

### ■ EVALUATION OF THE CYANOTIC NEONATE

**Cyanosis** is a physical sign characterized by blue mucous membranes, nail beds, and skin. Cyanosis results from an absolute concentration of deoxy-generated hemoglobin of at least 3.0 g/dL. Factors that influence whether cyanosis will appear include the hematocrit, which reflects the absolute concentration of hemoglobin, and the factors that affect the O<sub>2</sub> dissociation curve (pH, PCO<sub>2</sub>, temperature, level of 2,3-diphosphoglycerate, and ratio of adult to fetal hemoglobin). Cyanosis should not be confused with **acrocyanosis**, which is blueness of the extremities due to peripheral vasoconstriction noted in the first 24 to 48 hours of life. Neonates with acrocyanosis have pink mucosal membranes.

#### Differential Diagnosis

The causes of cyanosis in the newborn are of cardiac, pulmonary, neurologic, or hematologic origin. The incidence of structural heart disease is about 8 in 1000 live births, and severe congenital heart disease occurs in approximately 1 in 400 live births. Pulmonary disorders may lead to cyanosis as a result of primary lung disease, airway obstruction, or extrinsic compression of the lung. Neurologic causes of cyanosis include central nervous system dysfunction and respiratory neuromuscular dysfunction. Table 3-1 delineates the causes of cyanosis in the neonate.

#### Clinical Manifestations

##### *History and Physical Examination*

A complete birth history that includes maternal history; prenatal, perinatal, and postnatal complica-

tions; history of labor and delivery; and neonatal course should be obtained. Exactly when the child developed cyanosis is critical, because certain congenital heart defects present at birth, while others may take as long as one month to present themselves.

The initial physical examination should focus on the vital signs and cardiac and respiratory examinations, looking for evidence of right, left, or biventricular congestive heart failure and respiratory distress. Blue or dusky mucous membranes are consistent with cyanosis. Evaluate for rales, stridor, grunting, flaring, retractions, and evidence of consolidation or effusion on pulmonary examination. On cardiovascular examination, the precordial impulse is palpated, and the clinician should evaluate for systolic or diastolic murmurs, the intensity of S<sub>1</sub>, S<sub>2</sub> splitting abnormalities, and the presence of an S<sub>3</sub> or S<sub>4</sub> gallop, ejection click, opening snap, or rub. Examination of the extremities should focus on the strength and symmetry of the pulses in the upper and lower extremities, evidence of edema, and cyanosis of the nail beds. Hepatosplenomegaly may be consistent with right ventricular or biventricular heart failure.

#### Diagnostic Evaluation

The goal of the initial evaluation of the cyanotic neonate is to determine whether the cyanosis is cardiac or noncardiac in origin. An electrocardiogram (ECG), chest radiograph, and hyperoxia test should be performed. In addition, preductal and postductal oxygen saturation, and four extremity blood pressures should be documented.

A hyperoxia test should be carried out in neonates with a resting pulse oximetry reading less than 95%, visible cyanosis, or circulatory collapse. The hyperoxia test consists of obtaining a baseline right radial

■ TABLE 3-1

## Differential Diagnosis of Cyanosis in the Neonate

Cardiac	Pulmonary
Ductal-independent mixing lesions Truncus arteriosus Total anomalous pulmonary venous return without obstruction D-transposition of the great arteries <sup>a</sup>	<i>Primary lung disease</i> such as respiratory distress syndrome, meconium aspiration, pneumonia, or persistent pulmonary hypertension of the newborn
Lesions with ductal-dependent PBF Tetralogy of Fallot with pulmonary atresia <sup>b</sup> Ebstein's anomaly <sup>b</sup> Critical pulmonic stenosis Tricuspid valve atresia <sup>b</sup> with normally related great arteries <sup>b</sup> Pulmonic valve atresia with intact ventricular septum Heterotaxy <sup>b</sup>	<i>Airway obstruction</i> such as choanal atresia, vocal cord paralysis, or laryngotracheomalacia <i>Extrinsic compression of the lungs</i> such as pneumothorax, chylothorax, or hemothorax
Lesions with ductal-dependent SBF Hypoplastic left heart syndrome Interrupted aortic arch Critical coarctation of the aorta Critical aortic stenosis Tricuspid valve atresia with transposition of the great arteries <sup>b</sup>	<b>Neurologic</b> <i>CNS dysfunction</i> such as drug-induced depression of respiratory drive, postasphyxial cerebral dysfunction, or central apnea <i>Respiratory neuromuscular dysfunction</i> such as spinal muscular atrophy, infant botulism, or neonatal myasthenia gravis
	<b>Hematologic</b> Methemoglobinemia or polycythemia

<sup>a</sup>A patent ductus arteriosus may improve mixing, especially with an intact ventricular septum.

<sup>b</sup>Most forms.

PBF, pulmonary blood flow; SBF, systemic blood flow.

■ TABLE 3-2

## Interpretation of the Hyperoxia Test

	PaO <sub>2</sub> (mm Hg) at FiO <sub>2</sub> = 0.21 (% Saturation)	PaO <sub>2</sub> (mm Hg) at FiO <sub>2</sub> = 1.00 (% Saturation)	PaCO <sub>2</sub> (mm Hg)
Normal	70 (95)	>300 (100)	35
Pulmonary disease	50 (85)	>150 (100)	50
Neurologic disease	50 (85)	>150 (100)	50
Methemoglobinemia	70 (95)	>200 (100)	35
Cardiac disease			
Parallel circulation <sup>a</sup>	<40 (<75)	<50 (<85)	35
Mixing with restricted PBF <sup>b</sup>	<40 (<75)	<50 (<85)	35
Mixing without restricted PBF <sup>c</sup>	40–60 (75–93)	<150 (<100)	35

<sup>a</sup>D-Transposition of the great arteries with intact ventricular septum, D-transposition of the great arteries with ventricular septal defect.

<sup>b</sup>Tricuspid atresia with pulmonary stenosis or atresia, pulmonary atresia or critical pulmonary stenosis with intact ventricular septum, tetralogy of Fallot, or Ebstein's anomaly.

<sup>c</sup>Truncus arteriosus; total anomalous pulmonary venous return; single ventricle, hypoplastic left heart syndrome.  
PBF, pulmonary blood flow.

(preductal) arterial blood gas measurement with the child breathing room air,  $FiO_2 = 0.21$ , and then repeating the measurement with the child inspiring 100% oxygen,  $FiO_2 = 1.00$ . Interpretation of the hyperoxia test is delineated in Table 3-2. A  $PaO_2$  greater than 200 mm Hg on 100% oxygen makes congenital heart disease

very unlikely. A  $PaO_2$  less than 150 mmHg on 100% oxygen suggests a cardiac lesion characterized by complete mixing *without* restricted pulmonary blood flow. A  $PaO_2$  less than 50 mm Hg on 100% oxygen indicates a cardiac lesion with parallel circulation, or a mixing lesion *with* restricted pulmonary blood flow.

The PaO<sub>2</sub> should be measured directly via arterial puncture, though properly acquired transcutaneous oxygen monitor (TCOM) values for PaO<sub>2</sub> are also acceptable. **Pulse oximetry should not be used for interpretation of the hyperoxia test**, because a neonate given 100% inspired oxygen may have a PaO<sub>2</sub> of 80 mmHg with a pulse oximeter reading of 100% (abnormal), or a PaO<sub>2</sub> greater than 300 mmHg with a pulse oximeter reading of 100% (normal). If a cardiac cause is deemed likely, obtain an echocardiogram and a cardiology consultation.

Pulse oximetry should be documented at preductal and postductal sites to assess for differential or reverse differential cyanosis. If the preductal saturation is higher than the postductal saturation, differential cyanosis exists, which results when there are normally related great arteries and deoxygenated blood from the pulmonary circulation enters the descending aorta through a patent ductus arteriosus. Differential cyanosis is seen in persistent pulmonary hypertension of the newborn (PPHN) and in lesions with left ventricular outflow tract obstruction such as interrupted aortic arch, critical coarctation of the aorta, and critical aortic stenosis.

In rare cases of reverse differential cyanosis, the postductal saturation is higher than the preductal saturation. **This occurs only in children with transposition of the great arteries** with left ventricular outflow obstruction (i.e., critical coarctation of the aorta, interrupted aortic arch, critical aortic stenosis) or PPHN. Oxygenated blood from the pulmonary circulation enters the descending aorta through a patent ductus arteriosus.

When either the hyperoxia test or the preductal/postductal oxygen saturation measurement, or both, indicate cardiac disease, the chest radiograph and ECG may be used to delineate which cardiac structural defect is the most likely. The chest radiograph is obtained to determine the size of the heart and whether the pulmonary vascularity is increased or decreased. The ECG evaluates the heart rate, rhythm, axis, intervals, R-wave progression, and P-wave and ST/T wave morphology and helps determine if ischemia, atrial dilatation, or ventricular hypertrophy is present.

To differentiate among cyanotic congenital heart defects that present with a PaO<sub>2</sub> less than 50 mmHg on the hyperoxia test, the clinician should first examine the chest radiograph. If massive cardiac enlargement is noted, Ebstein's anomaly is the most likely diagnosis. Once massive cardiac enlargement

has been ruled out, the pulmonary vascularity becomes the focus. Increased pulmonary blood flow suggests the presence of D-transposition of the great arteries (D-TGA) with intact ventricular septum, whereas pulmonary edema is a manifestation of total anomalous pulmonary venous return with obstruction.

The remaining diagnoses (tricuspid atresia with normally related great arteries, pulmonic atresia with intact ventricular septum, critical pulmonic stenosis, and tetralogy of Fallot with or without pulmonary atresia) all produce decreased pulmonary vascularity and normal or only slightly enlarged heart size. These defects are differentiated by their axis on ECG and the presence or absence of a murmur. Tricuspid atresia with pulmonary stenosis or pulmonary atresia is noted for its superior axis, lying in the 270- to 0-degree quadrant. Critical pulmonic stenosis and pulmonary atresia with intact ventricular septum both have axes in the 0- to 90-degree quadrant. They are differentiated by the presence of the loud systolic ejection murmur heard from critical pulmonic stenosis. Similarly, tetralogy of Fallot and tetralogy of Fallot with pulmonary atresia both have axes in the 90- to 180-degree quadrant; they are distinguished from each other by the pulmonic stenosis murmur noted in tetralogy of Fallot.

## Treatment

Newborns with mixing lesions without adequate mixing (D-TGA with intact ventricular septum and restrictive patent foramen ovale) or defects that have ductal-dependent pulmonary blood flow or ductal-dependent systemic blood flow may require prostaglandin E<sub>1</sub> (PGE<sub>1</sub>) infusion to maintain patency of the ductus arteriosus until definitive surgical treatment can be accomplished. Rarely, the patient with congenital heart disease may become progressively more unstable after the institution of PGE<sub>1</sub> therapy. This clinical deterioration after institution of PGE<sub>1</sub> is an important diagnostic finding that identifies the congenital heart defect as one that has obstructed blood flow out of the pulmonary veins or left atrium. Lesions that have impaired blood flow from the left atrium include hypoplastic left heart syndrome with restrictive or intact foramen ovale, other variants of mitral atresia with restrictive foramen ovale, transposition of the great arteries with an intact ventricular septum and restrictive foramen ovale, and total anomalous pulmonary venous return with obstruction.

## KEY POINTS

1. The absolute concentration of deoxygenated hemoglobin, and not the ratio of oxygenated to deoxygenated hemoglobin, determines the presence of cyanosis.
2. Once cyanosis has been identified, stabilize the infant, quickly initiate the preliminary workup (chest radiograph, electrocardiogram, and hyperoxia test) and define whether the lesion is cardiac or noncardiac in origin.
3. If a cardiac lesion is suspected, determine whether the defect is ductal dependent for systemic or pulmonary circulation or a ductal-independent mixing lesion.
4. Once the infant is stabilized, obtain an emergent cardiology consultation, an echocardiogram, and, if indicated, begin PGE<sub>1</sub> therapy in preparation for surgical palliation or correction.

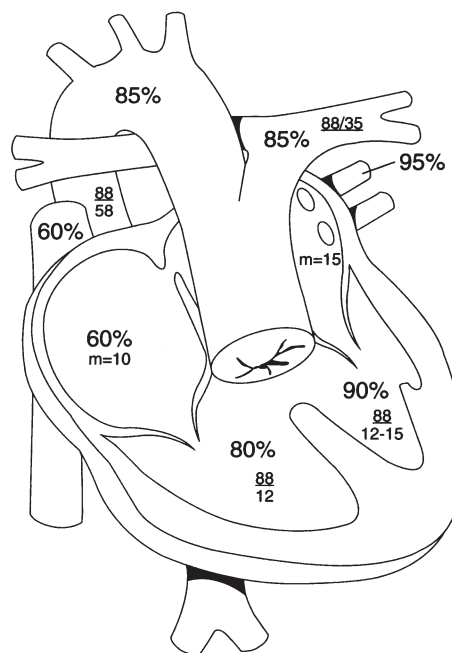
## ■ CYANOTIC CONGENITAL HEART DISEASE: DUCTAL-INDEPENDENT MIXING LESIONS

### Truncus Arteriosus

Truncus arteriosus (Figure 3-1) is a rare form of cyanotic congenital heart disease that consists of a single arterial vessel arising from the base of the heart from which arise the coronary, systemic, and pulmonary arteries. In this disorder, there is complete mixing of systemic and pulmonary venous blood in the truncus. This lesion, along with other conotruncal anomalies (tetralogy of Fallot, interrupted aortic arch, VSD, isolated arch anomalies, and vascular ring), is associated with microdeletion of chromosome 22 (22q11 deletion).

#### Clinical Manifestations

Moderate cyanosis is present at birth, and congestive heart failure develops in a matter of weeks as the pulmonary vascular resistance falls and shunting across the ventricular septal defect begins. On examination, a systolic ejection murmur is heard at the left sternal border, a widened pulse pressure is present, and bounding arterial pulses are palpated. There is a single loud second heart sound on cardiovascular exam. Seventy percent of children with truncus arte-



**Figure 3-1 • Truncus arteriosus (with right aortic arch).** Typical anatomic and hemodynamic findings include: (a) a single artery arises from the conotruncus giving rise to coronary arteries (not shown), pulmonary arteries, and aortic vessels; (b) abnormal truncal valve (quadricuspid shown) with stenosis and/or regurgitation is common; (c) right-sided aortic arch (occurs in approximately 30% of cases); (d) large conoventricular ventricular septal defect; (e) pulmonary artery hypertension with a large left-to-right shunt (note superior vena caval oxygen saturation of 60% and pulmonary artery oxygen saturation of 85%); (f) complete mixing (of the systemic and pulmonary venous return) occurs at the great vessel level.

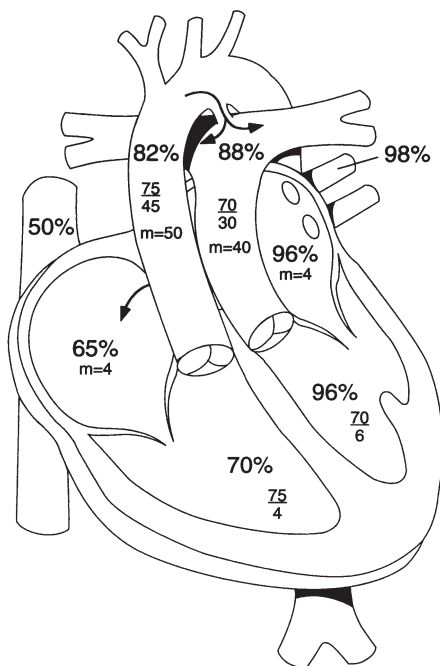
Cloherty JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

riosus have biventricular hypertrophy on ECG. On chest radiograph, marked cardiomegaly, increased pulmonary vascularity, and right aortic arch may be seen. DiGeorge's syndrome related to the 22q11 microdeletion may result in hypocalcemia.

#### Treatment

At most centers neonatal surgical repair is performed. Surgical repair involves closing the ventricular septal defect so the oxygenated blood in the left ventricle is baffled through the VSD to the truncal valve and a conduit is interposed between the right ventricle and pulmonary arteries, which are disconnected from the truncal vessel.





**Figure 3-2** • Transposition of the great arteries with an intact ventricular septum, a large patent ductus arteriosus (on PGE<sub>1</sub>) and atrial septal defect (status post balloon atrial septostomy). Note the following: (a) The aorta arises from the anatomic right ventricle, and the pulmonary artery from the anatomic left ventricle; (b) “transposition physiology,” with a higher oxygen saturation in the pulmonary artery than in the aorta; (c) “mixing” between the parallel circulations (see text) at the atrial (after balloon atrial septostomy) and ductal levels; (d) shunting from the left atrium to the right atrium via the atrial septal defect (not shown) with equalization of atrial pressures; (e) shunting from the aorta to the pulmonary artery via the ductus arteriosus; (f) pulmonary hypertension due to a large ductus arteriosus.

Cloherly JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

## D-Transposition of the Great Arteries

D-Transposition of the great arteries (Figure 3-2) accounts for 5% of congenital heart defects and is the most common form of cyanotic congenital heart disease presenting in the neonatal period. In this defect, the aorta arises anteriorly from the right ventricle, and the pulmonary artery rises posteriorly from the left ventricle. There are three basic variants: D-TGA with intact ventricular septum (60%), D-TGA with ventricular septal defect (20%), and D-TGA with ventricular septal defect and pulmonic stenosis (20%).

In this defect, the pulmonary and systemic circuits are parallel rather than in series. The systemic circuit (deoxygenated blood) is recirculated through the body, whereas the pulmonary circuit (oxygenated blood) recirculates through the lungs. A mixing lesion such as an atrial septal defect, ventricular septal defect, and/or patent ductus arteriosus that allows mixing of the systemic and pulmonary circulations is necessary for survival.

## Clinical Manifestations

Cyanosis is present from birth, the degree varying with the associated mixing lesions. In the absence of mixing lesions, there is pronounced cyanosis, right ventricular heave, and a single loud S<sub>2</sub> on examination. The presence of a systolic murmur indicates the presence of a VSD or pulmonic stenosis. The ECG is normal in the newborn; however, right-axis deviation and right ventricular hypertrophy are eventually seen. The chest radiograph reveals increased pulmonary vascular markings in D-transposition with or without ventricular septal defect, but if pulmonic stenosis is critical, decreased pulmonary vascular markings may be present. Cardiomegaly with “egg-shaped silhouette” is often seen on chest radiograph.

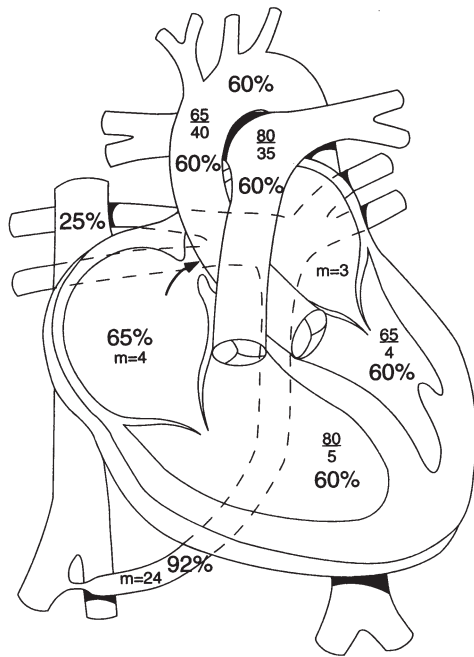
## Treatment

Initial management may include PGE<sub>1</sub> to keep the patent ductus arteriosus open and increase aorta (deoxygenated) to pulmonary artery (oxygenated) shunting. If needed, the Rashkind balloon atrial septostomy can be utilized to improve atrial mixing and relieve severe hypoxia. Surgical repair, utilizing the arterial switch procedure, is generally performed during the first week of life.

## Total Anomalous Pulmonary Venous Connection

Total anomalous pulmonary venous connection (TAPVC) (Figure 3-3) is a rare lesion in which the pulmonary venous return is directed to the right atrium either directly or indirectly through venous channels. There are four variants:

- **Supracardiac** (50% of cases): Blood drains via a vertical vein into the innominate vein or into the superior vena cava
- **Cardiac** (20% of cases): Blood drains into the coronary sinus or directly into the right atrium



**Figure 3-3 •** Infradiaphragmatic total anomalous pulmonary venous connection. Note the following: (a) pulmonary venous confluence does not connect with the left atrium, but descends to connect with the portal circulation below the diaphragm. This connection is frequently severely obstructed as shown; (b) obstruction to pulmonary venous return results in significantly elevated pulmonary venous pressures, decreased pulmonary blood flow, pulmonary edema and pulmonary venous desaturation (92%); (c) systemic to suprasystemic pressure in the pulmonary artery (in the absence of a patent ductus arteriosus, pulmonary artery pressures may exceed systemic pressures when severe pulmonary venous obstruction is present); (d) all systemic blood flow must be derived via a right-to-left shunt at the foramen ovale; (e) nearly equal oxygen saturations in all chambers of the heart (i.e., complete mixing at right atrial level), with severe hypoxemia (systemic oxygen saturation 60%) and low cardiac output (mixed venous oxygen saturation 25%).

Cloherty JP, Stark AR. Manual of Neonatal Care, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

- **Infradiaphragmatic** (20% of cases): Blood drains via a vertical vein into the portal or hepatic veins
- **Mixed** (10% of cases): Blood returns to the heart via a combination of the above routes

TAPVC can occur with or without obstruction. Obstruction occurs when the anomalous vein enters a vessel at an acute angle. The presence or absence of obstruction determines whether there is pulmonary venous hypertension and severe

cyanosis or increased pulmonary blood flow and mild cyanosis.

### Clinical Manifestations

Without obstruction, clinical findings are similar to those of an atrial septal defect. There is an active precordium with a right ventricular heave, a wide and fixed split  $S_2$  with a loud pulmonary component, and a systolic ejection murmur at the left upper sternal border. On chest radiograph, cardiomegaly is noted with increased pulmonary vascularity. On ECG, right-axis deviation and right ventricular hypertrophy are seen. A neonate with TAPVC with obstruction presents extremely cyanotic, tachypneic, and dyspneic. Examination reveals a right ventricular heave, a narrowly split  $S_2$ , and a ventricular gallop ( $S_3$ ).

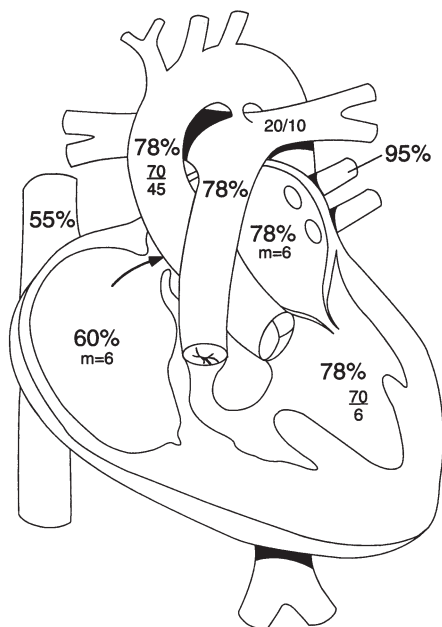
### Treatment

In TAPVC without obstruction, treatment of congestive heart failure is needed initially, and surgical redirection of aberrant vessels into the left atrium is necessary in the first month of life. In TAPVC with obstruction, the neonate should be taken to surgery emergently for correction.  $PGE_1$  should not be given because the patent ductus arteriosus adds more blood volume to an already flooded pulmonary circuit.

## ■ CYANOTIC CONGENITAL HEART DISEASE: LESIONS WITH DUCTAL-DEPENDENT PULMONARY BLOOD FLOW

### Tricuspid Atresia

Tricuspid atresia with normally related great arteries (Figure 3-4) is a rare defect that consists of complete absence of right atrioventricular connection, which leads to severe hypoplasia or absence of the right ventricle. Ninety percent of cases of tricuspid atresia have an associated ventricular septal defect. In children with tricuspid atresia with normally related great arteries, the ventricular septal defect allows blood to pass from the left ventricle to the right ventricular outflow and pulmonary arteries. The vast majority of patients with tricuspid atresia with normally related great arteries also have pulmonary stenosis. In tricuspid atresia the systemic venous



**Figure 3-4** • Tricuspid atresia with normally related great arteries and a small patent ductus arteriosus. Typical anatomic and hemodynamic findings include: (a) atresia of the tricuspid valve; (b) hypoplasia of the right ventricle; (c) restriction to pulmonary blood flow at two levels: a (usually) small ventricular septal defect and a stenotic pulmonary valve; (d) all systemic venous return must pass through the patent foramen ovale to reach the left ventricle; (e) complete mixing at the left atrial level, with systemic oxygen saturation of 78% (in  $FiO_2$  of 0:21), suggesting balanced systemic and pulmonary blood flow.

Cloherly JP, Stark AR. Manual of Neonatal Care, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

return is shunted from the right atrium to the left atrium through the patent foramen ovale or an atrial septal defect, and the left atrium and left ventricle handle both systemic and pulmonary venous return. Oxygenated and deoxygenated blood is mixed in the left atrium. Cyanosis is severe in the neonatal period and is proportionally related to the amount of pulmonary blood flow. In 30% of cases, there is transposition of the great arteries, which results in blood passing from the left ventricle through the ventricular septal defect to the right ventricular outflow and the ascending aorta. Tricuspid atresia with transposition of the great arteries is often associated with coarctation of the aorta or aortic arch hypoplasia. Unlike tricuspid atresia with normally related great arteries it is a cyanotic lesion with ductal dependent systemic blood flow.

### Clinical Manifestations

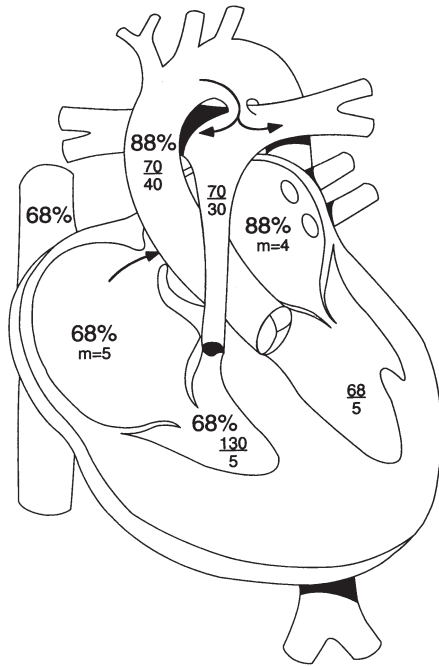
Neonates with tricuspid atresia with normally related great arteries present with progressive cyanosis, poor feeding, and tachypnea over the first 2 weeks of life. On cardiac examination, the harsh holosystolic murmur of a ventricular septal defect at the left lower sternal border and the continuous murmur of a patent ductus arteriosus may be heard. On ECG, there is a superior axis and left ventricular hypertrophy. Findings on chest radiograph include normal heart size and decreased pulmonary vascular markings.

### Treatment

A child with tricuspid atresia with normally related great arteries should have  $PGE_1$  started to maintain pulmonary flow, and a balloon atrial septostomy should be performed if the atrial defect is not adequate. Surgical management for tricuspid atresia involves placing a modified Blalock-Taussig shunt to maintain pulmonary blood flow. The modified Blalock-Taussig shunt is a Gortex conduit placed between the subclavian artery and the pulmonary artery. Ultimately, a cavopulmonary anastomosis (hemi-Fontan or bidirectional Glenn) is performed to provide stable pulmonary blood flow. In most centers, a modified Fontan procedure is performed to redirect the inferior vena cava and hepatic vein flow into the pulmonary circulation.

### Pulmonic Atresia with Intact Ventricular Septum

Pulmonic atresia with intact ventricular septum (Figure 3-5) is a rare defect consisting of pulmonary valvular and infundibular atresia and varying degrees of right ventricular and tricuspid valve hypoplasia. In this disorder, there is an obligate atrial shunt from right to left, and pulmonary blood flow is dependent on a patent ductus arteriosus. Since there is no pulmonary outflow, the right ventricle is hypertensive and there is often moderate to severe tricuspid regurgitation. Pulmonic atresia with intact ventricular septum may also be associated with coronary artery–myocardial sinusoid communication. The coronary arteries may be quite abnormal, with areas of stenosis or complete atresia. In some cases, coronary perfusion may be dependent on the hypertensive right ventricle. If the coronaries are right ventricle (RV) dependent, any palliative procedure that decompresses the right ventricle may lead to myocardial infarction and death.



**Figure 3-5** • Pulmonary atresia with intact ventricular septum (PA/IVS) in a neonate with a nonrestrictive patent ductus arteriosus while receiving PGE<sub>1</sub>. Typical anatomic and hemodynamic findings include: (a) hypertrophied, hypoplastic right ventricle; (b) hypoplastic tricuspid valve and pulmonary annulus; (c) atresia of the pulmonary valve with no antegrade flow; (d) suprasystemic right ventricular pressure; (e) pulmonary blood flow via the patent ductus; (f) right-to-left shunt at the atrial level with systemic desaturation. Many patients have significant coronary abnormalities with sinusoidal or fistulous connections to the hypertensive right ventricle or significant coronary stenosis (not shown).

Cloherty JP, Stark AR. Manual of Neonatal Care, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

### Clinical Manifestations

Neonates present at birth extremely cyanotic and tachypneic. Cardiac examination reveals a tricuspid regurgitation murmur in the left lower sternal border and the continuous murmur of a patent ductus arteriosus. On ECG, left ventricular hypertrophy and a leftward axis are seen. On chest radiograph, decreased pulmonary markings and left ventricular hypertrophy are seen.

### Treatment

PGE<sub>1</sub> should be started to ensure pulmonary blood flow initially. Prior to any surgery to provide more stable pulmonary flow, a cardiac catheterization must

be performed to assess the coronary arteries. If the coronary circulation is not RV dependent, then a right ventricle to pulmonary artery conduit or pulmonary valvotomy is performed to provide antegrade pulmonary blood flow. A modified Blalock-Taussig shunt is also typically performed to augment pulmonary blood flow further. Depending on the growth of the right ventricle and tricuspid valve, a single ventricle, one and a half ventricle, or two ventricle repair may be possible. If the coronary circulation is RV dependent, the RV is not decompressed and a modified Blalock-Taussig shunt is performed. After modified Blalock-Taussig shunt placement, patients with a right ventricle dependent coronary circulation are either listed for heart transplantation or staged to a Fontan palliation.

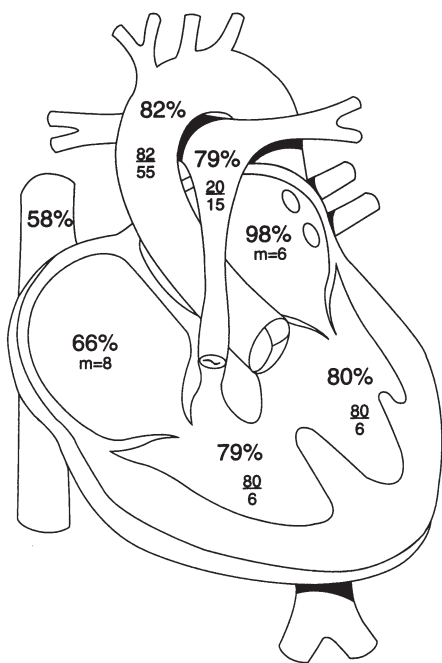
### Tetralogy of Fallot

Tetralogy of Fallot (Figure 3-6) is the third most prevalent cyanotic congenital heart lesion during the neonatal period and after the third week of life becomes the leading cause of cyanosis due to congenital heart disease in childhood. The four defects Fallot noted include an anterior malalignment ventricular septal defect, right ventricular outflow tract obstruction (50% infundibular stenosis, 20% pulmonary valve stenosis, and 30% infundibular stenosis and pulmonary valve stenosis), right ventricular hypertrophy, and an “overriding” large ascending aorta.

### Clinical Manifestations

Neonates with tetralogy of Fallot are cyanotic because of right-to-left shunting across the ventricular septal defect and decreased pulmonary flow. Shunting occurs when the combination of the pulmonary vascular resistance and the resistance created by the right ventricular outflow tract obstruction exceed the peripheral vascular resistance. The degree of cyanosis is proportional to the severity of the right ventricular outflow tract obstruction. Blood shunted from the aorta to the pulmonary artery through the patent ductus arteriosus provides additional pulmonary blood flow. Neonates present with cyanosis of varying severity and may have characteristic periodic episodic cyanosis and agitation. These episodes of cyanosis are known as “tet spells.” Tet spells are caused by an increase in right ventricular outflow tract resistance, leading to an increase in the right-to-left shunt. Such spells may last minutes to hours, may





**Figure 3-6 • Tetralogy of Fallot.** Typical anatomic and hemodynamic findings include: (a) an anteriorly displaced infundibular septum, resulting in subpulmonary stenosis, a large ventricular septal defect and overriding of the aorta over the muscular septum; (b) hypoplasia of the pulmonary valve, main and branch pulmonary arteries; (c) equal right and left ventricular pressures; (d) a right-to-left shunt at ventricular level, with a systemic oxygen saturation of 82%.

Cloherty JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

resolve spontaneously, or may lead to progressive hypoxia, acidosis, and death. On cardiac examination, a right ventricular heave is often felt and a loud systolic ejection murmur is heard in the left upper sternal border due to right ventricular outflow tract obstruction. The ECG reveals right atrial dilation and right ventricular hypertrophy, whereas the chest radiograph shows normal heart size with decreased pulmonary vascular markings. Twenty-five percent of children with tetralogy of Fallot have a right-sided aortic arch.

### Treatment

The treatment of tet spells is aimed at diminishing right-to-left shunting by increasing systemic vascular resistance and decreasing pulmonary vascular resistance. Tet spells may be treated with supplemental oxygen, vagal maneuvers, morphine sulfate, vasocon-

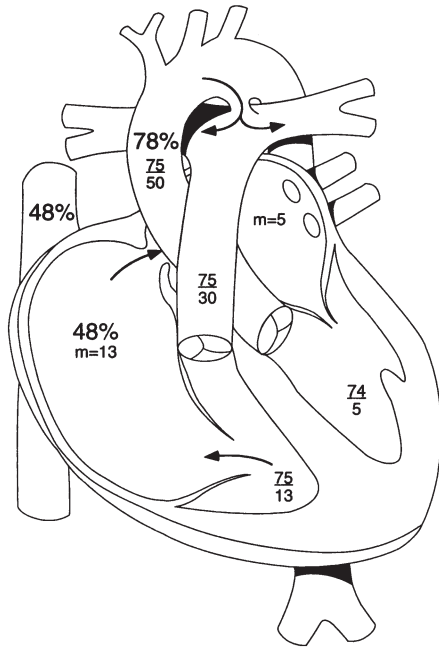
strictors, beta-blockers, and volume administration. Holding the infant over the shoulder and placing the child in a knee-chest position decreases preload and increases systemic vascular resistance. Morphine sulfate suppresses the respiratory center, stops hyperpnea, and dilates the pulmonary arteries. Vasoconstrictors raise the systemic vascular resistance, whereas beta-blockers are thought to minimize infundibular spasm. Volume is added to increase the systemic blood pressure, which minimizes right-to-left shunting. Metabolic acidosis must be corrected, because it increases pulmonary vascular resistance and thereby promotes right-to-left shunting across the ventricular septal defect. In most institutions, surgical repair is performed during the first 3 to 6 months of life, or after the first hypercyanotic episode (tet spell).

### Ebstein's Anomaly

Ebstein's anomaly (Figure 3-7) is an extremely rare anomaly in which the septal leaflet of the tricuspid valve is displaced into the right ventricular cavity and the anterior leaflet of the tricuspid valve is sail-like and redundant. This results in a portion of the right ventricle being incorporated into the right atrium. Functional hypoplasia of the right ventricle results, as well as tricuspid regurgitation or stenosis or both. A patent foramen ovale is present in 80% of neonates with the anomaly, and there is a right-to-left shunt at the atrial level. The right atrium is massively dilated, which may result in supraventricular tachycardia. Wolff-Parkinson-White (WPW) syndrome is associated with Ebstein's anomaly. In severe cases of Ebstein's anomaly, the majority of the pulmonary blood flow comes from the patent ductus arteriosus and not the right ventricle.

### Clinical Manifestations

Neonates with the severe form of the disease present with cyanosis and congestive heart failure in the first few days of life. The cardiac examination reveals a widely fixed split S<sub>2</sub>, and a tricuspid regurgitant murmur is heard at the left lower sternal border. The ECG reveals a right bundle branch block with right atrial enlargement. Delta waves due to WPW syndrome and supraventricular tachycardia may manifest themselves. Chest radiograph reveals extreme cardiomegaly with notable right atrial enlargement and decreased pulmonary vascular markings.



**Figure 3-7 •** Ebstein's anomaly (with large nonrestrictive ductus arteriosus). Typical anatomic and hemodynamic findings include: (a) inferior displacement of the tricuspid valve into the right ventricle, which may also cause subpulmonary obstruction; (b) diminutive muscular right ventricle; (c) marked enlargement of the right atrium due to "atrialized" portion of right ventricle as well as tricuspid regurgitation; (d) right-to-left shunting at the atrial level (note arterial oxygen saturation of 78%); (e) a left-to-right shunt and pulmonary hypertension secondary to a large patent ductus arteriosus supplying the pulmonary blood flow; (f) low cardiac output (note low mixed venous oxygen saturation in the superior vena cava).

Cloherty JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

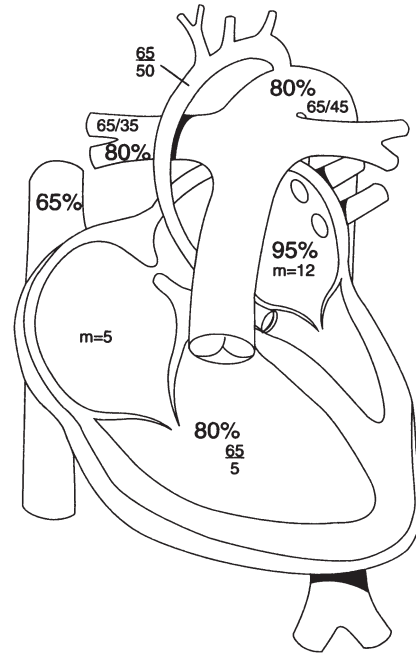
### Treatment

PGE<sub>1</sub> may help increase pulmonary blood flow. Congestive heart failure may be treated with digoxin and diuretics. Propranolol may be used to suppress supraventricular tachycardia if present. Surgical therapy to repair the abnormal tricuspid valve has had poor results.

## ■ CYANOTIC CONGENITAL HEART DISEASE: LESIONS WITH DUCTAL-DEPENDENT SYSTEMIC BLOOD FLOW

### Hypoplastic Left Heart Syndrome

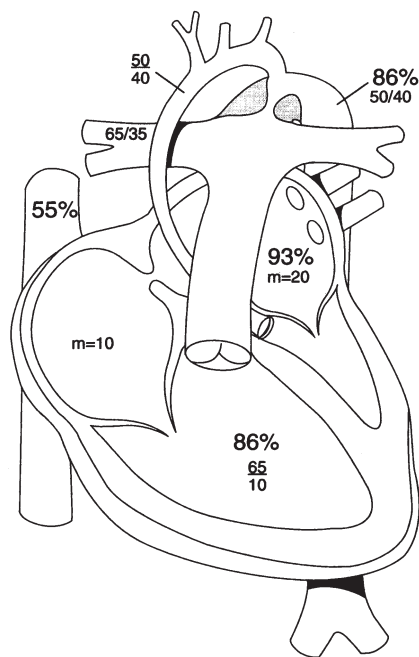
Hypoplastic left heart syndrome (HLHS) (Figures 3-8 and 3-9) is the second most common congenital



**Figure 3-8 •** Hypoplastic left heart syndrome in a 24-hour-old patient with falling pulmonary vascular resistance and a nonrestrictive ductus arteriosus. Typical anatomic and hemodynamic findings include: (a) atresia or hypoplasia of the left ventricle, mitral and aortic valves; (b) a diminutive ascending aorta and transverse aortic arch, usually with an associated coarctation; (c) coronary blood flow is usually retrograde from the ductus arteriosus through the tiny ascending aorta; (d) systemic arterial oxygen saturation (in FiO<sub>2</sub> of 0.21) of 80%, reflecting relatively balanced systemic and pulmonary blood flows—the pulmonary artery and aortic saturations are equal (see text); (e) pulmonary hypertension secondary to the nonrestrictive ductus arteriosus; (f) minimal left atrial hypertension; (g) normal systemic cardiac output (note superior vena cava oxygen saturation of 65%) and blood pressure (65/45).

Cloherty JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

cardiac lesion presenting in the first week of life and the most common cause of death from congenital heart disease in the first month of life. In this syndrome, there is hypoplasia of the left ventricle, aortic valve stenosis or atresia, mitral valve stenosis or atresia, and hypoplasia of the ascending aorta with discrete coarctation of the aorta. These lesions reduce or eliminate blood flow through the left side of the heart, causing an obligatory left-to-right shunt at the atrial level and a right-to-left shunt at the ductus arteriosus. Systemic flow is completely ductal dependent, and coronary perfusion is retrograde when aortic atresia or critical aortic stenosis is present.



**Figure 3-9** • Acute circulatory collapse following constriction of the ductus arteriosus in hypoplastic left heart syndrome. These neonates are typically in shock with poor perfusion, tachycardia, acidosis and in respiratory distress. Note (a) the low cardiac output (as evidenced by the low mixed venous oxygen saturation in the superior vena cava of 55%); (b) narrow pulse pressure; (c) elevated atrial and ventricular end-diastolic pressure—elevated left atrial pressure may cause pulmonary edema (note left atrial saturation of 93%); (d) significantly increased pulmonary blood flow, as reflected in an arterial oxygen saturation (in  $\text{FiO}_2$  of 0.21) of 86%.

Cloherly JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

### Clinical Manifestations

As the ductus closes, neonates with HLHS have severely diminished systemic blood flow and present in shock. They manifest signs of congestive heart failure with moderate cyanosis, tachycardia, tachypnea, pulmonary rales (from pulmonary edema), and hepatomegaly. Poor or absent peripheral pulses and vasoconstricted extremities are characteristic. The cardiac examination reveals an  $S_3$  and a loud single  $S_2$ . The ECG shows decreased R wave progression across the precordium. The chest radiograph reveals pulmonary edema.

### Treatment

$\text{PGE}_1$  should be started to maintain ductal-dependent systemic blood flow. No corrective

surgery is available. The stage I (or Norwood) palliation, which is performed in the first week of life, allows the majority of neonates to survive infancy. The stage I procedure involves amalgamation of the pulmonary artery and aorta to provide unobstructed systemic blood flow, atrial septectomy, and modified Blalock-Taussig shunt to provide restrictive pulmonary blood flow. After the stage I procedure, a cavopulmonary anastomosis is performed at 4 to 6 months of age and a modified Fontan procedure is generally performed at 2 to 4 years of age. Some centers do not perform the stage I palliation and proceed directly to heart transplantation.

### Interrupted Aortic Arch

There are three types of interrupted aortic arch (Figure 3-10): Type A is interruption beyond the left subclavian artery, type B is interruption between the left subclavian and left common carotid arteries, and type C is interruption between the left common carotid and the brachiocephalic arteries. In this anomaly, systemic blood flow is dependent on patency of the ductus arteriosus, which shunts blood from the pulmonary artery to the aorta. Interrupted aortic arch is often associated with DiGeorge's syndrome, due to the 22q11 microdeletion.

### Clinical Manifestations

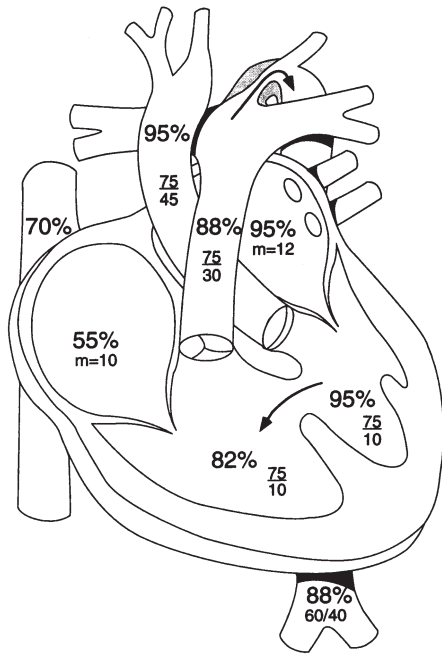
Pulmonary edema occurs almost immediately. The clinical presentation is similar to that of critical coarctation of the aorta.

### Treatment

$\text{PGE}_1$  therapy should begin immediately to maintain systemic blood flow via the right-to-left shunt through the patent ductus arteriosus. Emergent surgery is necessary to reanastomose the aortic segments.

## ■ ACYANOTIC CONGENITAL HEART DISEASE

Acyanotic cardiac defects that result in increased pulmonary blood flow include atrial septal defect, ventricular septal defect, patent ductus arteriosus, and common atrioventricular canal. Acyanotic lesions that result in pulmonary venous hypertension include coarctation of the aorta and aortic valve



**Figure 3-10** • Interrupted aortic arch with restrictive patent ductus arteriosus. Typical anatomic and hemodynamic findings include: (a) atresia of a segment of the aortic arch between the left subclavian artery and the left common carotid (the most common type of interrupted aortic arch—“type B”); (b) a posterior malalignment of the conal septum resulting in a large ventricular septal defect and a narrow subaortic area; (c) a bicuspid aortic valve occurs in 60% of patients; (d) systemic pressure in the right ventricle and pulmonary artery (due to the large, nonrestrictive ventricular septal defect); (e) increased oxygen saturation in the pulmonary artery due to left-to-right shunting at the ventricular level; (f) “differential cyanosis” with a lower oxygen saturation in the descending aorta due to a right-to-left shunt at the patent ductus. Note the lower blood pressure in the descending aorta due to constriction of the ductus; opening the ductus with PGE<sub>1</sub> results in equal upper and lower extremity blood pressures, but continued “differential cyanosis.”

Cloherty JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

stenosis. The acyanotic structural anomaly that results in relatively normal pulmonary blood flow is pulmonary valve stenosis.

## Atrial Septal Defects

Atrial septal defects account for 8% of congenital heart disease. There are three types of atrial septal defects:

- Ostium secundum defect, seen in the midportion of the atrial septum
- Ostium primum defect, located in the low atrial septum
- Sinus venosus defect, found at the junction of the right atrium and the superior or inferior vena cava

The degree of atrial shunting is dependent on the size of the ASD and the relative compliance of the ventricles in diastole. Since right ventricular diastolic compliance is usually greater than left ventricular diastolic compliance, left-to-right shunting occurs at the atrial level, thus increasing flow across the tricuspid and pulmonary valves and increasing pulmonary blood flow.

## Clinical Manifestations

Atrial septal defects are usually not associated with symptoms, although there may be a history of slow weight gain and frequent lower respiratory infections. On physical examination, the precordium is hyperdynamic, and a right ventricular heave is often present. A systolic ejection murmur in the pulmonic area and a mid-diastolic rumble in the lower right sternal border reflect the increased flow across the pulmonary and tricuspid valves. S<sub>2</sub> is widely and constantly split. On chest radiograph, the heart and main pulmonary artery are enlarged and pulmonary vascularity is increased. The ECG often shows right ventricular hypertrophy or right ventricular conduction delay. Right-axis deviation is often seen in secundum defects, whereas primum defects have characteristic extreme left-axis deviation. The amount of right ventricle and left atrium enlargement is directly proportional to the size of the left-to-right shunt. On echocardiogram, the defect can be visualized, and Doppler flow mapping demonstrates the direction of flow.

## Treatment

Spontaneous closure of small secundum ASDs is likely to occur in the majority of cases in the first year of life. Ostium primum and sinus venosus ASDs do not close spontaneously and must be addressed surgically. The symptomatic child with an ASD should have the defect closed as soon as possible. The timing of ASD repair in the asymptomatic infant or child is more controversial. In general, the defect should be repaired when circulatory arrest is not needed and when the likelihood of needing a blood transfusion



is low. After 6 months of age, both of these criteria are generally met. Subacute bacterial endocarditis prophylaxis is not recommended for secundum atrial septal defects but is indicated in primum and sinus venosus atrial septal defects.

## Ventricular Septal Defects

The ventricular septal defects are the most common congenital heart defect, accounting for 25% of all congenital cardiac lesions. The five types of ventricular septal defects are as follows:

- Muscular
- Inlet
- Conoseptal hypoplasia
- Conoventricular
- Malalignment

Muscular ventricular septal defects occur in the muscular portion of the septum and may be single or multiple and located in the posterior, apical, or anterior portion of the septum. The inlet VSD is an endocardial cushion defect and occurs in the inlet portion of the septum beneath the septal leaflet of the tricuspid valve. Conoseptal hypoplasia VSDs are positioned in the outflow tract of the right ventricle beneath the pulmonary valve. The conoventricular VSD occurs in the membranous portion of the ventricular septum. Malalignment VSDs result from malalignment of the infundibular septum.

When the VSD is non-restrictive, pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR) determine shunt flow. When the PVR is less than the SVR the shunt flow is left to right. Large defects eventually result in pulmonary hypertension, whereas small defects do not change PVR. The amount of left ventricular and left atrial dilatation is directly proportional to the size of the left-to-right shunt. Right ventricular hypertrophy occurs when pulmonary vascular resistance increases. If left untreated, the large VSD may result in elevated pulmonary arterial pressures and may lead to pulmonary vascular obstructive disease, and Eisenmenger's syndrome. In some cases of Eisenmenger's syndrome, the VSD shunt may reverse right to left. When the VSD is restrictive, shunt flow is left to right from the high pressure LV to the lower pressure RV.

### Clinical Manifestations

Clinical symptoms are related to the size of the shunt. A small shunt produces no symptoms, whereas

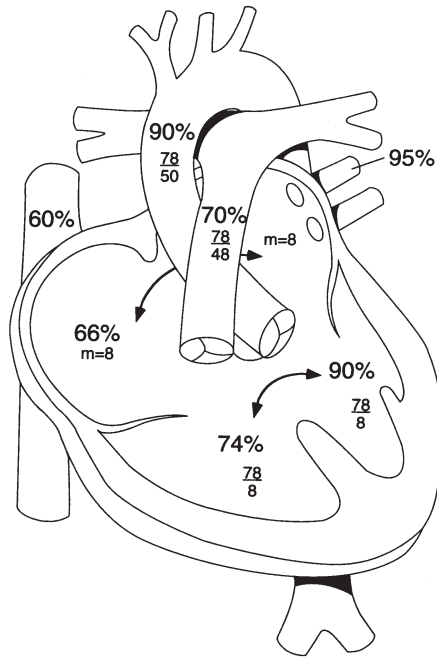
a large shunt without elevated pulmonary arterial pressures gives rise to congestive heart failure and growth failure. The patient with a large VSD with Eisenmenger physiology presents with shortness of breath, dyspnea on exertion, chest pain, and cyanosis. The smaller the defect, the louder the holosystolic murmur. As pulmonary vascular resistance increases, the holosystolic murmur shortens and the pulmonary component of  $S_2$  increases in intensity. In the presence of pulmonary vascular obstructive disease, a right ventricular heave, ejection click, short systolic ejection murmur, diastolic murmur of pulmonary valve insufficiency, and loud, single  $S_2$  are heard. Chest radiograph for small defects may be normal or show mild cardiomegaly and a slight increase in pulmonary vascularity, whereas in large left-to-right shunts cardiomegaly, increased pulmonary vascularity, and enlargement of the left atrium and left ventricle are seen. In small defects the ECG is normal, whereas with a large VSD, left atrial, left ventricular, or biventricular hypertrophy is seen. Right ventricular hypertrophy predominates when pulmonary vascular resistance is high. On echocardiogram, the defect can be visualized, and Doppler flow mapping demonstrates the direction of flow.

### Treatment

Most small VSDs close without intervention (40% by 3 years, 75% by 10 years), whereas the treatment for large VSDs is surgical closure before pulmonary vascular changes become irreversible. Congestive heart failure is treated with digoxin, diuretics, and an angiotensin-converting enzyme (ACE) inhibitor.

## Common Atrioventricular Canal

The common atrioventricular canal defect (Figure 3-11), results from deficiency of the endocardial cushions and results in an ostium primum ASD and inlet VSD with lack of septation of the mitral and tricuspid valves (common atrioventricular valve [CAVV]). In an **incomplete atrioventricular canal** defect, the CAVV leaflets attaches directly to the top of the muscular portion of the ventricular septum. As a result, there is no communication beneath the atrioventricular valves between the right and left ventricles. The communication at the atrial level is an ostium primum ASD. The mitral valve is cleft, and there may be some degree of mitral regurgitation. In **complete common atrioventricular canal**, there is a CAVV that is not attached to the muscular ventricular septum. As a result, there is a large inlet VSD located between the



**Figure 3-11** • Complete common atrioventricular canal. Typical anatomic and hemodynamic findings include: (a) large atrial and ventricular septal defects of the endocardial cushion type; (b) single, atrioventricular valve; (c) pulmonary artery hypertension (due to large ventricular septal defect); (d) bidirectional shunting (with mild hypoxemia) at atrial and ventricular level when pulmonary vascular resistance is elevated in the initial neonatal period. With subsequent fall in pulmonary vascular resistance, the shunt becomes predominantly left-to-right with symptoms of congestive heart failure.

Cloherty JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

CAVV and the top of the muscular ventricular septum. In this defect, there is a left-to-right shunt at the atrial (ostium primum ASD) and ventricular level (inlet VSD). Because of the increase in pulmonary blood flow, pulmonary hypertension and pulmonary vascular disease may develop over time.

### Clinical Manifestations

In complete common atrioventricular canal, congestive heart failure is seen early in infancy, with tachypnea, dyspnea, and poor feeding. On examination, a blowing holosystolic murmur is heard at the left lower sternal border due to the VSD and some degree of common atrioventricular valve regurgitation, and an  $S_2$  with a widely fixed split is heard due to the atrial septal defect. The ECG reveals left-axis deviation, right atrial dilation, and left atrial dilation. The clinical manifestations of the incomplete

common atrioventricular canal are the same as those described for an ostium primum ASD.

### Treatment

Surgical repair for complete common atrioventricular canal is usually done within the first year of life. Prior to surgical repair, congestive heart failure is treated with digoxin, diuretics, and an ACE inhibitor. Complete heart block occurs in 5% of patients undergoing repair, and residual mitral insufficiency is often seen.

### Patent Ductus Arteriosus

Patency of the ductus arteriosus accounts for 10% of congenital heart disease. There is a high incidence in premature neonates and a 2:1 female predominance. The ductus arteriosus connects the aorta and the left pulmonary artery just distal to the takeoff of the left subclavian artery from the aorta. The direction of flow through a large patent ductus arteriosus depends on the relative resistances in the pulmonary and systemic circuits. In the non-restrictive patent ductus arteriosus, as long as the systemic vascular resistance is greater than the pulmonary vascular resistance, a left-to-right shunt is present. If pulmonary vascular resistance rises above systemic vascular resistance, a right-to-left shunt develops.

### Clinical Manifestations

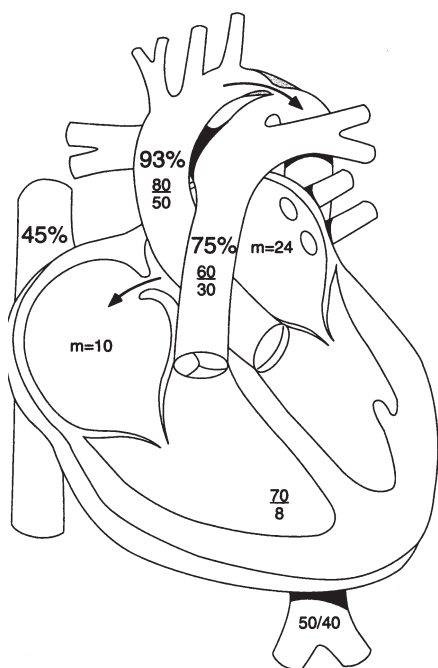
Symptoms are related to the size of the defect and the direction of flow. A small patent ductus arteriosus causes no symptoms. A large one with a left-to-right shunt may result in congestive heart failure, slowed growth, and repeated lower respiratory tract infections. Reversal of flow as a result of high pulmonary vascular resistance causes shortness of breath, dyspnea on exertion, and cyanosis. In a large shunt, bounding pulses, representing an aortic diastolic runoff, are palpable. The murmur, often referred to as a “machinery murmur,” is continuous beginning after  $S_1$ , peaks at  $S_2$ , and trails off during diastole. The chest radiograph of a large patent ductus arteriosus will show cardiomegaly, increased pulmonary vascularity, and left atrial and left ventricular enlargement. The neonate with a small patent ductus arteriosus has a normal ECG, whereas the neonate with a large patent ductus arteriosus and a generous left-to-right shunt shows left or biventricular hypertrophy. Right ventricular hypertrophy predominates on ECG in the presence of increased pulmonary vascular resistance. The patent ductus arteriosus is best seen on echocardiogram using Doppler flow mapping.

### Treatment

Indomethacin is often effective in closing the patent ductus arteriosus in the premature neonate by decreasing PGE<sub>1</sub> levels. A patent ductus arteriosus usually closes in the first month of life, but for those that do not, surgical ligation by thoracotomy or video-assisted thoracoscopic surgery, or coil embolization by catheterization is curative.

### Coarctation of the Aorta

Coarctation of the aorta (Figure 3-12) accounts for 8% of congenital heart defects and has a male-to-female predominance of 2:1. When coarctation of



**Figure 3-12** • Coarctation of the aorta in a critically ill neonate with a nearly closed ductus arteriosus. Typical anatomic and hemodynamic findings include: (a) “juxtaductal” site of the coarctation; (b) a bicommissural aortic valve (see in 80% of patients with coarctation); (c) narrow pulse pressure in the descending aorta and lower body; (d) a bidirectional shunt at the ductus arteriosus. As in critical aortic stenosis (see Fig. 3-13) there is an elevated left atrial pressure, pulmonary edema, a left-to-right shunt at the atrial level, pulmonary artery hypertension, and only a moderate (30-mm Hg) gradient across the arch obstruction. The low measured gradient (despite severe anatomic obstruction) across the aortic arch is due to low cardiac output.

Cloherly JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

the aorta occurs in a female, Turner’s syndrome must be considered. The obstruction is usually located in the descending aorta, at the insertion site of the ductus arteriosus. The aortic valve is bicuspid in 80% of cases, and mitral valve anomalies may also be present. The coarctation results in mechanical obstruction between the proximal and distal aorta and in increased left ventricular afterload. Congestive heart failure develops in 10% of cases in infancy.

### Clinical Manifestations

On examination, the femoral pulses are often weak and delayed relative to upper extremities—or are absent—and there is often upper extremity hypertension. Neonates with critical coarctation have ductal-dependent systemic blood flow and may present with circulatory collapse. Flow across the coarctation may produce a systolic ejection murmur heard at the apex. On chest radiograph, the aortic knob is enlarged; on ECG, right ventricular hypertrophy is seen in the neonate, and left ventricular hypertrophy is seen in the older child. The echocardiogram is used to visualize the defect and to check for abnormalities of the aortic valve, mitral valve, and left ventricular performance.

### Treatment

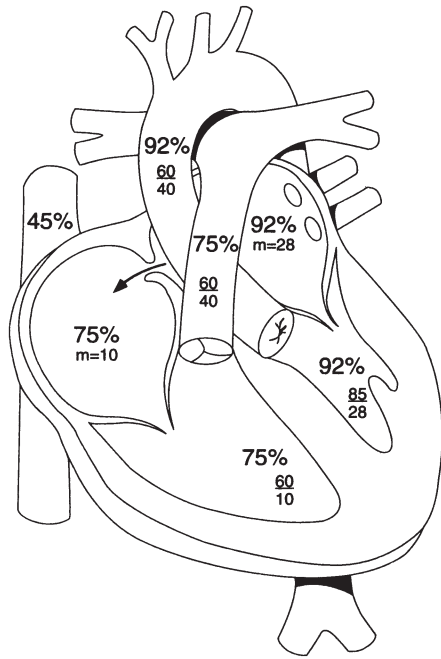
Palliation may be accomplished via balloon dilation angioplasty, stent placement, or by surgical end-to-end anastomosis, subclavian flap repair, patch repair, or graft placement.

### Aortic Stenosis

In aortic stenosis (Figure 3-13), the valvular tissue is thickened and often rigid. Most commonly, the valve is bicuspid, with a single fused commissure and an eccentric orifice. The stenotic valve produces a pressure gradient between the left ventricle and the aorta that results in left ventricular hypertrophy and, over time, decreased compliance and ventricular performance.

### Clinical Manifestations

The neonate with aortic stenosis may present with cardiovascular collapse or with a soft murmur. The level of symptomatology is related to the severity of the stenosis and the ventricular function. The neonate with critical aortic stenosis has ductal-dependent systemic blood flow and may present with circulatory collapse after the ductus closes. If ven-



**Figure 3-13** • Critical valvular aortic stenosis with a closed ductus arteriosus. Typical anatomic and hemodynamic findings include: (a) a morphologically abnormal, stenotic valve; (b) post-stenotic dilatation of the ascending aorta; (c) elevated left ventricular end diastolic pressure and left atrial pressures contributing to pulmonary edema (mild pulmonary venous and arterial desaturation); (d) a left-to-right shunt at the atrial level (note increase in oxygen saturation from superior vena cava to right atrium); (e) pulmonary artery hypertension (also secondary to the elevated left atrial pressure); (f) only a modest (25-mm Hg) gradient across valve. The low measured gradient (despite severe anatomic obstruction) across the aortic valve is due to a severely limited cardiac output, as evidenced by the low mixed venous oxygen saturation (45%) in the superior vena cava.

Cloherly JP, Stark AR. *Manual of Neonatal Care*, 4<sup>th</sup> ed. Philadelphia: Lippincott-Raven, 1998: 426.

tricular function is maintained, a harsh systolic ejection murmur is heard at the right upper sternal border and is preceded by an ejection click heard best at the left lower sternal border. If ventricular function is compromised, there may be significant stenosis with only a soft murmur appreciated. On chest radiograph, poststenotic dilatation of the ascending aorta is present, and in severe cases, pulmonary edema can be seen. The ECG may show left ventricular hypertrophy, and a strain pattern of ST depressions and inverted T waves may be seen. The valvular lesion, the degree of stenosis, and left ventricular function are all seen on echocardiogram.

### Treatment

If intervention is required, relief of the aortic valve gradient may be accomplished by open surgical valvotomy or by balloon valvuloplasty. Both surgical valvotomy and balloon valvuloplasty may result in progressive aortic regurgitation that may require aortic valve replacement with a mechanical, homograft, or autograft valve (Ross procedure).

### Pulmonic Stenosis

Pulmonic valve stenosis accounts for 5% to 8% of congenital heart defects. The pulmonary commissures are fused, the valve is domed and has a small central opening, and there is poststenotic dilatation of the main pulmonary artery. The valve is bicuspid or dysplastic in 10% of cases. Right ventricular hypertrophy occurs over time as the ventricle attempts to maintain cardiac output. In critical pulmonic stenosis, a decrease in the compliance of the right ventricle will increase right atrial pressure and may open the foramen ovale, producing a small right-to-left shunt.

### Clinical Manifestations

Most patients are asymptomatic. Severe to critical pulmonary stenosis may cause dyspnea on exertion and angina. Right-sided congestive heart failure is rare, except in infants with critical pulmonic stenosis who may have ductal-dependent pulmonary blood flow. Characteristically, the ejection click of pulmonic stenosis varies with inspiration, and a harsh systolic ejection murmur is heard at the left upper sternal border. In severe stenosis, a thrill and right ventricular heave are palpable. On chest radiograph, heart size and pulmonary vascularity are normal, but the pulmonary artery segment is enlarged. On ECG, the degree of right ventricular hypertrophy and right-axis deviation correlates with the degree of stenosis. The transvalvular gradient and the degree of right ventricular hypertrophy can be measured by echocardiogram.

### Treatment

Definitive treatment is accomplished by balloon valvuloplasty of the stenotic valve. Indications for pulmonary valvotomy include a right ventricular pressure greater than 50 mm Hg or symptoms of right-sided congestive heart failure.

Thus far, this chapter has focused on the evaluation of the cyanotic neonate and the most common



■ TABLE 3-3

## Classic Findings for the 10 Most Common Congenital Heart Lesions

Lesion	Presentation	Physical Examination	ECG	X-ray
Atrial septal defect	Murmur	Fixed split S <sub>2</sub>	Mild RVH	±CE, ↑ PBF
Ventricular septal defect	Murmur, CHF	Holosystolic murmur	LVH, RVH	+CE, ↑ PBF
Patent ductus	Murmur, ±CHF	Continuous murmur	LVH, ±RVH	±CE, ↑ PBF
AV canal defect	Murmur, ±CHF	Holosystolic murmur	“Superior” axis	± CE, ↑ PBF
Pulmonic stenosis	Murmur, ±cyanosis	Click, SEM	RVH	±CE, NL, or ↓ PBF
Tetralogy of Fallot	Murmur, cyanosis	SEM	RVH	±CE, ↓ PBF
Aortic stenosis	Murmur, ±CHF	Click, SEM	LVH	±CE, NL, PBF
Coarctation of aorta	Hypertension	↓Femoral pulses	LVH	±CE, NL, PBF
Transposition of the great arteries	Cyanosis	Marked cyanosis	RVH	±CE, NL, or ↑ PBF
Single ventricle	(Variable)	(Variable)	(Variable)	(Variable)

CE, cardiac enlargement; CHF, congestive heart failure; LVH, left ventricular hypertrophy; NL, normal; PBF, pulmonary blood flow; RVH, right ventricular hypertrophy; SEM, systolic ejection murmur.

cyanotic and acyanotic congenital heart defects. Before moving to acquired structural heart disease, functional heart disease, and arrhythmias, see Table 3-3, which lists the classic findings for the 10 most common congenital heart lesions.

## ■ ACQUIRED STRUCTURAL HEART DISEASE

### Rheumatic Heart Disease

Rheumatic heart disease results from single or multiple episodes of acute rheumatic fever. Mitral regurgitation is the most common lesion found. Aortic insufficiency is also commonly found with or without mitral regurgitation. Mitral stenosis is less common and usually is the end result of multiple attacks of acute rheumatic fever. Least common is aortic stenosis. The tricuspid and pulmonary valves are almost never affected. Symptoms are proportional to the degree of valvular damage. Rheumatic fever is discussed in Chapter 12.

### Kawasaki's Disease

Cardiac effects may include pericarditis, myocarditis, and transient rhythm disturbances. However, it is

the development of coronary artery aneurysms, with their potential for occlusion or rupture, that makes the disease life-threatening. Coronary artery aneurysms develop during the subacute phase (11th to 25th day) in about 30% of cases but regress in most patients. Early therapy with intravenous immunoglobulin decreases the incidence of coronary artery aneurysms to less than 10%. High-dose aspirin therapy lessens the likelihood of late aneurysms. The echocardiogram is used to assess ventricular function and visualize pericardial fluid and coronary artery aneurysms. A thorough discussion of Kawasaki's disease is found in Chapter 11.

### Endocarditis

#### Pathogenesis

Bacterial endocarditis is a microbial infection of the endocardium. Although it may occur on normal valves, bacterial endocarditis is much more likely to occur on congenitally abnormal valves, valves damaged by rheumatic fever, acquired valvular lesions (mitral valve prolapse), and prosthetic replacement valves as a consequence of turbulent blood flow. Factors that may precipitate bacterial endocarditis include a previous episode of endocarditis, dental manipulation or infection, instru-

mentation of the gastrointestinal or genitourinary tract, intravenous drug abuse, an indwelling central venous catheter, and prior cardiac surgery.

In children, alpha hemolytic streptococci (*Streptococcus viridans*) and *Staphylococcus aureus* are the most common etiologic agents. *S. viridans* accounts for approximately 67% of the cases, whereas *S. aureus* is present in about 20% of cases. When infection complicates cardiac surgery, *Staphylococcus epidermidis*, gram-negative bacilli, and fungi should be considered. Gram-negative organisms cause about 5% of cases of endocarditis in children and are more likely in neonates, immunocompromised patients, and intravenous drug abusers. Among the HACEK (*Haemophilus*, *Actinobacillus*, *Cardiobacterium*, *Eikenella*, *Kingella*) organisms, which are a rare cause of endocarditis, *Haemophilus influenzae* is the most common, frequently affecting previously damaged valves.

### Clinical Manifestations

Fever is the most common finding in children with bacterial endocarditis. Often, a new or changing murmur is auscultated. Children with endocarditis usually display nonspecific symptoms such as chest pain, dyspnea, arthralgia, myalgia, headache, and malaise. Embolic phenomena such as hematuria with red cell casts and transient ischemic attack or stroke may be present. Other embolic phenomena, such as Roth spots, splinter hemorrhages, petechiae, Osler nodes, and Janeway lesions, are relatively rare in children with bacterial endocarditis.

### Diagnostic Evaluation

Laboratory studies include a complete blood count, erythrocyte sedimentation rate (ESR), c-reactive protein (CRP), and urinalysis. Multiple blood cultures increase the probability of discovering the pathogen. Positive blood cultures, an elevated ESR, elevated CRP, hematuria, and anemia are most often found. The echocardiogram is used to define vegetations or thrombi in the heart.

### Treatment

Medical management consists of 6 weeks of intravenous antibiotics directed against the isolated pathogen. Surgery is indicated for endocarditis when medical treatment is unsuccessful, refractory congestive heart failure exists, or there are serious

embolic complications, myocardial abscess formation, or refractory prosthetic valve disease.

Prevention of endocarditis is necessary for high-risk patients. Antibiotic regimens to prevent endocarditis during dental, respiratory, gastrointestinal, or genitourinary procedures include oral amoxicillin or parenteral ampicillin and gentamicin prior to the procedure.

## KEY POINTS

1. Patients with congenitally abnormal valves, valves damaged by rheumatic fever, acquired valvular lesions (mitral valve prolapse), or prosthetic replacement valves are at increased risk for endocarditis.
2. Alpha hemolytic streptococci (*S. viridans*) and *S. aureus* are the most common etiologic agents in endocarditis.

## Coronary Artery Disease

Coronary artery disease is rare in childhood, but the atherosclerotic process appears to begin early in life. There is evidence that progression of atherosclerotic lesions is influenced by genetic factors (familial hypercholesterolemia) and lifestyle (cigarette smoking; high-cholesterol diet, high-saturated-fat diet). Because many lifetime habits are formed during childhood, the opportunity exists for prevention of coronary artery disease.

## FUNCTIONAL HEART DISEASE

### Myocarditis

Most cases of myocarditis in North America result from viral infection of the myocardium, predominantly enteroviruses (coxsackie B virus and echovirus). It is unclear whether myocardial damage from viral myocarditis results from direct viral invasion or an autoimmune antibody response.

### Clinical Manifestations

Depending on the degree of damage to the myocardium, patients may be asymptomatic and the diagnosis may be made only by finding ST- and T-

wave changes on an ECG done for an unrelated reason, whereas others may present with fulminant congestive heart failure. Common symptoms include fever, dyspnea, fatigue, and chest pain (usually due to a secondary pericarditis). Signs include tachycardia, evidence of congestive heart failure, and S<sub>3</sub> ventricular gallop. The ECG often reveals ST-segment depression and T-wave inversion, as well as arrhythmias and conduction defects. The chest radiograph varies from mild to marked cardiomegaly. Echocardiogram denotes dilated or hypocontractile ventricles, or both. Pericardial effusion may be present. Endomyocardial biopsy may be indicated in select cases to confirm diagnosis. Viral etiology should be evaluated by viral culture and PCR from the throat, stool, blood, and pericardial fluid, if present.

### **Treatment**

Therapy for patients with viral myocarditis is supportive to maintain perfusion and oxygenation. Treat ventricular arrhythmias, conduction abnormalities, and congestive heart failure as indicated. Intravenous immunoglobulin is given to minimize further damage to the myocardium. The prognosis for patients with myocarditis depends on the extent of myocardial damage.

## **Dilated Cardiomyopathy**

Dilated or congestive cardiomyopathy is characterized by myocardial dysfunction and ventricular dilatation. Although usually an idiopathic disorder, it can be caused by neuromuscular disease (Duchenne muscular dystrophy) or drug toxicity (anthracyclines). Dilation of the left ventricle results in congestive heart failure. An increase in left atrial pressure, pulmonary venous pressure, and pulmonary capillary wedge pressure results in pulmonary edema.

### **Clinical Manifestations**

Symptoms include dyspnea, orthopnea, and paroxysmal nocturnal dyspnea. Eventually, right heart failure with dependent edema occurs, and a pulsus alternans may be noted. On cardiac examination, a right ventricular heave and an S<sub>3</sub> gallop are found. The ECG reveals rhythm disturbances, left ventricular hypertrophy, and nonspecific ST- and T-wave ischemic changes. Ventricular function is evaluated by echocardiogram.

### **Treatment**

Medical therapy includes inotropic agents and vasodilators to improve myocardial contractility and to decrease the afterload on the weakened ventricle. Diuretics decrease preload and hopefully improve cardiac output by moving the dilated ventricle to a more favorable position on the Frank-Starling curve, and antiarrhythmic medications are used to control potentially fatal ventricular arrhythmias. If medical therapy fails, heart transplantation may be necessary.

## **Hypertrophic Cardiomyopathy**

Also known as idiopathic hypertrophic subaortic stenosis, hypertrophic cardiomyopathy is an autosomal dominant genetic disorder in which the ventricular septum is thickened, resulting in left ventricular outflow tract obstruction. In the thickened stiff left ventricle, diastolic function is well preserved, but systolic function is compromised. Abnormal motion of the mitral valve results in mitral insufficiency.

### **Clinical Manifestations**

Symptoms include dyspnea on exertion, chest pain, and syncope. There is often a bisferious pulse (double peaked) because ejection is hindered by septal obstruction, a ventricular gallop (S<sub>3</sub>), and murmurs indicative of mitral regurgitation and left ventricular outflow tract obstruction. ECG illustrates left-axis deviation, left ventricular hypertrophy, and possible ST- and T-wave changes consistent with ischemia or strain. The echocardiogram is diagnostic.

### **Treatment**

Therapy is centered around preventing fatal ventricular arrhythmias and decreasing the stiffness of the left ventricle with negative inotropic medications, such as calcium channel blockers, and beta-adrenergic blocking agents. The avoidance of competitive sports is essential because sudden death during exertion is a significant risk.

## **■ ARRHYTHMIAS**

Arrhythmias in children are much less common than in adults but can be just as life threatening. Arrhythmias result from disorders of impulse formation, impulse conduction, or both and are generally classified as follows.

## KEY POINTS

1. Most cases of myocarditis in North America result from viral infection of the myocardium.
2. Dilated or congestive cardiomyopathy is characterized by myocardial dysfunction and ventricular dilatation; it is usually idiopathic.
3. Therapy for dilated cardiomyopathy includes inotropic agents to improve left ventricular contractility and vasodilators to decrease afterload. Diuretics decrease preload, and antiarrhythmic medications are used to control potentially fatal ventricular arrhythmias.
4. In hypertrophic cardiomyopathy, the ventricular septum is thickened, resulting in left ventricular outflow tract obstruction.
5. Therapy for hypertrophic cardiomyopathy is centered around preventing fatal ventricular arrhythmias and decreasing the stiffness of the left ventricle with negative inotropic medications.

### *Bradycardias*

- Sinus node dysfunction
- Conduction block

### *Tachycardias*

- Narrow QRS
- Wide QRS

### *Premature Beats*

- Atrial
- Ventricular

Bradycardias are the result of either depressed automaticity or block of an impulse, whereas tachycardias or premature beats arise from abnormal impulse formation caused by enhanced automaticity, a reentrant circuit, or triggered activity. Arrhythmias may result from congenital, functional, or acquired structural heart disease; electrolyte disturbances (potassium, calcium, and magnesium); drug toxicity; poisoning; or an acquired systemic disorder. Table 3-4 lists etiologies predisposing children to arrhythmias.

## Bradycardias

As already stated, bradycardias result from sinus node dysfunction or conduction block. Bradycardias due to sinus node dysfunction include sinus bradycardia, junctional bradycardia, ectopic atrial

bradycardia, and sinus pauses. Bradycardias due to conduction block include first-degree heart block, second-degree heart block, and third-degree (complete) heart block. Second-degree heart block is further divided into Mobitz type I block (Wenckebach), Mobitz type II block, and fixed-ratio atrioventricular (AV) block.

### **Differential Diagnosis**

Figure 3-14 shows the rhythm strips of various bradycardias. **Sinus bradycardia** is associated with increased vagal tone, hypoxia, central nervous system disorders with increased intracranial pressure, hypothyroidism, hyperkalemia, hypothermia, drug intoxication (digoxin, beta-blockers, calcium channel blockers), and prior atrial surgery. It is also a normal finding in healthy athletic teenagers. The ECG reveals a normal P wave with normal AV conduction at rates less than 100 bpm in the neonate and 60 bpm in the older child. When sinus bradycardia becomes too slow, sinus pauses or escape rhythms may occur. The escape rhythms most often seen include ectopic atrial bradycardia or ectopic atrial rhythm, junctional bradycardia or junctional rhythm, or a slow idioventricular ventricular rhythm.

First-degree heart block usually results from slowing of atrioventricular conduction at the level of the AV node. It is associated with increased vagal tone, digoxin and beta-blocker administration, infectious etiologies (viral myocarditis, Lyme disease), hypothermia, electrolyte abnormalities (hypo/hyperkalemia, hypo/hypercalcemia, hypomagnesemia), congenital heart disease (ASD, atrioventricular canal defect, Ebstein's anomaly, TAPVC, and L-transposition of the great arteries or "corrected transposition"), rheumatic fever, and cardiomyopathy. First-degree AV block is characterized on ECG by PR interval prolongation for age and rate. The rhythm is regular, originates in the sinus node, and has a normal QRS morphology.

Second-degree heart block refers to episodic interruption of AV nodal conduction:

- **Mobitz type I (Wenckebach)** denotes progressive prolongation of the PR interval over several beats until a QRS is dropped. This cycle repeats itself often, although the number of beats in a cycle may not be constant. The QRS configuration is normal. Etiologies for this rhythm are the same as those for first-degree heart block.
- **Mobitz type II** is caused by abrupt failure of atrioventricular conduction below the AV node in the



■ TABLE 3-4

## Factors Predisposing to Dysrhythmias

## Congenital heart disease

- Supraventricular dysrhythmias: Ebstein's anomaly (may also present with WPW syndrome), atrial septal defects, atrial surgery, L-transposition of the great arteries, after Fontan operation
- Ventricular dysrhythmias: aortic valve disease, pulmonary valve disease, after tetralogy of Fallot repair, anomalous left coronary artery, RV dysplasia
- Heart block (varying degrees): after open-heart surgery (Ebstein's anomaly, L-transposition of the great arteries, common atrioventricular canal, VSD repair); congenital complete heart block (idiopathic, associated with maternal systemic lupus erythematosus, L-transposition of the great arteries)

## Isolated conduction system disorders

- WPW syndrome
- Prolonged QT interval syndromes

## Associated with systemic illness

- Infectious myocarditis
- Kawasaki's disease
- Idiopathic dilated or hypertrophic cardiomyopathy
- Friedreich's ataxia (atrial tachycardia or fibrillation)
- Muscular dystrophies (Duchenne, periodic paralysis)
- Glycogen storage diseases (Pompe's disease)
- Collagen vascular diseases (rheumatic carditis, systemic lupus erythematosus, periarteritis nodosa, dermatomyositis)
- Endocrine disorders (hyperthyroidism, adrenal dysfunction)
- Metabolic and electrolyte disturbances (hypomagnesemia, hyperkalemia, hypocalcemia, hypoxia)
- Lyme disease

## Drug toxicity

- Chemotherapeutic agents (anthracyclines)
- Tricyclic antidepressants
- Cocaine
- Digitalis, beta-adrenergic blockers, calcium channel blockers
- Asthma medications (sympathomimetics)

## Other causes

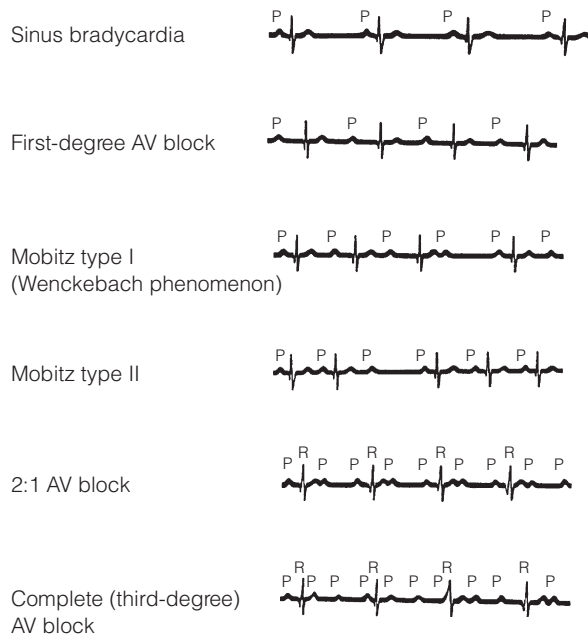
- Blunt chest trauma (myocardial contusion)
- Increased intracranial pressure

bundle of His-Purkinje fiber system. It is a more serious bradycardia than first-degree heart block or Wenckebach because it can progress to complete heart block. On ECG, there is sudden AV conduction failure with a dropped QRS after a normal P wave. No preceding PR interval prolongation is seen in normal conducted impulses.

- **Fixed-ratio AV block** is an arrhythmia in which the QRS complex follows only after every second (third or fourth) P wave, causing 2:1 (3:1 or 4:1) AV block. There is a normal PR interval in conducted beats. There is usually a normal or slightly prolonged QRS. Fixed-ratio block results from either AV node or His bundle injury, and intracar-

diac recordings are required to distinguish the site of injury. Patients may progress to complete heart block.

Third-degree heart block occurs when no atrial impulses are conducted to the ventricles. The atrial rhythm and rate are normal for the patient's age, and the ventricular rate is slowed markedly (40–55 bpm). If an escape rhythm arises from the AV node (junctional rhythm), the QRS interval is of normal duration, but if an escape rhythm arises from the distal His bundle or Purkinje fibers, the QRS interval is prolonged (idioventricular rhythm). Congenital complete AV block can be an isolated abnormality or can



**Figure 3-14** • Bradyarrhythmias.

be associated with L-transposition of the great arteries, atrioventricular canal defect, or maternal lupus erythematosus. Other causes include open-heart surgery (especially after large ventricular septal defect closure), cardiomyopathy, or Lyme disease. Newborns with congenital complete heart block may present with hydrops fetalis.

### Treatment

No intervention is necessary for sinus bradycardia if cardiac output is maintained. A management algorithm for sinus bradycardia is shown in Figure 3-15.

No treatment is necessary for first- or second-degree heart block (Mobitz type I). Mobitz type II, fixed-ratio AV block, and third-degree heart block all require pacemaker placement. In Mobitz type II and fixed-ratio AV block, prophylactic pacemaker insertion is essential to protect the patient should he or she progress to complete heart block with inadequate cardiac output away from medical care.

If the child with complete heart block is hemodynamically unstable, transcutaneous or transvenous pacing can be performed acutely, and permanent transvenous or epicardial pacemaker placement can be performed later. Third-degree heart block is managed with either ventricular demand pacing or AV sequential pacing. Figure 3-16 is a management algorithm for AV block.

## Tachyarrhythmias

Narrow-complex tachycardias have a QRS morphology similar or identical to that of normal sinus rhythm. They include most, but not all, SVTs (some SVTs have a widened QRS). Narrow-complex tachycardias may be due to increased automaticity or from a reentrant circuit. Narrow-complex tachycardias due to increased automaticity include sinus tachycardia, ectopic atrial tachycardia, junctional ectopic tachycardia, and atrial fibrillation. Narrow-complex tachycardias caused by reentrant mechanisms are categorized as orthodromic reentrant tachycardia (ORT) or antidromic reentrant tachycardia (ART). In ORT the SVT propagates down the AV node and up the bypass tract. Since the ventricles are depolarized in the normal fashion, down the AV node, the QRS complex is narrow. In ART the SVT propagates down the bypass tract and up the AV node. Since the ventricles are depolarized down the bypass tract, the QRS is widened. Narrow-complex AV reciprocating tachycardias include AV node reentrant tachycardia, WPW syndrome orthodromic tachycardia (accessory pathway not concealed on ECG—delta wave), orthodromic atrioventricular reciprocating tachycardia (accessory pathway concealed on ECG—no delta wave), sinoatrial reentrant tachycardia, and atrial flutter. Narrow-complex tachycardias are relatively well tolerated acutely.

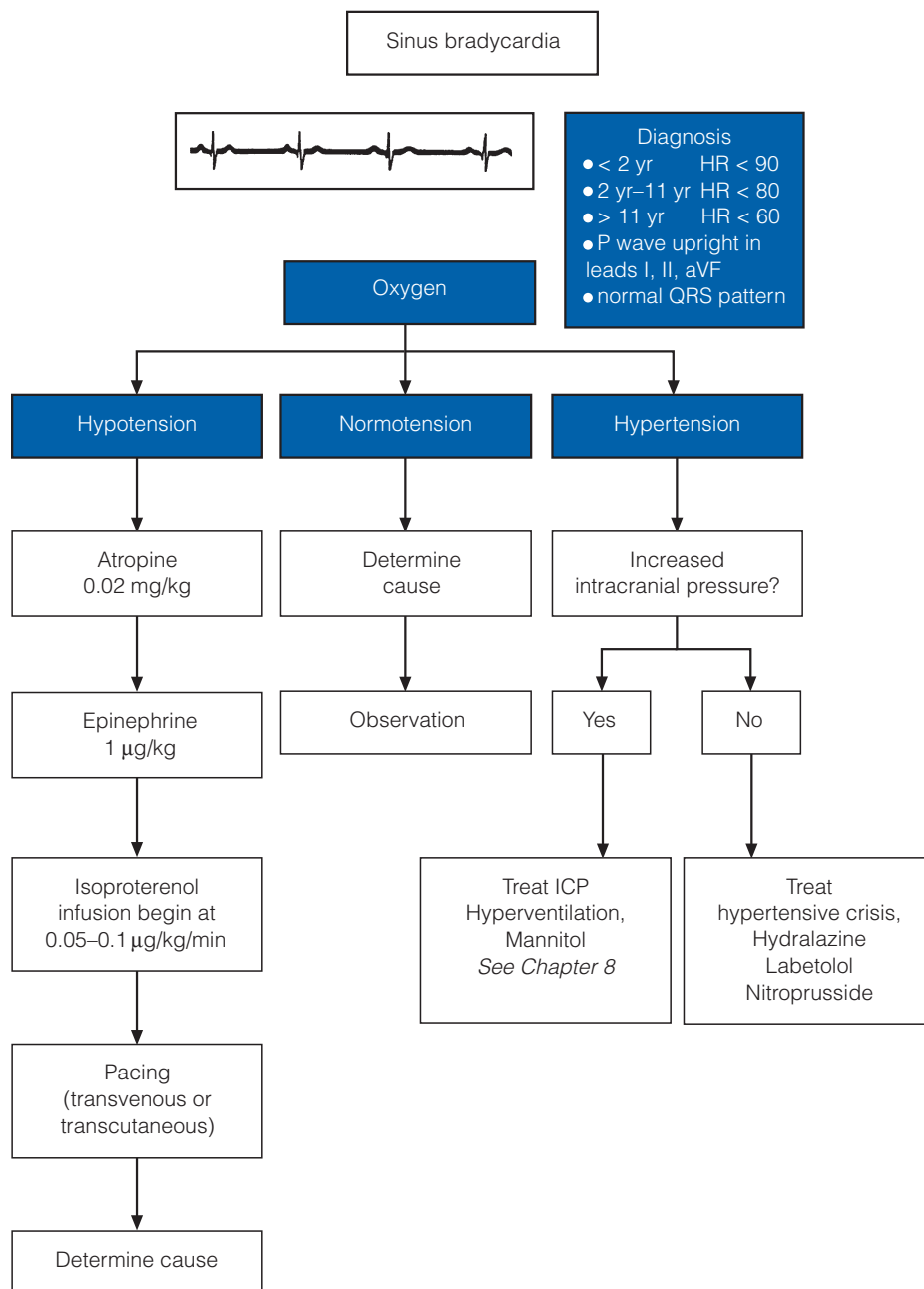
Conversely, wide-complex tachycardias, defined as tachycardias with a QRS more than 0.12 seconds, are a medical emergency. Wide-complex tachycardias include ventricular tachycardia, ventricular fibrillation, WPW syndrome antidromic reentrant tachycardia, and orthodromic SVT with aberrancy.

### Differential Diagnosis

Figure 3-17 shows the rhythm strips of the various tachycardias. The causes of tachyarrhythmia are as follows.

#### Narrow-Complex Tachycardias

- *Sinus tachycardia*: Fever, stress, dehydration, and anemia
- *ORT (most common non-sinus tachycardia SVT)*: Most cases result from a concealed bypass tract causing ORT, AV node reentrant tachycardia, WPW syndrome ORT, Ebstein's anomaly (associated with WPW syndrome), L-transposition of the great arteries
- *Atrial flutter*: Atrial surgery (D-TGA s/p Mustard/Senning procedure, ASD s/p repair Hemi-Fontan, Fontan), myocarditis, structural heart disease with



**Figure 3-15** • Management algorithm for sinus bradycardia.

dilated atria (Ebstein's anomaly, tricuspid atresia, rheumatic heart disease of the mitral valve), severe tricuspid regurgitation

- *Atrial fibrillation*: Most often seen with left atrial enlargement (rheumatic heart disease of the mitral valve, VSD, systemic to pulmonary artery palliative shunt placement); other causes that result in right atrial or biatrial enlargement include Ebstein's anomaly, WPW syndrome, and myocarditis

#### *Wide-Complex Tachycardia*

- *Ventricular tachycardia*: Congenital or acquired heart disease resulting in ventricular dilation or hypertrophy or ventricular suture line, drug ingestion, or WPW syndrome ART
- *Ventricular fibrillation*: Terminal rhythm that develops after hypoxia, ischemia, or high-voltage electrical injury; predisposing factors include WPW syndrome and long QT syndrome

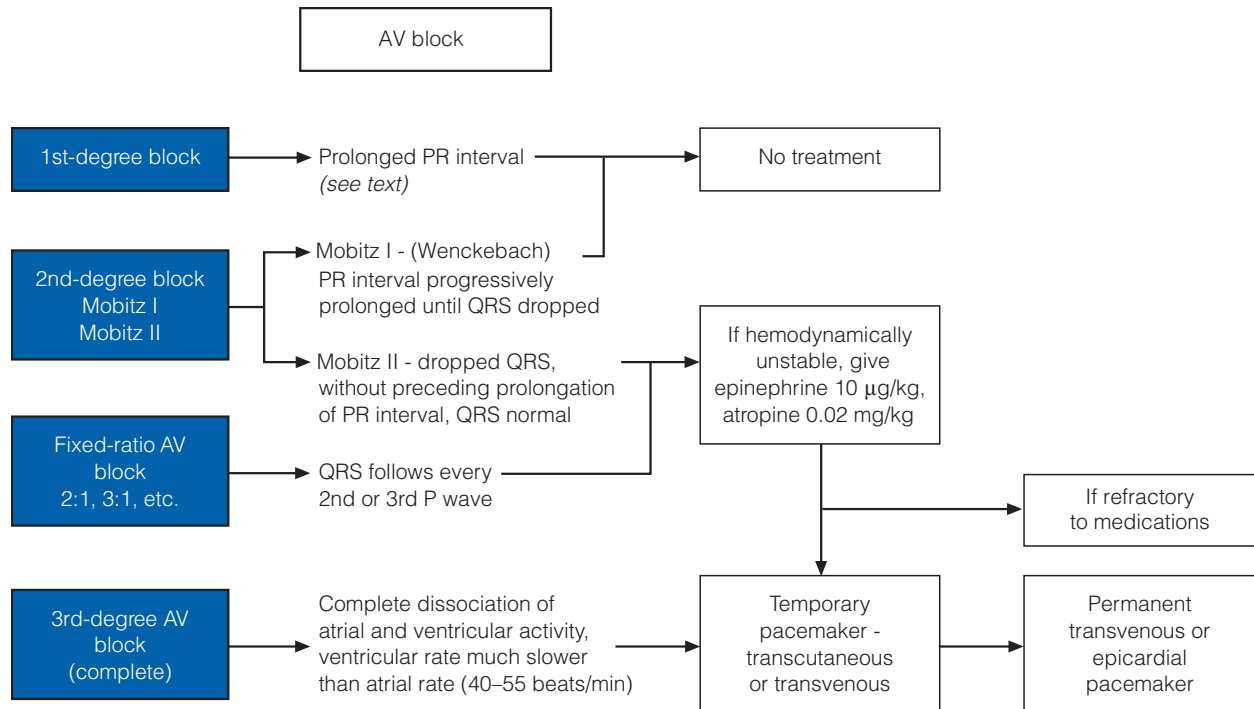


Figure 3-16 • Management algorithm for AV block.

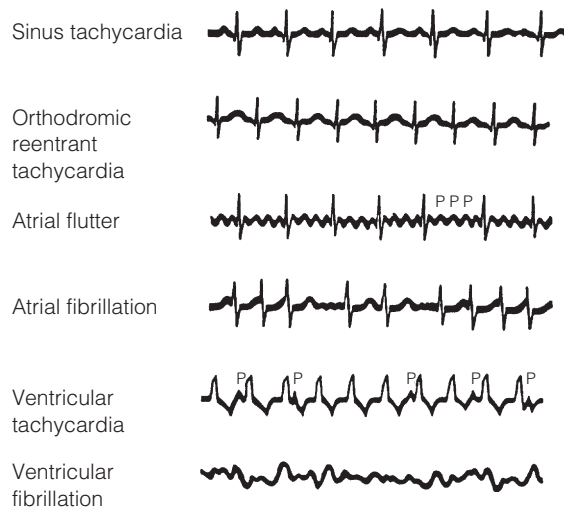


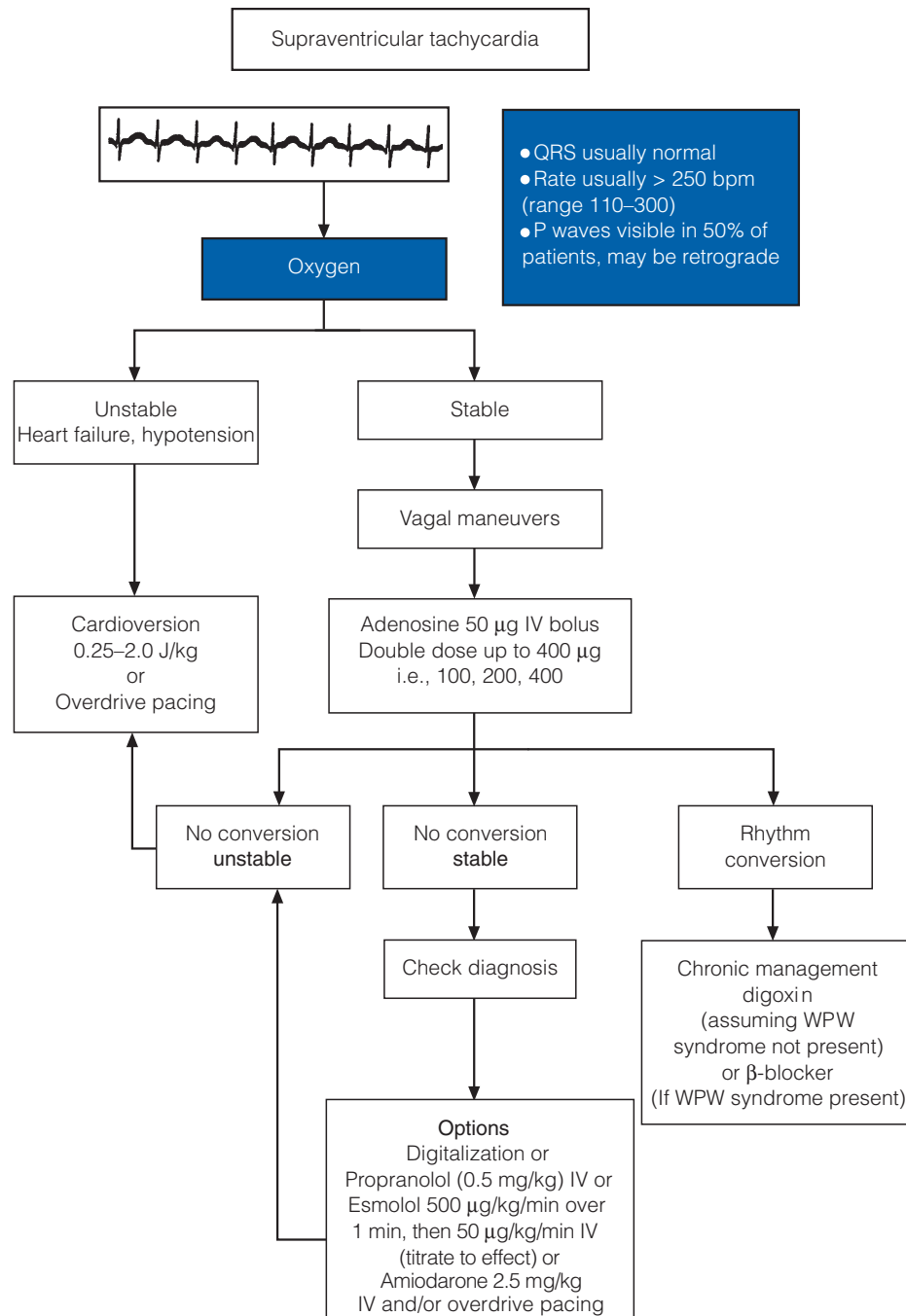
Figure 3-17 • Tachydysrhythmias.

## Treatment

### Narrow-Complex Tachycardia

Treatment of sinus tachycardia involves correcting the underlying cause of the tachycardia. Figure 3-18 outlines a management algorithm for supraventricular tachycardia. Treatment for stable narrow-complex

tachycardia progresses from vagal maneuvers to pharmacotherapy to cardioversion. Vagal maneuvers enhance vagal tone to slow conduction in the AV node and often result in termination of the arrhythmia. Vagal tone is increased in infants by applying ice to the face, and in older children through carotid massage; make sure to keep the infant's airway unobstructed when applying ice to the face. If vagal maneuvers are ineffective in stable narrow-complex tachycardia, adenosine is given to block the AV node to break a reentrant SVT whose circuit involves the AV node (AV node reentrant tachycardia, WPW syndrome with ORT, concealed bypass tract ORT). Adenosine will be ineffective on a narrow-complex tachycardia that results from increased automaticity or a reentrant mechanism that does not involve the AV node (sinus tachycardia, ectopic atrial tachycardia, junctional ectopic tachycardia, atrial flutter, or sinoatrial reentrant tachycardia). If adenosine returns the child to normal sinus rhythm and WPW is not suspected (no delta wave seen after conversion of tachycardia), the child is started on digoxin to reduce the risk of future events. If adenosine reveals WPW syndrome (delta wave noted after conversion of tachycardia), use a beta-blocker, because the use of digoxin can slow the AV node and speed up conduction over the accessory pathway in an antidromic fashion and



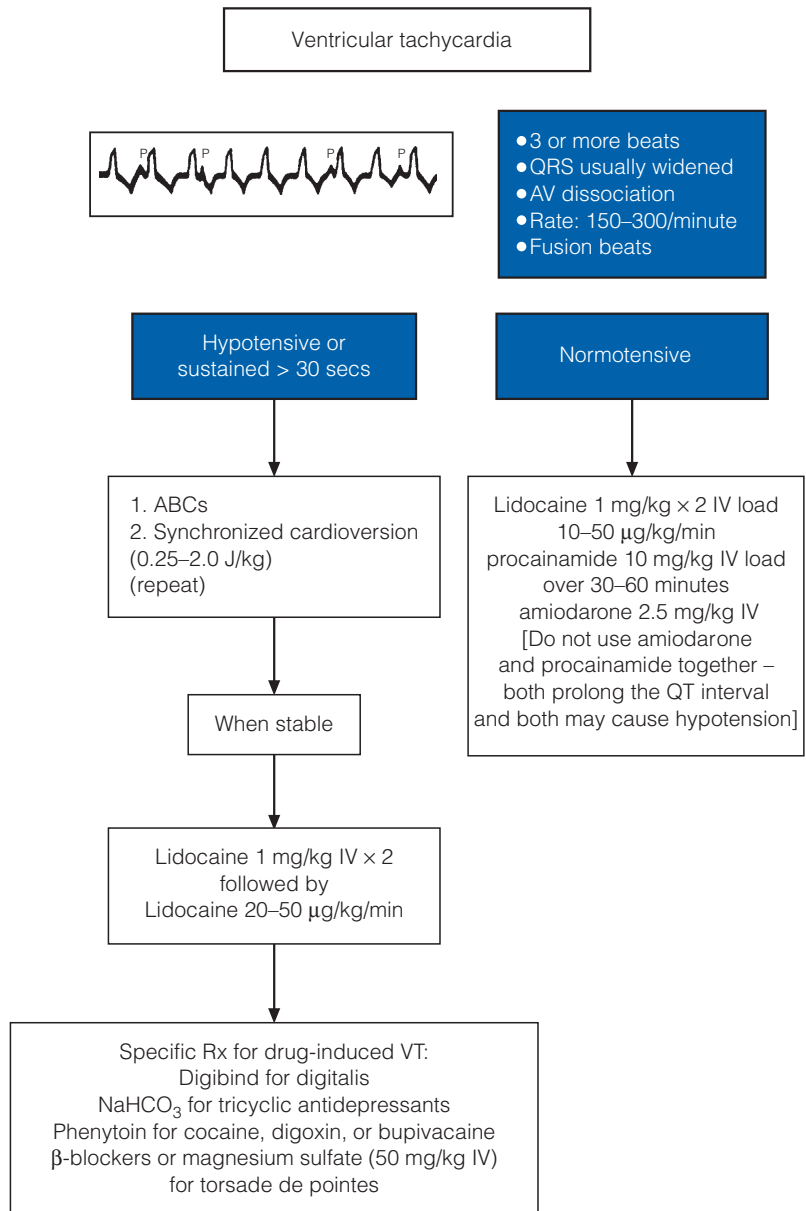
**Figure 3-18** • Management algorithm for supraventricular tachycardia.

result in ventricular fibrillation secondary to atrial fibrillation or some other fast atrial arrhythmia. For this reason, digoxin should be avoided to treat ORT associated with WPW syndrome. Propranolol is an effective and safe alternative to digoxin in ORT associated with WPW syndrome. When unstable narrow-complex tachycardia is present and the patient has

congestive heart failure or hypotension, cardioversion or transesophageal overdrive pacing is indicated. Synchronized cardioversion is required to avoid the inadvertent development of ventricular fibrillation.

In unstable atrial flutter, synchronized cardioversion or overdrive pacing is used when rapid intervention is necessary because of congestive heart



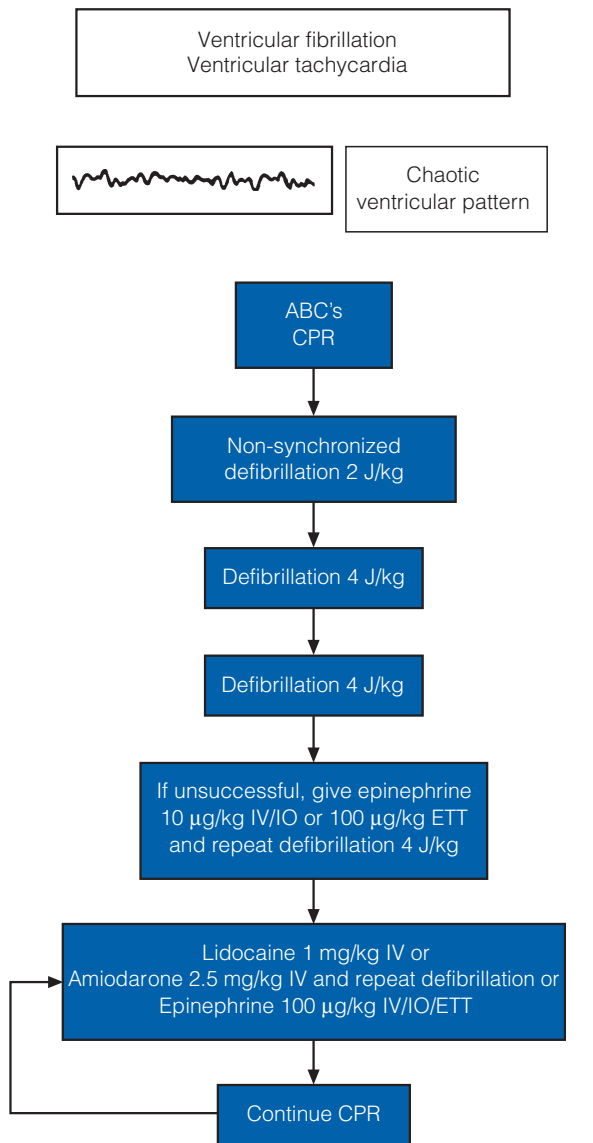


**Figure 3-19** • Management algorithm for ventricular tachycardia.

failure. Once cardioversion has occurred, digoxin, beta-blockers, procainamide, amiodarone, sotalol, or a quinidine/digoxin combination may be given to help prevent recurrences. If the child is hemodynamically stable, he or she should be loaded with digoxin and then given procainamide in an attempt to convert the arrhythmia. It is critical to load with digoxin before giving procainamide, because procainamide has vagolytic activity that could inadvertently increase the ventricular rate and cause acute hemodynamic deterioration.

If atrial fibrillation has been present for more than

a few days, anticoagulation is needed before converting the rhythm to decrease the risk of embolization of possible intra-atrial clots. An alternative to anticoagulation is transesophageal echocardiography to assess for clots. If no clots are seen, cardioversion may proceed, although with a slightly increased risk of thromboembolism relative to anticoagulation. Quinidine, procainamide, or amiodarone can be effective in pharmacologic conversion of atrial fibrillation, and quinidine and procainamide are good long-term maintenance drugs. Synchronized cardioversion converts most cases to sinus rhythm.



**Figure 3-20** • Management algorithm for ventricular fibrillation.

### Wide-Complex Tachycardia

Treat wide-complex ventricular tachycardia due to WPW syndrome with antidromic conduction or

orthodromic SVT with aberrancy as though the patient has ventricular tachycardia. Hypotensive or unresponsive patients should be treated immediately with cardiopulmonary resuscitation and synchronized cardioversion. After cardioversion, sinus rhythm can be maintained with intravenous lidocaine or amiodarone. Normotensive patients with acute-onset ventricular tachycardia can be treated with intravenous lidocaine or amiodarone in an attempt to break the arrhythmia without cardioversion.

Children with ventricular fibrillation should receive CPR and must be defibrillated with nonsynchronized cardioversion. Giving epinephrine may turn fine fibrillation into coarse fibrillation and allow successful defibrillation. The management algorithms for ventricular tachycardia and ventricular fibrillation/pulseless ventricular tachycardia are outlined in Figure 3-19 and Figure 3-20, respectively.

### KEY POINTS

1. Bradyarrhythmias with widened QRS complexes are likely to be escape rhythms from the His bundle or Purkinje system (idioventricular rhythm) and are at high risk for progression to complete heart block.
2. Symptomatic sinus bradycardia, second-degree heart block (Mobitz type II and fixed-ratio AV block), and third-degree heart block all need pacing.
3. Narrow-complex tachycardias tend to be well tolerated acutely, whereas wide-complex tachycardias are considered a medical emergency.
4. Treat wide-complex tachycardia due to SVT (WPW syndrome with ART or SVT with aberrancy) as though the patient has ventricular tachycardia.
5. When treating SVT, rule out WPW syndrome, because the treatment for WPW-associated SVT is different from that for non-WPW SVT.