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# Congenital Heart Disease in the Emergency Department

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*Children with congenital heart disease present to the emergency department (ED) at many stages of life. Some children present to the ED with the initial manifestations of their as-yet undiagnosed diseases, while others present to the ED with complications of their disorders or their therapies. This article reviews the pathophysiology involved in the newborn with congenital heart disease and familiarizes the reader with the nomenclature and known complications encountered in the care of these children.*

*—The Editor*

## Physiology of Cyanosis

Cyanosis is the visual recognition of discoloration due to the presence of reduced, or deoxygenated, hemoglobin. Cyanosis is classified as either central or peripheral. Central cyanosis is manifested by the presence of reduced hemoglobin in the capillaries and possibly arterioles and venules. Three to 5 gm/dL of dissociated hemoglobin are necessary before central cyanosis becomes visible.[1-3] Central cyanosis classically is visible in the lips and mucous membranes. In contrast to peripheral cyanosis, central cyanosis always is a manifestation of hypoxia.[4]

The level of hypoxia at which cyanosis becomes visible varies because it depends upon the hematocrit. The degree of hypoxemia required to cause cyanosis is related inversely to the hemoglobin and hematocrit. For example, a newborn with a hemoglobin of 20 gm/dL will be clinically cyanotic when 3 gm/dL of hemoglobin are reduced, or at an oxygen saturation of 85%. A child with a hemoglobin of 10 gm/dL will become visibly cyanotic when the oxygen saturation is 70%, when 3 gm/dL of the 10 gm/dL of hemoglobin becomes reduced.[2,4]

Peripheral cyanosis, by comparison, is a manifestation of poor peripheral perfusion. It is thought to be due to a relative decrease in the flow of oxygenated blood to the periphery with an increased oxygen extraction. Often, the lips and mucous membranes are spared. This inadequate perfusion does not have to occur in the presence of hypoxia. Clinically, often the child

with undiagnosed cyanotic cardiac disease will be hypoxic and will demonstrate both central and peripheral cyanosis upon initial presentation. Therefore, it will be difficult to distinguish central from peripheral cyanosis in the acutely ill child. This distinction, however, is more relevant in the clinically stable child. The stable child becomes symptomatic when oxygen delivery cannot meet oxygen demand.

## Initial Presentation of Critical Congenital Heart Disease

When faced with the hypoxic or cyanotic child, it is important for the emergency physician to consider the possibility of an undiagnosed form of congenital heart disease.[5] Children with congenital heart disease may be asymptomatic in the newborn nursery, yet may present to the ED as symptoms develop due to maturing physiology.[6] Newborn cardiovascular physiology changes through the first week of life, and possibly longer. Sometime early in the newborn period, the ductus arteriosis closes. This structure minimizes intrauterine pulmonary blood flow. It is unclear what acts as the physiologic signal to close. An asymptomatic child may become symptomatic when the ductus closes. The lesions that most commonly present at this time include coarctation of the aorta and hypoplastic left heart syndrome (HLHS). In addition, intrauterine pulmonary vascular resistance is relatively high. This resistance drops during the first few days after birth. As this resistance drops, right ventricular pressures drop, generating a pressure gradient within the heart that may alter intracardiac blood flow. The classic demonstration of this is how a new murmur develops outside the newborn period in the case of the child with a small ventricular septal defect.

Children symptomatic from their heart disease present to the ED with inadequate tissue oxygen delivery or congestive heart failure (CHF). As described, this inadequate oxygen delivery may be due to inadequate perfusion with oxygenated blood, or inadequate oxygenation of the blood delivered to the periphery, or both. The parents' reports of symptoms often are the same for both groups of children. Both will have feeding difficulty, tachypnea, and fussiness or agitation that may progress to lethargy. Physical examination often reveals a murmur, tachypnea, diaphoresis, and hepatomegaly. Depending on the particular cardiac lesion, there may be a difference in blood pressure between the right arm and the legs, or a difference in $PaO_2$ of preductal extremities (right arm) and postductal extremities (legs and left arm). The chest x-ray (CXR) is useful to determine degree of pulmonary vascularity and heart size.[7,8] An electrocardiogram (ECG) can exclude an arrhythmia as a cause for peripheral cyanosis and may suggest the cardiac etiology of the cyanosis.[7]

Once the clinician is suspicious a child may have heart disease, the next challenge is to classify the disease. Children with congenital heart disease present with a mixing lesion or reduced cardiac output from an obstruction, or some combination of the two. Transposition of the great vessels is the only lesion that does not fit smoothly into either of these categories. Despite this, use of these categories helps to simplify the presenting syndromes. Mixing lesions are either left-to-right (oxygenated blood mixes with deoxygenated blood and exits the heart into the pulmonary system; e.g., atrial septal defect), or right-to-left (deoxygenated blood mixes with oxygenated blood and exits the heart into the systemic circulation; e.g., tetralogy of Fallot). Left-to-right lesions can present with pulmonary edema, right heart failure, inadequate peripheral perfusion, and peripheral cyanosis. An example is a child with a VSD in pulmonary edema. Right-to-left lesions may present with central cyanosis and right heart failure, and may progress to central and peripheral cyanosis, as seen in children having a "tet-spell," or hypercyanotic episode. Symptomatic obstructive lesions can present due to aortic stenosis, coarctation, HLHS, or pulmonary atresia.[5,9]

Further classification beyond these broad categories will be difficult in the ED without the benefit of echocardiography. In addition, it may be difficult to determine whether a child's

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**Table 1. Glossary**

| | | |
|---|---|---|
| ASD | — | Atrial septal defect |
| AV canal | — | Atrioventricular septal defect |
| B-T shunt | — | Blalock-Taussig shunt (subclavian artery to branch pulmonary artery shunt) |
| Central shunt | — | Anastomosis from ascending aorta to main pulmonary artery |
| CHD | — | Congenital heart disease |
| DORV | — | Double outlet right ventricle |
| HLHS | — | Hypoplastic left heart syndrome |
| PDA | — | Patent ductus arteriosis |
| PGE1 | — | Prostaglandin E1 |
| Potts anastomosis | — | Shunt from descending aorta to left pulmonary artery |
| TGA | — | Transposition of the great arteries/ vessels |
| VSD | — | Ventricular septal defect |
| Waterston shunt | — | Anastomosis from ascending aorta to main or right pulmonary artery |

cyanosis is indeed a primary central or peripheral cyanosis. Fortunately, diagnostic and management principles remain similar within and across each category. Because initial management principles are the same for any suspected heart disease, the most difficult decision is how aggressively to consider congenital heart disease in the sick infant. In the hypoperfused newborn with hepatomegaly and normal or decreased pulmonary vascularity, the clinician quickly will consider cardiac causes. However, in the case of the acyanotic child with pulmonary congestion, the diagnosis may be more challenging.

## Hypoxia

When faced with a hypoxic newborn, the clinical difficulty is to determine the etiology of the hypoxia. Hypoxia has either a pulmonary or primary cardiac etiology—the difficulty is to decide the primary cause. If there are no respiratory symptoms and the CXR demonstrates a paucity of pulmonary vascularity, as may be seen with pulmonary stenosis and a closed ductus arteriosis, the etiology likely is cardiac. If there are respiratory symptoms or the CXR is consistent with pulmonary edema or diffuse, multilobar pneumonia (as may be seen with group B streptococcal pneumonia), then the decision is more difficult.[5] Is this child hypoxic from pneumonia or did the child's ductus arteriosis close, and now the child no longer is perfusing the periphery while developing pulmonary edema from a sudden increase in pulmonary blood flow (e.g., aortic stenosis or HLHS)?

Classically, the hyper-oxygenation test is used to separate pulmonary causes from cardiac causes of hypoxia in the newborn. A child has an arterial blood gas drawn while he or she is breathing 100% oxygen. If severe pneumonia is present, oxygenation is a diffusion problem, so hyper-oxygenation can elevate arterial oxygenation significantly. Conversely, in the case of congenital heart disease where arterial and venous mixing occurs, oxygenation is limited by the degree of mixing and is not altered significantly by hyper-oxygenation. Unfortunately, when evaluating the arterial oxygenation after hyper-oxygenation, there is no "magic number" to determine whether the child has pulmonary or cardiac disease. However, a child presenting with central cyanosis from congenital heart disease should not be able to achieve a $PaO_2$ of 150 mmHg.[3]

## Treatment for Suspected Congenital Heart Disease in the Newborn in Shock

Initial treatment of suspected undiagnosed congenital heart disease in the child in shock is the same for all lesions. Children who present acutely ill typically present due to a "tet spell" or the closing of the ductus arteriosis. Children with tetralogy of Fallot often have a murmur in the nursery, so they infrequently present to the ED for initial diagnosis. Other lesions that more commonly present to the ED include HLHS, coarctation of the aorta, and transposition of the great vessels. These lesions are called "ductal-dependent lesions" because they become symptomatic when the ductus arteriosis closes. Therefore, ductal-dependent lesions are best treated with prostaglandin E1 (PGE1) infusion. Prostaglandin infusion typically is started at 0.05 mcg/kg/min and increased until clinical improvement occurs. Common complications include apnea, hyperthermia, hypotension, and irritability.[10,11] Complication rates of prostaglandin infusion are unknown.

The apnea associated with PGE1 use does not occur with predictable timing, although it is more common at higher doses. Therefore, many centers prefer to electively intubate infants prior to transfer, after a prostaglandin infusion has been started.

## Anatomy of Congenital Heart Disease

Following are descriptions of some of the more complex congenital heart lesions, their presentations, and initial management.

**Ventricular Septal Defect.** Ventricular septal defect (VSD) represents 20% of all congenital cardiac lesions. There are four types: 1) perimembranous, the most common, occurs under the aortic valve; 2) subpulmonic; 3) atrioventricular (AV) canal, an endocardial cushion defect that never closes spontaneously; and 4) muscular, which has the highest spontaneous closure rate.

Children with VSDs usually present outside the newborn period, most commonly between 2 and 6 weeks of age. Children often are asymptomatic without a murmur in the nursery until pulmonary vascular resistance decreases. This decrease in pulmonary vascular resistance is a normal physiologic event and occurs during the first weeks of life. As this drop occurs, right ventricular pressure decreases, allowing left-to-right shunting across a ventricular septal defect. Children will present with symptoms of increased pulmonary blood flow: pulmonary edema, dyspnea (especially while eating), sweating with feeding, poor weight gain, and hepatomegaly.

VSD presentation depends on the size of the defect. Defects are determined by the Qp (pulmonary flow):Qs (systemic flow) ratio. A VSD that is small has a Qp:Qs of less than 1.5, one that is moderate $1.5 < Qp:Qs < 2.0$, and a severe VSD may have a

ratio greater than 2.0. Small ratios have only slightly elevated right ventricular pressures. A small VSD may have a holosystolic murmurs. For larger defects, there is an increased right ventricular impulse. The point of maximal impulse (PMI) is shifted laterally to the left. If there is a large shunt, there can be a mid-diastolic mitral flow murmur that creates a "gallop rhythm." The character of the murmur depends upon the jet across the VSD. A small VSD with normal pulmonary vasculature will result in a high pressure jet across the VSD and, hence, a loud murmur. A large VSD or high pulmonary pressures will result in a small pressure drop across the VSD and, hence, a soft murmur.

The larger the defect, the greater the initial left-to-right flow. In moderate defects with left-to-right shunts, the left ventricle and atrium are overloaded, leading to dilation and failure. Larger defects initially have a left-to-right shunt. However, flow may progress to the Eisenmeger complex, or right-to-left shunting, secondary to a high pulmonary resistance that develops in response to the chronically elevated right-sided pressures. The presence of this complex predisposes patients to pulmonary hemorrhage, cerebral abscess, and cerebral thrombosis.

Ancillary tests are not particularly helpful in the initial ED presentation, except to establish the presence of pulmonary edema on chest x-ray. Studies usually are normal in small defects. Larger defects show enlarged cardiac silhouette on chest x-ray (CXR) and evidence of ventricular hypertrophy on ECG.

**Patent Ductus Arteriosis.** The patent ductus arteriosis (PDA) is the distal portion of the sixth aortic arch.[12-14] It closes within days of delivery. The incidence is greater in premature births—8/1000 vs. 1/1000 in pre-term and full-term infants, respectively.

The continuous murmur of a PDA is heard best at the left infraclavicular border. The murmur begins at or just after the first heart sound, continues through systole, and wanes late in diastole. Rarely, infants may present with CHF due to a large PDA. This pulmonary edema occurs when children have a high flow shunt across the ductus. However, children present asymptomatic with a murmur.

The ECG is of limited value. Abnormalities include: left or biventricular hypertrophy and right axis deviation. The CXR rarely shows cardiomegaly except in the aforementioned large defects.

**Tetralogy of Fallot.** The anatomy of tetralogy of Fallot includes: 1) obstruction to right ventricular outflow (subvalvular pulmonic stenosis); 2) right ventricular hypertrophy; 3) ventricular septal defect; and 4) dextroposition or overriding of the aorta.[12,15,16] Pulmonary artery anatomy is variable in children with tetralogy of Fallot.[13] In cases of severe right ventricular outflow tract obstruction, pulmonary atresia may result with variable forms of hypoplastic branch and distal pulmonary arteries. The pulmonary defect most commonly is a hypoplastic valve annulus with marked pulmonary stenosis. These patients have a dominant right-to-left shunt. Blood flow is normal until reaching the right ventricle. The underlying pulmonary stenosis shunts the blood to the aorta via a VSD. With persistent shunting, the child has deoxygenated arterial blood and subsequent cyanosis. The extent of the stenosis dictates the level of cyanosis. Patients with moderate pulmonary outflow obstruction can be "pink," as there is enough pulmonary blood flow to oxygenate hemoglobin.

Cyanosis is the most obvious presentation of tetralogy. While this is common at 1 year of age, newborns can have limited right ventricular obstruction, decreasing the right-to-left shunt.[12,14] Over time, the neonate's pulmonary stenosis becomes more severe, limiting pulmonary blood flow.

Children with tetralogy of Fallot have stereotypical symptoms. Parents often will notice their children are dyspneic on exertion or crying. Although children with tetralogy of Fallot usually are operated on by 1 year of age, there is some historical information about children who have not had surgery. Infants and toddlers will play and then sit or lie down.[18] Older children assume the classical squatting position. Within minutes, the infant or child with dyspnea will resolve and he/she will resume physical activity.

The cardiac physical exam shows a loud and harsh systolic murmur heard throughout the precordium, loudest at the left sternal border. Often in neonates the murmur is subtle or non-existent. The murmur is due to turbulence over the right ventricular outflow tract and is less prominent with large right-to-left shunts. In the cases of pulmonary atresia, a continuous diastolic murmur is noted.[12] The first heart sound is normal; however, the second is single and loud. Older patients with prolonged hypoxia are likely to have clubbing.

ECG will show right ventricular hypertrophy (RVH) and right atrial hypertrophy (RAH), except in the infant where these findings are not pathologic. On echocardiogram, the right ventricular obstruction, overriding aorta, and VSD are visible. The chest radiographs in older children with tetralogy of Fallot demonstrate a boot-shaped heart. This may not be seen in infants because of the presence of the thymus. Pulmonary blood flow in infants and children typically is reduced. However, normal or increased pulmonary blood flow may be evident, depending upon the degree of right outflow tract obstruction. A right-sided aortic arch is evident in 5% of children with tetralogy of Fallot. Kazim, in a pair of articles, notes an 11% risk of tracheal anomalies in children with tetralogy of Fallot.[19,20] Difficulties with intubation were noted in three of 44 patients in one series.

In the first two years of life, "tet" spells (paroxysmal hypercyanotic attacks) may occur. The episodes may last hours. These spells are more frequent in the summer and with infections.[13] The child becomes initially hyperpneic and restless. The subsequent hypoxia causes peripheral vasodilation, resulting in an increase in pulmonary vascular resistance and a subsequent decrease in systemic vascular resistance, which increases the right-to-left shunt. This creates a vicious cycle where the child gets more agitated, leading to a continuous decrease of oxygenated blood. Severe cases can progress to syncope, permanent cognitive impairment, and death.

Children who present with a "tet" spell should be placed in a knee-chest position and given supplemental oxygen. Attempting to calm the child may abort a mild or early attack. If these therapies fail, subcutaneous morphine (maximum dose 0.1 mg/kg) can be administered. Phenylephrine and intravenous volume infusion may improve symptoms. Metabolic acidosis needs to be watched

carefully, as it causes increased pulmonary vascular resistance and worsening of the existing right-to-left shunt.

Children with tetralogy of Fallot may present in shock because of a tet spell, pulmonary atresia, or shunting due to the VSD. The presentation may worsen after the closing of a patent ductus arteriosis. In the case of the cyanotic neonate with suspected tetralogy, PGE1 at a rate of 0.05 mcg/kg/min is given. Prompt transfer to a center that either has experienced echocardiographers or the ability to perform catheterization is crucial.

**Transposition of the Great Vessels.** In transposition of the great vessels, the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. Venous return to the heart is normal. This anatomy causes systemic deoxygenated blood to flow from right ventricle to the aorta, while oxygenated pulmonary venous blood is returned to the lungs.[21,22] This creates a parallel system that is dependent on a patent foramen ovale and patent ductus arteriosis for some form of mixing. In nearly all patients, anatomic left atrial pressure (physiologic right atrial pressure) is higher than the right, allowing for some oxygenated blood to enter the systemic circulation.

It is rare for a patient to present to the ED with transposition and an intact ventricular septum, as these patients are cyanotic upon closure of the PDA. These children typically become symptomatic within the first hours of life, but occasionally not for the first days of life. In those patients with other congenital cardiac defects, (i.e., VSD or a continued PDA) the presentation of cyanosis may be delayed for weeks.[12,14,21] A patent ductus allows for mixing of the deoxygenated blood to flow into the pulmonary circuit. In those with a VSD, a physiologic right-to-left shunt exists, allowing for oxygenation of blood. In the neonate, a PDA initially can be beneficial by allowing for mixing. However, CHF occurs similar to a patient with a VSD, as there is increased flow from the aorta to pulmonary arteries that cannot withstand the increased pressures. The combination of transposition, VSD, and pulmonary stenosis can mimic tetralogy of Fallot. Therefore, these patients are more likely to have a delayed cyanotic presentation, and usually have more severe hypercyanotic spells. (For further discussion see the description of tetralogy of Fallot.)

On physical exam, TGA patients have a loud single S2 secondary to the anterior aortic valve and a posterior pulmonary valve. In most patients with TGA and an intact ventricular septum there is no murmur. Although there is increased right ventricular pressure, hepatomegaly is rare. ECGs provide limited information. The QRS is rightward; however, all neonates initially have this. In those patients with a VSD, the rightward forces may remain into childhood, at which time they would indicate pathology.

As in other newborns with severe cyanosis suspicious for cardiac origin, a prostaglandin infusion should keep a PDA open or re-open a recently closed ductus arteriosis, which will re-establish arterial mixing. This will temporize the child's condition until transfer to a tertiary center.

**Hypoplastic Left Heart Syndrome.** This describes neonates with either hypoplasia or absence of the left ventricle.[23] Up to 84% have concomitant aortic and mitral valve atresia. The incidence is rare, occurring in 0.036% of live births, and with a slight male predominance.[24]

Initially, patients with hypoplastic left ventricles can appear asymptomatic. Pulmonary venous blood flow passes into the right atrium via a left-to-right shunt (ASD). This blood mixes with deoxygenated caval blood and enters the pulmonary circulation where it either returns to the left atrium or enters the systemic circulation via the PDA. In neonates, the presentation and severity of hypoplastic left ventricle are dependent on a patent ductus arteriosis.[12,23,24]

Neonates with HLHS present critically ill to the ED. Although the majority of neonates are diagnosed in utero or within 24 hours of delivery, those with delayed closure of the ductus can present several days to weeks post delivery.[12,13] Symptoms are those of poor perfusion, including: CHF, cyanosis, hepatomegaly, and either weak or absent pulses. Often there is a pan-systolic murmur.

The emergency physician should start PGE1 on all patients with suspected HLHS. If time allows, an echocardiogram can be performed to confirm the diagnosis; however, there should not be a delay in administering PGE1. Oxygen causes vasoconstriction of the ductus and pulmonary arteries; therefore, inspired oxygen concentration should be decreased as tolerated once the ductus arteriosis is open. Prompt transfer to a center that is capable of either the staged reconstruction procedure or transplantation is indicated.

**Coarctation of the Aorta.** Constriction of the aorta can occur at any point along its length. However, in 95% of patients it exists distal to the subclavian artery at the ductus arteriousus.[12,14,25] Males are two times more likely to have coarctation than females. There is a 50% incidence of bicuspid aortic valve (and may be as high as 90%), as well as mitral valve involvement.[12,14]

Classically, coarctation presents with pulse differences between the patient's arms and legs. The femoral, posterior tibial, and dorsalis pedis are markedly diminished. Often the child has upper extremity hypertension. There may be a pulse oximetry differential between pre-ductal and post-ductal extremities. In the neonate, where the most severe cases present, the child will be tachypneic, cyanotic, and possibly in CHF. These symptoms occur in response to the closing of the ductus, which reveals the extent of the obstruction.

Once the child is several months old, the presentation is less dramatic. Hypertension usually is the first finding. It is crucial to measure lower extremity blood pressure to note if there is a decrease in pressure in lower extremities. In normal children, the blood pressure taken by cuff in the legs is equal to or greater than in the arms. If the left arm pressure is lower than the right, the coarctation is likely above the origin of the left subclavian. If a murmur is present, it is heard over the left infraclavicular area and between the scapula. A continuous murmur indicates the child likely has collateral flow. On CXR, an enlarged aortic knob may be noted. Rib notching is a late radiographic finding. The ECG may show signs of LVH with or without a strain pattern.

If pediatric cardiology is available, a two-dimensional echocardiogram can confirm the diagnosis. It is important for the

emergency physician to be suspicious of coarctation when the systolic blood pressure of an infant is greater than 140 mmHg.[26] In older children, consider coarctation when systolic hypertension is noted.

In the neonate who presents critically ill, reperfusion of distal tissue and correction of metabolic acidosis is necessary. PGE1 is given to keep the ductus open.[12,13]

**Tricuspid Atresia and Pulmonary Atresia.** Tricuspid atresia and pulmonary atresia are two lesions that present with decreased pulmonary blood flow. Right ventricular size and pulmonary artery diameter is dependent upon in-utero pulmonary blood flow. Therefore, patients with tricuspid atresia often have decreases in right ventricular size and pulmonary artery diameter. Tricuspid stenosis or atresia causes obstruction to flow at the level of the right atrium. A patent foramen ovale, patent ductus arteriosis, or a ventricular septal defect is necessary for pulmonary blood flow.[12,13] The patent foramen ovale allows a right-to-left shunt that bypasses the atretic area. In patients without a VSD, the right ventricle is hypoplastic and the pulmonary valve is atretic. Within hours of birth the patient will be cyanotic from decreased pulmonary blood flow. Physical examination may reveal hepatomegaly and hypoxia unresponsive to oxygen. CXR usually reveals decreased pulmonary blood flow and occasionally reveals a right aortic arch.

Prostaglandin infusion is necessary to open the ductus in children who suddenly develop cyanosis and shock.

**Anomalous Left Coronary Artery**. Normally, the left coronary artery exists immediately after the aortic valve. An anomalous coronary artery can originate from several anatomic locations. In the most common form of this congenital lesion, the left coronary artery arises from the pulmonary artery. After birth, pulmonary artery pressure decreases, and perfusion of the myocardium is inadequate. Infants may present with intermittent pain, classically occurring after the exertion of feeding. Other symptoms include failure to thrive, diaphoresis, or dyspnea.[27] This form of anomalous left coronary artery is a common cause of myocardial infarction in the infant.[28] If not symptomatic as an infant, these patients often die from sudden death in young adulthood. In those cases, the ECG often reveals ST segment elevation and inverted T waves in the lateral leads.

There are rare cases of right-to-left coronary artery anastomosis when the left coronary artery arises from the pulmonary artery. This leads to heart failure due to the increased pressure of the left ventricle that causes blood flow from the right coronary to the left and into the pulmonary artery. In another variant of anomalous left coronary artery syndrome, the left coronary artery courses between the pulmonary artery and aorta. As young adults, sudden death occurs because exercise increases cardiac output, which results in arterial dilation causing compression of the anomalous left coronary artery and resultant ischemia.

**Double Outlet Right Ventricle.** Double outlet right ventricle (DORV) is a rare lesion that exists when the aorta and pulmonary artery arise from the right ventricle. Systemic perfusion is a combination of pulmonary and systemic venous blood, since both drain into the right ventricle. Severity and presentation depend upon the anatomic relationships of the great vessels and the ventricular septal defect. In these complicated lesions, presentation may mimic that of transposition of the great vessels, ventricular septal defect, or tetralogy of Fallot. The presentation is usually in the newborn period, but may be delayed.

## Surgical Procedures

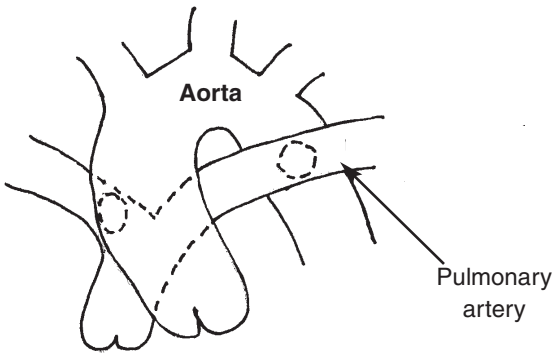
Following are descriptions of some of the more common surgical procedures performed in children with congenital heart disease, and their common complications.

**Atrial Septal Defects (ASD).** Surgical closure of an atrial septal defect involves either patching or primarily suturing the ASD. Complications may arise if flow from the vena cava or pulmonary vein is obstructed by the repair, or if the SA node is injured during repair. Venous obstruction may lead to either hepatic congestion or pulmonary edema. The most common delayed complications are atrial arrhythmias, either atrial fibrillation or flutter. Infrequently, bradycardia has been reported. These arrhythmias are more common in older patients who have atrial septal defect repairs.[29,30]

**Ventricular Septal Defects (VSD).** Many small VSDs will close spontaneously. Those that are unlikely to close spontaneously may be repaired, as they fail medical therapy. The goals of medical therapy are to control CHF, allowing the child to grow to facilitate the surgical repair. Indications for surgery include uncontrolled CHF, poor weight gain in infancy, or developing pulmonary hypertension. The type of surgical repair depends upon the type and size of the VSD. Some defects can be accessed through the tricuspid valve, some through the pulmonary artery, some through the aortic valve, and some require right ventriculotomy. Surgical risks include residual VSD, resultant aortic insufficiency, and electrocardiographic abnormalities. Residual complete atrioventricular block is uncommon after repair.[31] The most common residual electrocardiographic abnormality usually is a right bundle-branch block. Long-term risk of arrhythmia is unknown; however, no sudden deaths occurred after corrected VSD in the series of studies on sudden death in post-operative survivors of congenital heart disease.[32] However, in the series of patients with residual pulmonary hypertension, cases of sudden death have occurred.[33] Therefore, although the long-term risk of arrhythmia is real, the true incidence is not known. Pre-existing pulmonary hypertension may cause late complications, but, otherwise, these patients do very well with only mild exercise intolerance. In fact, patients with no residual defect post-operatively do not require endocarditis prophylaxis after six months. Most survivors of VSD repair are asymptomatic.

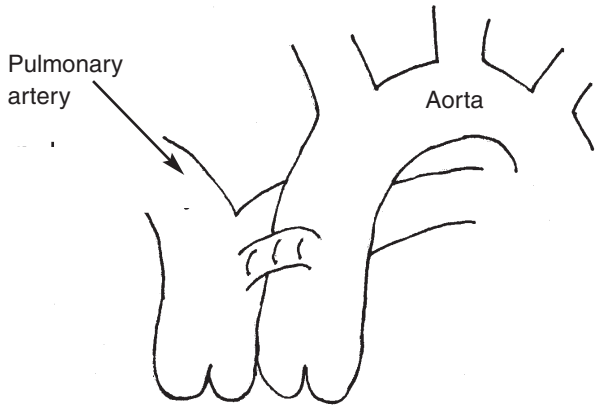
**Atrioventricular Septal Defects.** Atrioventricular septal defects (AV-canal) have a varied degree of clinical severity. Patients can range from almost asymptomatic to severe CHF. Clinical presentation depends upon the size of the defects and the amount of regurgitation from the AV-valves. Additionally, the timing of surgery, the type of surgical repair and the risks of surgery are related to the severity of the initial anatomy, especially if there is pulmonary hypertension from left-to-right shunting, pulmonary outflow tract obstruction (tetralogy of Fallot physiolo-

**Figure 1. Waterston and Potts Shunts**




Examples of the Waterston shunt (ascending aorta to right pulmonary artery) and the Potts shunt (descending aorta to left pulmonary artery)

**Figure 2. Central Shunt**




Central shunt (ascending aorta to main pulmonary artery)

gy), or complex congenital heart disease. Surgical techniques continue to improve; therefore, there is little information on the long-term complications of patients undergoing surgical repair. Atrial arrhythmias or heart failure can occur post-operatively if the patient has a residual ventricular septal defect, pulmonary hypertension, or residual AV valve regurgitation. Heart block requiring a pacemaker can occur post-operatively. Artificial AV-valves may be necessary if functional valves cannot be fashioned surgically.

## Shunt Procedures

Temporizing shunt procedures are performed if the lesion results in cyanosis due to inadequate pulmonary blood flow. These temporizing procedures allow delay of definitive correction until the child is larger, which makes the final procedure easier to perform.The three main types of aorta to pulmonary artery shunts are Potts, Waterston, and central shunts. The Potts anastomosis is between the descending aorta and the left pulmonary artery. (*See Figure 1.*) The Waterston procedure is an anastomosis between the ascending aorta and the right or main pulmonary artery. (*See Figure 1.*) The central shunt uses a graft from the ascending aorta to the pulmonary artery. (*See Figure 2.*)

The most common shunt is a modified Blalock-Taussig (B-T) shunt or an anastomosis between a subclavian artery and an ipsilateral pulmonary artery. In the classical B-T shunt, the subclavian artery is sacrificed for this procedure. Currently, the modified B-T shunt is performed by using a Gore-Tex conduit between the subclavian and pulmonary artery so that the subclavian artery is spared. (*See Figure 3.*) This shunt may be performed in cases of pulmonary atresia, tricuspid atresia, or tetralogy of Fallot.[34] Other shunts commonly used are aorta to pulmonary artery shunts.

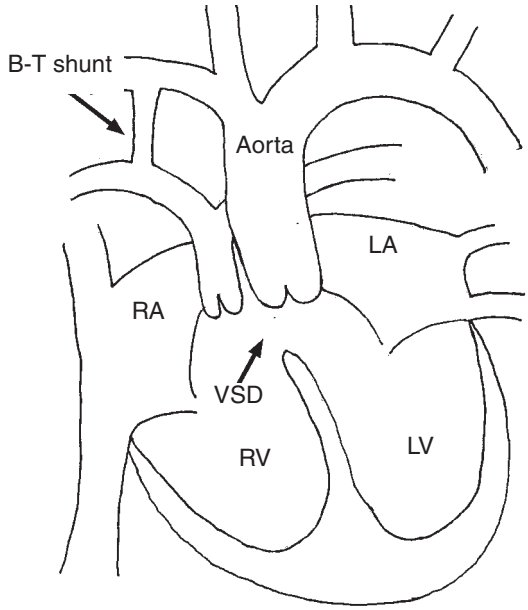
Complications of B-T shunts can include injury to the phrenic or recurrent laryngeal nerves or injury to the sympathetic chain with resultant Horner syndrome. Currently, the ipsilateral vertebral artery is ligated during the modified B-T shunt to prevent a basilar steal phenomenon.[35] Complications also can arise from the shunt size. If the shunt is too small, the child may have residual cyanosis from inadequate pulmonary blood flow. If the shunt is too large, pulmonary edema can result. Excessive flow complications after B-T shunts usually are limited by the size of the subclavian artery. Excess pulmonary blood flow is the most common complication after a modified B-T shunt, occurring in 28% of patients in one series.[34] As with all procedures, stenosis at the anastomosis may occur.[36] Additionally, a thrombosis may develop in shunts formed with grafts. The shunt may clot when a child gets dehydrated as a complication of a viral illness, such as respiratory syncytial virus (RSV) or rotavirus. An uncommon complication recently noted is an aneurysm of the shunt that was diagnosed as an infiltrate on CXR.[37]

Complications of Potts, Waterston, and central shunts usually depend on the relative size of the shunt. A shunt that is too small may not relieve cyanosis, while one that is too large may produce increased pulmonary blood flow, which may lead to pulmonary hypertension.[38,39] These shunts rarely are used today because of these complications.

## Pulmonary Artery Banding

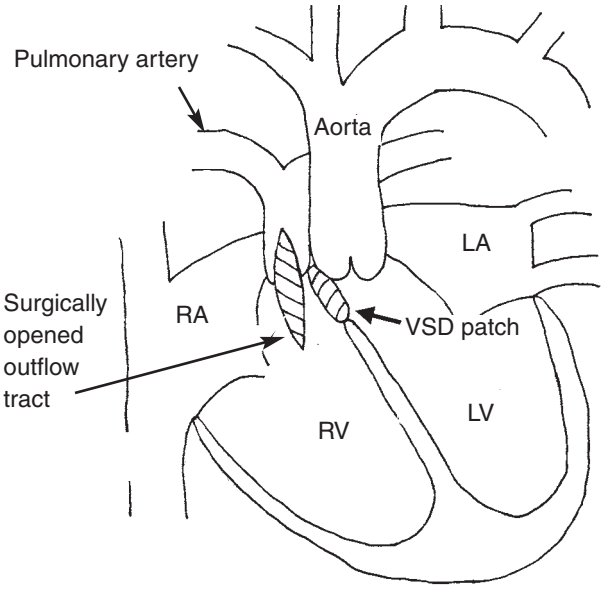
Pulmonary artery hypertension may develop within the first years of life for children with increased pulmonary blood flow from a left-to-right shunt. Large left-to-right shunts can occur in children with ASD, VSD, or an atrioventricular-canal defect. If a complicated definitive repair is anticipated, a temporizing pulmonary artery band may be placed. This band is, essentially, a noose that fits around the pulmonary artery to decrease the diameter. The goal of this treatment is to decrease pulmonary blood flow by increasing the pulmonary outflow tract resistance, which delays the development of pulmonary hypertension that may worsen outcome after definitive repair. This procedure is not performed as often as in the past for two reasons. First, prior surgi-

**Figure 3. Blalock-Taussig Shunt**




This illustration shows a Blalock-Taussig shunt (allowing flow from right pulmonary artery to right subclavian artery) in a patient with tetralogy of Fallot.

**Figure 4. Corrected Tetralogy of Fallot**




This illustration shows a corrected tetralogy of Fallot with a VSD patch and a right ventricular outflow tract patch to increase pulmonary artery flow.

cal techniques could lead to sub-aortic stenosis. Second, improved technique has allowed for definitive surgery in smaller children, eliminating the need for palliative procedures.

## Tetralogy of Fallot

Initial definitive repair of tetralogy of Fallot is preferred over an initial palliative procedure with delayed definitive repair. However, in some patients, a palliative shunt procedure (i.e., B-T shunt) may be needed before this definitive repair. The surgical repair of tetralogy of Fallot essentially involves decreasing pulmonary outflow tract obstruction and patching the VSD. Subtle or gross variations in anatomy can affect surgical outcome of this repair. The largest factor that determines surgical options is the size of the native pulmonary arteries. If they are atretic with a large subvalvular stenosis, a ventriculotomy and Gore-Tex outflow conduit will be necessary. A transatrial VSD repair may be performed in cases of minimal pulmonary stenosis with a full-size main pulmonary artery. This is a less risky procedure than a formal ventriculotomy. (*See Figure 4.*)

CHF may develop postoperatively in cases where there is a residual intracardiac shunt, a significant systemic to pulmonary collateral artery, or a residual systemic to pulmonary palliative shunt. Right ventricular dysfunction can occur from persistent pulmonary outflow obstruction, pulmonary hypertension from a palliative procedure, pulmonary regurgitation through a conduit or dysfunctional pulmonary valve, or volume overload from a residual VSD. This dysfunction usually is a postoperative phenomenon. No data exists on long term complications from this increased right ventricular work. Conduction abnormalities and arrhythmias may occur postoperatively.[40] The bundle of His may be injured during repair, leaving a residual right bundle-branch block, bifascicular block, or complete AV block. A pacemaker is necessary in the case of complete AV block.[31] Patients with persistent premature ventricular contractions should have a cardiology evaluation, especially if these are multifocal or provoked by exertion. These patients are at increased risk for sudden death from arrhythmias.[32]

## Coarctation Repair

The three main surgical repairs of coarctation are: resection of coarctation shelf with end-to-end anastomosis of the descending aorta; subclavian flap aortoplasty; and synthetic patch aortoplasty. All three have a significant risk of restenosis. Restenosis may occur at the suture line, as with the end-to-end anastomosis, or along the length of the patch, as with the subclavian flap. The resection with end-to-end anastomosis may have a restenosis rate of up to 60%.[41] Survivors of coarctation repair also are at an increased risk of sudden death from arrhythmias.[32] The ECG of survivors may have a persistent pattern of LVH, even without increased left ventricular pressure.

## Arterial Switch

Arterial switch repair is the procedure most commonly used for transposition of the great vessels. After some initial poor results, surgical technique has improved to make this the procedure of choice in children with uncomplicated transposition. At birth, the aorta and coronary arteries arise from the anatomic right ventricle, and the pulmonary artery arises from the anatomic left ventricle. In the arterial switch repair, the aorta and coronaries are switched to the left ventricle, and the pulmonary artery is switched to the right ventricle. Long-term complication rates

after this procedure are unknown. Long-term complications include sudden death, branch pulmonary artery stenosis, left ventricular failure, and caval vein thrombosis.[42] The most difficult portion of the procedure is moving the coronary arteries during the surgery. Myocardial infarction can occur if the arteries are kinked or injured during surgery and coronary artery stenosis can occur after arterial switch.[43] Long-term risk of myocardial ischemia after this procedure is unknown. The other possible complication is stenosis at the anastomosis of the pulmonary artery to the right ventricle and at the anastomosis of the aorta to the left ventricle.

## Atrial Baffle Procedures

The Mustard and Senning procedures were the most effective palliative procedures for transposition of the great vessels prior to development of a feasible arterial switch procedure. Both of these procedures rely on baffles placed in the atria to redirect flow across the atrial septum into the correct ventricle, generating an atrial level switch. The anatomic right ventricle continues to pump systemic blood, while the anatomic left ventricle pumps pulmonary blood. The true complication rate of these procedures is unknown. Although they have been performed since the 1960s, modifications and low late complication rates make true complication rates difficult to determine. Possible complications include syncope,[44] atrial arrhythmias, junctional rhythms, and thrombosis of venous drainage of either the pulmonary or systemic flow (baffle obstructions or stenosis). Atrial arrhythmias include atrial fibrillation, atrial flutter, and sick-sinus syndrome. Sudden death may be associated with the development of atrial flutter.[45] Thrombosis or stenosis of systemic venous drainage may lead to superior or inferior vena cava syndrome, ascites, protein losing enteropathy, and peripheral edema.

## Rastelli Procedure

Complex transpositions, such as those with sub-pulmonary stenosis with a ventricular septal defect, may require a Rastelli procedure instead of an arterial switch. The Rastelli procedure involves placing a patch to shunt blood from the left ventricle, across the VSD, and out the aorta. Second, the proximal pulmonary artery is ligated as it arises from the left ventricle, and a homograft or conduit is constructed from the right ventricle to the pulmonary artery. (*See Figure 5.*) Post-operative complications include right ventricular hypertension if conduit or branch PA stenosis develops. Patients having this procedure also may be at risk for ventricular arrhythmias and conduction abnormalities. The incidence and timing of this complication are unknown.

## Fontan

The Fontan procedure essentially bypasses the right ventricle to provide pulmonary blood flow. In the typical Fontan procedure, the superior vena cava drains predominantly into the right pulmonary artery. The inferior vena cava drains predominantly into the right atrium then directly flows into the left pulmonary artery. (*See Figure 6.*) There are several variations of this procedure, based upon initial anatomy and right ventricular size. This procedure is a surgical option for any patient with single ventricle physiology (tricuspid atresia, severe Ebstein's anomaly, HLHS, and some forms of complex congenital heart disease). The Glenn shunt, a superior vena cava to pulmonary artery anastomosis, is a staging shunt on the path to a Fontan.

Short-term complications of the Fontan include thromboembolism, thrombosis, and stensosis of the anastomosis.[46,47] Long-term complications include increased central venous pressure, fluid retention with ascites, atrial arrhythmias, pulmonary effusions, pericardial effusions, sudden death, and a protein-losing enteropathy with peripheral edema.[45,48] A cavopulmonary isolation procedure is performed, which has decreased the incidence of some of these complications. In this modification, the right atrium is bypassed and the vena cava are anastamosed directly (using external conduits) to the pulmonary artery, decreasing atrial arrythmias and pulmonary effusions.[48] Some patients will require a pacemaker to control atrial arrhythmias.[45,49] Laboratory studies can show a mildly hypercoaguable state and cholestasis.[48,50] One series found thrombotic complications in 25% of long-term survivors within four years of surgery,[46] but the presence of an activated coagulation system has not been demonstrated to be a risk factor. Seventy-five percent of thrombi were on the peripheral venous or pulmonary arterial portion of the circulation, and 25% were systemic arterial thrombi.[46]

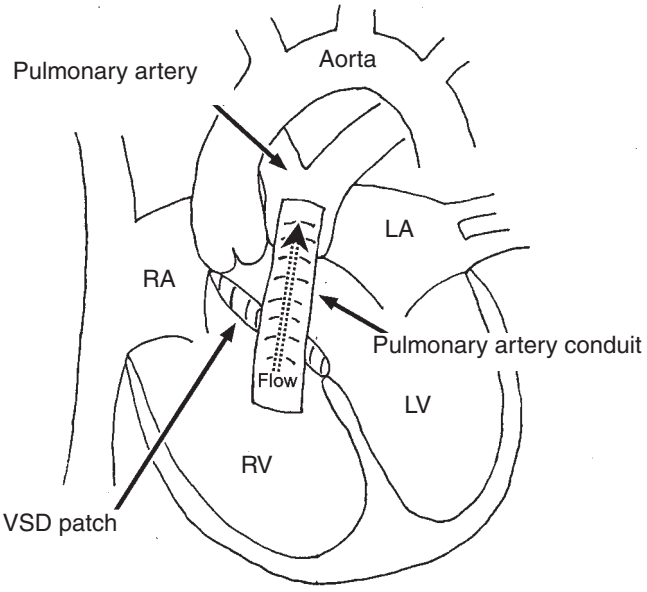
## Staged Reconstruction or Norwood

Children with HLHS or complex left-sided cardiac lesions have difficult therapeutic choices. Possible options include no therapy, orthotopic cardiac transplant, or the Norwood procedure. As experience has increased with the Norwood, more cardiologists are considering the Norwood a possible surgical option.

The staged reconstruction is used for HLHS or complicated left-sided cardiac lesions. The first stage involves creating a neoaorta, essentially a truncus arteriosis. The branch PAs are disconnected and perfused through a modified B-T shunt. The native, full-sized, main pulmonary artery is used to construct an aortic arch from the atretic aorta that is perfused by the full-sized right ventricle. (*See Figure 7.*)

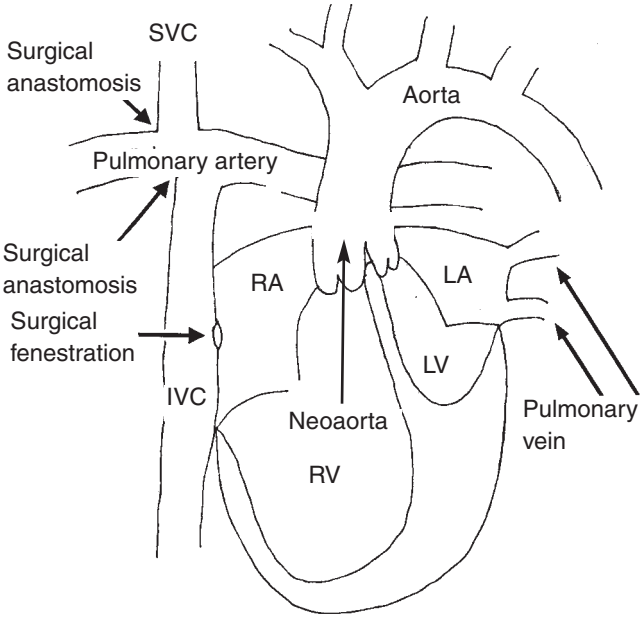
The second stage involves either a Glenn shunt or hemi-Fontan procedure with the removal of the aortopulmonary shunt constructed in stage one. (*See Figure 8.*) The inferior vena cava and right atria are not yet isolated from the systemic perfusion. This second stage is performed at approximately 6 months of age. The superior vena cava is disconnected from the heart and anastomosed to the right pulmonary artery. As with an isolated Fontan, noted complications include pleural effusions, pericardial effusions, phrenic nerve palsy, and transient superior vena cava syndrome.[51-54] The third and final stage, performed at approximately 18 months, involves completion of the Fontan, so that systemic venous blood drains directly to the pulmonary system. Pulmonary venous blood enters the left atrium, crosses the open atrial septum into the right atrium, enters the right ventricle, then exits through the constructed neoaorta. This technique is evolving, and long-term acute complications are unknown.

**Figure 5. Rastelli Procedure**




This illustration shows transposition of the great arteries and a ventricular septal defect after a Rastelli procedure (VSD patch and a right ventricle to pulmonary artery conduit to allow pulmonary flow).

**Figure 6. Hypoplastic Left Heart Syndrome after Fenestrated Fontan**




Shown above is hypoplastic left heart syndrome after a fenestrated Fontan (IVC and SVC to right pulmonary artery, with a surgical fenestration into the right atrium) that was performed after a Norwood procedure (surgical creation of a neoaorta and of a Blalock-Taussig shunt).

## Heart Transplantation

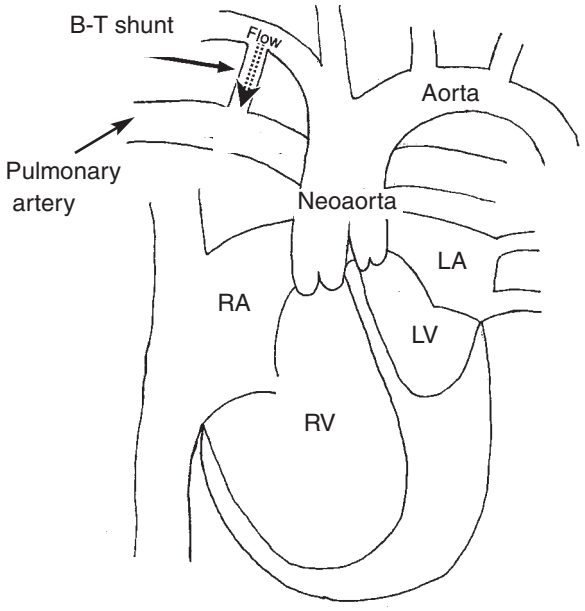
Cardiac transplantation in the pediatric population is due to cardiomyopathy unresponsive to medical management or congenital anomalies unresponsive to palliative procedures. Complications from transplantation include rejection, infection, and toxicity from immunosuppression. Rejection may present as cardiac failure, but also may present with mild vague symptoms, including fever, fatigue, vomiting, and diarrhea. Focal infections can include pneumonia, mediastinitis, and wound infections. As with all transplants, the greatest risk of bacterial infections is in the first month post-transplant. After that, viral infections predominate. Fungal infections are relatively infrequent, but still important, causes of infection. Transplant patients are at an increased risk of neoplasm; approximately 8% in one study, with lymphoma being the most common neoplasm seen.[55] The long-term complication of orthotopic heart transplantation is early-onset coronary artery disease.

## Endocarditis

Children with congenital heart disease are at an increased risk of endocarditis. Unfortunately, the classic symptoms of Roth spots, Osler's nodes, Janeway lesions, and splinter hemorrhages are all quite uncommon. The most common symptoms are fever, petechiae, malaise, embolic events, and a new or changing murmur. Other nonfocal symptoms can occur, including: splenomegaly, heart failure, gastrointestinal symptoms, and arthralgias. Fever usually is low-grade. The enlarged spleen usually is nontender. Arthralgias usually involve the large joints. Unfortunately, laboratory findings are not very sensitive. Possible laboratory abnormalities include elevated erythrocyte sedimentation rate, anemia, and hematuria.

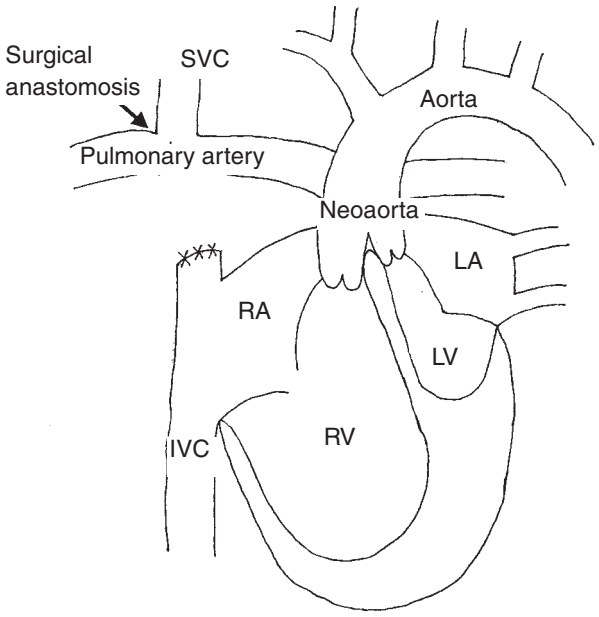
The data from one study demonstrate that endocarditis is uncommon in patients with surgically corrected septal defects or right-sided corrected lesions.[56] In fact, only one patient with a corrected VSD developed endocarditis. However, that patient also had a residual defect after surgery. The patients at highest risk for endocarditis were those who had surgical procedures on the aorta or aortic valve. Another study reviewed the cases of endocarditis in all patients with congenital heart disease.[57] In 87 of 214 with endocarditis, a pre-disposing event could be identified. Dental procedures preceded 42 cases of endocarditis; skin infections preceded another 10 cases. The study reported that patients received appropriate endocarditis prophylaxis in half of those cases of endocarditis related to dental procedures. Currently, endocarditis prophylaxis is recommended for all patients with corrected congenital heart disease. Exceptions include patients with isolated secundum ASD and VSD and PDA who underwent complete correction more than 6 months prior and have no residual shunting.[58] It is unknown which ED procedures have indications for prophylaxis. Dental procedures have been clearly defined. However, lacrimal duct probing causes a transient bacteremia in 17% of children. Cetta reviewed survey data of physicians caring for patients with congenital heart disease who had tattooing or piercing. Although only 6% took appropriate prophylaxis, and local skin infections occurred in 23% of patients, there were no cases of endocarditis.[59] There is no clear data on endocarditis risk after facial trauma, open frac-

**Figure 7. Hypoplastic Left Heart Syndrome after Norwood Procedure**




This illustration shows hypoplastic left heart syndrome after a Norwood procedure (creation of a neoaorta and a Blalock-Taussig shunt).

**Figure 8. Hypoplastic Left Heart Syndrome after Glenn Shunt**




The illustration above shows hypoplastic left heart syndrome after a Glenn shunt (SVC to right pulmonary artery anastomosis) performed after a Norwood procedure.

tures, or traumatic lacerations. Routine endocarditis prophylaxis is currently is not recommended for uncomplicated and clean skin lacerations.

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**In Future Issues:** **Urinary Tract Infection**

## CME Instructions

Physicians participate in this continuing medical education program by reading the article, using the provided references for further research, and studying the questions at the end of the article. Participants should select what they believe to be the correct answers, then refer to the list of correct answers to evaluate their knowledge. To clarify confusion surrounding any questions answered incorrectly, please consult the source material. *After completing this activity, you must complete the evaluation form that will be provided at the end of the semester and return it in the reply envelope provided to receive a certificate of completion.* When your evaluation is received, a certificate will be mailed to you.

## Physician CME Questions

51. To what does peripheral cyanosis refer?
    A. Visible cyanosis of the lips and face
    B. Poor peripheral oxygenation
    C. Inadequate distal perfusion
    D. Hypoxia

52. Children with which of the following lesions present infrequently to the ED for initial diagnosis?
    A. Hypoplastic left heart syndrome
    B. Coarction of the aorta
    C. Tetralogy of Fallot
    D. Transposition of the great vessels

53. At what age do children typically present with VSD?
    A. 2-6 days old
    B. 2-6 weeks old
    C. 12-14 months old
    D. 6 months old

54. Prostaglandin infusion may cause which of the following?
    A. Hypothermia
    B. Lethargy
    C. Apnea
    D. Tachycardia
    E. Hypertension

## *Emergency Medicine Reports* CME Objectives

*To help physicians:*

- quickly recognize or increase index of suspicion for specific conditions;
- understand the epidemiology, etiology, pathophysiology, and clinical features of the entity discussed;
- be educated about how to correctly perform necessary diagnostic tests;
- take a meaningful patient history that will reveal the most important details about the particular medical problem discussed;
- apply state-of-the-art therapeutic techniques (including the implications of pharmaceutical therapy discussed) to patients with the particular medical problems discussed;
- understand the differential diagnosis of the entity discussed;
- understand both likely and rare complications that may occur;
- and provide patients with any necessary discharge instructions.

55. What causes the intraventricular conduction delay seen on ECGs of patient's with a repaired tetralogy of Fallot?
    A. The VSD interrupts the conduction system.
    B. Recurrent "tet spells" cause ischemia with conduction delay.
    C. Pulmonary hypertension causes the conduction delay.
    D. The bundle of His may be injured during repair.
    E. The hypoplastic right ventricle has an abnormal conduction system.

56. After complete repair, patients with which heart lesion no longer require endocarditis prophylaxis?
    A. VSD
    B. B-T shunt
    C. Tetralogy of Fallot
    D. Transposition of the great vessels
    E. Coarctation of the aorta

57. Which of the following describes hypoplastic left heart syndrome?
    A. The aorta arises from the right ventricle and the pulmonary artery from the left ventricle.
    B. Neonates with either hypoplasia or absence of the left ventricle
    C. Constriction of the aorta
    D. The aorta and pulmonary artery arise from the right ventricle.

58. Complications from B-T shunts include which of the following?
    A. Injury to the phrenic or recurrent laryngeal nerves
    B. Residual cyanosis from inadequate pulmonary blood flow if the shunt is too small
    C. Excess pulmonary blood flow
    D. Pulmonary edema from a shunt that is too large
    E. All of the above

59. Which of the following is true of VSD repair?
    A. Small VSDs will not close spontaneously.
    B. Type and size of VSD does not influence surgical repair.
    C. Indications for surgery include uncontrolled CHF, poor weight gain in infancy, and developing pulmonary hypertension.
    D. All patients still require endocarditis prophylaxis six months after surgery.

60. The most common symptoms of endocarditis are fever, petechiae, malaise, embolic events, and a new or changing murmur.
    A. True
    B. False

### CME Answers

| | | | |
|---|---|---|---|
| 51. | C | 56. | A |
| 52. | C | 57. | B |
| 53. | B | 58. | E |
| 54. | C | 59. | C |
| 55. | D | 60. | A |

Emergency Medicine Reports
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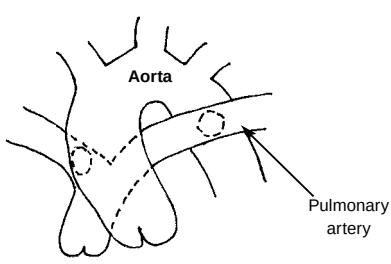
# Pediatric Congenital Heart Disease

## Glossary

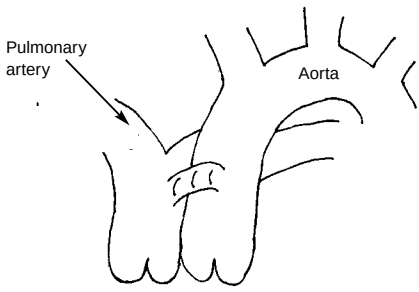
| | | |
|---|---|---|
| ASD | — | Atrial septal defect |
| AV canal | — | Atrioventricular septal defect |
| B-T shunt | — | Blalock-Taussig shunt (subclavian artery to branch pulmonary artery shunt) |
| Central shunt | — | Anastomosis from ascending aorta to main pulmonary artery |
| CHD | — | Congenital heart disease |
| DORV | — | Double outlet right ventricle |
| HLHS | — | Hypoplastic left heart syndrome |
| PDA | — | Patent ductus arteriosis |
| PGE1 | — | Prostaglandin E1 |
| Potts anastomosis | — | Shunt from descending aorta to left pulmonary artery |
| TGA | — | Transposition of the great arteries/ vessels |
| VSD | — | Ventricular septal defect |
| Waterston shunt | — | Anastomosis from ascending aorta to main or right pulmonary artery |

## Waterston and Potts Shunts



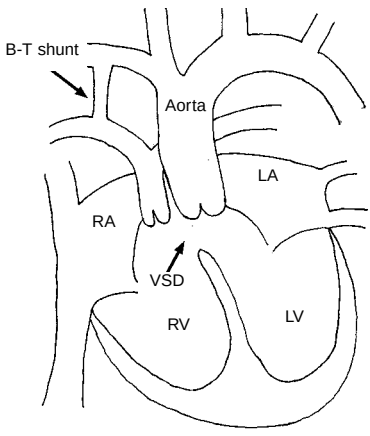

Examples of the Waterston shunt (ascending aorta to right pulmonary artery) and the Potts shunt (descending aorta to left pulmonary artery)

## Central Shunt



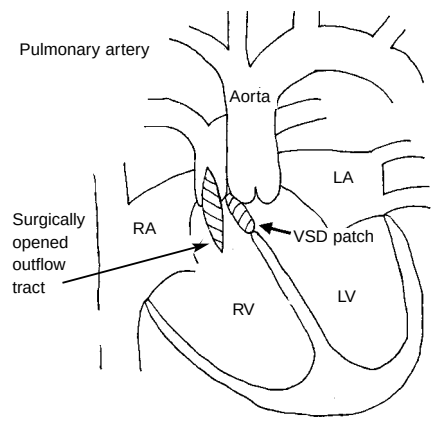

Central shunt (ascending aorta to main pulmonary artery)

## Blalock-Taussig Shunt



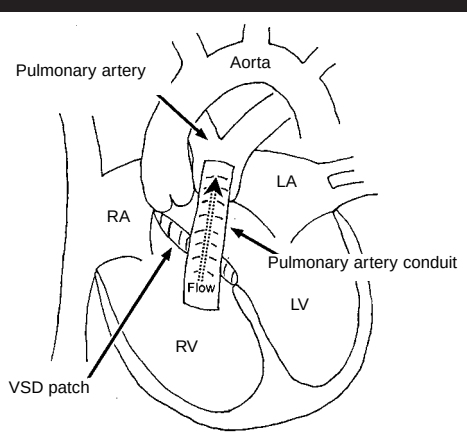

This illustration shows a Blalock-Taussig shunt (allowing flow from right pulmonary artery to right subclavian artery) in a patient with tetralogy of Fallot.

## Corrected Tetralogy of Fallot



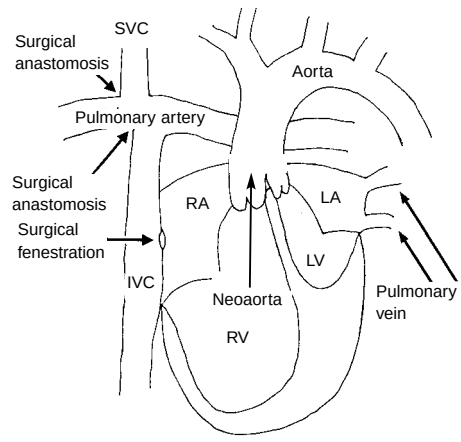

This illustration shows a corrected tetralogy of Fallot with a VSD patch and a right ventricular outflow tract patch to increase pulmonary artery flow.

## Rastelli Procedure



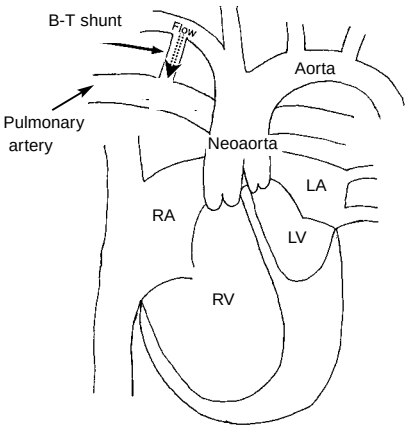

This illustration shows transposition of the great arteries and a ventricular septal defect after a Rastelli procedure (VSD patch and a right ventricle to pulmonary artery conduit to allow pulmonary flow).

## Hypoplastic Left Heart Syndrome after Fenestrated Fontan



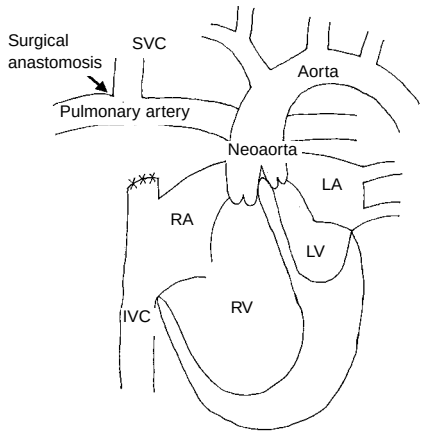

Shown above is hypoplastic left heart syndrome after a fenestrated Fontan (IVC and SVC to right pulmonary artery, with a surgical fenestration into the right atrium) that was performed after a Norwood procedure (surgical creation of a neoaorta and of a Blalock-Taussig shunt).

## Hypoplastic Left Heart Syndrome after Norwood Procedure




This illustration shows hypoplastic left heart syndrome after a Norwood procedure (creation of a neoaorta and a Blalock-Taussig shunt).

## Hypoplastic Left Heart Syndrome after Glenn Shunt




The illustration above shows hypoplastic left heart syndrome after a Glenn shunt (SVC to right pulmonary artery anastomosis) performed after a Norwood procedure.

Supplement to *Emergency Medicine Reports*, March 10, 2003: "Congenital Heart Disease in the Emergency Department." *Authors:* **William A. Woods, MD, FAAP,** Assistant Professor of Emergency Medicine and Pediatrics, University of Virginia, Charlottesville; **Edward Ullman, MD,** Attending Physician, Department of Emergency Medicine, Beth Israel Deaconess Medical Center—Harvard Medical School, Boston, MA; and **Deborah Schutte, MD,** Pediatric Cardiologist, Cook Children's Hospital, Fort Worth, TX.