggest a respiratory disease, while an increase of PaO_2 of less than 80 mmHg would require evaluation for cyanotic CHD. Thus, persistent hypoxia refractory to 100% oxygen supply would indicate cyanotic CHD rather than a primary pulmonary disease. It should be noted that the hyperoxia test should be utilized only in cyanotic infants to differentiate cyanosis from pulmonary vs. cardiac etiology. It should not be used in infants who are in shock and therefore appear gray but have SpO_2 above 90%. These patients should

be suspected of ductal-dependent left-sided obstructive lesion in whom 100% oxygen administration could be detrimental.

The most important first step in managing a neonate presenting within the first week of life with either shock or cyanosis necessitates that the ED physician considers a ductal-dependent cardiac lesion and initiates the administration of an alprostadil drip to improve patency of the DA.

Case Presentation 2

A 3-month-old, full-term male presents to the ED due to tachypnea and poor feeding for the last 12 h. His vital signs are temperature 37 °C, HR 170 beats per minute, RR 50 breaths per minute, BP 78/50 mmHg, and oxygen saturation 94% on room air. On examination he is fussy and showing signs of respiratory distress. Extremities are cool with prolonged capillary refill time. He has grunting, retractions, and wheezing bilaterally. Liver edge is palpable at 4 cm below the costal margin. His cardiac examination is significant for a gallop. Capillary blood gas reveals pH 7.28, PCO_2 20 mmHg, sodium bicarbonate 11 mEq/L, and lactate 5.8 mmol/L. Chest radiograph shows cardiomegaly and left lung atelectasis (Fig. 1.2). The EKG is obtained (Fig. 1.3). What is the most likely diagnosis? What are the risks of albuterol, 100% oxygen, and fluid boluses?

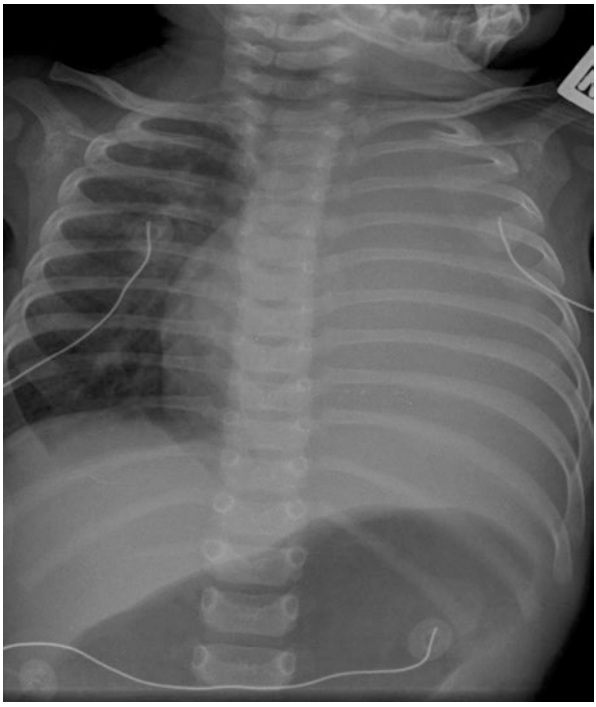

Fig. 1.2 Chest radiograph showing marked cardiomegaly and left lung atelectasis

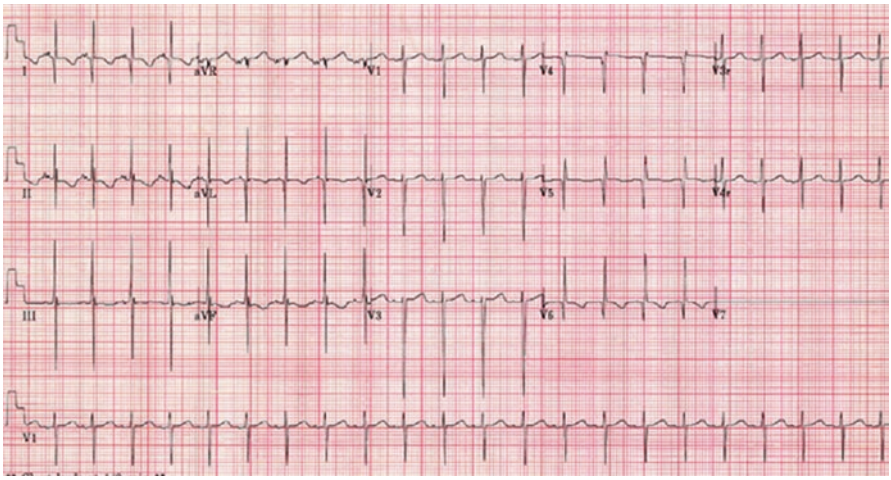

Fig. 1.3 Electrocardiogram showing deep Q waves in left-sided chest leads

The infant in this vignette is presenting with signs of uncompensated heart failure and systemic hypoperfusion. Infants 1 month to 6 months of age with undiagnosed congenital heart disease usually present to the ED with signs of heart failure due to decreasing pulmonary vascular resistance (PVR). In lesions with large intracardiac left-to-right shunts (large ventricular septal defects, atrioventricular canal defects, large patent DA), the continued decline in PVR results in increasing pulmonary blood flow. This increased pulmonary blood flow may cause the child to present with volume overload and signs and symptoms of congestive heart failure (poor feeding, tachypnea, grunting, retractions, cardiac wheezing, tachycardia, hepatomegaly, gallop, metabolic acidosis with respiratory alkalosis, and cardiomegaly on chest radiography). These infants often show evidence of hyperdynamic circulation with bounding pulses and increased pulse pressure.

The infant in this vignette exhibits cardiogenic shock with peripheral vasoconstriction and decreased pulse pressure. EKG shows myocardial infarction evidenced by deep Q waves in left-sided leads. This clinical picture is pathognomonic of ALCAPA which is typically unmasked during the early infancy period. As PVR and PAP decrease, the coronary perfusion pressure is compromised resulting in myocardial ischemia and infarction. Symptoms of a failing left ventricle and congestive heart failure progressing to shock develop around 2–4 months of age. The diagnostic clues include the signs of shock and the deep Q waves on left-sided leads on EKG.

An infant who presents with heart failure may be misdiagnosed as having viral bronchiolitis, bacterial pneumonia, or asthma. These infants may be treated with fluid boluses due to tachycardia or concern for sepsis. This may worsen the clinical status of a child in shock from congestive heart failure. Similar to Case Presentation 1, a careful history and examination to differentiate the etiology of shock must be undertaken with attention to tachypnea, gallop rhythm, jugular venous distension (JVD), hepatomegaly, and weak pulses. This assessment could prevent the inappropriate excessive administration of fluids to a child in CHF. The use of oxygen in an infant

with CHF can be of particular harm as oxygen itself is a pulmonary vasodilator and may worsen left-to-right shunting and further increase pulmonary overcirculation and worsen pulmonary edema. Finally, administration of albuterol with resultant tachycardia in a child whose respiratory symptoms are secondary to myocardial dysfunction will lead to increased oxygen demands on an already compromised heart.

Acute management of infants with CHF includes optimizing hemodynamic status, decreasing oxygen consumption, maintaining optimal preload, decreasing afterload, and improving contractility. Decreasing oxygen consumption is achieved by taking away the work of breathing by sedation and mechanical ventilation, achieving atrioventricular synchrony, and avoiding β-agonists bronchodilators and other medications that may cause tachycardia. Maintaining optimal preload may require an isotonic intravenous fluid bolus. This requires frequent assessments of the clinical response to fluid. Excessive fluid administration will not be well tolerated and may worsen pulmonary edema. Improving contractility with the addition of inotropic support may be necessary.

Case Presentation 3

A 16-year-old female presents to the ED with difficulty breathing, emesis, and progressive listlessness. She has had a previous ED visit for progressive wheezing and shortness of breath 7 days ago. She was given an albuterol inhaler and dexamethasone. Despite the use of his inhaler every 6 h, her shortness of breath and wheezing worsened. Her temperature is 38.2 °C, HR 118 beats per minute, RR 23 breaths per minute, BP 102/70 mmHg, and SpO_2 92% on RA. On examination, she has mild respiratory distress and is taking shallow, quick breaths. Her lung fields are notable for diffuse crackles and wheezing. Her cardiac examination reveals tachycardia with a gallop rhythm and capillary refill of 3 s. Her rhythm strip suggests sinus tachycardia. She is given an albuterol treatment and fluid bolus of 20 mL/kg of normal saline. During the breathing treatment and fluid bolus, she suddenly develops hypotension and worsening respiratory distress. Repeat vital signs are as follows: HR 145 beats per minute, RR 45 breaths per minute, and BP 72/42 mmHg. She is somnolent, with rapid breathing and diffuse wheezing. Cardiac exam reveals tachycardia with poor peripheral pulses. She has hepatomegaly 4 cm below costal margin. CXR reveals pulmonary infiltrates and cardiomegaly (Fig. 1.4.) What are the diagnostic approach and management strategies in this patient?

This child is presenting with signs of acute decompensated heart failure. Similar to the infant in the earlier vignette, this child has signs and symptoms of heart failure including difficulty breathing, wheezing, tachycardia, gallop, hepatomegaly, and cardiomegaly. The differential diagnosis includes dilated cardiomyopathy (due to genetic causes, chronic hypertension, etc.), large pericardial effusion, and myocarditis. This child was previously healthy but had a preceding viral illness 7 days prior to the ED visit suggesting myocarditis. Myocarditis may occur at any age although studies indicate that infants and teenagers are more commonly affected. Common pitfalls include misdiagnosis as asthma or bacterial pneumonia and initiation of steroids or β-agonist bronchodilator therapy which will be harmful. Immediate management is centered around optimizing cardiac output (contractility, preload, and afterload) and transfer to a tertiary care pediatric cardiac center.

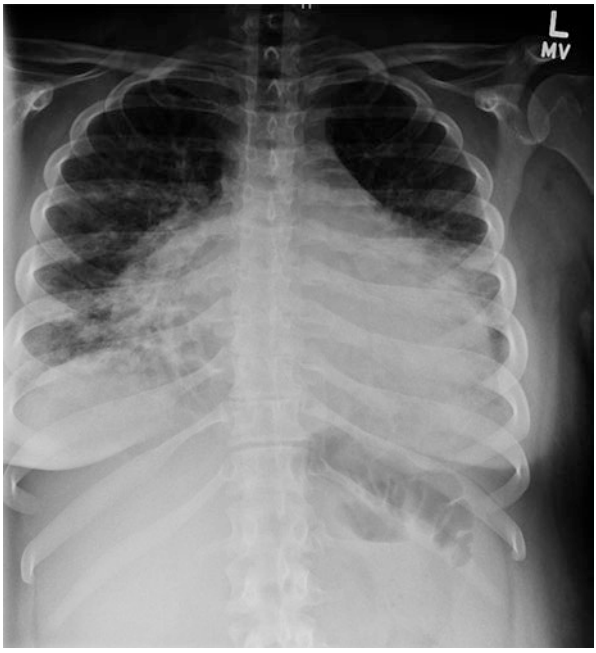

Fig. 1.4 Chest radiograph showing marked cardiomegaly and pulmonary edema

ED Management

The goals of ED management are to stabilize the airway, obtain vascular access, and support the circulation and in neonates and infants to establish and maintain patency of the ductus arteriosus. Age and physical examination findings will be the most helpful clues toward diagnosis in the initial assessment period (Table 1.3). Alprostadil infusion should be considered in infants <1 month who present in shock or cyanosis where ductal-dependent lesion is strongly suspected based on history and clinical findings. While echocardiography is necessary to make the diagnosis, awaiting echocardiography to initiate therapy may result in wasting valuable time to open the ductus and may contribute to mortality. Side effects of alprostadil include hypotension from systemic vasodilation, apnea, and fever. Blood pressure should be closely monitored, and hypotension should be treated with judicious intravascular fluid expansion. Most of these infants require prophylactic intubation and mechanical ventilation during transport.

Neonates and infants with shock, respiratory distress, or profound cyanosis may require positive pressure ventilation. Unlike older children and adults, noninvasive positive pressure (NIPPV) will rarely be an option. More likely the patient will need a definitive airway via endotracheal intubation. Prior to intubation ED physicians should prepare for further deterioration due to cardiopulmonary interactions after intubation, the effects of intubation medications, and supplemental oxygen on the pulmonary vascular bed. Pulmonary arterial reactivity and risk for pulmonary hypertensive episodes in response to medications and the unavoidable peri-intubation trauma are important considerations.

Positive pressure ventilation (PPV) has advantages and disadvantages due to cardiopulmonary interactions. Disadvantages include peri-intubation trauma, decrease in venous return leading to decreased RV preload, and increase in RV afterload. The advantages of PPV include decrease in LV afterload, limitation of left-to-right shunting, improvement in pulmonary edema, and reduction in work of breathing. The goal of PPV is to minimize the risks and maximize the benefits.

Careful selection of intubation medications is important. Many sedatives decrease systemic vascular resistance (SVR) and are myocardial depressants which contribute to hypotension, reduced aortic diastolic pressure, and poor cardiac output. Yet, sedation is needed to minimize the risk of pulmonary hypertensive crisis and for safe intubation. Intubation agents should be chosen thoughtfully and are discussed in detail in Chap. 7. In addition, supplemental oxygen should be considered similar to a drug that can have desirable effects (pulmonary vasodilation in infants at risk for pulmonary hypertension) and undesirable effects in other situations (constriction of the DA, pulmonary vasodilation, and worsening of left-to-right shunting).

Summary

ED presentations of pediatric heart disease are age and lesion specific. Children with cardiac emergencies present with respiratory distress, shock, or cyanosis. The likelihood of a given disease entity depends on the age at presentation. Tachycardia, sweating, poor feeding, cyanosis, hepatomegaly, wheezing, and cardiomegaly may be important diagnostic clues. Common pitfalls are misdiagnosis as other respiratory disorders commonly encountered in children such as asthma, bronchiolitis, and pneumonia and conditions such as sepsis and dehydration. Routinely employed ED therapeutic agents such as oxygen, bronchodilators, fluid administration, etc. may be potentially harmful in this patient population, and a high index of suspicion is necessary to manage these complex patients in the acute setting.

Clinical Pearls

- Neonates with ductal-dependent lesions typically present early within the first weeks to a month of life.
- Initiation of alprostadil (PGE1) should not be delayed in a neonate who presents in shock or with cyanosis.
- Infants with large left-to-right shunts and ALCAPA present to the ED with acute heart failure as pulmonary vascular resistance drops between 1 and 6 months of age.
- Myocarditis and arrhythmias may occur at any age.
- Goals of management include stabilizing the airway, initiating alprostadil therapy promptly when indicated, optimizing preload, decreasing afterload, and improving contractility.
- Acute management of heart failure is based on optimizing preload, reducing systemic vascular resistance, and reducing myocardial oxygen consumption.

Board Exam Questions

1. A 3-week-old male born at full term presents to the ED with tachypnea. He was well until yesterday when he stopped feeding. On physical examination he is irritable and tachypneic with a respiratory rate of 60 breaths per minute with a HR of 170 beats per minute. He is afebrile and right upper extremity blood pressure is 70/50 mmHg. Extremities are cool with delayed capillary refill and femoral pulses are difficult to palpate. Oxygen saturations are 94% on room air. What is the most important next step in managing this patient?
   (A) Intubation and mechanical ventilation
   (B) Systemic antibiotics
   (C) Albuterol treatment with administration of 100% oxygen
   (D) **Initiate alprostadil (PGE-1) infusion**
2. The neonate from question 1 has an echocardiogram performed in the ED showing severely depressed LV function and no evidence of a patent ductus arteriosus. The next most appropriate treatment for this child is to initiate:
   (A) Calcium chloride infusion
   (B) Vasopressin infusion
   (C) Continuous albuterol inhalation
   (D) **Epinephrine infusion and reassess end organ perfusion**
3. The neonate from question 1 most likely has which one of the following diagnoses?
   (A) Total anomalous venous return (TAPVR)
   (B) Anomalous coronary artery from the left pulmonary artery (ALCAPA)
   (C) Myocarditis
   (D) **Critical coarctation of the aorta**
4. A 4-month-old with known unrepaired atrioventricular septal defect presents to the ED with poor feeding and respiratory distress. On arrival to her bedside, you see a small infant with tachypnea, retractions, and nasal flaring. Vital signs show a T 37 °C, HR 160 beats per minute, RR 60 breaths per minute, BP 80/40 mmHg, and SpO_2 92% on high-flow nasal cannula at 8 L per minute with 70% FiO_2. On respiratory examination, the liver is 4 cm below the costal margin, and a systolic murmur over the precordium and bilateral wheezes are heard. The nurse asks you if FiO_2 should be raised because of the low SpO_2. Which of the following is the best response?
   (A) Increase the FiO_2 to 80%
   (B) Increase the FiO_2 to 100% and prepare for intubation
   (C) Increase in FiO_2 will not help because of right-to-left shunting
   (D) **Titrate the FiO_2 downwards as long as SpO_2 remains > 90%**

Conflicts of Interest *Disclosures*: The authors have not disclosed any potential conflicts of interest.

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2 Recognizing, Stabilizing, and Managing Children with Heart Failure in the Emergency Department and Other Acute Care Settings

Matthew J. O'Connor, Robert E. Shaddy, and Robert D. Ross

Clinical Vignette

A previously healthy 9-year-old boy was admitted from the emergency department to the general pediatric unit after a week of daily episodes of vomiting, diarrhea, decreased oral intake, and decreased energy and activity. In the emergency department, his blood pressure for age was normal, although tachycardia was present (heart rate 155/min). Cardiac auscultation revealed a gallop versus splitting of the second heart sound. He was given a 500 mL IV fluid bolus, admitted for rehydration, and discharged 24 h later with a diagnosis of gastroenteritis.

He was readmitted to the hospital 1 week later with ongoing vomiting, new epigastric abdominal pain exacerbated by activity, and dyspnea exacerbated by laying supine. Blood pressure was normal, but he continued to be tachycardic. A chest radiograph revealed cardiomegaly and pulmonary edema. He was given an IV fluid bolus and again admitted to the general pediatric unit. Examination after admission was notable for hepatomegaly. His B-type natriuretic peptide concentration was markedly elevated, at 8578 pg/mL. An echocardiogram subsequently revealed a dilated left ventricle and severely diminished left ventricular systolic function, with an ejection fraction (EF) of 15%. He was transferred to the intensive care unit and started on diuretics and inotropic drugs.

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Clinical Vignette Comment

The clinical course of this patient highlights the fact that heart failure (with a subsequent diagnosis of dilated cardiomyopathy in the patient described above) in children may initially present with symptoms manifested primarily through other organ systems (e.g., gastrointestinal, respiratory). Given the high incidence of viral, self-limited gastrointestinal and respiratory illnesses in the community, particularly in children, such symptoms are likely to be attributed to common causes. Children with heart failure are often evaluated several times before the diagnosis is established.

Introduction

The clinical syndrome of heart failure (HF) is defined by the American Heart Association (AHA) as symptoms or signs attributable to inadequate ventricular filling or ejection of blood [1]. This definition encompasses HF with reduced ejection fraction (EF), or systolic dysfunction, as well as HF with preserved EF, frequently referred to as diastolic dysfunction. This latter entity is uncommon in children and is not discussed here. Although HF is generally uncommon in unselected populations of children, it often initially presents in acute care settings, such as the emergency department or urgent care clinic [2]. For this reason, clinicians working in such environments need to know the symptoms, signs, causes, and initial management of HF in children.

Definition of Pediatric Heart Failure

There is no formal definition of HF in children, although several professional societies have published diagnostic classifications for various HF syndromes in children [3, 4]. The definition of HF proposed by the AHA for adults can be applied to children. The different presentations of HF are often classified with various staging criteria, such as the AHA/American College of Cardiology HF staging classification, which includes elements of risk classification, symptomatology, and treatment, and the New York Heart Association (NYHA) HF criteria, which are symptom-based [1] (Table 2.1). Given the unique developmental characteristics of children, however, these classifications may not be as relevant. The original Ross criteria classified HF in children by their developmental status [5] (Table 2.2), and recently, other parameters have been added, such as serum natriuretic peptide concentrations and the results of exercise testing (for older children and adolescents).

Table 2.1 Heart failure classification systems in adults

ACCF/AHA stages of heart failure		NYHA functional classifications	
A	At high risk for HF but without structural heart disease or symptoms of HF	None	
B	Structural heart disease but without signs or symptoms of HF	I	No limitation of physical activity. Ordinary physical activity does not cause symptoms of HF
C	Structural heart disease with prior or current symptoms of HF	I	(as above)
		II	Slight limitation of physical activity. Comfortable at rest, but ordinary physical activity results in symptoms of HF
		III	Marked limitation of physical activity. Comfortable at rest, but less than ordinary activity causes symptoms of HF
		IV	Unable to carry on any physical activity without symptoms of HF or symptoms of HF at rest
D	Refractory HF requiring specialized interventions	IV	Unable to carry on any physical activity without symptoms of HF or symptoms of HF at rest

ACCF American College of Cardiology Foundation, *AHA* American Heart Association, *HF* Heart failure, *NYHA* New York Heart Association
Reprinted from reference [1], with permission from Wolters Kluwer

Table 2.2 Original and Revised Ross classifications for heart failure in children

Original Ross classification
Class I: Asymptomatic
Class II: Mild tachypnea or diaphoresis with feeding in infants; dyspnea on exertion in older children
Class III: Marked tachypnea or diaphoresis with feeding in infants; prolonged feeding times with growth failure resulting from HF; marked dyspnea on exertion in older children
Class IV: Symptoms such as tachypnea, retractions, grunting, or diaphoresis at rest
Revised Ross classification • Defined age groups (0–3 months, 4–12 months, 1–3 years, 4–8 years, 9–18 years) • Ten clinical variables, each scored 0, 1, or 2 – Score 0–5: **Class I** – Score 6–10: **Class II** – Score 11–15: **Class III** – Score 16–20: **Class IV** • Clinical variables – Feeding (infants—3 years), growth (1–8 years), nausea/vomiting (9–18 years) – Breathing – Respiratory rate – Heart rate – Perfusion – Hepatomegaly – NT-proBNP – Ejection fraction – AV valve insufficiency – Maximum percent oxygen consumption (9–18 years)

Reprinted and adapted from reference [6], with permission (Springer publication)

Causes of Heart Failure in Children

Heart failure in children has several causes, and an in-depth discussion of each is beyond the scope of this chapter. Broadly speaking, HF can be caused by primary heart muscle disease (i.e., cardiomyopathy), toxic or inflammatory conditions (i.e., myocarditis, sepsis), acquired heart disease (i.e., Kawasaki disease, rheumatic fever), and congenital conditions (i.e., congenital heart disease, metabolic disease, or inborn errors of metabolism; Table 2.3). The probability a particular diagnosis depends in

Table 2.3 Causes of heart failure in children

Neonates and infants
Congenital heart disease
Obstructive lesions
Aortic stenosis
Aortic arch obstruction
After closure of PDA in critical left-sided heart obstruction
Volume overload
Left-to-right shunts (VSD, AV canal, PDA, truncus arteriosus)
Valvular dysfunction
Atrioventricular valve regurgitation
Semilunar valve regurgitation
Complex defects
Single ventricle
Pulmonary hypertension (right ventricular failure)
Cardiomyopathy
Primary (dilated, noncompaction)
Secondary (arrhythmia, hypothyroidism, hypoglycemia, sepsis, hypoxemia)
Inflammatory
Myocarditis
Kawasaki disease
Metabolic disease (inborn errors of metabolism, mitochondrial disease)
Children and adolescents
Unoperated congenital heart disease (all defects)
Status post surgery for congenital heart disease
Residual defects
Residual atrioventricular or semilunar valve regurgitation
Ischemic injury
Failure of systemic ventricle in single-ventricle congenital heart defects
Pulmonary hypertension (right ventricular failure)
Cardiomyopathy
Primary (dilated, noncompaction; less commonly hypertrophic, restrictive)
Secondary (arrhythmia, sepsis, toxins/chemotherapy, muscular dystrophy, Friedreich ataxia)
Inflammatory
Myocarditis
Kawasaki disease
Metabolic

PDA Patent ductus arteriosus, *VSD* Ventricular septal defect, *AV* Atrioventricular

large part on age at presentation. For example, metabolic disease and congenital heart disease are relatively common causes in infants, whereas cardiomyopathies and myocarditis tend to predominate in adolescents. However, these are broad guidelines, and it is important to recognize that most causes of heart failure can present in any age group. A more in-depth discussion of HF causes and epidemiology in children can be found in several recent reviews, important original papers, and book chapters [6–9].

Pathophysiology

Failure of myocardial pump function decreases cardiac output, which leads to decreased tissue oxygen delivery and metabolic stress in end organs and tissues. However, the clinical syndrome of HF is not simply an imbalance between tissue oxygen supply and demand caused by myocardial dysfunction; rather it encompasses a broad array of compensatory and adaptive responses that involve a number of metabolic, hormonal, and neutrally mediated pathways. An in-depth discussion of these mechanisms is beyond the scope of this chapter, but a simplistic characterization follows. Decreased tissue oxygen delivery leads to the release of endogenous catecholamines, which when released in the circulation act to stimulate the myocardium and increase cardiac output through increasing cardiac contractility and heart rate. While initially effective, this response eventually proves maladaptive by way of increasing myocardial oxygen consumption and myocardial calcium concentration, both of which lead to cellular damage and fibrosis. In the setting of decreased cardiac output, there is a resultant increase in extracellular fluid volume. In the renal circulation, this leads to the activation of the renin-angiotensin-aldosterone pathway, which has a number of downstream effects including natriuresis and diuresis, but also activates metabolic pathways that lead to myocardial fibrosis and scarring, a process referred to as remodeling. While alteration of the abovementioned pathways is not critical in the acute management of decompensated HF, they are important in the therapy of chronic HF. For example, blockade of the renin-angiotensin-aldosterone system with angiotensin-converting enzyme inhibitors and the mineralocorticoid receptor antagonist aldosterone are mainstays of the management of chronic HF. Further discussion of the various mechanisms involved in HF can be found in several excellent reviews [10, 11].

Clinical Presentation

The clinical presentation of children with HF in the acute setting varies with its cause, the age of the child, and the timing of disease onset (acute versus chronic), in addition to several other factors. As highlighted in the opening clinical vignette, practitioners must recognize that HF in children often masquerades as common childhood maladies, particularly in the early stages of disease, which of course can have serious outcomes. For example, wheezing caused by left atrial and pulmonary venous hypertension and resultant transudation of fluid into the interstitial space between alveoli in left ventricular dilation and systolic dysfunction could readily be

mistaken for reactive airway disease, asthma, or bronchiolitis. In addition, vague gastrointestinal complaints, such as vomiting, abdominal pain, and fussiness during feeding are common in infants and children with HF [2]. Despite the high index of suspicion needed to discriminate children with HF from more common, self-limited conditions, all children presenting with common signs and symptoms should not be suspected of having HF or undergo targeted evaluation for it. A detailed history and physical examination with appropriate imaging and laboratory studies, however, should be able to distinguish children with self-limited conditions from those with more serious underlying disease. In particular, HF should be considered in children with repeated presentations of the same signs and symptoms or those who have not responded to usual therapies.

Heart failure is often associated with systemic or pulmonary venous congestion and impaired cardiac output. However, the balance of these factors varies among patients, as do compensatory responses. Conceptually, children with HF can be placed into one of four categories, depending on the adequacy of systemic perfusion ("warm" or "cold"), as well as the presence or absence of venous congestion ("wet" or "dry") [12]. These four categories include "wet and warm" (the most common presentation), "dry and warm," "dry and cold," and "wet and cold." The appropriate category can be determined rapidly at the bedside by physical examination without the need for diagnostic testing. For example, a patient may conceptually be described as "wet" on the basis of peripheral edema, hepatomegaly, or the presence of tachypnea or rales on auscultation of the lungs. The distinction between "cold" and "warm" may be made by assessing the quality and amplitude of the peripheral pulses and duration of the capillary refill time. In addition, as discussed below, placing patients into one of these four categories can help guide initial therapy.

Diagnostic Evaluation

History

The first steps in diagnosis are the history and physical examination. Children with acute HF often have prodromal symptoms suggesting a viral infection, with lethargy, poor oral intake, and varying degrees of dyspnea. Fever may be present in the setting of viral myocarditis, rheumatic fever, or Kawasaki disease. Children with more chronic and indolent symptoms, such as those of the dilated cardiomyopathies, often have vague symptoms, such as slowly worsening fatigue, weight loss or gain, and dyspnea on exertion. Heart failure may occur in children and young adults with certain forms of congenital heart disease, particularly those with prior palliative surgery for single-ventricle physiology (Fontan operation) and survivors of other complex surgical reconstructions (e.g., atrial switch procedures [Mustard/Senning] for transposition of the great arteries). Children with a history of cancer who have received chemotherapy, particularly anthracycline agents, are at risk for the development of HF. In these patients, HF may become apparent many years after receiving chemotherapy. A pediatric cardiologist should be involved early when HF is suspected.