The Sick Neonate With Cardiac Disease

Jennifer F. Anders, MD,
Karen A. Schneider, RSM, MD, MPH

The approach to the infant with a cardiac emergency begins with recognition of the unstable or critically ill child and proceeds rapidly into stabilization and provision of immediate therapies. Support of oxygenation, ventilation, and circulation will precede identification of specific cardiac lesions. The emergency clinician can use clinical findings, chest x-ray, and electrocardiographic information to plan emergent intervention. Infants in the first days of life who present with circulatory collapse secondary to obstruction of pulmonary or systemic blood flow (ductus dependent) conditions can be stabilized with prostaglandin E infusion. The more common presentation of cardiac disease in the first month of life is congestive heart failure. Infants with congestive heart failure require respiratory support, careful fluid management, and inotropic support.

Keywords: congenital heart disease; prostaglandin
Infant respiratory reserves are limited, and neonates with cardiac lesions frequently show signs of respiratory distress. Both cyanotic and noncyanotic heart disease will include respiratory distress. Oxygen will likely be initiated immediately upon arrival of the distressed neonate. Children whose hypoxia resolves after administration of supplemental oxygen should continue to receive oxygen by face mask or nasal cannula. Hypoxia that fails to resolve with administration of high-flow supplemental oxygen likely represents cyanotic congenital heart disease. Hyperoxegenation of infants with mixing lesions can create complications. For most cyanotic congenital heart disease lesions, a reasonable goal for oxygen saturation is 75% to 85%. Rapid onset of intense cyanosis, particularly in the first 2 weeks of life, strongly suggests the presence of ductal dependent cyanotic congenital heart disease. Supplemental oxygen should be used sparingly in these infants, as high oxygen levels will speed closure of the ductus arteriosus, which may precipitate rapid decline.

Venous access should be obtained rapidly in any sick neonate. For a child with symptoms of shock, including decreased level of consciousness or agitation, diminished pulses, or prolonged capillary refill, bolus isotonic fluids should be provided by intravenous or intraosseous route. An initial bolus of isotonic fluids at 10 mL/kg will support circulation with minimal risk of worsening cardiac function in a neonate. As basic resuscitation measures proceed, the clinician will simultaneously turn to diagnostic considerations. History taking should include the maternal medical history and prenatal exposures. Table 1 lists prenatal risk factors for congenital heart disease. The differential diagnosis of neonatal cardiac emergencies requires an understanding of fetal and transitional circulation.

**FETAL AND TRANSITIONAL CIRCULATION**

The 3 unique structures in fetal circulation are the ductus venosus, foramen ovale, and the ductus arteriosus. The umbilical vein transports oxygenated blood from the placenta through the ductus venosus to the inferior vena cava where it joins the deoxygenated blood from the lower body. This mixed blood enters the right atrium where it is directed through the foramen ovale to the left atrium, thus bypassing the right ventricle (RV) and the pulmonary circulation. The blood traverses from the left atrium to left ventricle (LV) to the aorta. Meanwhile, deoxygenated blood from the upper body is carried by the superior vena cava into the right atrium and is directed through the tricuspid valve into the RV. With the next contraction, this blood is pushed into the pulmonary artery, but the pulmonary system is vasoconstricted, so only about 10% of blood enters the pulmonary circulation. The remainder of the blood takes the path of least resistance into the ductus arteriosus, a conduit into the descending aorta and the lower part of the body eventually landing in 1 of the 2 umbilical arteries and returning to the placenta.

With the infant’s first breath, an increase in PaO₂ results in a rapid decrease in pulmonary vascular resistance. As a result, blood preferentially flows from the right side of the heart to the low-resistance pulmonary circulation. Cessation of low-resistance placenta circulation immediately raises the systemic vascular resistance. With systemic vascular resistance higher than pulmonary resistance, the ductus arteriosus flow reverses, with blood now flowing from the aorta to the pulmonary artery. Over hours to days, the ductus arteriosus constricts and closes. The high flow and, thus, high pressure into the left atrium enables the foramen ovale to close. The RV thins as pulmonary resistance falls, whereas the left ventricle wall begins to thicken once exposed to higher systemic vascular resistance.

### IDENTIFICATION OF CARDIAC EMERGENCIES

Cardiac emergencies in infants can be categorized as structural or nonstructural. Structural defects can be further defined as either volume overload or obstructive (pressure overload) states. Nonstructural emergencies include arrhythmias (bradycardia and tachycardia) and disorders of myocardial function.

---

**TABLE 1. Prenatal (maternal) exposures as risk factors for congenital cardiac disease.**

<table>
<thead>
<tr>
<th>Exposure</th>
<th>Risk Factors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alcohol</td>
<td>ASD, VSD</td>
</tr>
<tr>
<td>Lupus</td>
<td>Complete heart block</td>
</tr>
<tr>
<td>Valproic acid</td>
<td>Coarctation of aorta, hypoplastic left ventricle,</td>
</tr>
<tr>
<td></td>
<td>AS, pulmonary atresia</td>
</tr>
<tr>
<td>Retinyl acid</td>
<td>Conotruncal anomalies</td>
</tr>
<tr>
<td>Diabetes</td>
<td>Hypertrophic cardiomyopathy, VSD, conotruncal</td>
</tr>
<tr>
<td>Phenylketonuria</td>
<td>VSD, ASD, PDA, coarctation of aorta</td>
</tr>
<tr>
<td>Rubella</td>
<td>PDA, peripheral pulmonic stenosis</td>
</tr>
</tbody>
</table>

Abbreviations: AS, aortic stenosis; ASD, atrial septal defect; PDA, patent ductus arteriosus; VSD, ventricular septal defect.
Several reliable online resources are available with diagrams and additional description of the full spectrum of congenital heart disease lesions. An example is the site maintained by the Johns Hopkins Helen Taussig Heart Center at www.pted.com.2

**STRUCTURAL DEFECTS: VOLUME OVERLOAD CONDITIONS**

When a left to right shunt exists, fully oxygenated blood from the left side of the heart circulates back to the right side of the heart, and forms a repeat circuit, through the pulmonary circulation. Examples of left to right shunt conditions include atrial septal defect (ASD), ventricular septal defect (VSD), atrioventricular (AV) canal defects, partial anomalous pulmonary venous return, and patent ductus arteriosus (PDA). The volume of blood passing through the shunt and, thus, the severity of the condition depends on differences between pulmonary and systemic vascular resistance. As neonatal circulation transitions, these pressures are labile, and thus, rapid changes in condition can occur. The decline of pulmonary vascular resistance in neonates occurs over hours to weeks. If a septal defect is present, the left to right shunt will be minimal soon after birth when the pulmonary vascular resistance is high, but as pulmonary vascular resistance falls, the shunt volume will increase, and the infant will develop right side of the heart overload and symptoms of congestive heart failure. Another example of volume overload is regurgitant valves. This is most commonly seen in patients with AV canal defects. Ebstein anomaly is an isolated regurgitation of the tricuspid valve.

The principal negative consequence of volume overload in neonates is poor pulmonary function. Increased vascular volume decreases pulmonary compliance, increasing work of breathing required for effective ventilation. At the same time, fluid leaks from the vasculature into the alveolar spaces (pulmonary edema) and impedes oxygenation. The clinical appearance of an infant with a volume overload state is hypoxia and respiratory distress; frequent signs include tachypnea, retractions, and wheezing. Infants will usually have decreased tolerance for feeding and often fail to keep pace with growth percentiles. In addition, volume overload and high right atrial pressures rapidly lead to hepatic congestion and hepatomegaly.

**Ventricular Septal Defect**

Ventricular septal defect is, by far, the most common congenital cardiac malformation, representing about 25% of congenital heart disease.3 The amount of shunting depends on the size of the defect and pulmonary and systemic vascular resistance. Defects less than 0.5 cm in diameter are termed restrictive because the small communication does not allow for significant left to right shunt. Defects greater than 1 cm in diameter are termed nonrestrictive and are associated with left to right shunting. Severity depends on the size of the defect and the infant’s pulmonary and systemic vascular resistance. In addition to the clinical features of volume overload, neonates with VSDs usually have an audible murmur. In very large defects, the flow across the defect may lack the turbulence necessary to generate an audible murmur. When present, a VSD murmur is best heard along the lower left sternal border and is described as harsh and holosystolic. Chest x-ray will show cardiomegaly, with prominence of both ventricles, left atrium, and pulmonary artery as well as pulmonary edema with increased pulmonary vascular markings. Electrocardiographic findings are nonspecific, showing biventricular hypertrophy.

**Atrial Septal Defect**

Atrial septal defects are described in 3 types: secundum, primum, or sinus venosus. A patent foramen ovale is not considered an ASD because the flap structure of the foramen does not usually allow shunting. As with VSDs, the degree of shunting depends on the size of the defect and the pulmonary and systemic resistance. Volume overload conditions should not become clinically apparent in the neonatal period, but prolonged stress on the RV can lead to eventual right sided heart failure later in life. In general, ASD should not cause significant illness in an infant, but presence of an ASD in conjunction with certain right side of the heart obstructive lesions (tricuspid atresia, transposition of the great arteries [TGA]) can allow enough shunting for the infant to survive past the newborn period without detection. Clinical signs of congestive heart failure are less likely than with VSD, and emergent presentation in the neonatal period is unlikely. A systolic murmur may be heard at the left upper sternal border, and the second heart sound (S2) may be widely split and fixed. Chest radiograph findings are not expected, and ECG findings are not apparent in the neonatal period.

**Partial Anomalous Pulmonary Venous Return**

Partial anomalous pulmonary venous return describes a portion of the pulmonary venous system
that does not connect to the left atrium but instead joins the systemic venous return into the right atrium. This circulatory loop creates a left to right shunt and overcirculation of the right side of the heart and pulmonary system. The degree of shunting depends on the proportion of pulmonary veins involved. Clinical signs are those common to volume overload conditions, including hypoxia, respiratory distress, and hepatomegaly. No murmur is associated. Chest radiograph may show lack of pulmonary venous markings. If the anomalous vein enters the inferior vena cava, a crescent shadow may be visible along the right border of the heart. This finding is known as Scimitar syndrome. Electrocardiographic findings are nonspecific during the neonatal period.

**Total Anomalous Pulmonary Venous Return**

Total anomalous pulmonary venous return is a more severe version where all pulmonary veins return to the systemic venous return system instead of emptying to the left atrium. This entry may be directly into the right atrium, into the superior vena cava, the inferior vena cava, or the veins may even descend below the diaphragm and enter the portal vein. The result is a total mixing lesion of oxygenated and deoxygenated blood and a lack of flow into the systemic circulation. An ASD must also be present for the neonate to survive. Infants with ASD will present with volume overload states. Chest x-ray findings include cardiomegaly and increased pulmonary vascular markings. The LV is poorly developed, and an ECG will show right ventricular hypertrophy.

**Endocardial Cushion Defect**

Endocardial cushion defects refer to a range of conditions (including complete AV canal, AV septal defects) in which one or more portions of the AV septum fails to form and/or the AV valves form abnormally. Presentation of these conditions range from very mild (ASD) to severe forms (complete AV canal). Complete AV canal is frequently associated with trisomy 21. Valve defects with regurgitation add to volume overload. Clinical signs are those common to volume overload conditions, including hypoxia, respiratory distress, and hepatomegaly. Auscultatory findings with this condition are variable, depending on the exact defect, but frequently include a wide fixed split of S2 and pulmonary ejection murmur. Infants with mitral insufficiency will have an apical holosystolic murmur that radiates to the axilla. Chest x-ray will show varying degrees of cardiomegaly. Electrocardiographic findings may include superior oriented QRS axis, RV hypertrophy, right bundle-branch block, left ventricular hypertrophy, or prolonged PR interval.

**Truncus Arteriosus**

A truncus arteriosus refers to an anomaly where the aorta and pulmonary artery arise as a single vessel directly above a large VSD, allowing total mixing of deoxygenated and oxygenated blood. As pulmonary vascular resistance falls, blood will preferentially flow into the pulmonary system, thus creating left to right shunting and volume overload. Truncus arteriosus is usually clinically apparent at the time of birth with cyanosis, but presentation can be delayed if pulmonary vascular resistance is adequately high. Chest x-ray will show cardiomegaly. Electrocardiogram will show biventricular hypertrophy and may lack the right-axis deviation expected in a neonate.

**Patent Ductus Arteriosus**

The normal ductus arteriosus differs from the aorta and pulmonary artery in that its middle layer contains circularly arranged smooth muscle. If the ductus arteriosus fails to close in a full-term infant in the first weeks of life, there is likely a deficiency of both the mucoid endothelial layer and muscular media of the vessel, and this vessel will never close on its own. The extent of left to right shunt depends on the ratio of pulmonary to systemic pressures and the size of the PDA. A large PDA can lead to congestive heart failure and failure to thrive. Clinical characteristics of PDA can be striking. Infants with a large PDA may have a wide pulse pressure and bounding arterial pulses due to runoff of blood into the pulmonary artery during diastole. A thrill may be felt along the second left intercostal space with radiation to the left clavicle, down the left sternal border, or to the apex. The defining characteristic of a PDA is that it has been described as a machinery-like continuous murmur that begins soon after the first heart sound, crescendos through systole and wanes through diastole. Chest x-ray may show a large pulmonary artery with increased pulmonary vascular markings. Electrocardiographic findings are nonspecific. If the PDA shunt is large, the ECG may have signs of left ventricular or biventricular hypertrophy.

**Conotruncal Anomalies: Double-Outlet RV, Double-Inlet Left Ventricle, and Tetralogy of Fallot**

The conotruncal anomalies include a variety of complex congenital heart disease. Numerous
variations of these conditions exist, and a detailed description is not necessary for emergency care. These conditions are marked by mixing of oxygenated and deoxygenated blood, usually with cyanosis apparent from birth, and may have increased or decreased pulmonary blood flow. Infants with increased pulmonary blood flow will present with signs of volume overload and congestive heart failure. Infants with decreased pulmonary blood flow will present with cyanosis. Tetralogy of Fallot is the most common of these conditions and frequently presents in the emergency department. Tetralogy of Fallot consists of 4 specific anomalies: (1) pulmonary stenosis, (2) VSD, (3) aorta overriding both the right and left ventricle outflow tracts, and (4) right ventricular hypertrophy. The combination of pulmonary stenosis and VSD creates a right to left shunt and cyanosis. Wide variability is seen in the degree of pulmonary stenosis. Infants with critical pulmonic stenosis are dependent on the PDA for blood flow to the lungs and will present with profound cyanosis when the ductus closes. Infants with mild stenosis of the pulmonary valve may be acyanotic ("pink Tet") and present with intermittent cyanosis or signs of heart failure. Clinical features common to neonates with tetralogy of Fallot include a loud, harsh systolic murmur at the lower left sternal border. Approximately 50% have an associated thrill. Chest x-ray findings include cardiomegaly and right ventricular hypertrophy with decreased vascular markings (Figure 2). This appearance is sometimes compared with a boot or a wooden shoe. An ECG should show right-axis deviation and right ventricular hypertrophy.

Infants with tetralogy of Fallot frequently present with cyanotic episodes or "Tet spells." These episodes commonly occur in early morning after waking and may be provoked by exercise or vigorous crying. The infant becomes fussy, cyanotic, and tachypneic and may proceed to syncope. Spells represent a temporary increase in right to left shunting secondary to decreased pulmonary blood flow. On cardiac auscultation, a previously noted murmur may be less prominent or absent because of the reduction of pulmonary blood flow. Therapy for these spells is directed at calming the infant and increasing pulmonary blood flow through a variety of methods: decreasing pulmonary vascular resistance (oxygen and morphine are potent pulmonary vasodilators) and increasing systemic venous return (knees-to-chest positioning or abdominal compression). In a prolonged spell, the infant may develop metabolic acidosis requiring treatment with sodium bicarbonate. β-Adrenergic blockade has been shown to be helpful especially in infants with severe crying and tachycardia. Drugs that increase systemic vascular resistance such as phenylephrine will increase right ventricular outflow, decrease right to left shunt, and thus, improve symptoms.

**Ebstein Anomaly**

Ebstein anomaly is a rare defect of the tricuspid valve where the tricuspid valve is displaced downward into the RV. The tricuspid valve is regurgitant, the right atrium is dilated and hypertrophied, and the RV is hypoplastic and dysfunctional. In addition, Wolff-Parkinson-White (WPW) syndrome is commonly associated with this anomaly. Neonates may present with cyanosis and right to left shunting. Clinical features of Ebstein anomaly include hepatomegaly and features of right sided heart failure. Massive dilation of the right atrium can cause respiratory distress by bronchial obstruction. Classic description of cardiac auscultation includes findings of split first heart sound and S2 as well as the presence of third heart sound and fourth heart sound. The tricuspid regurgitation murmur may be heard at the lower left sternal border as a soft systolic murmur. Extreme cardiomegaly can be seen on chest x-ray of neonates with Ebstein anomaly. Possible ECG findings include both short PR interval in 20% and long PR interval in 40%; right atrial hypertrophy and right bundle-branch block should be present. Neonates who present with decompensated Ebstein anomaly have an extremely high risk of neonatal death. In severely cyanotic infants, this is a ductus-dependent lesion. Immediate therapy with prostaglandin infusion, vasopressor infusion, and correction of metabolic acidosis will likely be necessary to maintain the infant before surgical intervention.

**STRUCTURAL HEART DEFECTS: OBSTRUCTIVE CONDITIONS**

Obstructive lesions present in 2 main ways. The more dramatic form is of critical interest to the emergency physician and will present in the early neonatal period with cardiovascular collapse because blood is unable to flow into the pulmonary or systemic circulation. These lesions may be present from the moment of birth or may not be clinically apparent until the ductus arteriosus closes. Incomplete forms of obstructive lesions will create pressure overload states that may not be apparent in the first weeks of life but will eventually lead to
ventricular hypertrophy or dysfunction. Table 2 lists the ductus dependent cardiac lesions.

### Pulmonary Valve Stenosis

Pulmonary outflow tract obstruction (critical pulmonary stenosis and pulmonary atresia) presents soon after birth with cyanosis and right sided heart failure. Pulmonary blood flow is supplied exclusively by retrograde flow through the PDA from the aorta to the pulmonary artery. Clinical presentation may initially be cyanotic because of right to left shunting of deoxygenated blood through the foramen ovale or signs of severe heart failure with hepatomegaly and peripheral edema. If they are not previously identified by these signs, infants will present with cardiovascular collapse when the ductus arteriosus closes. Chest x-ray findings are nonspecific, with cardiomegaly and decreased pulmonary vascular markings. Electrocardiogram shows LV hypertrophy because of a hypoplastic right ventricle and a relatively large left ventricle. Critical pulmonary stenosis requires emergency treatment with prostaglandin E₁ infusion.

### Coarctation of the Aorta

Presentation of coarctation is highly variable based on degree of narrowing. In the critical form, coarctation presents at time of closure of the ductus arteriosus. Less severe forms can present months to years later with ventricular hypertrophy and systemic hypertension. Neonates with critical coarctation appear pale with circulatory collapse and severe metabolic acidosis. Coarctation usually occurs in the descending aorta after the origin of the right and left subclavian arteries. Clinical findings that support the diagnosis of coarctation include diminished or absent femoral pulses, differential pulse oximetry readings on preductal (upper body) and postductal (lower body) sites or lower body cyanosis, and blood pressure differential between upper and lower extremities. A short systolic murmur may be heard at third or fourth intercostal space along the left sternal border. Chest x-ray may show cardiomegaly. Electrocardiogram is usually normal in the first weeks of life but, later, may show RV hypertrophy or right bundle-branch block.

### Hypoplastic Left Heart Syndrome

Hypoplastic left heart syndrome (HLHS) includes a constellation of anomalies including mitral and aortic valve atresia and hypoplasia of the aortic root. Systemic blood flow is supplied exclusively via the PDA. Newborns can have surprisingly few symptoms at birth then present suddenly with cardiovascular collapse as the ductus closes. Chest x-ray findings are nonspecific. Electrocardiographic findings of HLHS include peaked P waves and RV hypertrophy. Without surgical intervention, HLHS is uniformly fatal in the neonatal period. Emergent care is directed at supporting flow through the ductus arteriosus with prostaglandin infusion and supportive critical care to allow the infant to survive to surgery.

### Transposition of the Great Vessels

In transposition of the great vessels, alternatively known as TGAs, the origin of the 2 great vessels is switched. The aorta arises from the RV, thus carrying deoxygenated blood back to the body. The pulmonary artery arises from the LV and carries oxygenated blood back to the lungs. Transposition of the great arteries with an intact atrial septum will present at birth with profound cyanosis. If an ASD or patent foramen ovale is present and allows adequate mixing, it is possible that a child could be only mildly symptomatic at birth, but when the ductus arteriosus begins to close, the child will become more cyanotic and rapidly collapse.

### Tricuspid Atresia

Tricuspid atresia blocks outflow from the right atrium into the RV. Deoxygenated blood must shunt right to left through the foramen ovale. Flow to the pulmonary system is limited to left to right shunt through the ductus arteriosus. Cyanosis is usually obvious at birth, but some infants will have intermittent hypoxia only. Neonates will present with cyanosis as the ductus closes. Chest x-ray findings are subtle, without cardiomegaly, but pulmonary vascular markings are usually decreased. Hypoplasia of the RV is reflected in the ECG with superior QRS axis, right atrial hypertrophy and left atrial hypertrophy, and LV hypertrophy.
NONSTRUCTURAL CARDIAC EMERGENCIES IN NEONATES

Arrhythmia
Bradycardia in newborns occurs most frequently secondary to other illness such as hypoxia, hypoglycemia, hypothermia, and sepsis. Congenital complete AV block is most often seen in infants born to mothers with systemic lupus erythematosus or other collagen vascular disease. Electrocardiogram should reveal a junctional escape rhythm and rate of 60 to 80. If the infant is symptomatic because of bradycardia, congenital complete AV block is treated with pacing. Tachyarrhythmias such as supraventricular tachycardia can present in the early neonatal period, and are often associated with structural congenital heart disease. Arrhythmias are discussed elsewhere in this issue.

Myocardial Dysfunction
A wide variety of conditions can lead to myocardial dysfunction in neonates. Signs of congestive heart failure and poor contractility can be seen in infants with overwhelming sepsis or hypoxia secondary to respiratory disorders. More esoteric possibilities include inborn errors of metabolism, viral myocarditis, and congenital cardiomyopathy.

Anomalous Left Coronary Artery
The normal origin of the coronary arteries is from the coronary ostia at the base of the aorta. In this rare condition, the left coronary artery arises instead from the pulmonary artery. The myocardium is therefore supplied with deoxygenated blood at relatively lower pulmonary vascular pressures. As pulmonary resistance declines in the first weeks of life, coronary perfusion slows, and myocardial ischemia and infarction occur in early infancy. This condition has been suggested as a possible etiology of colic and as a potential mimic of bronchiolitis.8,9 Infants typically present with signs of dilated cardiomyopathy and congestive heart failure: feeding intolerance, respiratory distress with wheezing, and hepatomegaly. Chest x-ray will reveal cardiomegaly and pulmonary edema in late stages. Before infarction and failure, ECG will show classic ischemic changes such as ST elevations and T-wave inversions in the precordial leads. Late ECG findings are consistent with old infarction, particularly deep and wide Q waves in the precordial leads. The diagnosis is made by echocardiogram. Treatment is supportive, pending urgent surgical reimplantation of the coronary arteries.10

DIAGNOSTIC TESTING AND THERAPY

Interpretation of Chest Radiograph
A chest radiograph should be obtained in the infant with suspected cardiac disease. A short list of simple assessments by the emergency physician can narrow the differential diagnosis: measurement of heart size, assessment of pulmonary vascular markings, and presence of a right-sided aortic arch. Most classic descriptions of common lesions use combinations of

Figure 1. Chest radiograph of infant with tetralogy of Fallot. Boot shape of the heart results from right ventricular hypertrophy.
TABLE 3. Stepwise approach to chest x-ray interpretation in infants with suspected cardiac disease.

<table>
<thead>
<tr>
<th>Question</th>
<th>Answer</th>
</tr>
</thead>
<tbody>
<tr>
<td>Is there obvious cardiomegaly?</td>
<td>If yes, increases likelihood of congenital heart disease or cardiac failure. Predominantly volume overload states. Massive cardiomegaly suggests Ebstein anomaly or cardiomyopathy.</td>
</tr>
<tr>
<td>Is there evidence of increased pulmonary vascular markings?</td>
<td>Increased pulmonary vascular markings suggests a pulmonary overflow state, with inadequate systemic flow. Conversely, lack of expected pulmonary vascular markings suggests obstruction of pulmonary circulation.</td>
</tr>
<tr>
<td>Is there a right-sided aortic arch?</td>
<td>Tetralogy of Fallot, truncus arteriosus, tricuspid atresia</td>
</tr>
<tr>
<td>Is there normal abdominal situs?</td>
<td>Situs inversus suggests TGAs.</td>
</tr>
</tbody>
</table>

Interpretation of Infant ECG

Principles of electrocardiography are the same regardless of age. The sinoatrial node located in the right atrium is the pacemaker for the heart. The sinoatrial node sends out an electric impulse that simultaneously depolarizes the right and left atria, producing a P wave. As the electrical impulse passes through the AV node, the conduction slows, producing the PR interval. The electrical impulse then travels through the right and left branches of the Bundle of His to depolarize the ventricular muscle, thus producing the QRS complex. The T wave is created by the repolarization of the ventricles.

The normal neonatal heart rate ranges between 110 and 150 but varies greatly and increases with crying, fever, or activity. From birth through the first month of life, the RV muscle is thicker than the left ventricle; thus, the ECG tracing reveals right-axis deviation and right ventricular hypertrophy. The normal infant QRS axis is right and anterior (+135 to +180), whereas R-wave amplitude is higher in the right precordial leads (V1 and V2) and the S-wave amplitude peaks in the left precordial leads (V5 and V6). T waves are typically inverted in all precordial leads in infants (Figure 2). In infants older than 3 days, an upright T wave in V1 is highly unusual and suggests right ventricular hypertrophy. Presence of a superior axis (−1 to −180) suggests the presence of an AV canal or tricuspid atresia (Figure 3). Table 4 lists additional ECG abnormalities that can be seen in neonatal cardiac disease.

Laboratory Findings

A limited number of laboratory studies may be specifically useful in the evaluation and treatment for infants with congenital heart disease. It is likely that most sick neonates will undergo a standard comprehensive laboratory evaluation to eliminate other diagnostic considerations such as infectious and metabolic diseases. From common screening laboratory testing, renal and hepatic functions are the most likely to be abnormal in poorly perfused neonates. Arterial blood gas measurement may provide specific diagnostic information and should be pursued in neonates with suspected cardiac disease. The hyperoxia test measures PaO2 in the right radial artery while the neonate is receiving 100% oxygen. Demonstration of arterial oxygen content less than 150 mm Hg suggests a cardiac mixing lesion. In addition, measurement of arterial pH may prompt specific therapy for metabolic acidosis. Lactate levels may be useful to evaluate the impact of the low perfusion state and guide critical care therapies. Troponin levels have been shown to correlate with myocardial injury and in neonates with severity of volume overload with PDA but are not a specific marker and are also elevated in neonatal respiratory distress syndromes.11-13

Echocardiography

For an infant with actual or impending circulatory collapse, an echocardiogram should be obtained as urgently as possible to define the lesion and guide surgical or medical therapy. The emergency provider proficient with the focused assessment with sonography in trauma (FAST) examination may be comfortable using bedside ultrasound to obtain a subxyphoid view of the heart, but it cannot substitute for echocardiogram by an experienced examiner.

Therapeutic Interventions

The crux of advanced therapeutic management of the infant with cardiogenic shock is balance of pulmonary and systemic blood flow. For infants with...
Figure 2. Infant ECG with right ventricular hypertrophy. Signs of right ventricular hypertrophy include larger than normal amplitudes of R and inverted T waves in right precordial leads.
Figure 3. Electrocardiogram of infant with complete AV canal defect. Note the superior axis deviation. QRS is downgoing in aVF, indicating electrical axis “away” from the lower body.
ductal dependent lesions, this includes supporting patency of the ductus arteriosus.

**Prostaglandin**

Prostaglandin E₁ (PGE₁) infusion should be considered immediately when infants present with severe hypoxia or shock in the first 1 to 2 weeks of life. A list of ductus-dependent lesions that may benefit from PGE₁ infusion is found in Table 2. However, the decision to initiate PGE₁ infusion should be made on clinical grounds and not wait for definitive diagnosis. Prostaglandin E₁ halts closure of the ductus arteriosus and can result in rapid clinical stabilization of the infant with a ductus-dependent lesion. Initial dosing includes a bolus of 0.1 mg/kg followed by continuous infusion at a rate of 0.05 to 0.1 μg/kg per minute. Prostaglandin infusion does not require central venous access but does require a dedicated intravenous site to ensure continuous infusion.

Prostaglandin infusion has several frequent and severe adverse effects. Apnea occurs in 12% of neonates receiving PGE₁, and for that reason, elective intubation should be considered before transport of an infant on prostaglandin infusion. Other potentially life-threatening adverse effects of prostaglandin infusion include bradycardia, hypotension, and seizures. Fever occurs in about 10% and may complicate clinical diagnosis by raising suspicion of sepsis. Flushing, or cutaneous vasodilatation, also occurs in about 10%.

**Ionotropes and Vasopressors**

Several inotropic agents may be useful for emergent stabilization of the infant with cardiogenic shock. Choice of agent should be based on the relative balance of pulmonary and systemic blood flow. Ideally, the ratio of pulmonary flow to systemic flow should be 1:1. Pulmonary and systemic blood flow exists as a zero-sum equation; when the pulmonary circuit is overcirculated, the systemic circulation is hypoperfused and vice versa. In addition to any effects on cardiac contractility or heart rate, available inotropes act to increase or decrease systemic vascular resistance. When blood is shunted toward the pulmonary circulation, the goal of therapy is to increase systemic circulation, and thus, the preferred inotropic agents are those that will reduce systemic vascular resistance such as milrinone and dobutamine. When blood is

---

**TABLE 4. Electrocardiographic interpretation for infants with suspected cardiac disease.**

<table>
<thead>
<tr>
<th>Assess rate</th>
<th>Rate &lt;80 beat/min</th>
<th>Bradycardia</th>
<th>Congenital AV block, hypoxia, hypothermia</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Rate &gt;160 beat/min</td>
<td>Tachycardia</td>
<td>Sinus tachycardia, ventricular tachycardia, Supraventricular tachycardia</td>
</tr>
<tr>
<td>Axis determination</td>
<td>QRS upright in lead I and downward in aVF</td>
<td>Right-axis deviation</td>
<td>Expected in healthy infants</td>
</tr>
<tr>
<td></td>
<td>QRS upright in aVF</td>
<td>Superior axis</td>
<td>Endocardial cushion defect or tricuspid atresia</td>
</tr>
<tr>
<td>Assess PR interval</td>
<td>The PR interval is most easily measured in lead II</td>
<td>PR interval long</td>
<td>Ebstein anomaly, hypoxia, digitalis toxicity, hyperkalemia</td>
</tr>
<tr>
<td></td>
<td>PR interval short</td>
<td>Severe right ventricular hypertrophy, transposition of great vessels, single ventricle, mirror image dextrocardia</td>
<td></td>
</tr>
<tr>
<td>Assess QRS</td>
<td>Q waves present in V₁ not in V₆</td>
<td>Ventricular hypertrophy (left, right or bi-ventricular hypertrophy), anomalous left coronary artery, idiopathic hypertrophic subaortic stenosis</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Deep Q waves</td>
<td>Ventricular hypertrophy</td>
<td></td>
</tr>
<tr>
<td></td>
<td>QRS amplitude large</td>
<td>Endocardial cushion defect, coarctation of the aorta</td>
<td></td>
</tr>
<tr>
<td>Assess T waves</td>
<td>Tall or peaked T waves</td>
<td>Hyperkalemia, left ventricular hypertrophy</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Low or flat T waves</td>
<td>Normal newborn, hypoglycemia, myocardial ischemia</td>
<td></td>
</tr>
</tbody>
</table>
shunted away from the pulmonary circulation, the goal of therapy is to decrease systemic circulation, and thus the preferred inotropes are those that will raise systemic vascular resistance such as high-dose dopamine and epinephrine. Inhaled nitrous oxide and sildenafil are 2 pharmacologic agents that act on pulmonary vascular resistance, although they are unlikely to be initiated in the emergency department setting.

**Sodium Bicarbonate**

Use of sodium bicarbonate to treat metabolic acidosis is fraught with controversy. Extreme acidosis places further stress on the heart and brain. However, the weight of current evidence suggests that administration of bicarbonate does more harm than good in neonates with metabolic acidosis secondary to hypoperfusion. Use of bicarbonate has been shown to increase troponin levels in children with acute renal failure and to depress myocardial function in animal models.\(^\text{15,16}\) In addition, bicarbonate does not cross the blood brain barrier, whereas carbon dioxide crosses freely. Therefore, administration of bicarbonate may paradoxically worsen cerebral acidosis. There is consensus that the initial priority must be on correction of respiratory acidosis and support of circulation.\(^\text{17}\)

**SUMMARY**

Emergent care of the neonate with congenital heart disease includes recognition of an unstable condition and standard resuscitative measures to stabilize. Airway management with intubation and ventilatory support should be strongly considered for critically ill infants. For infants with suspected ductal-dependent lesions, prostaglandin infusion should be initiated as rapidly as possible. The emergency physician can use standard chest radiography and ECG information to narrow the diagnostic possibilities, but emergent echocardiography and pediatric cardiology consultation are indicated. If interfacility transport is required for access to subspecialty services, the stability of the neonate and capabilities of the transport team should be carefully considered, with particular assessment of need for prophylactic intubation before transport.

**REFERENCES**