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Cardiac Emergencies in the First Year of Life

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The presence of a distressed or obtunded infant in any adult or pediatric emergency department can prove to be a challenging process in airway management, vascular access, and decision making. Cardiac emergencies, as well as a number of other diseases, can present in this manner. It is essential to accurately diagnose and expeditiously care for these potentially complicated cardiac patients. Diagnosis can be difficult because of a number of nonspecific elements in the history and physical exam. However, by developing an effective strategy in dealing with these patients, the emergency department management of these individuals can be completed in an efficient and prompt manner.

The most challenging scenarios of cardiac emergencies in the first year of life include cyanotic episodes, congestive heart failure, cardiogenic shock or collapse, and arrhythmias. All of these emergent presentations can be the result of either the initial presentation of disease or as a known complication of an already diagnosed cardiac lesion.

In approaching cardiac emergencies, cardiac disease can be divided into structural disease, conduction abnormalities, and acquired illnesses. While recognizing that many lesions can be a combination of many defects, structural congenital heart disease can be divided into cyanotic and acyanotic categories. The cyanotic category can be further subdivided into increased and decreased pulmonary blood flow. Division of the acyanotic category is based on left-to-right shunting and left ventricular outflow obstruction. Conduction abnormalities can be congenital or the result from a new-onset illness. Acquired heart disease includes cardiomyopathies, myocarditis, pericarditis, endocarditis, and Kawasaki's disease.

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A cyanotic patient suggests that there is cyanotic congenital heart disease with shunting from the right to the left. In a patient with cardiogenic shock or collapse (the result of outflow obstruction and pump failure), the infant may appear mottled, ashen, and gray. A patient with left-to-right shunting and congestive heart failure can appear to be normal in color [1–7]. This article will discuss the cardiac emergencies that may present within the first year of life.

Basic pathophysiology

There are a number of changes that occur within the cardiovascular system in the transition from a fetus to a newborn. The placenta functions as the pulmonary system for the fetus, as oxygenated blood is transferred from the placenta to the fetus via the umbilical vein. At birth, blood then travels through a now lower resistance pulmonary system for oxygenation with closure of the shunts that were used between the pulmonary and systemic circulations (foramen ovale, ductus arteriosus, ductus venosus). Expansion of the lungs and the elimination of fluid from the lungs cause dilatation of the pulmonary vasculature, which then leads to a decrease in pulmonary resistance and increased pulmonary blood flow. Oxygenation of the blood through the pulmonary system leads to the closure of the umbilical vessels, the ductus arteriosus, and the ductus venosus. Decreased pulmonary artery resistance and subsequent increased systemic resistance changes the flow though the atria, with pressures now higher in the left atria than the right, resulting in the closure of the foramen ovale [8,9].

Cyanosis

Cyanosis is seen when desaturated blood is present in the capillary beds. Deoxygenated hemoglobin is blue and the presence of cyanosis means that there is 3 to5 mg/dL of deoxyhemoglobin in the blood. This corresponds with a room air oxygen saturation of 70% to 85% [10,11]. Because the oxygen carrying capacity is based on the amount of hemoglobin available to carry oxygen, an infant who is polycythemic and cyanotic is still able to deliver oxygen to tissues as opposed to an anemic infant who may not appear cyanotic but is not able to deliver oxygen to tissues.

It is important to differentiate between central and peripheral cyanosis as the evaluation and treatment differ based on the underlying cause. There are a number of different causes for central cyanosis. These include central nervous system (CNS) depression, pulmonary disease, and cardiac disease as well as sepsis and metabolic disease and toxic ingestions. Peripheral cyanosis is the result of acrocyanosis, exposure to cold, and decreased peripheral perfusion.

Factors to keep in mind when assessing cyanosis are the arterial oxygen saturation, the oxygen binding capacity (hemoglobin), and the arteriovenous oxygen difference [10].

Cyanotic heart disease

There are five well-known cyanotic congenital heart lesions—also known as the ''Terrible Ts.'' They are Tetralogy of Fallot (TOF), Transposition of the Great Arteries (TGA), Tricuspid Atresia (TA), Total Anomalous Venous Return (TAPVR), and Truncus Arteriosus.

Tetralogy of Fallot

Tetralogy of Fallot is the most common form of cyanotic congenital heart disease in the post infancy period and represents up to 10% of all congenital heart disease [12,13]. Tetralogy of Fallot consists of four basic lesions. The lesions are a large ventricular septal defect (VSD), right ventricular outflow obstruction (from pulmonic stenosis), an overriding aorta, and right ventricular hypertrophy. Two of the lesions will determine the extent of the disease pathophysiology. There must be right ventricular outflow obstruction and the VSD must be large enough to equalize pressures in both of the ventricles.

The extent of obstruction of the right ventricular outflow track will determine the amount of cyanosis present in the patient. Systolic pressures are equally balanced in the right and left ventricle because of the nonrestrictive VSD. There will be a left-to-right shunt, a bidirectional shunt, or a right-to-left shunt depending on the extent of the right ventricular outflow tract obstruction. If the pulmonic stenosis is severe, there will be a right-to-left shunt with subsequent cyanosis and decreased pulmonary blood flow. If there is mild pulmonic stenosis, a left-to-right shunt will occur resulting in an acyanotic Tetralogy of Fallot.

In addition to cyanosis, the physical exam may show a systolic thrill at the lower and middle left sternal border. A loud and single S2, an aortic ejection click, and a loud grade 3 to 5/6 systolic ejection murmur in the middle to lower left sternal border will also be found. A continuous patent ductus arteriosus (PDA) murmur may also be present.

The ECG will show right axis deviation (RAD) and right ventricular hypertrophy (RVH).

A boot-shaped heart with a main pulmonary artery segment is characteristic of the cyanotic Tetralogy of Fallot. The heart size is normal with decreased pulmonary vascular markings. Acyanotic Tetralogy of Fallot will have chest x-rays similar to that of moderate VSDs.

Transposition of the great arteries

Transposition of the great arteries represents around 5% to 8% of congenital heart disease and is the most common cyanotic heart lesion in the newborn period [14]. There are many variations of the disease, with the underlying factor being that the aorta originates from the right ventricle and that the main pulmonary artery has origins in the left ventricle. Within these

two distinct circulatory systems, the main pulmonary artery has a significantly higher oxygen saturation than the aorta, with hyperoxemic blood traveling through the pulmonary system and hypoxic blood traveling within the systemic system.

The presence of a VSD, atrial septal defect (ASD), or PDA is essential to survival, because the mixing of the circulations is the only way of providing oxygenated blood to the systemic system. A VSD can be found in approximately 20% to 40% of patients.

With progressive closure of the PDA, cyanosis becomes more prevalent. Hypoxia and acidosis result from the suboptimal mixing of oxygenated and deoxygenated blood.

Congestive heart failure is a common presentation in the first week of life, with dyspnea and feeding difficulties in addition to the cyanosis. If the interventricular septum is intact, these patients will be the critically ill. The severe arterial hypoxemia will not respond to the administration of oxygen. Acidosis as well as hypocalcemia and hypoglycemia are common. They will respond well to PGE1 infusion and, ultimately, a Rashkind balloon septostomy. If there is a VSD or large PDA, these patients will not be as cyanotic but will present with congestive heart failure and obstructive pulmonary disease.

There will be a loud, single S2. If there is a VSD, a systolic murmur can be heard. Otherwise, there are no specific auscultatory findings.

The ECG will show right axis deviation (RAD) and right ventricular hypertrophy (RVH).

The egg-shaped heart with a narrow mediastinum is the characteristic chest x-ray. There is cardiomegaly with increased pulmonary vascular markings (Fig. 1).

Echocardiogram will show two circular structures instead of the circle and sausage pattern of normal great arteries.

Total anomalous pulmonary venous return

TAPVR represents around 1% of congenital heart disease [15]. The pulmonary veins bring the blood from the lungs to the right atrium instead of the left atrium. TAPVR is generally divided into four groups, depending on where the pulmonary veins drain. In the supracardiac type (50%) the common pulmonary vein attaches to the superior vena cava. In the cardiac type (20%) the common pulmonary vein empties into the coronary sinus. In the infracardiac/subdiaphragmatic type (20%), the common pulmonary vein empties into the portal vein, ductus venosus, hepatic vein, or inferior vena cava. A mixed type is seen in 10% of the lesions, which is a combination of any of the types. An ASD or patent foramen ovale is necessary for mixing of the blood.

Pulmonary venous return is delivered to the right atrium, and there is mixing of the pulmonary and systemic circulations. Blood flow then travels to the left atrium through the ASD and to the right ventricle. Systemic

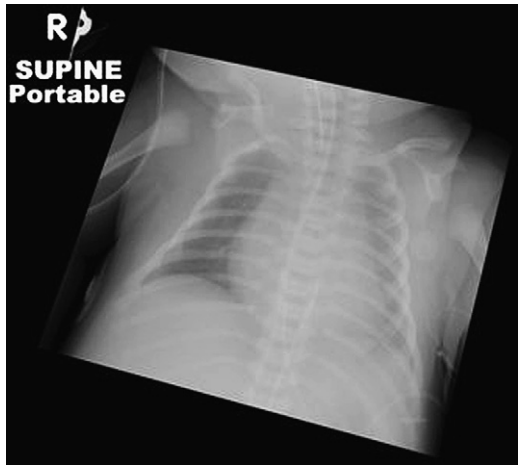

Fig. 1. Chest radiograph of TGA with cardiomegaly and increased vascular markings.

arterial desaturation occurs as the result of mixing of pulmonary and systemic blood. Pulmonary blood flow determines the amount of desaturation of systemic arterial blood. If there is no obstruction to pulmonary venous return, there is minimal desaturation of the systemic blood. If there is obstruction to pulmonary venous return, there is significant cyanosis. With the blood from both the pulmonary and systemic circulations pumped by the right ventricle, there can be volume overload, with subsequent right ventricular and atrial enlargement.

In a patient without pulmonary venous obstruction, there can be a history of frequent pneumonias and growth difficulties. Patients will frequently present with a congestive heart failure presentation with tachypnea, tachycardia, and hepatomegaly, in addition to slight cyanosis. There will be a hyperactive right ventricular impulse, with a split and fixed S2. A grade 2 to 3/6 systolic ejection murmur is at the upper left sternal border, with a mid diastolic rumble at the left lower sternal border.

The ECG will show right axis deviation, right ventricular hypertrophy, and right atrial enlargement (Fig. 2).

Chest x-ray will exhibit significant cardiomegaly with increased pulmonary vascular markings (Fig. 3). The characteristic ''snowman sign'' is found in infants older than 4 months.

In those patients with TAPVR and pulmonary venous obstruction, cyanosis and respiratory distress dominate the presentation. There can be minimal cardiac exam findings aside from a loud and single S2 and gallop rhythm. A murmur is usually not found.

The ECG will also show right axis deviation and right ventricular hypertrophy and the chest radiograph will have a normal heart silhouette with lung fields consistent with pulmonary edema.

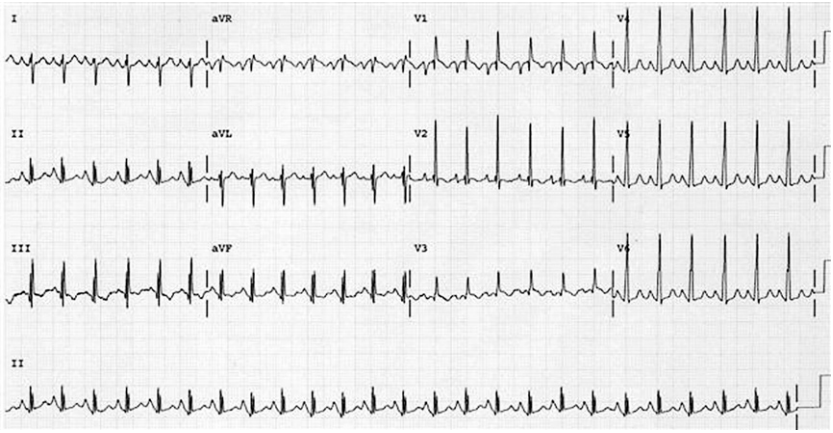

Fig. 2. ECG of TAPVR with right atrial enlargement, right ventricular hypertrophy.

Tricuspid atresia

Tricsupid atresia represents 1% to 2% of congenital heart disease in infancy [16]. There is no tricuspid valve and there is underdevelopment of the right ventricle and pulmonary artery. Therefore, pulmonary blood flow is decreased. With no flow across the right atrium to the right ventricle, the right atrium needs a right-to-left shunt to empty, making an ASD, VSD, or PDA essential for survival. The great arteries are transposed in 30% of the cases, with a VSD and no pulmonic stenosis. In 50% of cases there is normal artery anatomy, with a small VSD and pulmonic stenosis.

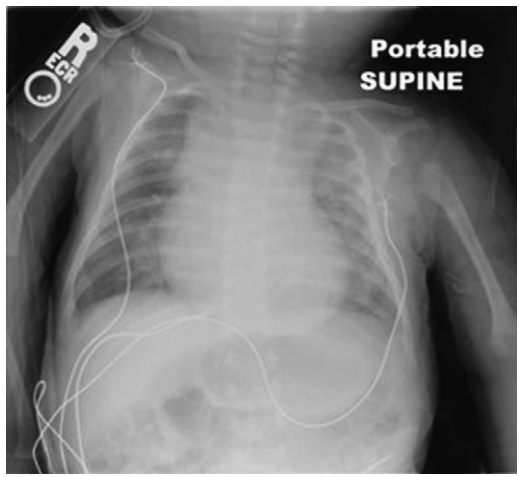

Fig. 3. Chest radiograph of TAPVR with cardiomegaly and increased vascular markings.

There will be right atrial dilatation and hypertrophy because all systemic venous return is shunted from the right atrium to the left atrium. Enlargement of the left atrium and ventricle occurs because of the work of handling both systemic and pulmonary returns.

The amount of cyanosis is inversely related to the amount of pulmonary blood flow.

Severe cyanosis, tachypnea, and poor feeding are common presentations. There is a single S2. The murmur is a grade 2 to 3/6 systolic regurgitant murmur from the VSD and is heard best at the left lower sternal border. There can also be a continuous murmur of a PDA. Hepatomegaly can be found with congestive heart failure.

The ECG has a superior QRS axis, along with right atrial hypertrophy (RAH), left atrial hypertrophy (LAH) and left ventricular hypertrophy. The chest radiograph will show a normal to slight increase in heart size along with decreased pulmonary vascular markings.

Truncus arteriosus

Truncus arteriosus is seen in less than 1% of all congenital heart disease [17]. All of the pulmonary, systemic, and coronary circulations result from a single arterial trunk. A large VSD is associated with this, as well as abnormalities of the coronary arteries.

DiGeorge syndrome (hypocalcemia, hypoparathyroidism, absence or hypoplasia of the thymus, chromosomal abnormalities) is often seen with truncus arteriosus. Pulmonary blood flow can be normal, increased, or decreased, depending on the type of truncus arteriosus.

There is a direct relationship between the amount of pulmonary blood flow and the degree of systemic arterial oxygen saturation. Cyanosis is prevalent with decreased pulmonary blood flow, and is minimal with increased pulmonary blood flow. Congestive heart failure can be seen with increased pulmonary blood flow. The left ventricle has to deal with significant volume overloads.

Usually within the first weeks of life, the patient will present with congestive heart failure and cyanosis. There will be a loud regurgitant 2 to 4/6 systolic murmur at the left sternal border, sometimes associated with a high-pitched diastolic decrescendo murmur or a diastolic rumble. The S2 will be single and accentuated.

The ECG will usually show bilateral ventricular hypertrophy and the chest radiograph will have cardiomegaly with increased pulmonary vascular markings.

Acyanotic heart disease

Left-to-right shunt lesions include ventricular septal defects, atrial septal defects, patent ductus arteriosus, and endocardial cushion defects. This

group comprises almost 50% of all congenital heart disease [18]. Left-to-right shunt lesions have blood shunted from the systemic system into the pulmonary system. The high pulmonary vascular resistance in the neonate controls the amounts shunted but once pulmonary vascular resistance starts to drop in the first few weeks of life, pulmonary blood flow and pressures will increase. The extent of the lesion is directly related to the degree of pulmonary vascular blood flow. More blood flow will lead to chamber enlargement, and increased pulmonary vascular pressures and subsequent signs of congestive heart failure.

Atrial septal defects

Atrial septal defects comprise up to 10% of all congenital heart disease [19]. In infancy this connection from the left to right atria has the potential for causing problems in about 10% of patients [14]. If there is a large defect, or if there are associated defects, there will be considerable left-to-right shunting and subsequent overload of the pulmonary circulation. Some defects will close spontaneously but larger defects will require surgical intervention.

Difficulty feeding and difficulty gaining weight are common complaints.

The cardiac exam will have a widely split and fixed S2, with a grade 2 to 3/6 systolic ejection murmur at the upper left sternal border, sometimes associated with a mid-diastolic rumble.

ECG findings include right axis deviation and right ventricular hypertrophy or right bundle branch block.

Chest radiograph will have cardiomegaly with increased pulmonary vascular markings.

Ventricular septal defects

Ventricular septal defects are the most common type of congenital heart disease. Seen in approximately 25% of all congenital heart disease cases [20], ventricular septal defects allow for mixing of blood in the ventricles. The extent of the defect determines the degree of disease. Small defects will have minimal impact, as compared with large defects, which will cause pulmonary hypertension and congestive heart failure. Large VSDs have volume and pressure overload in the right ventricle as well as volume overload in the left atrium and left ventricle.

In larger VSDs, poor weight gain along with delayed development are common. Congestive heart failure and cyanosis are frequent presentations.

The exam will have a grade 2 to 5/6 systolic murmur (holosystolic) heard best at the left lower sternal border. A systolic thrill or diastolic rumble can also be present with a narrowly split S2.

ECG findings in a moderate VSD will show left atrial hypertrophy and left ventricular hypertrophy. In a larger VSD there will be left and right

ventricular hypertrophy and left atrial hypertrophy. The chest radiograph can show cardiomegaly as well as increased pulmonary vascular markings.

Patent ductus arteriosus

Seen in 10% of all congenital heart disease, the ductus arteriosus remains patent and does not close as it ordinarily would [18]. The degree of the left-to-right shunting is dependent on the lesion length and diameter and pulmonary vascular resistance. The larger the left-to-right shunt, the more symptomatic the patient will be. Ordinarily, in healthy patients the ductus arteriosus will close within 15 hours after birth and then will completely seal around 3 weeks of age, becoming the ligamentum arteriosum. Hypoxia and prematurity have a tendency to keep the ductus arteriosus patent.

If the defect is large, as with all left-to-right shunts, signs of congestive heart failure will be present.

Physical exam will be remarkable for a grade 1 to 4/6 continuous machinery like murmur heard best at the left upper sternal border. A diastolic rumble can also be present as well as bounding peripheral pulses.

ECG findings can show left and right ventricular hypertrophy in large PDAs.

Chest radiograph will have cardiomegaly and increased pulmonary vascular markings.

Endocardial cushion defect

When the endocardial cushion does not develop properly, there will be defects to the atrial septum, the ventricular septum, and the atrioventricular valves. Complete defects involve the entire endocardial cushion and will have atrial and ventricular septal lesions and a common atrioventricular valve. Incomplete or partial defects have atrial involvement with an intact ventricular septum. There can also be variations of both complete and incomplete lesions. A history of failure to thrive, and multiple respiratory tract infections are common. Endocardial cushion defects represent around 3% of congenital heart disease and almost two thirds have the complete form [18]. Down's syndrome is strongly associated with the complete form of endocardial cushion defects.

Left-to-right shunting is directly dependent on the extent of the defects, with complete lesions presenting with congestive heart failure early from volume overload in both the left and right ventricles.

Cardiac exam will be remarkable for a hyperactive precordium, a systolic thrill, a loud holosystolic regurgitant murmur, and a loud and split S2.

The ECG will show a superior QRS axis with RVH, right bundle branch block (RBBB), and left ventricular hypertrophy, along with a prolonged PR interval (Fig. 4).

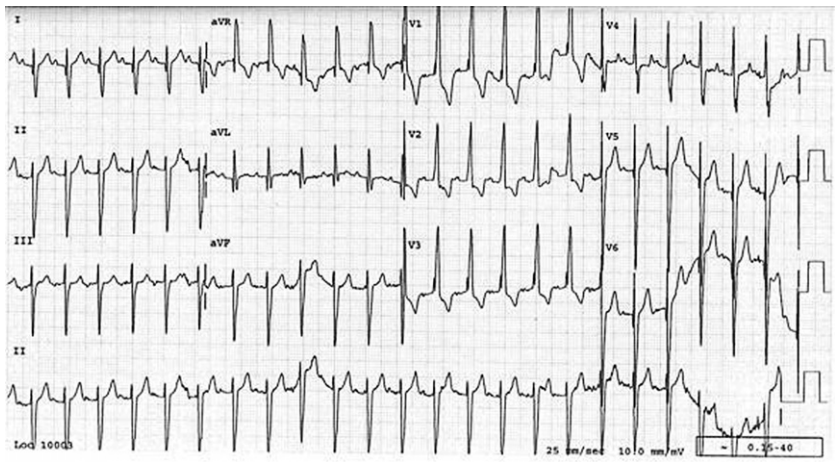

Fig. 4. ECG of CAVC (common AV canal) or endocardial cushion defect with superior QRS axis.

Coarctation of the aorta

Coarctation of the aorta represents 8% to 10% of congenital heart disease and is seen in males in a 2:1 ratio [21]. There is congenital narrowing of the aorta, in the upper thoracic aorta in the region of the ductus arteriosus. The extent of illness is a factor of the degree of narrowing, the length of the narrowing and the presence of other cardiac defects. If the right ventricle supplies the descending aorta via the PDA in fetal life, infants will be symptomatic early. Many other cardiac defects are present such as a VSD, PDA, and aortic hypoplasia and collateral circulation is underdeveloped.

The PDA is able to temporarily negate the obstructive effects of the coarctation obstruction. Additionally, the PDA can maintain blood flow to areas distal to the obstruction. When the PDA eventually closes, the development of pulmonary hypertension and subsequent pulmonary venous congestion leads to congestive heart failure.

Tachypnea, feeding difficulties, and minimal urine output along with shock and metabolic acidosis are common presentations. When presenting in congestive heart failure, there will be a loud gallop, a murmur may or not be present, and pulses will be weak.

The ECG will show RVH or RBBB. There will be significant cardiomegaly as well as pulmonary edema on chest radiograph (Fig. 5). In older children, the appearance of notching of the first rib, also known as the ''3 Sign'' may be present.

The presence of decreased pulses in the lower extremities is key in the diagnosis of a coarctation. Comparison of the right upper extremity blood pressures and pulse oximeter readings with the lower extremity aids in the

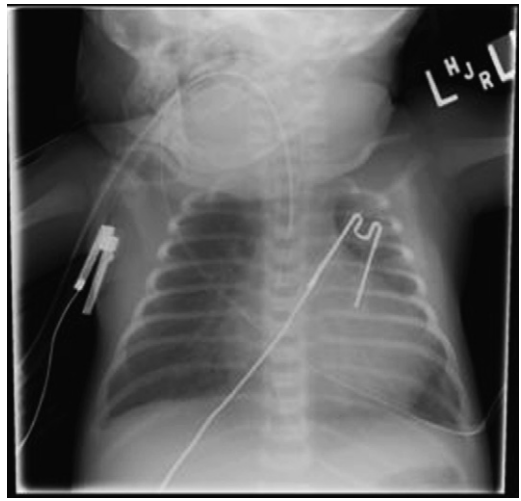

Fig. 5. Chest radiograph of coarctation with cardiomegaly and pulmonary edema.

diagnosis. If the patient is in significant shock, however, pressures can be decreased everywhere.

Hypoplastic left heart syndrome

Hypoplastic left heart syndrome (HLHS) includes hypoplasia of the left ventricle and hypoplasia of the ascending aorta and aortic arch. There can be atresia or marked stenosis of the mitral and aortic valves. The left atrium is also underdeveloped. The ultimate result is that of minimal left ventricular outflow [22].

In utero, the pulmonary vascular resistance is higher than the systemic vascular resistance. The right ventricle (through the right-to-left shunt of the ductus arteriosus) and the elevated pulmonary vascular resistance are able to keep a normal perfusion pressure to the descending aorta and systemic fetal system. The hypoplastic left ventricle does not contribute. An ASD allows the left atrium to decompress. All systemic blood flow is dependent on the ductus arteriosus. After birth, significant problems occur. Systemic vascular resistance is now greater than pulmonary vascular resistance, reversing the pressure system. The patent ductus arteriosus now begins to gradually close. With the nonfunctioning left side and increased systemic vascular resistance, cardiac output falls and aortic pressure drops. This leads to circulatory shock and metabolic acidosis. Increased pulmonary blood flow leads to an increase in left atrial pressure and subsequent pulmonary edema.

These patients appear listless, dusky with tachypnea. There is a single heart sound with a systolic ejection murmur and diminished pulses. The

ECG will show right atrial enlargement, right ventricular hypertrophy, and peaked P waves. The chest radiograph will show cardiomegaly.

Aortic stenosis

Aortic stenosis is seen in 6% of congenital heart disease, with a 4:1 ratio in males [23]. The stenosis will be at the valvular, supravalvular, or subvalvular level, with the degree of obstruction determining the severity of disease in the patient. Those with severe obstruction (approximately 10% to 15%) will present with congestive heart failure in infancy [24]. Left ventricular hypertrophy will develop with severe stenosis. The most common type of aortic stenosis is a bicuspid aortic valve. William Syndrome has supravalvular stenosis in addition to elfin facies, mental retardation, and pulmonary artery stenosis.

The physical exam will be remarkable for a systolic thrill in the region of the upper right sternal border, suprasternal notch, or carotid arteries. There can be an ejection click. The murmur will be a rough or harsh systolic murmur grade 2 to 4/6 at the right intercostal space or left intercostal space with transmission to the neck.

In cases of severe aortic stenosis, the ECG will show left ventricular hypertrophy. If there is resultant congestive heart failure, the chest radiograph will show cardiomegaly.

Anomalous origin of the left coronary artery (ALCAPA Syndrome, Bland-White-Garland Syndrome)

In anomalous origin of the left coronary artery (also known as ALCAPA or Bland-White-Garland Syndrome), the left coronary artery has origins in the pulmonary artery instead of the aorta. When pulmonary artery pressure diminishes in the second to third month of life, there will be decreased perfusion of the left ventricle, resulting in a distressed patient with cardiomegaly and congestive heart failure. There may or may not be a murmur consistent with mitral regurgitation [25,26].

The ECG will show myocardial infarction with abnormally deep and wide Q waves, inverted T waves, and ST segment changes in the precordial leads (Fig. 6). The chest radiograph will be most likely show cardiomegaly. An echocardiogram will help in the diagnosis, with an aortogram if necessary.

Acquired disease

Inflammatory diseases of the heart are grouped under carditis. Included in this group are myocarditis, pericarditis, and endocarditis (along with valvulitis).

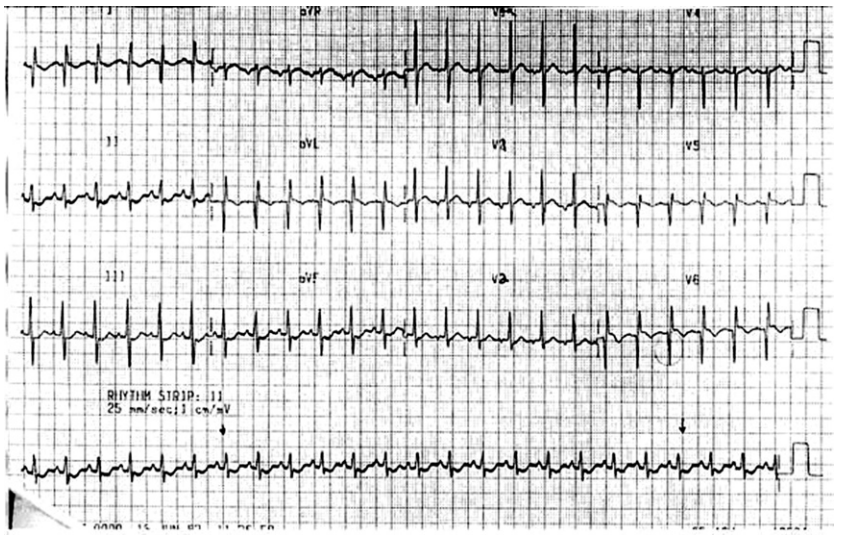

Fig. 6. ECG of anomalous origin of the left coronary artery (ALCAPA) with deep and wide Q waves, inverted T waves, and ST segment changes.

Myocarditis

There are a number of different etiologies in myocarditis. Infectious and autoimmune, as well as toxin-mediated processes can contribute to the inflammatory response in the myocardium [27,28].

Viruses, such as adenovirus, coxsackievirus, echovirus, mumps, and rubella, are the most commonly associated infectious agents. Nonviral causes such as protozoans (Chaga's Disease seen in South America) also cause myocarditis. Less frequently, bacteria, rickettsia, fungal, mycobacteria, and other parasites can be etiologic agents.

Kawasaki's disease and acute rheumatic fever as well as collagen vascular disease can also be seen with myocarditis. Toxic myocarditis is the result of drug ingestion.

Infants may present with vomiting, decreased activity, poor feeding, and congestive heart failure, with tachycardia, tachypnea, a gallop rhythm, and decreased heart tones.

There are no specific lab tests for myocarditis. Erythrocyte sedimentation rate, white blood cell count, myocardial enzymes, and cardiac troponin will be normal or elevated. Troponin levels are thought to be more sensitive than cardiac enzymes [29]. Chest radiograph will show cardiomegaly and, depending on the extent of the disease, pulmonary venous congestion.

ECG abnormalities are common but are nonspecific. There will be tachycardia, low QRS voltages, flattened or inverted T waves with ST-T wave changes, and prolongation of the QT interval. Arrhythmias such as premature contractions are also seen.

Echocardiogram studies will show dilatation of the heart chambers and decreased left ventricular function. The echocardiogram will also help to evaluate myocardial contractility and the presence of a pericardial effusion. Radionuclide scanning and endomyocardial biopsies can help in confirming the disease.

The mortality rate in symptomatic neonates with acute viral myocarditis can be significant. Management of myocarditis revolves around identifying an etiologic agent and, if identified, treating that suspected agent, treating the congestive heart failure, and controlling the arrhythmias. Rest, supplemental oxygen, rapid-acting diuretics like furosemide, and rapid-acting inotropic agents such as dopamine and dobutamine are mainstays in treatment along with the use of angiotensin-converting enzyme inhibitors like captopril. Digoxin is used cautiously because of its potential to induce arrhythmias. In Kawasaki's disease, high-dose immunoglobulins have been beneficial. Other treatment modalities, such as immunosuppressive agents and corticosteroids (except in severe rheumatic carditis) are not universally accepted.

Pericarditis

Inflammation of the pericardium is the hallmark of pericarditis. The most common cause in infancy is a viral etiology such as coxsackie, echovirus, adenovirus, or influenza. Viral pericarditis is usually associated with a viral myocarditis, with the myocarditis being the more prominent entity. Bacterial causes include *Staphylococcus aureus, Streptococcus pnuemoniae, Haemophilus influenzae, Neisseria meningitides,* and streptococci as well as tuberculosis. Acute rheumatic fever, collagen vascular disease, and uremia can also cause a pericarditis. Postpericardiotomy syndrome is seen in patients who have had cardiac surgery involving interruption of the pericardium.

Since the pericardium is a fixed space, the extent of symptoms and signs of disease will be determined by the rate of accumulation of fluid and by the health of the myocardium.

If the myocardium is normal and fluid accumulation is slow, then the patient will tolerate the pericarditis better than if there was underlying myocardial injury with a slow collection of fluid or if there was a rapid collection of a large amount of fluid.

If pericardial tamponade were to occur, the heart, to improve hemodynamics would increase heart rate (improves cardiac output), increase systemic vascular resistance (offset hypotension), and improve diastolic filling by systemic and pulmonary venous constriction.

There is usually a predisposing illness in the history, with an upper respiratory infection or, in the case of a bacterial pericarditis, a pneumonia, empyema, osteomyelitis, pyelonephritis, or tonsillitis.

A pericardial friction rub is diagnostic. A murmur may not be found and the heart will be hypodynamic.

On ECG there will be a low-voltage QRS complex. Early in the disease, ST segments will be elevated everywhere except in V1 and aVR. Later in the disease, ST segments will return to normal and the T waves will flatten or invert. A chest radiograph will show cardiomegaly, with the heart in a water-bottle shape.

Echocardiogram is the key to establishing the presence of an effusion. Additionally, the echo can also evaluate for cardiac tamponade, as it will show the collapse of the right atrial wall or the right ventricular wall in diastole.

To treat pericarditis or pericardiocentesis, surgical intervention is essential, especially if an infectious etiology is suspected. Multiple blood cultures are also indicated as well as standard fluid studies. In milder cases not requiring drainage or antibiotics, antivirals, or antifungals, nonsteroidal anti-inflammatory drugs can be used to treat the discomfort.

In postpericardiotomy syndrome, which can affect as many as 30% of pediatric patients who undergo cardiovascular surgery involving the pericardium, the patients will present with fever, irritability, and a pericardial friction rub anywhere from a month to a few months postoperatively. The etiology is thought to be autoimmune [30].

In cardiac tamponade with signs of tachycardia, tachypnea is an immediate concern.

Chest radiograph will have cardiomegaly and pleural effusion. ECG will have ST segment elevation and flat or inverted T waves. The most helpful test is an echocardiogram because this will assess the amount of pericardial effusion as well as the presence of cardiac tamponade.

Endocarditis

Congenital heart disease is a significant risk factor in infective endocarditis. It is thought that turbulent flow from pressure gradients leads to endothelial damage and thrombus formation. Transient bacteremia then seeds the damaged areas. With the exception of a secundum ASD, all congenital heart diseases and valvular heart diseases are prone to endocarditis, especially if there is any artificial material within the heart (prosthetic heart valve or graft). Common bacterial causes include *S viridans*, enterococci, and *S aureus* as well as fungal and bacteria such as Eikenella, Cardiobacterium [31].

In infancy, endocarditis is rare and is associated with open-heart surgery. The usual presentation is with fulminant disease and a septic appearance. A heart murmur and fever are always present. Embolic phenomena tend to be seen more in the adult population.

Using the Duke Criteria for Infective Endocarditis, a patient must have two major criteria or one major criterion with three minor criteria or five minor criteria. Major criteria include two separately obtained positive blood cultures growing the typical microorganisms and an echocardiogram with

endocardial involvement such as an intracardiac mass on a valve, abscess, partial dehiscence of a prosthetic valve, or new valvular regurgitation. Minor criteria include predisposing conditions, fever, vascular phenomena (emboli, hemorrhages, Janeway lesions), and immunologic phenomena (glomerulonephritis, Osler's nodes, Roth spots, rheumatoid factor), microbiological evidence (positive blood culture not meeting major criteria), and echocardiographic findings (not meeting major criteria).

While an echocardiogram identifying valvular vegetation is helpful in the evaluation, the echocardiogram is not 100% sensitive or specific. Because of this, a negative echocardiogram does not exclude endocarditis. A more definitive diagnosis is made by obtaining a positive blood culture. The isolation of a specific microorganism is key to determining antibiotic therapy. Treatment regimens may take place for weeks to be certain that the microorganism has been eliminated.

Kawasaki's disease

Kawasaki's disease (mucocutaneous lymph node syndrome) is a self-limiting generalized systemic vasculitis of indeterminate etiology. Fever, bilateral nonexudative conjunctivitis, erythema of the mucous membranes (lips, oral mucosa), rash, and extremity changes are the hallmarks of the disease. It is among the most common systemic vasculitic illnesses along with Henoch-Schoenlein Purpura. Kawasaki's primarily affects infants and younger children, and can occur in endemic or community-wide epidemic forms [32].

Coronary artery aneurysms or ectasia have been found in 15% to 25% of untreated children with Kawasaki's [33]. These coronary artery lesions can lead to myocardial infarction, sudden death, or ischemic heart disease [34,35].

In the acute phase of Kawasaki's, there can be involvement of all parts of the heart—the pericardium, the myocardium, the endocardium, the valves, and the coronary arteries. The cardiac exam can show a hyperdynamic precordium, tachycardia, a gallop, and a flow murmur or regurgitant pansystolic murmur. Depressed myocardial function can present as cardiogenic shock. The ECG will show nonspecific ST and T wave changes, a prolonged PR interval, or arrhythmia.

The classic Kawasaki's patient will present with fever greater than or equal to 5 days' duration, and at least four of the primary physical criteria, which include involvement of the extremities, the skin, the conjunctivae, the lips and mouth, and the cervical lymph nodes. The extremity changes include erythema to the palms and soles, with induration and desquamation to the fingers and toes. There can be an extensive erythematous rash that is usually a nonspecific diffuse maculopapular rash. Sometimes early desquamation in the perineal region can occur. Bilateral conjunctival injection

involving the bulbar conjunctivae is seen around the time of the fever. There can be erythema; peeling, cracking, or bleeding from the lips and mouth; a strawberry tongue; and diffuse erythema of the mucosa of the oropharynx. The cervical lymphadenopathy is generally unilateral, and usually one node is greater than 1.5 cm in diameter.

Lab findings include thombocytosis (appears in second week, peaking in third week), leukocytosis, and anemia. Thrombocytopenia in active disease is a risk factor for coronary aneurysms. There is elevation of the C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR). Serum transaminases can be moderately elevated. Gammaglutamyl transpeptidase (GGT) is elevated in a majority of patients.

In the younger patient, an incomplete or atypical presentation is common [36]. Diagnosis is often made by echocardiogram findings of coronary artery abnormalities [37].

Pharmacologic management of the acute phase of Kawasaki's includes aspirin and intravenous immunoglobulin (IVIG). High-dose aspirin at 80 to 100 mg/kg per day dosed four times a day along with IVIG have an additive anti-inflammatory effect [32]. Length of treatment with aspirin is variable. IVIG is thought to have a generalized anti-inflammatory effect and is dosed at 2 g/kg in a single infusion. Best results are seen when IVIG is started within the first 7 to 10 days of illness.

Cardiomyopathies

Cardiomyopathies affect the heart muscle and are divided into three categories. They are hypertrophic, dilated, or congestive and restrictive (Fig. 7).

In hypertrophic cardiomyopathies, there is significant ventricular muscular hypertrophy and increased ventricular contractility but these factors limit or reduce ventricular filling.

An autosomal dominant link has been documented [38]. The left ventricle is relatively stiff and affects diastolic ventricular filling. The physical exam is notable for a sharp upstroke of the arterial pulse [39]. There can be a systolic ejection murmur or holosystolic murmur.

The ECG will show left ventricular hypertrophy, ST and T wave changes, deep Q waves, and decreased R waves. The chest radiograph may show a globular heart or cardiomegaly.

Dilated or congestive cardiomyopathies have ventricular dilatation with diminished contractility. This is the most common form of cardiomyopathies and results from infectious or toxic etiologies. They will present with evidence of congestive heart failure. A significant S3 will be found on exam.

Restrictive cardiomyopathies limit diastolic filling of the ventricles. This is the least common form and results from noncompliant ventricular walls that have been subject to an infiltrative process such as a glycogen storage disease.

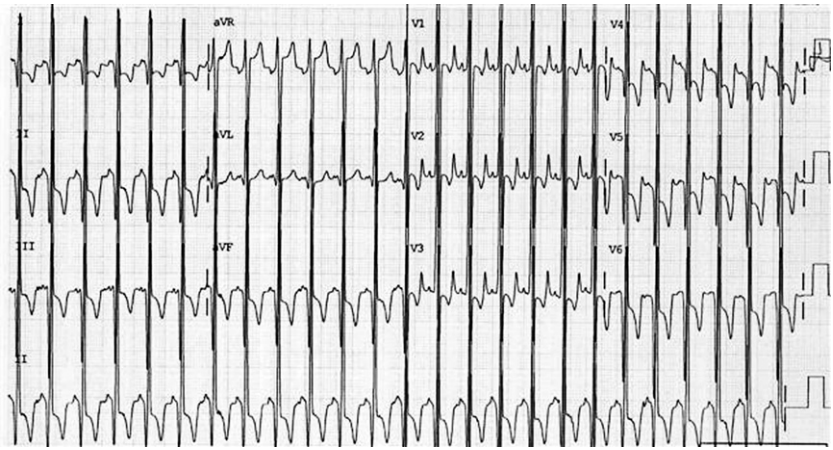

Fig. 7. ECG of hypertrophic cardiomyopathy with increased voltages throughout.

Arrhythmias

Damage, from either congenital or acquired causes, to cardiac structure will predispose the patient to arrhythmias. There can be congenital abnormalities to the conduction system, injured conduction pathways from surgery or postinflammatory changes, or irritation to the conduction system from injured myocardium. Arrhythmias have their origins in the atrial or ventricular conduction systems.

The most common arrhythmia is paroxysmal SVT [40,41]. The usual cause is idiopathic. The majority of patients with SVT have normal hearts, with 23% having congenital heart disease and 22% with Wolff-Parkinson-White (WPW) syndrome [42].

WPW is associated with congenital heart disease, such as transposition of the great arteries. WPW is a preexcitation syndrome with an accessory pathway between the atria and ventricles.

SVT is a narrow complex tachycardia with a rate ranging from 220 to 280 beats per minute in the 1-year age group. The determination of sinus tachycardia and a reentrant tachycardia must be made before the initiation of therapy. In this age group, pulse rate will linearly increase with body temperature, at a ratio of 10 beats per minute per °C increase in body temperature [43].

The ECG in SVT will show a regular rhythm with no beat-to-beat variability and a heart rate greater than 220 beats per minute in the infant. P waves can be present but are usually not. In most cases, the QRS complex is narrow. In a hemodynamically unstable SVT, immediate synchronized cardioversion with 0.5 to 1.0 J per kilogram should be done. In a hemodynamically stable SVT, vagal maneuvers can be initiated. Applying a bag of ice water to the face for 15 to 30 seconds can be used. Adenosine is the drug

of choice. Adenosine acts by temporarily blocking conduction at the AV node, thereby interrupting the reentrant circuit. Because the drug is rapidly metabolized, IV access as close to the heart is ideal, with the drug delivered via a rapid intravenous injection. Constant cardiorespiratory monitoring should be in place. Initial dosing of adenosine is 0.1 mg/kg. If there is no response, the next dose should be doubled. The maximum dosing is 0.25 to 0.35 mg/kg (Fig. 8A, B). Verapamil should not be used in the patient younger than 1 year because of the potential for hypotension and cardiovascular collapse [44].

In WPW there is a ventricular preexcitation pathway because of an accessory pathway between the atria and ventricles [3]. There is a short PR interval, a prolonged QRS duration, and delta waves (Fig. 9). Slowing the conduction through the atrioventricular node can allow another pathway to become dominant.

In a WPW-induced SVT, adenosine can cause atrial fibrillation, which can then lead to ventricular fibrillation. This underscores the need for always having resuscitation material at the bedside whenever dealing with arrhythmias.

Sick Sinus Syndrome is usually the result of cardiac surgery involving the atria or can be from myocarditis. The sinus node no longer acts as the primary pacemaker of the heart or functions at a significantly slower rate. This leads to marked sinus bradycardia, sinus arrest with a junctional escape, atrial flutter, fibrillation, or SVT.

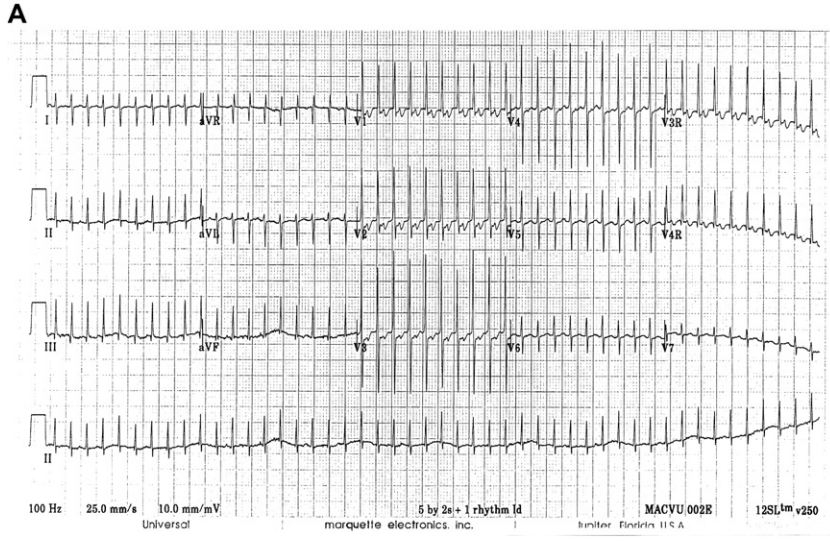

Fig. 8. (*A*) ECG of supraventricular tachycardia (SVT) in a 19-day-old. (*B*) Rhythm changes after adenosine.

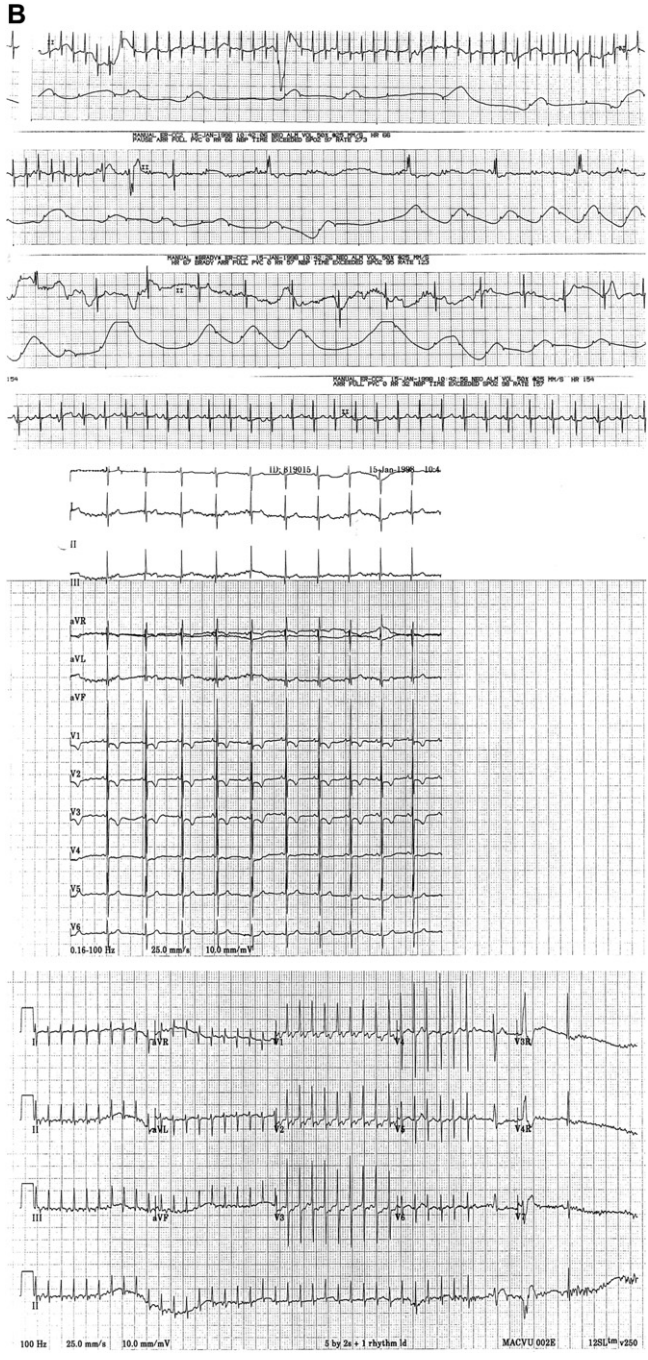

Fig. 8 (*continued*).

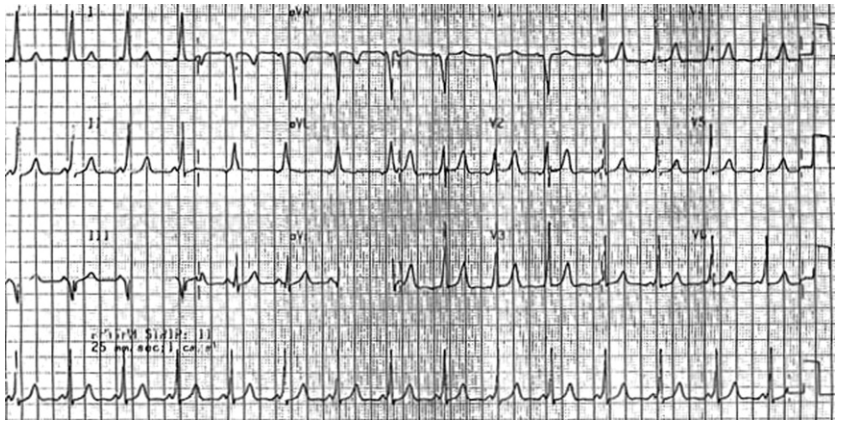

Fig. 9. ECG of WPW with delta waves.

AV block is found when there is an interruption of the conduction of the normal sinus impulse and the subsequent ventricular response. There are first-degree, second-degree, and third-degree blocks.

The first-degree block has a prolonged PR interval because of delayed conduction through the AV node. This is the result of a cardiomyopathy, congenital heart disease, postcardiac surgery, or digitalis toxicity or can be found in healthy patients.

In a second-degree block, not all of the P waves are followed by QRS complexes. The Mobitz Type I Wenckebach phenomenon has a PR interval that gets progressively longer until the QRS complex is completely dropped. The block is at the AV node level and can be attributed to myocarditis, cardiomyopathy, surgery, congenital heart disease, or digitalis toxicity. The Mobitz Type II block has similar etiologies but the block is at the Bundle of His. AV conduction is either all or none. There is potential for a complete block to develop. In two-to-one or three-to-one blocks, the block is at the level of the AV node, but can also be at the Bundle of His.

Third-degree or complete heart blocks have independent atrial and ventricular activity. There are regular P waves at a normal heart rate for age. The QRS complexes are also regular but at a slower rate than the P waves. The usual presentation in infancy is congestive heart failure. Congenital complete heart blocks have a normal QRS complex duration and can be found in patients with a structurally normal heart. A history of maternal lupus or connective tissue disease such as Sjogen's Syndrome predispose a patient to complete heart block (Fig. 10). It is thought that there is transplacental passage of autoimmune antibodies affecting the atrioventricular node [45]. Acquired complete heart blocks are the result of cardiac surgery but can also be attributed to cardiomyopathies and myocarditis and have a prolonged QRS duration.

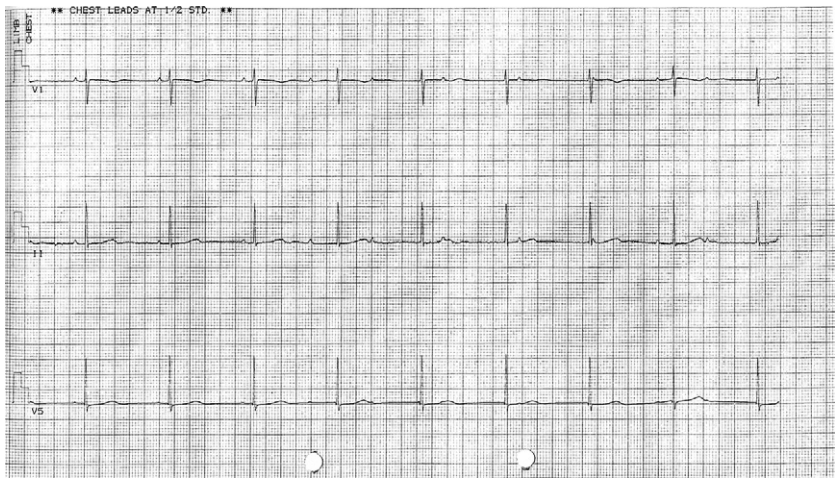

Fig. 10. ECG of complete heart block, patient's mother with lupus.

If asymptomatic, no intervention is indicated. If symptomatic, atropine, isoproterenol or temporary transvenous ventricular pacing are sometimes required.

Surgical repairs

The surgical repair of congenital heart disease continues to progress, with some lesions now repaired in the neonatal period, and most lesions repaired in the first couple of months of life. There are still patients, however, who may appear in the emergency department with no prior surgery, palliative surgery, or corrective surgery. These patients may have a less than optimal nutritional status, can be on multiple medications, or can be exhibiting post-operative complications such as a dysrhythmia or post pericardiotomy syndrome. Also a shunt could develop stenosis.

A Blalock-Taussig shunt is used in the Tetralogy of Fallot. This shunt joins the subclavian artery to the ipsilateral pulmonary artery. The modified Blalock-Taussig shunt uses a Gore-Tex shunt and requires less dissection, is not dependent on the vessel length, and has decreased shunt failure [46].

The Rastelli procedure is done in older patients, and is used in severe Tetralogy of Fallot with significant right ventricular outflow tract obstruction. There is patch closure of the VSD, with the placement of a conduit from the right ventricle to the pulmonary artery.

The Mustard and Senning operations were used in the Transposition of the Great Arteries and functioned at the atrial level. The Mustard operation was an atrial switch using prosthetic material for an intra-atrial baffle, while the Senning operation used native material for an intra-atrial baffle.

Because of atrial dysrhythmias and the inability of the right ventricle to function as a normal left ventricle in later life, these procedures were discontinued. The Arterial Switch, which has now replaced the Mustard and Senning, corrects the TGA at the great artery level. The aortic trunk is attached to the left ventricle and the pulmonic trunk is attached to the right ventricle.

The Fontan operation is done in HLHS, tricuspid atresia, and HRHS. This shunt is a cavocaval baffle to pulmonary artery anastomosis. Systemic venous return is redirected to the pulmonary artery.

The bidirectional Glenn (cavopulmonary shunt) or hemi-Fontan operation anastomoses the superior vena cava to the right pulmonary artery and is performed in patients with HLHS and HRHS. The bidirectional Glenn operation is usually done at 6 months of age, and the hemi-Fontan at 1.5 years of age.

The Norwood operation, performed in the neonatal period, is a palliative procedure in HLHS [47]. The hypoplastic aorta is reconstructed using an aortic or pulmonary artery allograft, the main pulmonary artery is divided, a Gore-Tex shunt is placed on the right to establish pulmonary blood flow, and the atrial septum is excised to provide interatrial mixing [48].

Complications that may be seen in the postoperative patient include dysrhythmias, obstruction of the surgical grafts or conduits, endocarditis, myocardial ischemia or postpericardiotomy syndrome.

Management of acute issues

Cardiac emergencies in the first couple of weeks of life will involve cyanosis and shock. The ductal-dependent lesions dominate this group and preserving ductal patency is crucial in managing these patients. While many of these patients will be diagnosed in the newborn nursery, the advent of earlier newborn discharges increases the chances that the patient will present to the emergency department for the initial diagnosis.

Cyanotic or hypoxemic episodes are seen in patients with congenital heart disease (usually Tetralogy of Fallot). They will present with hyperpnea, irritability, and increasing cyanosis along with a decreased intensity of the underlying heart murmur. A decrease in systemic vascular resistance or increased resistance to the right ventricular outflow tract increases right-to-left shunting, causing hyperpnea and, then, increased systemic venous return. This causes increased right-to-left shunting through the VSD.

To manage a ''tet spell'' the patient should be placed in a knee-chest position. Morphine sulfate (0.1 to 0.2 mg/kg subcutaneously [SC] or intramuscularly [IM]) will stop the hyperpnea. Oxygen may or may not help because the issue is to improve pulmonary blood flow. Sodium bicarbonate (1 mEq/kg IV) can treat the acidosis. Propanolol (0.01 to 0.2 mg/kg IV over 5 minutes) can be beneficial. Phenylephrine (0.02 mg/kg IV) can help to increase

systemic vascular resistance. Ketamine (1 to 3 mg/kg IV) can also increase systemic vascular resistance and provide sedation.

Tricuspid Atresia, Transposition of the Great Arteries, Total Anomalous Pulmonary Venous Return, Truncus Arteriosus, Hypoplastic Right Heart Syndrome, and Pulmonary Atresia can all present with cyanosis or shock in the first couple of weeks of life. Cyanosis or congestive heart failure will be the usual presentation of Tetralogy of Fallot. Shock will be the initial presentation for Hypoplastic Left Heart Syndrome, Aortic Stenosis, and Coarctation of the Aorta.

The key to dealing with the ductal-dependent lesions is to start intravenous prostaglandin E1 (PGE1). Decreasing pulmonary vascular resistance will help in left-to-right shunting and increasing pulmonary blood flow. The initial dose of PGE1 is 0.05 μg/kg/min. If at all possible, consultation with pediatric cardiology as well as the critical (neonatal or pediatric) care staff is beneficial. Apnea and hypotension are potential complicating side effects of PGE1 so management of the airway is essential as well as determining that the patient is not possibly septic. Additionally, the side effect of fever can cloud the potential sepsis picture. In certain variants of TAPVR, PGE1 can actually exacerbate the symptoms. Supplemental oxygen can hasten the closure of the ductus arteriosus, so this must be used with caution.

Acyanotic lesions that are dependent on ductal flow will present with cardiogenic shock.

Those lesions with critical left heart obstruction such as HLHS, aortic stenosis, and coarctation of the aorta depend on the ductus to maintain systemic perfusion. Poor perfusion, diminished pulses, and pallor are common, and the presentation can mimic sepsis. If central cyanosis is present, a response to oxygen may not take place or the patient may become worse.

Airway management is paramount, as mechanical ventilation can increase pulmonary vascular resistance [49]. Increasing right-to-left shunting over the PDA will improve systemic perfusion. Volume assists in treating the acidosis and fluid deficits. Vasopressors can be initiated if decreased ventricular function is evident.

Patients with critical right heart obstruction such as Tetralogy of Fallot and pulmonic stenosis are also ductal dependent. Airway management is a primary concern. IV prostaglandins are also key in the management, especially with oxygen saturations less than 70%. Decreasing pulmonary vascular resistance will help in left-to-right shunting and increasing pulmonary blood flow.

Congestive heart failure in the first year of life is generally associated with congenital heart disease but can also be the result of acquired disease such as myocarditis, arrhythmias, sepsis, and respiratory and metabolic diseases. Pressure overload, volume overload, decreased inotropic function, and rhythm abnormalities can all be factors in causing congestive heart failure. Cardiac congenital abnormalities that have predisposition to presenting with congestive heart failure include left ventricular outflow obstruction

(such as coarctation of the aorta and aortic stenosis) and volume overload (left-to-right shunts, VSDs, TAPVR). Endocardial cushion defects with complete involvement and AV valve insufficiency will present acutely ill in the first couple of months of life.

Difficulty feeding, tachypnea, tachycardia, cardiomegaly, hepatomegaly, and rales are all common findings. Prolonged feeding times with diaphoresis can function as a stress test for the infant. Pulmonary diseases can also present in the same fashion as cardiac disease. Supplemental oxygen may not help in differentiating between the two. Echocardiogram is much more definitive.

To treat congestive heart failure, inotropic assistance is important. Modification of preload (end diastolic volume roughly equivalent to the intravascular volume), afterload, contractility, and heart rate all play roles. Cardiac output is determined by heart rate multiplied by stroke volume. In the under 1-year-old, heart rate is the primary method of increasing cardiac output.

Airway management is important and should take precedence, as a stabilized airway and mechanical ventilation can prevent respiratory decompensation. Elevation of the head of the patient can help to decrease pulmonary blood volume. Morphine sulfate assists in treating agitation. Bicarbonate can be used in severe acidosis.

If immediate intervention is needed, dopamine and dobutamine are appropriate choices.

Dopamine is started at a continuous infusion at 5 to10 μg/kg/min. There should be a rapid response to the chronotropic effects with increases in heart rate and blood pressure and urine output. Dobutamine is also started as a continuous infusion at the same dosing. Dobutamine has less of an arrhythmic potential and chronotropic effect than dopamine and because of its vasodilatory effect, reduces afterload. Dobutamine should be used with caution in the less than 1 year of age population. Dobutamine will improve cardiac output without increasing blood pressure so if there is severe hypotension, dobutamine may be a better choice as an adjunct rather than primary agent [4,50].

Amrinone (0.5 mg/kg IV over 3 minutes) and milrinone (loading dose of 10 to 50 μg/kg IV over 10 minutes) can also be considered as potential aids in treating congestive heart failure. They do not increase the heart rate but have inotropic and vasodilator properties.

Digoxin is the inotrope of choice in the nonacute setting. Digoxin improves cardiac contractility and subsequently increases cardiac output. Care must be taken with dosing regimens. Diuretics such as furosemide promote diuresis.

Summary

The diagnosis and management of cardiac emergencies in the first year of life can be challenging and complicated. By reviewing the pathophysiology

of the heart and circulation, one can be more prepared for these difficult scenarios.

Early presentations will usually be the result of ductal-dependent lesions and will appear with cyanosis and shock. Later presentations will be the result of volume overload or pump failure and will present with signs of congestive heart failure. Acquired diseases will also present as congestive heart failure or arrhythmias.

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