Congenital Heart Disease

Kriti Puri, MD,* Hugh D. Allen, MD,* Athar M. Qureshi, MD*[†]

*Department of Pediatrics, [†]CE Mullins Cardiac Catheterization Laboratories, The Lillie Frank Abercombie Section of Cardiology, Texas Children's Hospital, Baylor College of Medicine, Houston, TX

Education and Practice Gaps

Congenital heart disease (CHD) is present in about 9 of every 1,000 live-born children. [1][2][3][4][5] Children with CHD are surviving longer, and better understanding of the long-term complications of CHD is continuously emerging. Hence, it is important to be comfortable with the primary care requirements for these children, including physical manifestations prior to surgery and interventional cardiac catheterizations, as well as those concerning for potential need for reintervention, the latest recommendations for endocarditis prophylaxis, respiratory precautions and immunization considerations, and close monitoring of development and behavior. In this article, we will discuss the common types of cyanotic ("blue") and acyanotic ("pink") CHD and the role of the primary care physician in the health care of these children before and after surgery and interventional cardiac catheterizations.

ABBREVIATIONS

ASD	atrial septal defect
AVSD	atrioventricular septal defect
CHD	congenital heart disease
CHF	congestive heart failure
CoA	coarctation of the aorta
EKG	electrocardiogram
HLHS	hypoplastic left heart syndrome
IAA	interrupted aortic arch
IVC	inferior vena cava
LV	left ventricle
mBTT	modified Blalock-Taussig-Thomas
PAPVD	partial anomalous pulmonary venous drainage
PDA	patent ductus arteriosus
RV	right ventricle
SBE	subacute bacterial endocarditis
TAPVD	total anomalous pulmonary venous drainage
VSD	ventricular septal defect

Objectives   After completing this article, readers should be able to:

1. Describe newborn pulse oximetry screening for congenital heart disease (CHD) and clinical features of CHD during the newborn period.

2. Describe the clinical presentations and briefly outline management strategies of infants and children with different types of CHD.

3. Discuss single-ventricle palliation by using hypoplastic left heart syndrome as the model lesion.

4. Describe the evaluation and primary care management factors (including endocarditis prophylaxis, immunizations, and exercise restriction) in children with different types of CHD.

NEWBORN PRESENTATION OF CRITICAL CONGENITAL HEART DISEASE

Newborns with critical congenital heart disease (CHD) may present with symptoms of cyanosis, congestive heart failure (CHF), poor pedal pulses, or a failed newborn CHD pulse oximetry screen prior to discharge. CHD lesions that are dependent on a patent ductus arteriosus (PDA) to support flow either to the systemic circulation or to the pulmonary circulation will manifest with shock or

cyanosis, respectively, when the PDA starts to close. CHD lesions that allow shunting of blood to the pulmonary circulation manifest with progressive symptoms of pulmonary overcirculation as the pulmonary vascular resistance decreases.

It may be challenging to distinguish cardiac disease from pulmonary disease or sepsis in a newborn with cyanosis and/or respiratory distress. Chest radiography and assessment of response to oxygen and/or positive pressure ventilation as clinically indicated help to elucidate the cause. Lack of response to 100% oxygen for at least 10 minutes (hyperoxygenation) indicates that the etiologic origin for the clinical picture is likely cardiac, and further cardiac workup is indicated. Hyperoxygenation will help infants with pulmonary parenchymal etiologic origins and those with pulmonary hypertension as causes of cyanosis. A cyanotic newborn who is otherwise well appearing may also have a hemoglobinopathy, such as methemoglobinemia. However, chest radiography and echocardiography performed to rule out cyanotic CHD are indicated in a cyanotic infant who is not responsive to hyperoxygenation.

Features of CHF in a newborn include tachypnea, increased work of breathing, tachycardia, and hepatomegaly. While tachypnea and increased work of breathing may also indicate pulmonary disease or an inborn error of metabolism, hepatomegaly at examination suggests cardiac involvement. It is useful to percuss the top of the liver, as well as palpate its edge to estimate the liver span (normal newborn liver span is 4.5 to 5.0 cm). A chest radiograph showing cardiomegaly and pulmonary vascular congestion in an infant with respiratory distress and normal saturations in all 4 extremities is indicative of a left-to-right shunting process. These physical examination and chest radiographic findings in a newborn prompt a differential diagnosis of obligate shunts, such as arteriovenous malformations. In comparison, these examination and chest radiographic features in a 2- to 8-week-old infant with decreased pulmonary resistance suggest a dependent shunt, such as a large ventricular septal defect (VSD). N-terminal pro–B-type natriuretic peptide may be considered to help distinguish between noncardiac and cardiac causes of respiratory distress. (6) Severe pulmonary hypertension may also cause hepatomegaly and right ventricle (RV) dysfunction. Hyperoxygenation will help reduce the pulmonary vascular resistance and ameliorate the pulmonary hypertension; however, in these instances, it will worsen shunting and pulmonary overcirculation in left-to-right shunt lesions.

Weaker femoral pulses in a child who has otherwise strong radial pulses is concerning for coarctation or interruption of the aorta. Weak femoral pulses in the setting of

lethargy and other signs of poor perfusion and normal saturations are concerning for critical obstructive lesions at the level of the aortic valve or the aortic arch that require rapid prostaglandin initiation. Sepsis and inborn errors of metabolism are other situations that must be evaluated and/or empirically treated in a sick newborn who has poor systemic output; however, cardiac evaluation with an electrocardiogram (EKG), chest radiograph, and echocardiogram must be simultaneously initiated.

The pulse oximetry screen for critical CHD in newborns was approved as a part of the Routine Universal Screening Program in 2011 and has been adopted in 46 states and the District of Columbia. (5)(7)(8) The most common protocol used is shown in Fig 1 (variations exist, described elsewhere [8]). This screen is intended to target detection of types of critical CHD that will manifest in the newborn period and require early intervention, including coarctation of the aorta (CoA), interrupted aortic arch (IAA), tetralogy of Fallot, truncus arteriosus, tricuspid atresia, total anomalous pulmonary venous drainage, double-outlet RV, Ebstein anomaly, pulmonary atresia, transposition of the great arteries, hypoplastic left heart syndrome (HLHS), and other single-ventricle lesions. A failed newborn screen may also indicate other disease processes, such as pulmonary hypertension, primary pulmonary parenchymal or interstitial disease, or hemoglobinopathies. Regardless, a failed screening result is an indication for prompt evaluation, including chest radiography, EKG, and echocardiography (supervised and evaluated by a pediatric cardiologist). If cardiac investigations or a pediatric cardiologist are not easily available in such a situation, initiation of prostaglandin infusion with close monitoring of the airway (due to risk of apnea with prostaglandin initiation) and prompt transport to a higher-level center are recommended.

ACYANOTIC HEART DISEASE

Atrial Septal Defects

Atrial septal defects (ASDs) comprise 7% to 10% of CHD. (1)(2)(3)(4) Based on their embryological origin, they can be classified as secundum ASDs (the most common type, due to a deficiency in the central part of the septum primum that develops from the middle of the atrial cavity and grows inferiorly to meet the endocardial cushion), primum ASDs (due to deficient proliferation of the endocardial cushion), sinus venosus ASDs (defects near the superior vena cava and right upper pulmonary vein, truly a form of partial anomalous pulmonary venous drainage), and other rarer
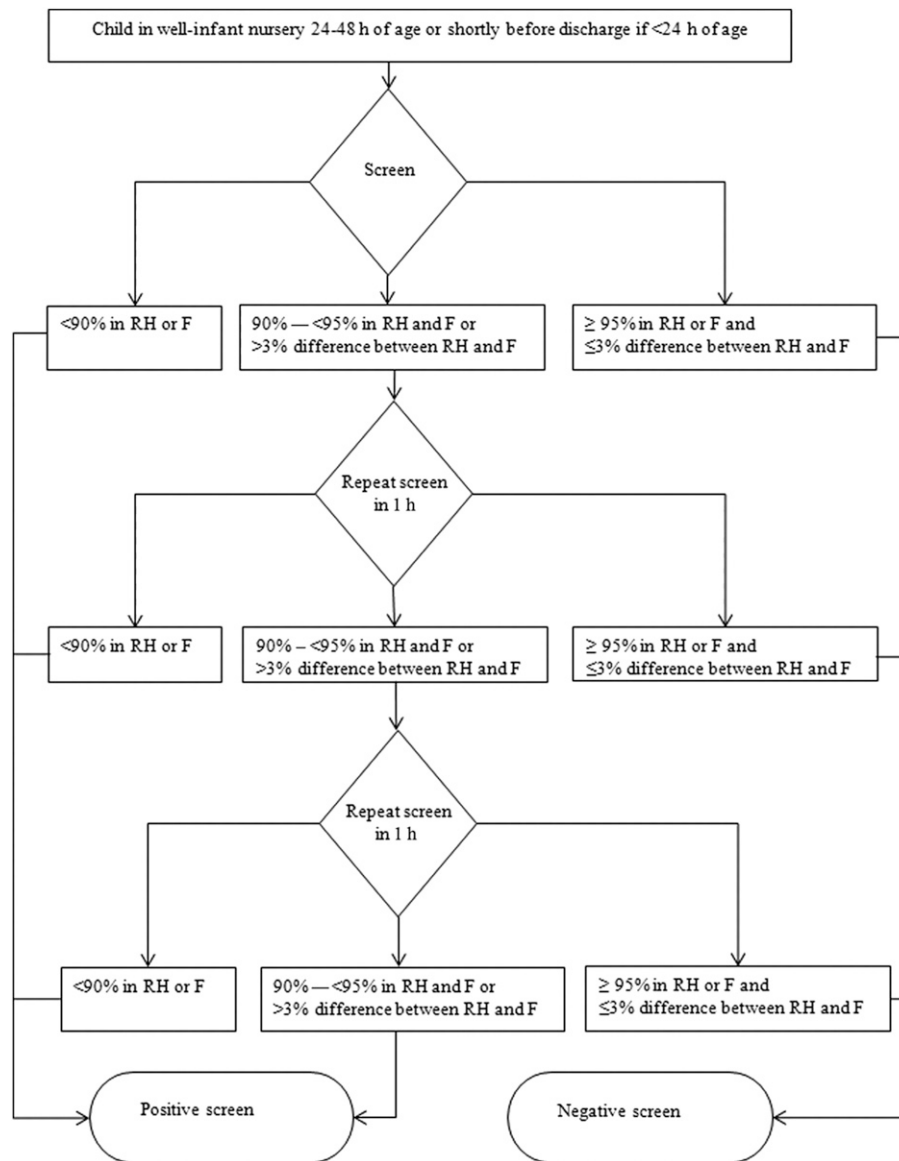
Figure 1. Flowchart shows the protocol for newborn pulse oximetry screening for critical congenital heart disease. F=foot, RH=right hand. Image obtained with permission from Kemper AR, Mahle WT, Martin GR, et al. Strategies for implementing screening for critical congenital heart disease. *Pediatrics.* 2011;128:e1259.

forms of ASDs. Shunting of blood from left to right at the atrial level leads to increased diastolic blood volume in the RV, which causes right-sided chamber dilation.

Clinical Manifestations and Diagnosis. ASDs are typically discovered when a murmur is heard at a regular 4- to 6-month infant well-child visit. The murmur is loudest over the pulmonic region and is associated with a fixed splitting of the S_2 during different phases of respiration and a loud S_1. The murmur heard is from increased blood flow across the pulmonary valve because of the greater volume of blood in the right side of the heart (relative pulmonic stenosis). An additional diastolic murmur may be heard at the left lower sternal border from excess flow across the tricuspid valve. There may be left precordial bulging due to the enlarged RV being present during cartilaginous rib development. Patients with ASDs are usually asymptomatic but may have some fatigue. They are followed up with Doppler echocardiography to monitor enlargement of the right atrium and/or RV, which, if present, indicates hemodynamic significance. (9) Infants with an ASD manifesting with features of CHF (tachypnea, increased work of breathing, frequent pauses in feeding, failure to thrive, profuse sweating while feeding, decreased activity levels) must be investigated for an additional lesion such as a PDA, VSD with pulmonary

arterial stenosis, or a left-sided obstructive lesion (cor tria-triatum, coarctation, aortic or mitral stenosis).

Management. Secundum ASDs can be repaired either surgically by using a patch or a primary suture closure of the defect or in the cardiac catheterization laboratory by using an ASD closure device. (10)(11) ASDs that are too large or do not have appropriate anatomy usually require surgical repair. (12) There has been no significant difference shown in studies comparing the procedures, and there is also no difference observed clinically. Typically, in clinical practice, patients with hemodynamically significant ASDs are referred for intervention around 3 to 4 years of age. Rarely do ASDs have to be closed earlier.

Ventricular Septal Defects

VSDs are the most common CHD lesion and are present in 50% to 60% of all children with CHD. (1)(2)(3)(4) VSDs develop from defective formation of the interventricular septum and are classified on the basis of their location in the septum relative to the atrioventricular valves and the right and left ventricular outflow tracts. Perimembranous VSDs (deficiency in a fibrous part of the septum at the base of the heart) are the most common type, constituting about 80% of all VSDs, followed by doubly committed juxta-arterial (deficiency in the infundibular septum, between the pulmonary and aortic valves), muscular (in the trabecular part of the septum), and inlet (deficiency in the septum inferior to the perimembranous region and above the level of cordal attachments of the atrioventricular valves). Size of the VSD is defined relative to the area of the aortic valve: small (less than one-third of the area), moderate (one-third to two-thirds of the area), and large (more than two-thirds of the area).

Clinical Manifestations and Diagnosis. A newborn with a VSD may not initially have a murmur; however, as the pulmonary resistance decreases with age, an S_1-coincident pansystolic murmur can be heard the loudest over the left lower sternal border. A diastolic rumble at the apex may be heard from excess flow across the mitral valve. A child with a hemodynamically significant VSD presents with features of pulmonary overcirculation and CHF. A chest radiograph shows cardiomegaly with pulmonary vascular congestion (Fig 2), while EKG shows left or biventricular hypertrophy. As the child grows, some perimembranous VSDs can get occluded by aneurysmal tissue, and muscular VSDs can become smaller in size with muscular growth. One must be wary of the perimembranous defect that becomes smaller owing to prolapse of the aortic valve.

Management. Children with VSDs are regularly followed up by a pediatric cardiologist. Children with CHF symptoms are temporized with diuretics—usually furosemide,
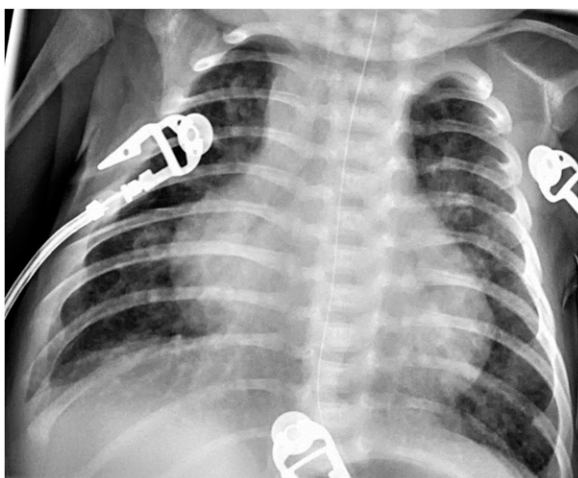

Figure 2. Frontal chest radiograph in a patient with a ventricular septal defect shows features of congestive heart failure, including cardiomegaly and increased pulmonary vascular congestion, as well as splaying of the bronchi, which is indicative of left atrial enlargement.

sometimes chlorothiazide, and/or spironolactone—and may require frequent evaluation of electrolyte levels. Patients with failure to thrive may need up to 125 to 150 kcal/kg per day of caloric intake through fortified formula. Close, reliable, and accurate monitoring of weight, with attention to using the same weighing scale and reinforcement of education about proper mixing of formula, are critical goals to be reviewed during the weight-check visits at the pediatrician's office. With current surgical success in treating these lesions, there is little need for prolonged medical treatment. Surgical patch repair of the VSD is performed either in a symptomatic infant or in a toddler who has enlargement of the left atrium or ventricle at echocardiography. Sometimes perimembranous and doubly committed juxta-arterial VSDs can be associated with prolapse of the right or noncoronary cusp of the aortic valve through the defect, which may cause progressive aortic insufficiency. These VSDs are repaired even in asymptomatic patients. Some VSDs (eg, muscular VSDs) can be closed with devices in the cardiac catheterization laboratory. After VSD repair, these patients need to have continued follow-up of weight and nutrition to ensure regaining of growth curves.

Atrioventricular Septal Defects

Atrioventricular septal defects (AVSDs), (also known as *atrioventricular canal defects*), constitute 5% of CHD. (1)(2)(3)(4) About 50% of patients with AVSDs have Down syndrome. AVSDs arise from defects in the proliferation and fusion of the endocardial cushions and are classified on the basis of the commitment of the common atrioventricular valve to the ventricles, the presence or absence of separate orifices, and the anatomy of the subvalvar

attachments. Most commonly, patients have a primum type of ASD, as well as an inlet-type VSD component, and in addition, they may have a lack of complete apposition or coaptation (so-called cleft) in the mitral valve.

Clinical Manifestations and Diagnosis. Patients with AVSD typically have a systolic ejection or holosystolic murmur from the VSD component. However, if the VSD is large and there is equalization of pressures in the ventricles (or if pulmonary hypertension is present), a murmur may not be heard. Additionally, these patients may have murmurs of atrioventricular valve regurgitation, characterized as an S_1-coincident systolic murmur, heard the loudest over the apex in the case of left-sided regurgitation and the loudest over the right lower sternal border in the case of right-sided regurgitation. A diastolic rumble may be heard, similar to that heard in patients with VSDs. An EKG in these children shows left, right, or biventricular hypertrophy and may have a northwest axis (negative axes in leads I, II, III, and aVF), which is suggestive of a diagnosis of AVSD [Fig 3]). These children may also present with features of CHF and failure to thrive, with chest radiographic findings as previously described (in the VSD section). Severe atrioventricular valve regurgitation, creating more preload, will exacerbate earlier CHF symptoms.

Management. Patients with AVSD with an additional diagnosis of Down syndrome may remain in the hospital for a prolonged period of time as a neonate, owing to feeding difficulties. All patients with AVSD are closely followed up, since they typically require surgical intervention around 4 to 6 months of age or earlier, if they have severe CHF or failure to thrive. The diuretic and nutritional management and close monitoring for CHF is similar to that in patients with VSD. While the surgical mortality rate in the current surgical era is less than 3%, the need for reintervention owing to atrioventricular valve regurgitation or stenosis remains about 10% to 15% at the 5-year postoperative mark. (13)(14)

Patent Ductus Arteriosus

An arterial duct is a normal fetal connection between the aorta and the pulmonary artery that is present in all newborns. It closes functionally within 24 hours and anatomically within 3 to 4 weeks in most patients. It may remain patent longer in some patients, especially infants born prematurely.

Clinical Manifestations and Diagnosis. The examination of patients with PDA varies with their age. In a newborn with higher pulmonary resistance and pressures, the PDA may not shunt much blood and may not be audible. In an
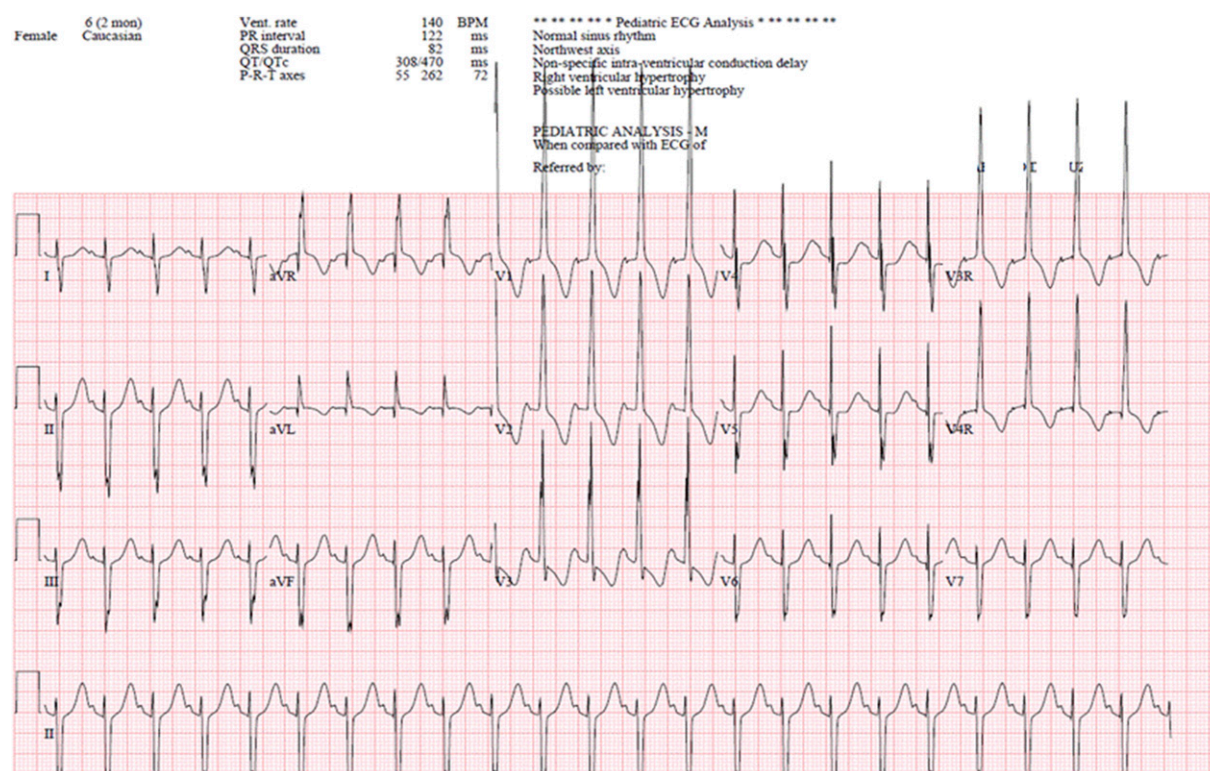

Figure 3. Electrocardiogram of a patient with atrioventricular septal defect shows biventricular hypertrophy, as well as the northwest axis deviation, with negative QRS axes in leads I and aVF.

older infant, however, one can detect a loud systolic or continuous murmur of blood shunting left to right during the entire cardiac cycle, which is loudest over the left precordium. In an older patient with a hemodynamically significant PDA, one may be able to detect a diastolic rumble of transmitral flow into an enlarged left ventricle (LV). The left-to-right shunting caused by the PDA may also lead to features of CHF. Chest radiographs illustrate features of pulmonary overcirculation and cardiac enlargement, EKG can show LV hypertrophy and ST-segment changes of ischemia (beware of this finding in a newborn, as it implies a very large shunt), and Doppler echocardiograms can demonstrate left atrium and LV enlargement, characteristics of flow, and anatomy of the PDA.

Management. Ibuprofen and indomethacin have been used to medically close PDAs in premature infants. Acetaminophen is also being studied as an alternative medical therapy. (15) A symptomatic or hemodynamically significant PDA is a class I indication for intervention, with either device occlusion in the interventional catheterization laboratory or surgical ligation. (9) In a center with interventional cardiac catheterization expertise, device occlusion is the first-line approach for an older infant or child, with excellent results and extremely low complication rates comparable to those of surgical ligation. (16) In babies with low birth weight and those with extremely low birth weight, personnel at some centers lean toward surgical ligation, often at the bedside. However, successful PDA device occlusion has been reported in small neonates, and the most recent series demonstrates successful catheter-based intervention in neonates as small as 700 g. (17) Both procedures have similarly high success rates and a complication rate of less than 1%.

Patients with left-to-right shunt lesions (eg, with a VSD, AVSD, PDA, and, in very rare instances, an ASD) that present late may have pulmonary hypertension. Pediatricians may encounter these scenarios when examining patients who have not had access to medical care (eg, patients from some developing countries or areas of the United States that are medically underserved). The degree of pulmonary hypertension due to pulmonary vascular obstructive disease may be prohibitive for repair. Some of these patients will be treated with agents that lower pulmonary hypertension and may later in life go on to develop Eisenmenger syndrome (when pulmonary vascular resistance exceeds systemic vascular resistance, resulting in a reversal of the shunt) and its associated sequelae.

Aortic and Pulmonic Valve Stenosis

Aortic stenosis constitutes 5% to 8% of all CHD, and pulmonary stenosis constitutes about 8% to 10% of all CHD. (1)(2)(3)(4) The degree of stenosis is categorized as critical if the blood flow through the respective valve is insufficient and requires additional contribution from the PDA. These valves undergo aberrant tissue resorption during in utero development, which leads to thickened dysplastic valves that do not open or close normally. Hence, they create obstruction to blood flow and may also be regurgitant. Aortic stenosis may be associated with a bicuspid or even a unicuspid aortic valve, as well as other levels of obstruction in the left-sided circulation, including CoA, subaortic obstruction, and mitral stenosis. A bicuspid aortic valve has been found to be associated with aortic root dilation due to an aortopathy and requires close follow-up, even if there is no stenosis across it.

Clinical Manifestations and Diagnosis. Patients present with an ejection systolic murmur that is loudest over the respective valvar region and may also have an ejection click (if stenosis is moderate or mild). The aortic click is heard at the apex and right upper sternal border and does not vary with respiration, whereas the pulmonary click heard at the left lower and upper sternal borders will be loudest when the patient exhales. The EKG may show ventricular hypertrophy of the RV in cases of pulmonic stenosis and of the LV in cases of aortic stenosis. Doppler echocardiograms help estimate the velocity of blood flow across the valve. Based on the continuity equation, the narrower the valve area, the faster the flow velocity. This flow velocity is then used to estimate the pressure gradient across the stenotic valve by using a modified Bernoulli principle (peak velocity in meters per second squared, times 4).

Management. Patients with critical aortic stenosis or pulmonic stenosis meet a class I indication to undergo a catheter-based balloon valvuloplasty procedure in the newborn period to ameliorate ductal dependence. (9)(18) For noncritical stenosis, the time of intervention is determined by the presence of peak gradient of ≥50 mm Hg obtained across the aortic valve (≥40 mm Hg in the setting of ventricular dysfunction) or ≥40 mm Hg across the pulmonary valve at echocardiography or during cardiac catheterization, on the basis of the American Heart Association guidelines (class I indication [9][19]). Patients with aortic stenosis may undergo a surgical valvuloplasty, depending on the valve anatomy, annulus size, and experience and preference of the personnel at the center. There has been no reported significant difference in survival or rate of aortic valve replacement between patients undergoing balloon valvuloplasty and those undergoing surgical valvuloplasty, although patients undergoing balloon valvuloplasty may have a greater frequency of reintervention. (10)(20)(21) Patients undergoing balloon valvuloplasty of the aortic valve in the first year after

birth typically need reintervention in about 50% of the critical aortic stenosis cases and 33% of the noncritical aortic stenosis cases, necessitating long-term follow-up for restenosis and/or regurgitation. (18)(19) Patients undergoing balloon valvuloplasty of the pulmonary valve may not require another procedure for several years or ever again; however, they do require regular cardiology re-evaluation and Doppler echocardiography to assess the need for re-intervention due to restenosis and/or regurgitation. (9)

CoA and Interruption of the Aorta

CoA is a narrowing at the isthmus of the arch of the aorta (with concomitant narrowing of the transverse arch in some cases), constituting 5% to 8% of CHD. (1)(2)(3)(4) IAA is the most extreme end of this spectrum, with a discontinuation of the arch and distal continuation through a PDA past the point of interruption. IAA accounts for about 1.5% of CHD and is classified according to the site of the interruption: type A is interrupted at the isthmus distal to the left subclavian artery, type B is interrupted between the origins of the left subclavian and left common carotid arteries (the most common type and frequently associated with chromosome 22 abnormalities), and type C is interrupted between the origins of the innominate and left common carotid arteries (the most rare type). About 80% of patients with IAA have an associated VSD.

Clinical Manifestations and Diagnosis. Severe and critical CoA and IAA may be detected at the time of newborn screening with a lower saturation level at the postductal site or via clinical suspicion on the basis of poor pedal or femoral pulses at the first newborn visit to the pediatrician. These patients may present in extremis at 2 to 3 weeks of age, in shock, with feeble pulses, lethargy, poor feeding, decreased urine output, and metabolic acidosis. While sepsis remains the most common cause of this constellation of symptoms, an echocardiogram should be performed for CoA and/or IAA if the shock is unresponsive to fluid resuscitation. Mild CoA may be detected on the basis of a gradient of more than 20 mm Hg between the upper- and lower-extremity blood pressures (carefully performed with proper technique) during infancy. A harsh systolic murmur that is loudest over the back may be heard. CoA may also be found in young adolescents undergoing workup for hypertension. There may not be a clinically significant blood pressure gradient in older patients with CoA because they develop collateral vessels over time that supply blood distal to the narrowing. In the case of collateralization, one can detect a continuous murmur of the flow in these vessels over the chest wall.

Management. Newborns with severe or critical CoA and IAA are dependent on prostaglandin infusion to keep the PDA open until the time of surgical repair. Some children (particularly older children) are candidates for intervention in the cardiac catheterization laboratory, with balloon angioplasty and stent placement for the coarctation (Videos 1, 2). (22)(23) After repair, patients are followed up closely in the cardiology clinic to monitor for recurrence of CoA and persistent hypertension and continue to receive long-term follow-up at 1–2-year intervals.

CYANOTIC CHD

Tetralogy of Fallot

Tetralogy of Fallot is the most common cyanotic CHD, accounting for 5% of all CHD. (1)(2)(3)(4) It develops from anterior malalignment of the interventricular septum, which leads to a VSD, as well as overriding of the VSD by the aorta. There is a narrowing of the pulmonary outflow tract due to the septal deviation, and this causes RV outflow (infundibular) obstruction and consequent RV hypertrophy (Videos 3, 4).

Clinical Manifestations and Diagnosis. Patients with tetralogy of Fallot have a harsh ejection systolic murmur heard over the pulmonic area, indicating pulmonic stenosis. There may be a single second sound. Higher saturations indicate less RV outflow obstruction. Patients who do not receive a diagnosis prenatally may present for the first time as children having a hypercyanotic spell in a period of agitation, fever, or other concurrent illness. Agitation and crying increase pulmonary vascular resistance, while also increasing the heart rate. Owing to a subsequently shorter diastolic period, ventricular filling is less, which adds to the obstruction to the RV outflow from its hypertrophic muscle bundles. "Tet spells" may become a vicious cycle and require a

Video 1. Aortogram in a teenager with coarctation of the aorta demonstrates narrowing of the descending aorta.
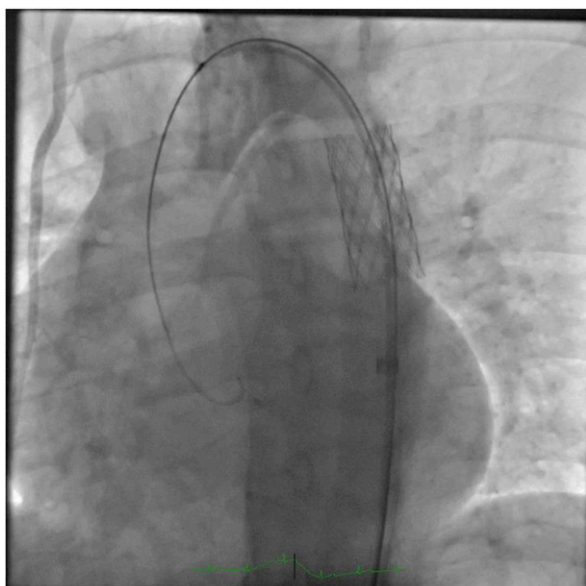
Video 2. Aortogram obtained after angioplasty in a patient with coarctation of the aorta who underwent stent placement in the cardiac catheterization laboratory. The narrowing is abolished with stent placement.

combination of heart rate control and vascular resistance manipulation to break. The murmur during a hypercyanotic spell or "Tet spell" becomes softer because of decreased pulmonary flow. Iron deficiency anemia will accelerate the onset of these spells.

Management. The time of presentation and intervention for tetralogy of Fallot is determined by the degree of RV outflow obstruction and the limitation of pulmonary blood flow. (24) These clinical indicators are the oxygen saturation levels and the development of hypercyanotic spells. The Doppler echocardiographic indicators are the pressure gradients observed across the RV outflow tract. Children with saturations less than 80% or those having hypercyanotic spells are scheduled for surgery. Acute "Tet spells" are managed by first helping to calm the patient (eg, handing the child to the parent to hold, allowing the child to feed, or administering sedation if needed), placing the child in a knees-to-chest position (to increase systemic vascular resistance; this may be achieved by having the parent hold the baby in her arms and cradling the knees and chest together), initiating oxygen (preferably through the least noxious route for the child, for example, blow-by oxygen or use of a face mask), administering a bolus of intravenous fluids, and, finally, intravenous metoprolol (to slow the heart rate) and phenylephrine (to increase systemic vascular resistance). Some patients will require anesthesia (on the way to the operating room). Surgical management includes palliative procedures like a modified Blalock-Taussig-Thomas (mBTT) shunt to help provide consistent pulmonary blood flow in the setting of significant stenosis or atresia of the pulmonary valve. Complete repair of the heart comprises closure of the VSD, as well as resection of the RV obstruction, resulting in

Video 3. Echocardiogram in a patient with tetralogy of Fallot. A 4-chamber view of the heart is shown, with the left side of the patient on the reader's right. The ventricular septal defect (VSD) and the right ventricle (RV) hypertrophy are demonstrated. The aorta arises from the left ventricle and overrides the VSD. As the video plays, it sweeps through the heart anteriorly to show the pulmonary artery, which appears narrow. The hypertrophied muscle bundles of the RV are also seen, especially in the subvalvar region, which come very close together in systole. This creates a setup for dynamic obstruction to blood flow out of the RV.

normal saturations. Complete repair is replacing the palliative approach in many centers. Even after complete tetralogy of Fallot repair, patients require lifelong cardiology follow-up, since their pulmonary valve may become more regurgitant (depending on the surgical approach and the intervention on the valve annulus itself), which can lead to RV dilation. Patients may also develop arrhythmias because of the RV dilation and/or hypertrophy and scarring (as a result of the pulmonary valve regurgitation, as well as the primary surgery), with an increase in risk of sudden cardiac death in pediatric patients with QRS duration longer than 170 ms on the EKG. (25)(26) Cardiac magnetic resonance imaging is used to estimate the volume of the RV to determine the time for pulmonary valve replacement (catheter-based or surgical). While complete repair of tetralogy of Fallot ensures that these children have normal saturations, most patients have a prolonged QRS duration and features of right bundle block on the EKG, owing to the intervention on the interventricular septum.

Transposition of the Great Arteries

Transposition of the great arteries is the second most common cyanotic CHD, accounting for about 2% of all CHD. (1)(2)(3)(4) It is the most common cyanotic heart disease manifesting in the first week after birth. There is ventriculoarterial discordance, with the aorta arising from the RV (usually anterior to the pulmonary artery) and the pulmonary artery arising from the LV. Hence, the systemic and pulmonary circulations are in parallel, with systemic venous (deoxygenated) blood returning to the right atrium, the RV, and going out the aorta again. There is mixing of blood at the atrial level through a patent foramen ovale or an ASD or at the ventricular level through a VSD (35% to 40% of transposition of the great arteries).

Clinical Manifestations and Diagnosis. Newborns with transposition of the great arteries present with cyanosis within the first 12 hours after birth and are not responsive to oxygen or mechanical ventilation. The presence of a VSD may delay presentation. Chest radiography may show a narrow mediastinal silhouette due to the orientation of the great vessels and thymic regression, and EKG results may be normal or show RV hypertrophy. There is usually no murmur at examination; however, there may be a single S_2 with a loud aortic component due to the orientation of the great vessels.

Management. Once the diagnosis is established, these patients require reparative surgery to switch the great vessels to the appropriate ventricles, known as the *arterial switch procedure*. (27) They may need respiratory support with oxygen, mechanical ventilation, and initiation of prostaglandin

E1 until the time of surgery. Prostaglandin allows the PDA to remain open, which shunts blood from the aorta into the pulmonary circulation. This increases the amount of blood returning to the left atrium and mixing at the atrial level, as long as the foramen is open. Most patients undergo a catheter-based procedure called *balloon atrial septostomy* to help create or enlarge the ASD to allow more mixing while awaiting surgery. If cyanosis persists despite an open PDA, a fluid bolus or inotropic support may help improve mixing and systemic arterial saturations. After repair, these patients require regular cardiology follow-up for life. The first arterial switch procedure was performed in 1975, and since the patients undergoing this procedure are followed up for progressively longer periods of time, data about the long-term complications are emerging. Depending on the study, 5% to 30% of these patients have been reported to require reintervention at 25-year follow-up for a variety of reasons, most commonly regurgitation of the neoaortic valve (more than 75%), supravalvar pulmonary stenosis (more than 75%), and coronary artery disease (5% to 8%). (28)(29)

Truncus Arteriosus—Common Arterial Trunk

Truncus arteriosus accounts for 2% to 5% of CHD and manifests early in the neonatal period. (1)(2)(3)(4) It develops from lack of formation of the aorticopulmonary septum; hence, the common truncal outflow does not divide into an aorta and a main pulmonary artery. There is always an associated VSD, and there is a common truncal valve, most commonly either tricuspid or quadricuspid. About one-third of the patients with truncus arteriosus have a 22q11.2 deletion (DiGeorge or velocardiofacial syndrome).

Clinical Manifestations and Diagnosis. Patients with truncus arteriosus present within the first 48 hours after birth with symptoms of profound pulmonary overcirculation. On examination, they have a systolic murmur heard the loudest over the left sternal border, as well as a loud S_2. Multiple sounds may be heard that temporally coincide with the timing of the S_2. Their general physical examination may also demonstrate features of 22q11.2 deletion, including a small mouth (micrognathia), cleft lip and/or palate, and flat cheek bones (malar flattening). (30)

Management. Symptoms of pulmonary overcirculation may be managed with diuresis and fluid restriction. These children undergo surgical repair in the first 2 weeks after birth. (31) The truncal outflow and the truncal valve are committed to the aorta, and the branch pulmonary arteries are committed to a conduit placed from the RV to the pulmonary artery. These children should have normal saturations after repair. They continue to require close cardiology

follow-up over the years as they outgrow their RV-to–pulmonary artery conduit (only 50% are free of RV-to–pulmonary artery conduit reintervention at 10 years), and their truncal valve (now their committed "neoaortic" valve) may become regurgitant (only about 23% are free of reintervention for the truncal valve at 10 years). (31) Other than their cardiac concerns, they should also undergo 22q11.2 deletion testing by way of chromosomal microarray analysis and a genetics consultation, regardless of whether the typical facies are present. (30) Serum electrolyte testing for calcium levels should also be performed in all cases while awaiting chromosomal microarray analysis results to rule out hypocalcemia that may be seen in patients with 22q11.2 deletion.

Total and Partial Anomalous Pulmonary Venous Drainage

Abnormal return of the pulmonary veins to the systemic veins or the right atrium comprises about 1% of CHD. (1)(2)(3)(4) Anomalous pulmonary venous return is defined as *(i)* partial anomalous pulmonary venous drainage (PAPVD) when at least 1 pulmonary vein returns to the left atrium and *(ii)* total anomalous pulmonary venous drainage (TAPVD) when none of the pulmonary veins return to the left atrium. TAPVD can be further classified on the basis of the site of return of the anomalous drainage to the systemic veins—supracardiac (into the superior vena caval system), intracardiac (into the coronary sinus or right atrium), or infracardiac (into the inferior vena cava [IVC] or hepatic venous system). Blood returning from the pulmonary veins to the right side of the heart causes right atrial hypertension, with right-to-left shunting across a patent foramen ovale and/or an ASD. In TAPVD, all the venous return to the heart returns to and mixes in the right atrium before shunting across the interatrial septum or coursing across the tricuspid valve. Hence, the saturations in each of the cardiac chambers are the same.

Clinical Manifestations and Diagnosis. The number of veins returning anomalously and the degree of obstruction determine the rate and severity of the manifestation. Physiologically, TAPVD can be categorized as *(i)* obstructed, in which case the pulmonary venous return is impeded, which can lead to pulmonary venous hypertension and pulmonary edema, or *(ii)* unobstructed, in which case the anomalous drainage causes cyanosis due to mixing after return to the right side of the heart but does not cause respiratory distress in the first days after birth. The newborn with obstructed TAPVD presents with respiratory distress and cyanosis within 12 to 24 hours after birth, with a classic chest radiographic finding of *(a)* "whiteout" of the lung fields

due to backing up of blood flow prior to the site of the obstruction of pulmonary vein drainage and *(b)* a small heart on chest radiographs. The respiratory distress does not respond to oxygen and may potentially be worsened by starting prostaglandin as more blood starts shunting to the pulmonary circulation at the ductal level, to the point of pulmonary edema or lung bleeding.

PAPVD (which may or may not be associated with an ASD) typically manifests later in life in childhood or adolescence with signs similar to those of an ASD, with an ejection systolic murmur that is loudest over the pulmonic region owing to the increased blood returning to the right side of the heart. EKG shows right atrium and RV enlargement, and Doppler echocardiographic findings confirm the diagnosis.

Management. Obstructed TAPVD constitutes about one-third of the cases and is a surgical emergency. (30)(31) Children with unobstructed TAPVD can be followed up by a cardiologist for a brief time as outpatients with a definite plan for early surgical repair, since obstruction can develop over time and manifests similarly with respiratory distress. After repair of TAPVD, patients continue to be closely monitored as outpatients, since pulmonary venous obstruction can recur, with reported rates around 15%. (32)(33) Evaluation of the anastomosis site at echocardiography is especially important. Recurrent pulmonary venous obstruction manifests as respiratory distress, tachypnea, and wheezing that is not responsive to bronchodilation. These patients require chest radiography and echocardiographic evaluation if they have not undergone a recent study to assess their pulmonary venous drainage.

Stable patients with PAPVD are scheduled for elective surgical repair (if indicated) and are followed up as an outpatient in the cardiology clinic every 1 to 2 years.

### HLHS, Tricuspid Atresia, and Single-Ventricle Palliation

HLHS is the fourth most common form of cyanotic CHD, comprising severe stenosis or atresia of the mitral and aortic valves. (1)(2)(3)(4) Owing to decreased flow from the left atrium into the LV, as well as from the LV into the aorta, the LV is hypoplastic, and there is associated hypoplasia of the ascending aorta, as well (Video 5). This lesion is PDA dependent for systemic circulation, as the descending aorta continues from the PDA insertion, and the coronary arteries, innominate arteries, left common carotid arteries, and left subclavian arteries are supplied primarily by retrograde flow from the PDA. Prostaglandin infusion is required to maintain PDA patency. A corresponding lesion on the right side of the heart is a hypoplastic RV in the setting of severe tricuspid stenosis or atresia and severe

Video 4. Echocardiogram in a patient with tetralogy of Fallot. The doming, narrow pulmonary valve, and infundibular narrowing from muscle below the pulmonary valve in the right ventricular outflow tract can be seen.

pulmonary stenosis or atresia. The absence of 2 normal-sized ventricles, the absence of a normal or repairable atrioventricular connection (in the form of severe stenosis or atresia of the mitral or tricuspid valves), the absence of a repairable outflow tract in some situations (severe stenosis or atresia of the pulmonary or aortic valve), or any combination of these conditions indicates that patients should undergo single-ventricle palliation. (34) These lesions are dependent on
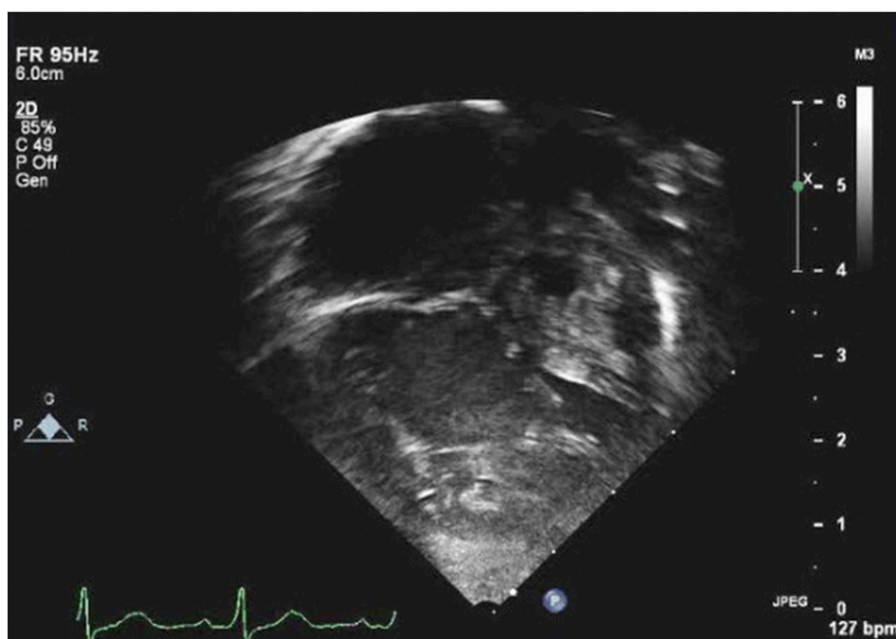
Video 5. Echocardiogram in a patient with hypoplastic left heart syndrome. A 4-chamber view of the heart is shown with the left side of the patient on the reader's right. The diminutive left-sided chambers can be seen, with the hypoplastic left atrium and the hypoplastic left ventricle, as well as compensatory dilation and hypertrophy of the right ventricle.

a PDA maintained by prostaglandin to ensure cardiac output into both systemic and pulmonary circulations.

The subtypes of these lesions have variable physiology and are beyond the scope of this article. We will proceed to discussing the stages of palliation by using HLHS as a model.

The first stage is to help stabilize both cardiac outputs, either by surgically constructing a "neoaorta" by using the main pulmonary artery tissue to supplement the ascending aorta (the Norwood procedure), creating an mBTT shunt (or RV-to–pulmonary artery conduit) to supply blood flow to the pulmonary arteries, and creating an ASD. This first stage may also be performed in a "hybrid fashion" in the cardiac catheterization laboratory and/or operating room (the details are beyond the scope of this article).

The second stage, known as the *Glenn procedure*, involves anastamosing the superior vena cava to the pulmonary arterial system to direct part of the deoxygenated venous blood directly to the lungs. This also allows take-down of the mBTT shunt (or RV-to–pulmonary artery conduit), as there is consistent pulmonary blood flow through the Glenn connection.

The final step of this palliation (the Fontan procedure) is to connect the IVC to the pulmonary arterial system as well, to allow all the deoxygenated blood to go straight to the lungs without mixing in the heart first (Fig 4). The Fontan connection may be fenestrated in some cases, creating a connection between the IVC–pulmonary arterial anastomosis and the pulmonary venous atrium. This may be especially useful as a "pop off" for blood passively returning from the IVC to the pulmonary circulation in cases of high pulmonary vascular resistance.

Clinical Manifestations and Diagnosis. The presentation of single-ventricle lesions depends on the adequacy of cardiac output. Systemic cardiac output is dependent on the ductal blood flow. In patients with HLHS, when the duct starts to close, the infant begins to have signs of poor systemic perfusion with feeble pulses, poor urine output, shock, and mottling, in addition to pulmonary overcirculation, seen as tachypnea and respiratory distress. Chest radiographs will show pulmonary vascular congestion, in addition to possible cardiomegaly due to right ventriculomegaly (as a result of the RV effectively handling systemic as well as pulmonary cardiac output).

Management. The first-stage surgical intervention is performed within the first 2 weeks after birth. Saturations after the first-stage palliation can vary from 80% up to 95%, depending on the amount of pulmonary blood flow. A continuous or pansystolic murmur is easy to appreciate in a child with an mBTT shunt, and a systolic ejection
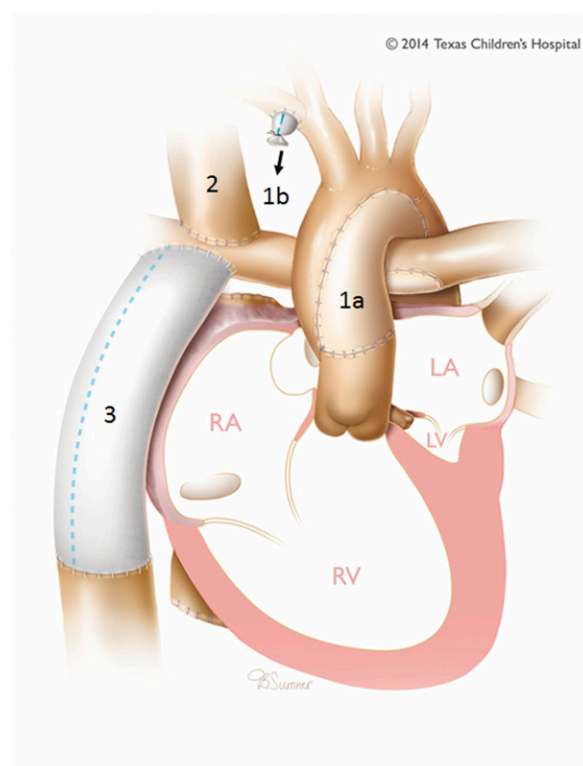

© 2014 Texas Children's Hospital

Figure 4. Diagram shows the shunts and sites of anatsamoses for single-ventricle palliation by using hypoplastic left heart syndrome as a model. *1a* indicates the reconstructed neoaorta by using the hypoplastic ascending aorta, as well as the normal main pulmonary artery, to ensure stable systemic outflow. *1b* indicates the site of the modified Blalock-Taussig-Thomas shunt to direct blood flow from the right subclavian artery into the pulmonary circulation. This is taken down at the time of the second stage of palliation—the Glenn anastomosis. This involves the creation of an anastomosis between the superior vena cava and the right pulmonary artery, *2*. The final step in this palliation is the Fontan anastomosis, *3*. This disconnects the remaining deoxygenated blood coming in through the inferior vena cava and redirects it through the Fontan conduit to the pulmonary circulation. This final step eliminates any mixing and completely separates the oxygenated and deoxygenated blood, ensuring normal saturations. LA=left atrium, LV=left ventricle, RA=right atrium, RV=right ventricle. Printed with permission from Texas Children's Hospital.

murmur is heard with an RV-to–pulmonary artery conduit. As these children grow older and bigger, the relative size of their mBTT shunt (or RV-to–pulmonary artery conduit) limits the amount of pulmonary blood flow for their body surface area, and their saturations start to drift down into the 70% range. These patients are very closely followed up until they undergo the Glenn procedure. The period between the Norwood and Glenn procedures (the "interstage period") is a particularly vulnerable time. An intercurrent stress such as respiratory syncytial viral pneumonia can be devastating.

The Glenn procedure is typically performed around 4 to 6 months of age. By this age, the pulmonary vascular resistance decreases to the physiological minimum, and the branch pulmonary arteries grow adequately, so that a stable and durable superior vena cava–pulmonary anastomosis

can be created. After the Glenn procedure, the saturations are in the 80% to 90% range, and these patients generally do not have any murmurs. In some patients, an S₁-coincident systolic murmur of atrioventricular valve regurgitation may be heard.

After the Glenn procedure, as the children grow older, their oxygen saturations trend downward, as the proportion of their cardiac output from the lower part of their body that returns through the IVC increases relative to the proportion from the upper body and the head that returns through the superior vena cava. These patients are followed up as outpatients on a semiannual basis until their final Fontan palliation at around 2 to 4 years of age, depending on trends of saturation and the prevalent clinical practice at each institution. Hence, accurate documentation of saturations at the interim pediatric office visits is extremely helpful in case a downward trend is noted. After the Fontan palliation, the patients are expected to be fully saturated in the case of a nonfenestrated Fontan procedure or around 90% to 95% saturated in the case of a fenestrated Fontan procedure. These children are still at risk of developing right-sided heart failure or systemic ventricular heart failure because they have a single ventricle driving their entire circulation. Hence, accurate assessment and documentation of history of CHF symptoms, such as tachypnea, poor exercise tolerance, decreasing appetite, diarrhea, chronic cough, or pedal edema, are important for these patients during their pediatric clinic visits, especially if the parents have a new concern.

GENERAL PRINCIPLES OF IMMUNIZATION, SUBACUTE BACTERIAL ENDOCARDITIS PROPHYLAXIS, AND POSTOPERATIVE AND CARDIAC CATHETERIZATION CARE

All patients undergoing cardiopulmonary bypass should not receive live vaccines for 2 weeks before or 6 weeks after cardiopulmonary bypass. Cytokine and immunologic markers return to normal by 2 months after surgery, and the vaccination schedule can be resumed. (35)(36) Most children should receive the influenza vaccine. Children younger than 12 months with unrepaired cyanotic CHD or those receiving diuretic therapy for CHF in acyanotic CHD should receive respiratory syncytial viral prophylaxis with palivizumab. (37) Further, for children who meet the criteria for respiratory syncytial viral prophylaxis after a palliative surgery involving cardiopulmonary bypass, a postoperative dose of 15 mg/kg palivizumab should be considered.

Children undergoing cardiac surgery or interventional catheterization procedures to repair a lesion or occlude a defect require subacute bacterial endocarditis (SBE) prophylaxis for 6 months after the procedure and indefinitely if residual defects are present around a patch and/or a device. All children with unrepaired cyanotic CHD require SBE prophylaxis. Children who have prosthetic or bioprosthetic valves or conduits implanted percutaneously or surgically should receive lifelong SBE prophylaxis. Children with a prior history of endocarditis or who have valvular heart disease in the context of heart transplantation also require SBE prophylaxis. (38)

After cardiac surgery via median thoracotomy, patients observe sternal precautions for 6 to 8 weeks. (39)(40) The precautions vary from center to center, but the general principles include (i) no prone position or "tummy time" unless they can roll and be prone voluntarily, (ii) close supervision when playing with other children and avoiding crowded places, and (iii) avoiding picking up or lifting the patient by the arms or armpits and instead trying to "scoop up" the patient. The patient can continue to be in an age- and weight-appropriate car seat after cardiac surgery. After the period of sternal restrictions, the patient is allowed to exercise as tolerated. After a cardiac catheterization, most children should be ambulating within 2 to 6 hours and should be back to full activities within 3 to 5 days.

Children with CHD are at risk for developmental delay and disability, with greater complexity of CHD lending a higher risk. (41) The American Heart Association guidelines for neurodevelopmental screening for these patients help stratify these patients into low- and high-risk groups. Neonates or infants that require open-heart surgery (cyanotic and acyanotic CHD), children with cyanotic CHD (regardless of open-heart surgery during the neonatal or infant period), and patients with CHD accompanied by certain comorbidities (such as prematurity, known genetic syndrome, prolonged hospitalization, or cardiopulmonary resuscitation) are at increased risk. These children should be directly referred to a developmental pediatrician, while all other patients with CHD should still continue regular assessment and surveillance of neurodevelopmental status with their pediatrician.

Finally, the transition of care to an adult CHD specialist is an important issue for children with CHD as they enter young adulthood. (42) Adult CHD is a growing field of practice and expertise; however, many pediatric cardiologists continue to follow up their patients during adulthood as well, especially if there is a constraint of adult providers comfortable with complex CHD. From the pediatrician's perspective, these children should continue to be followed up in the pediatrician's office until the typical age when the office transitions care to adult providers. The process of

transition to adult CHD specialists is performed by the cardiologist, along with detailed communication of the CHD-related diagnoses and past procedures. Owing to limitations in the familiarity of adult cardiac surgeons with CHD, as well as adult interventional cardiologists, most of the long-term procedures for these patients are performed by pediatric CHD specialists.

Summary

1. On the basis of clinical consensus and research observation, newborns with ductal-dependent congenital heart disease (CHD) may present with cyanosis or shock as the patent ductus arteriosus starts to close, indicating a need for prostaglandin to maintain output to the pulmonary or systemic circulation. (5)(7)(8)

2. On the basis of research evidence, as well as clinical consensus, children with acyanotic shunting lesions can present with features of congestive heart failure and require nutritional and diuretic optimization until the time of early surgical correction. (1)(2)(9)

3. On the basis of research evidence, as well as clinical consensus, children with cyanotic heart disease may undergo early corrective surgery or, in some instances, undergo palliation with a shunt (modified Blalock-Taussig-Thomas shunt) to supply pulmonary blood flow prior to their corrective surgery. (22)(32)

4. On the basis of strong clinical evidence, children who undergo single-ventricle palliation are at risk for developing complications at any stage of the palliation. (31)

5. On the basis of clinical evidence, as well as clinical consensus, although most children with corrected CHD have a normal quality of life, lifelong cardiology follow-up is needed for all of these patients to monitor the development of any long-term complications and/or sequelae. (13)(14)(20)(24)(27)(30)(39)

To view teaching slides that accompany this article, visit http://pedsinreview.aappublications.org/content/38/10/471.supplemental.

Congenital Heart Disease

Kriti Puri, MD

Hugh D. Allen, MD

Athar M. Qureshi, MD

Pediatrics in Review

American Academy of Pediatrics
DEDICATED TO THE HEALTH OF ALL CHILDREN®

References and Suggested Readings for this article are at http://pedsinreview.aappublications.org/content/38/10/471.

Additional Resources for Pediatricians

AAP Textbook of Pediatric Care, 2nd Edition
• Chapter 234: Congenital and Acquired Heart Disease - https://pediatriccare.solutions.aap.org/chapter.aspx?sectionid=135605095&bookid=1626

Point-of-Care Quick Reference
• Congenital and Acquired Heart Disease - https://pediatriccare.solutions.aap.org/content.aspx?gbosid=165499

Parent Resources from the AAP at HealthyChildren.org

• Challenges Faced by Parents of Children with Congenital Heart Disease: https://www.healthychildren.org/English/health-issues/conditions/heart/Pages/Challenges-Faced-by-Parents-of-Children-with-Congenital-Heart-Disease.aspx

• Pulse Oximetry Screening to Detect Newborn Critical Congenital Heart Disease: https://www.healthychildren.org/English/ages-stages/baby/Pages/Newborn-Pulse-Oximetry-Screening-to-Detect-Critical-Congenital-Heart-Disease.aspx

For a comprehensive library of AAP parent handouts, please go to the *Pediatric Patient Education* site at http://patiented.aap.org.

PIR Quiz

1. A 15-day-old male neonate presents with tachypnea, tachycardia, and hepatomegaly but no heart murmur. The neonate originally passed the newborn pulse oximetry heart screen. A chest radiograph shows cardiomegaly and increased pulmonary markings. There are normal oxygen saturations in all 4 extremities. Which of the following is the most likely diagnosis in this patient?
   A. Arteriovenous malformation.
   B. Inborn error of metabolism.
   C. Large ventricular septal defect.
   D. Tetralogy of Fallot.
   E. Truncus arteriosus.

2. A female newborn with Down syndrome receives a diagnosis of atrioventricular septal defect. She was evaluated by the genetics service, and her chromosome study findings are pending. Her newborn screening result is normal, with no evidence of hypothyroidism. Physical examination findings are significant for typical Down syndrome features. Heart examination demonstrates a holosystolic murmur. Which of the following is the most appropriate next step in the care of this patient?
   A. Monitor the patient for possible feeding difficulties prior to discharge.
   B. Restrict caloric intake to 75 cal/kg per day.
   C. Restrict total fluid intake to three-fourths maintenance levels.
   D. Start furosemide prophylactically to prevent congestive heart failure (CHF).
   E. Perform surgical intervention prior to discharge from the nursery if the baby is stable, with no signs of CHF.

3. A male full-term neonate born via spontaneous vaginal delivery is admitted to the nursery. The initial newborn examination performed in the first hours after birth yielded unremarkable findings, and the baby was allowed to room in with mom. At 24 hours after birth, he was noted to have mild tachypnea and poor feeding. Physical examination findings were significant for poor lower-extremity pulses when compared to the upper extremities and a harsh systolic murmur that was heard loudest over the back. Newborn oximetry screening showed a saturation of 89% in the lower extremities, compared to 94% in the upper extremities. Four extremity blood pressures showed a difference of 20 mm Hg between the upper and lower extremities. In addition to starting intravenous fluids and prostaglandin infusion, which of the following is the next best step in management of this patient?
   A. Cardiac catheterization.
   B. Chest radiography.
   C. Echocardiography.
   D. Electrocardiography.
   E. Serum renin level.

4. You are treating a female neonate in the well-baby nursery who is cyanotic 24 hours after birth. At physical examination she has micrognathia, malar flattening, and a harsh systolic murmur, with maximal intensity at the left sternal border. Which of the following is the most appropriate next step in the care of this baby?
   A. Bolus with 10 mL/kg normal saline, followed with 2 times a maintenance rate of 10% dextrose in one-half normal saline (D10%-NS/2) solution.
   B. Genetics consult, chromosome microarray, and serum calcium level.
   C. Immediate pulmonary balloon dilation.
   D. Stat administration of digoxin.
   E. When medically stable, conduit creation between the left ventricle and the pulmonary artery.

5. A male newborn was delivered via emergency cesarean delivery due to fetal heart decelerations. Apgar scores were 5 and 6 at 1 and 5 minutes, respectively. The baby was placed on 100% oxygen at birth. Eight hours after birth, he was noted to have tachypnea and respiratory distress, as well as poor systemic perfusion with feeble pulse, poor urine output, shock, and mottling. Chest radiographs showed pulmonary vascular congestion. Echocardiography showed cardiomegaly, hypoplastic left heart syndrome, tricuspid atresia, and a single ventricle. Which of the following is the most appropriate next step in the care of this patient?

    A. Balloon atrial septostomy, followed by reparative surgery to switch the great vessels.
    B. Fontan procedure, followed by a Glenn procedure and the modified Blalock-Taussig-Thomas (mBTT) shunt.
    C. mBTT shunt at 4 to 6 months of age.
    D. Normal saline bolus of 20 mL/kg.
    E. Prostaglandin infusion until staged repair is performed.

Updated Information & Services	including high resolution figures, can be found at: http://pedsinreview.aappublications.org/content/38/10/471
References	This article cites 40 articles, 17 of which you can access for free at: http://pedsinreview.aappublications.org/content/38/10/471#BIBL
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): **Medical Education** http://classic.pedsinreview.aappublications.org/cgi/collection/medical_education_sub **Journal CME** http://classic.pedsinreview.aappublications.org/cgi/collection/journal_cme **Cardiology** http://classic.pedsinreview.aappublications.org/cgi/collection/cardiology_sub **Cardiac Surgery** http://classic.pedsinreview.aappublications.org/cgi/collection/cardiac_surgery_sub **Cardiovascular Disorders** http://classic.pedsinreview.aappublications.org/cgi/collection/cardiovascular_disorders_sub
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://classic.pedsinreview.aappublications.org/site/misc/Permissions.xhtml
Reprints	Information about ordering reprints can be found online: http://classic.pedsinreview.aappublications.org/site/misc/reprints.xhtml

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™

The online version of this article, along with updated information and services, is located on the World Wide Web at:

Data Supplement at:

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™