Cardiac Evaluation of the Newborn

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KEYWORDS
• Term newborn cardiovascular examination
• Term newborn with congenital heart disease (CHD)
• Common types of neonatal congenital heart disease • Pulse oximetry screening
• Critical congenital heart disease (CCHD)

KEY POINTS
• Although congenital heart defects can be diagnosed using fetal cardiac ultrasonography, some defects can be challenging to identify.
• Even with a careful complete physical examination, some infants seem normal and are discharged home undiagnosed.
• The persistence of fetal channels can mask the presence of critical congenital heart disease, and the rather short postpartum hospital stay contributes to the diagnostic challenges.
• It is essential for the examiner to use all physical examination skills, including inspection, palpation, and auscultation, and to perform more than one physical assessment before discharge or shortly thereafter.
• The recent introduction of Pulse Oximetry Screening has been an extremely helpful adjuvant in assisting with the diagnosis of CCHD.

CARDIAC EVALUATION OF THE NEWBORN

The approach to the cardiac evaluation of a newborn can be challenging. As a result, many pediatricians report that they often feel uncomfortable when it comes to differentiating the normal from the abnormal state with regard to a newborn’s

Declaration of conflict of interest: Both R.L. Bucciarelli, MD and D.J. Fillipps, MD attest that they have no conflicts of interest to declare in relation to the materials and information provided in this article.

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http://dx.doi.org/10.1016/j.pcl.2014.11.009
0031-3955/15/$ – see front matter Published by Elsevier Inc.
cardiovascular examination. It is the authors’ goal for this article to provide the reader with the background knowledge that will make the cardiac evaluation of newborns less intimidating and assist the general pediatrician in understanding, detecting, and treating a newborn with congenital heart disease (CHD).

CHD is the most common congenital disorder in newborns, occurring in approximately 8 out of 1000 live births, and is responsible for almost 30% of infant deaths related to birth defects. Of those children with CHD, about 1 in 4 (25%) babies born with a heart defect will have critical CHD (CCHD), defined as needing intervention within the first year of life.¹⁻³

Although CHD can be diagnosed using fetal cardiac ultrasonography, some defects can be challenging to identify. Similarly, even with a careful complete physical examination, some infants seem normal and are discharged home undiagnosed. The persistence of fetal channels can mask the presence of CCHD, and the rather short postpartum hospital stay contributes to the diagnostic challenges. Thus it is essential for the examiner to use all physical examination skills, including inspection, palpation, and auscultation, and to perform more than one physical assessment before discharge or shortly thereafter. The recent introduction of pulse oximetry screening (POS) has been an extremely helpful adjuvant in assisting with the diagnosis of CCHD before signs of decompensation occur.⁴

**Initial Evaluation**

The first step in the assessment of the newborn infant’s cardiovascular system is a careful review for conditions that are associated with an increased risk of CHD (Table 1). The presence of any of these factors should raise the index of suspicion, but a complete physical examination should be performed regardless.⁵⁻⁷

**Inspection and Palpation of the Skin and Mucous Membranes**

The color of the skin and briskness of capillary refill can be indicators of the adequacy of oxygenation and cardiac output. The mucous membranes of a normal newborn should be pink. This is usually checked by looking at the tongue and lips. When light

<table>
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<th>Table 1 Common conditions associated with CHD</th>
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<td><strong>Maternal</strong></td>
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<td>Diabetes</td>
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<td>Influenza or flulike symptoms</td>
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<td>Maternal alcohol/medication use</td>
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<td>Multifetal pregnancy</td>
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*Abbreviations: TORCH, toxoplasmosis, other agents, rubella, cytomegalovirus, herpes simplex; VACTERL, vertebral, anal, cardiac, tracheal, esophageal, renal, and limb.*

*Data from Refs. ¹⁻³,⁹*
pressure is applied to the skin or nail beds, normal color should return within 3 to 4 seconds after the pressure is released (capillary refill time).

Acrocyanosis is usually described as cyanosis of the distal portions of the extremities but can be seen around the mouth and in the nail beds. However, the mucous membranes generally remain pink. Acrocyanosis is common in newborns and is normal. It can be caused by vasomotor instability, vasoconstriction caused by cold, or polycythemia. The degree of acrocyanosis can be related to the level of hematocrit and is most obvious with a central hematocrit of 65% or greater. Acrocyanosis is increased with crying and fades when sleeping. It is usually uniformly distributed in the arms and legs but may have an asymmetric pattern being more obvious in certain extremities. However, distinct differences in appearance between upper and lower parts of the body should raise concern and be investigated. Determination of a central hematocrit and peripheral hemoglobin saturation can be helpful. With acrocyanosis caused by polycythemia, the hematocrit will be elevated and hemoglobin saturation will be 90% to 95%. A normal hematocrit and/or abnormal hemoglobin saturation should prompt further investigation.

Central cyanosis is always abnormal. This condition may be caused by primary pulmonary disease or CCHD, which restricts pulmonary blood flow (PBF) (Box 1). Cyanosis caused by pulmonary disease is often responsive to the administration of oxygen. Central cyanosis caused by CCHD does not change significantly when patients are placed in an oxygen-enriched environment.

Mottling or pallor can be a sign of diminished cardiac output as blood is shunted away from the skin to support more central organs and tissues. The capillary refill time is prolonged, and a significant metabolic acidosis may be present. There are both common cardiac and noncardiac causes of compromised cardiac output, and they must be investigated (Box 2).

**Assessment of Peripheral Pulses**

Palpation of the brachial and femoral pulses is an essential element of the cardiovascular examination of the newborn. On first palpation, the examiner should assess the pulse rate (normal between 100 and 160 beats per minute) and rhythm (consistent rhythm without irregular beats). Although it is acceptable to palpate the pulses in each extremity separately, it is a good practice to also palpate one femoral pulse simultaneously with each of the brachial pulses. This practice gives one the opportunity to not only assess the pulse amplitude but also the timing of the pulses in the arms and the legs. Delays in the timing of the pulses between the upper and lower extremities is suggestive of abnormality in the aortic arch.

**Box 1**

**Congenital heart lesions producing a decrease in PBF**

- PS with intact ventricular septum
- PA with intact ventricular septum
- Tetralogy of Fallot (PS/PA with VSD)
- Tricuspid atresia
- Hypoplastic right heart syndrome
- Epstein anomaly of the tricuspid valve

**Abbreviations:** PA, pulmonary atresia; PS, pulmonary valve stenosis; VSD, ventricular septal defect.
After noting the rate and timing of the pulses, their amplitude and character should be noted. Amplitude of the peripheral pulses is frequently graded as 0 to 4+, with 0 being not palpable and 4+ being bounding.

**Assessment of the Breathing Pattern**

The normal respiratory pattern of a term neonate is regular and effortless. The breath sounds are usually very quiet and distant and may frequently be detected only with close observation and auscultation. Babies generally show an abdominal breathing pattern because of their weak chest wall muscles and diaphragm-directed breathing. A common additional pattern known as **periodic breathing** may also be noted. Periodic breathing is a normal variation of breathing found in premature and full-term infants. The examiner will notice pauses in breathing for less than 10 seconds, followed by a series of rapid, shallow breaths. Breathing then returns to normal without any stimulation or intervention.8

Changes in respiratory rate and effort are frequently the result of changes in lung compliance, a reflection of the stiffness of the lung and its ability to change volume in relation to a change in pressure.9 Various types of CCHD can have a profound effect on pulmonary compliance, leading to predictable signs on physical examination.

When PBF is reduced, there is less blood in the lungs and compliance increases. Breathing becomes easier. Diminished PBF also produces hypoxemia, which stimulates the respiratory center in the brain stem and results in hyperpnea, effortless tachypnea with increased tidal volume. The baby seems cyanotic but has no significant respiratory distress. Because of the hyperpnea, PaCO₂ is often low. Effortless tachypnea in a cyanotic baby is the hallmark of CCHD caused by right heart obstructive lesions.6,8

CCHD that increases PBF increases the amount of blood in the lungs, leading to pulmonary edema and congestive heart failure (CHF). Pulmonary compliance is reduced; work of breathing is increased leading to tachypnea, grunting, alar flaring, and intercostal retractions. Because cardiac output is frequently diminished, the skin is mottled with increased capillary refill time. Often a mixed metabolic and respiratory acidosis is present. The possibility of CCHD should be considered in any term infant presenting with significant respiratory distress but whose history and initial evaluation does not support primary pulmonary disease.10

**Box 2**

**Common causes of compromised cardiac output**

- Sepsis
- Hypoplastic left heart syndrome
- Myocardial dysfunction with tricuspid regurgitation caused by asphyxia
- Great vein of Galen aneurysm
- Sustained supraventricular tachycardia
- Inborn errors of metabolism

**Inspection, Palpation, and Auscultation of the Heart**

Similar to the examination of the lungs, the normal cardiac activity in the term neonate is barely visible with inspection and the precordium is usually quiet to palpation. A slight parasternal lift can usually be seen and palpated along the left sternal boarder, secondary to the normal right ventricular dominance seen in term newborns.
Auscultation of the heart should be performed using a high-quality stethoscope with a small bell and diaphragm. The examination is best performed by starting at the base of the heart (second left and right intercostal spaces), moving down the sternal boarder to the apex. Both the bell and diaphragm should be used while the examiner concentrates on heart rate, rhythm, cardiac sounds, and any murmurs heard. The normal heart rate of a term neonate is usually between 100 and 160 beats per minute. However, heart rates may exceed 180 when the baby is crying and can go as low as 70 when in a deep sleep. The rhythm should be regular without extra or skipped beats. The examiner may notice a predictable change in rate associated with respirations, the sinus arrhythmia, which is entirely benign.

Normal heart sounds are generated by closure of the tricuspid and mitral valves (S1) and by closure of the pulmonary and aortic valves (S2). As PBF increases after birth, the pulmonary valve closes later causing audible splitting of S2. A split S2 indicates the presence of 2 normally positioned great vessels, suggests that the pulmonary vascular resistance is low, and indicates the presence of 2 normal ventricles separated by a ventricular septum. Thus the presence of splitting strongly suggests normal cardiac anatomy; however, its absence does not imply the presence of CHD. Occasionally extra heart sounds (S3, S4, or a summation S3/S4) can be heard. Although these sounds can be heard in some normal situations, their presences should prompt further evaluation.

**Cardiac Murmurs**

Murmurs are caused by turbulent blood flow, either by increased flow through normal vessels and valves or normal flow through abnormal valves, vessels, or septal defects. Murmurs are graded 1 to 6 and described as to whether they occur in systole or diastole. Attention should be also given to the quality of the murmurs (harsh, smooth, or vibratory) and whether or not they are only heard in a small area or radiate more widely throughout the chest. There are numerous common innocent murmurs that can be detected in newborns. The pulmonary flow murmur is probably one of the most common. It is heard best in the upper left sternal border (ULSB) and transmits well to both sides of the chest, axilla, and the back. It is usually soft and generally not louder than grade 2/6 intensity. Another innocent murmur is the transient systolic patent ductus arteriosus (PDA) murmur, which is audible at the ULSB and in the left infraclavicular area during the first days of life. It is thought to be caused by a closing ductus arteriosus and usually disappears shortly after the first day. Frequently a transient grade 1-2/6 murmur of tricuspid regurgitation can be heard in the fourth intercostal space to the right of the sternum. Transient tricuspid regurgitation is most commonly heard in stressed newborns with mild to moderate pulmonary hypertension and generally resolves before discharge as pulmonary vascular resistance falls. Finally a soft, vibratory, low-frequency murmur is often best heard at the left lower sternal boarder, which may persist for some time. This murmur is also benign and often referred to as merely a functional murmur.

Pathologic murmurs heard in the neonatal period are often of grade 3 to 6 intensity. They are usually harsh in quality, occur in systole, and can be heard all over the chest and into the back. These murmurs are frequently associated with CCHD.

**Examination of the Abdomen**

When assessing for CCHD, it is also important to inspect and palpate the abdomen. A neonate with cyanosis most likely will have a normal abdominal examination. However, infants with overcirculation of the lungs and respiratory distress may develop abdominal distention caused by aerophagia, which can further compromise respiratory mechanics. These infants would benefit from gastric decompression.
with CHF may have liver enlargement and, if a metabolic acidosis is present, may develop an ileus with diminished or absent bowel sounds on auscultation.

**Blood Pressure**

Determination of blood pressure in both arms and at least one leg is important in the evaluation of an infant suspected of having CCHD. The pattern of variation in pressure between the extremities can suggest the presence of significant CCHD (Table 2).

### COMMON CONGENITAL HEART DEFECTS

In this section, the authors discuss only the most common congenital heart defects. It is not important that the examiner arrive at the correct anatomic diagnosis in their assessment of a newborn with suspected CCHD. Rather, it is important to recognize the general presenting signs of CCHD and how to stabilize the infant until further evaluation can be accomplished.

**Uncomplicated Congenital Heart Defects**

Simple atrial septal defects (ASD), ventricular septal defects (VSD), endocardial cushion defects, and a patent ductus arteriosus (PDA) do not significantly affect the cardiopulmonary physiology of the newborn and, although possible, are not usually associated with symptoms in the first few days after birth. The blood crossing these defects is usually small in volume and nonturbulent, producing little to no murmur. With simple ASDs, even very large ones, the amount of blood that goes from the left atrium to the right atrium is very limited until several months after birth, when the right ventricular muscle becomes thinner, more compliant, and can accommodate additional blood. With VSDs, flow depends on the reduction in pulmonary vascular resistance, which occurs over the first 3 to 6 weeks after delivery. This evolution in the pulmonary vascular bed occurs more rapidly in premature infants, leading to earlier identification and an increased likelihood of symptoms. One exception to this rule is a very small muscular VSD that may be heard within the first days of life because flow through the defect is turbulent. Although the murmur may be a grade 2–3/6, it is very short in duration, smooth in character, and mid to high frequency. These infants should be asymptomatic.

**Lesions Causing Decreased Pulmonary Blood Flow**

The lesions discussed in this section and presented in Box 1 all have a significant degree of obstruction of blood flow into and/or out of the right ventricle.

**Pulmonary valve stenosis and pulmonary valve atresia with an intact ventricular septum**

In pulmonary valve stenosis (PS), the pulmonary valve is thickened and only allows a jet of blood to pass into the lungs. Because this jet is turbulent, it creates a loud,
harsh systolic murmur at the base of the heart in the second intercostal space along the left and right sternal boarder. The murmur is also well heard along the back. Because the valve does not close properly, $S_2$ is single and diminished. With severe pulmonary stenosis, there is often massive tricuspid regurgitation, producing a grade 3 to 6 (murmur plus a thrill) at the third to fourth intercostal space along the right sternal margin. If there is no opening to the valve at all (pulmonary atresia [PA]), the obstruction is complete. The pulmonary valve and the main pulmonary arteries are underdeveloped. The baby is deeply cyanotic, but no murmur is heard over the precordium. There may be a faint (grade 2-3), smooth systolic murmur of a PDA heard along the ULSB and under the left clavicle. In this instance, the infant’s entire PBF depends on the ductus. Because the tricuspid valve is competent, the pressure in the right ventricle is greater than systemic levels. Blood flow into the ventricle is minimal and leaves the chamber through the myocardium sinusoids, which drain into the coronary system. As a result, the right ventricle is small, underdeveloped, and nonfunctional. This combination of lesions is also known as the hypoplastic right heart syndrome (Fig. 1). However, when PS or PA exists in association with tricuspid valve incompetence, the pressure in the right ventricle is very low, allowing blood to enter during ventricular diastole and then flow retrograde into the right atrium during ventricular systole. This antegrade/retrograde flow creates enough volume variation to allow near-normal development of the right ventricle. Babies with PS/PA may

![Fig. 1. Hypoplastic right ventricle with pulmonary atresia. An infant with severe PS or PA and a competent tricuspid valve (A) may have a severely underdeveloped and nonfunctional right ventricle. The infant is deeply cyanotic with little to no audible murmur. However, if the tricuspid valve is better developed and incompetent (B), there is both antegrade and retrograde flow into and out of the right ventricle, which remodels the ventricular wall, resulting in a much larger, functional chamber. In this situation there will be a loud murmur (grade iv/vi) along the right lower sternal boarder. A palpable thrill may also be present. Ductus A, Ductus Arteriosus; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle. (Adapted from Krovetz LJ, Gessner IH, Schiebler GL. Handbook of pediatric cardiology. 2nd edition. Baltimore: University Park Press; 1979. p. 301; with permission.)](image-url)
develop a large right atrium and are at risk for developing supraventricular tachycardia (SVT), which is briefly discussed later.

**Pulmonary valve stenosis/pulmonary valve atresia with a ventricular septal defect**

This combination of lesions is one of the most common types of CHD and is also known as tetralogy of Fallot (TOF). The 4 elements of TOF are PS or PA, VSD, overriding aorta, and right ventricular hypertrophy (RVH) (Fig. 2). RVH is not obvious because right ventricular dominance is common in the term neonate. The presenting signs depend on the degree of pulmonary obstruction. Severe obstruction produces deep cyanosis, whereas minor degrees of stenosis may affect color only slightly, hence the term *Pink Tetralogy*. In addition to PS, there is usually narrowing below the pulmonary valve, which is called muscular infundibular stenosis. S₂ is single because of PS and subvalvular stenosis. The VSD is always large with little restriction of flow such that blood flows easily from the right ventricle to the left ventricle with little turbulence, generating no murmur and allowing normal ventricular development. The aorta straddles the ventricular septum (overrides) and receives blood from both ventricles. The murmur heard in an infant with TOF is similar to the murmur of the PS. If tetralogy exists with pulmonary atresia, there may be no murmur at all or only the faint murmur of a PDA, which supplies all of the PBF.

**Dextro-transposition of the great vessels**

D-transposition of the great vessels (TGV) is one of the more common defects. In this case, the main pulmonary artery arises from the left ventricle and the aorta from the left atrium; hence, the pulmonary artery is fed by the right ventricle and the aorta by the left. As a result, the oxygenated blood is directed to the systemic circuit instead of the pulmonary circuit.

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*Fig. 2. Cyanotic TOF. Note the presence of PS with additional muscular narrowing in the infundibular region. The aorta is overriding the ventricular septum and receives blood from both the right and left ventricle. A, aorta; LA, left atrium; LV, left ventricle; PT, pulmonary trunk; PS, pulmonary valve stenosis; RA, right atrium; RV, right ventricle. (Adapted from Krovetz LJ, Gessner IH, Schiebler GL. Handbook of pediatric cardiology. 2nd edition. Baltimore: University Park Press; 1979. p. 288; with permission.)*
right ventricle. The volume of PBF is normal; but because the origin of the great vessels is switched, oxygenated blood merely recirculates to the lungs and deoxygenated blood recirculates to the body. Mixing of the circulations only occurs across the atrial septum and the PDA. There are usually no murmurs because there is no turbulent flow. S₂ is single because the pulmonary artery is malpositioned and hidden by the aorta. Most frequently TGV occurs with an intact ventricular septum, presenting with deep cyanosis. However, it can also be associated with a VSD or a VSD and PS. It then takes on the characteristics of the other lesions described throughout this section.

**Lesions Causing Increased Pulmonary Blood Flow**

Lesions that cause increased PBF almost always involve obstruction to flow on the left side of the heart (Box 3). These lesions can quickly produce severe CHF because they often involve pressure overload of the left ventricle associated with the obstruction and volume overload of the right ventricle caused by an associated ASD and VSD. It is important to consider the possibility of left heart obstruction in any term neonate who has a period of well-being and then develops respiratory distress, a mottled appearance of the skin, with hypotension and shock. Unlike defects associated with right-sided lesions, left-sided lesions create turbulent flow and demonstrate increased heart activity with loud systolic murmurs. Careful attention to the pattern of blood pressure and pulse can give the examiner insight into the location of the lesion (see Table 2).

**Aortic valve stenosis**

Patients with mild, uncomplicated aortic valve stenosis (AS) usually do not have difficulty as newborns. However, more significant degrees of stenosis, so-called critical AS, cause symptoms at an early age. They present with a loud harsh systolic murmur at the base of the heart to the right of the sternum, radiating well into the carotids. Blood pressure and pulses are normal with mild disease but are uniformly diminished in all extremities with critical AS. S₂ is single because of delayed aortic valve closure. Extra sounds (ejection clicks and S₃–S₄) may be heard. CHF can develop quickly in infants with critical AS.

**Coarctation of the aorta and coarctation of the aorta with a ventricular septal defect**

A discrete, isolated coarctation of the aorta does not usually cause symptoms in the first few days of life. It is often detected on follow-up examination when upper

<table>
<thead>
<tr>
<th>Box 3</th>
<th>Congenital heart lesions producing an increase in PBF</th>
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<tr>
<td>Atrial septal defect</td>
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<td>Ventricular septal defect</td>
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<tr>
<td>Endocardial cushion defect</td>
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<td>Aortic valve stenosis</td>
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<tr>
<td>Aortic valve atresia</td>
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<tr>
<td>Hypoplastic left heart syndrome</td>
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<tr>
<td>Discrete coarctation of the aorta</td>
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<td>Long segment coarctation of the aorta with VSD</td>
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<td>Total anomalous pulmonary venous return</td>
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<td>Single ventricle, double inlet left ventricle, and double outlet right ventricle</td>
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extremity hypertension is noted in both arms and diminished blood pressure and pulses are present in the legs. The coarctation itself does not produce any audible murmurs; however, an abnormal aortic valve is often associated with coarctation and could be the cause of an AS murmur.

However, a coarctation of the aorta associated with a VSD frequently produces symptoms within the first few days after birth. The coarctation produces pressure overload of the left ventricle, and blood flowing through the VSD causes volume overload of the right ventricle leading to early CHF. Coarctation with a VSD often has a long segment narrowing of the aorta, which classically occurs after the origin of the left common carotid artery and before the origin of left subclavian artery. This area can be so hypoplastic that it is completely obstructed, creating an interruption of the aortic arch. There is a marked increase in precordial activity. Loud murmurs and signs of CHF with significant respiratory distress are obvious. There is usually a profound metabolic acidosis. Significant hypocalcemia can be present because the area of the aortic arch hypoplasia is also associated with the embryologic origin of the parathyroid glands, which are important in calcium homeostasis (Di George syndrome).

The pulse pattern in long segment coarctation may be helpful. The right arm blood pressure and pulse will be normal to elevated, whereas the left arm and lower extremity pulses and blood pressures are diminished or absent (see Table 2).

**Hypoplastic left heart syndrome**

The counterpart to the hypoplastic right heart syndrome is the hypoplastic left heart syndrome (Fig. 3). It may involve severe mitral stenosis or atresia, hypoplastic or absent left ventricle, severe AS or atresia, hypoplastic aortic arch, and long segment coarctation of the aorta. The entire systemic blood flow is supplied through a PDA. When the PDA is functioning, symptoms may be minimal. But when the PDA constricts, CHF with shock and metabolic acidosis occurs suddenly. Intervention must be quick and decisive.

![Fig. 3. Hypoplastic left heart syndrome. Note the atretic aortic valve and the hypoplastic, nonfunctional left ventricle. Ductal closure results in severe limitation of systemic blood flow, leading to profound shock. Coronary Art, coronary artery; Ductus Art, Ductus arteriosus; LA, left atrium; LV, left ventricle; PDA, patent ductus arteriosus; RA, right atrium; RV, right ventricle. (Adapted from Krovetz LJ, Gessner IH, Schiebler GL. Handbook of pediatric cardiology. 2nd edition. Baltimore: University Park Press; 1979. p. 346; with permission.)](image-url)
**Total anomalous pulmonary venous return**

With total anomalous pulmonary venous return, the pulmonary veins do not attach to the left atrium directly. Rather they take one of 3 persistent fetal pathways to return to the right atrium. Once in the right atrium, oxygenated blood then crosses the foramen ovale into the left atrium and then out the left ventricle to the body. When the persistent fetal channels are nonrestrictive, signs may be minimal and presentation can be delayed for days to weeks. However, when there is obstruction within these fetal pathways or within the pulmonary venous system, pulmonary venous hypertension and CHF develops rapidly.

**SPECIAL CONSIDERATIONS**

**Conditions Causing Central Cyanosis Without Congenital Heart Disease**

Several common conditions can mimic CHD by causing central cyanosis and should be considered in the evaluation of the cyanotic newborn (Box 4). Infants with neurologic depression can be cyanotic because of central nervous system–induced hyperventilation. In addition to hypoxemia and cyanosis, the PaCO₂ is frequently elevated. Patients with rare hemoglobinopathies are cyanotic because the abnormal hemoglobin cannot load oxygen. Because PaCO₂ is a measure of oxygen dissolved in plasma, it is normal. However, hemoglobin saturation, a measure of the oxygen contained within the red cell, is extremely low. Inborn errors of metabolism can also cause cyanosis, acidemia, or CHF.

**Arteriovenous Malformation of the Great Vein of Galen**

The vein of Galen is located under the cerebral hemispheres and drains the anterior and central regions of the brain into the sinuses of the posterior cerebral fossa. A vein of Galen aneurysmal malformation (AVM) is formed early in gestation, and the amount of blood crossing the AVM can become massive. The vein dilates and obstructs the third ventricle, causing significant hydrocephalus. Because CHF secondary to the AVM occurs in utero, babies with this AVM can present as nonimmune hydrops fetalis with cardiomegaly, pleural effusions, and ascites at delivery. Auscultation for a bruit over the anterior fontanel and over the temporal bones in term infants with CHF within the first hours after delivery can help make the diagnosis.

<p>| Box 4 |</p>
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<td>Sepsis</td>
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<td>Pulmonary hypertension</td>
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<td>Hypoglycemia</td>
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<tr>
<td>Methemoglobinemia</td>
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<tr>
<td>Nonimmune hydrops fetalis</td>
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<td>Inborn errors of metabolism</td>
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_Abbreviation: CNS, central nervous system._
DYSRHYTHMIAS

Sinus tachycardia and sinus bradycardia are common in term neonates and are benign. All ventricular complexes are preceded by a normal P wave originating at the sinus node and are positive in the electrocardiogram in lead I. Sinus tachycardia can have rates close to 200 beats per minute during crying. Sinus bradycardia can have rates as low as 70 beats per minute during sleep. Although these rates are concerning to the observer, they are benign as long as they change with activity and the pulse oximetry during the variations is normal. Sometimes with deep sleep and bradycardia, the P wave on an electrocardiogram (ECG) or cardiac monitor will have a different appearance or may be absent. This is an escape rhythm, usually junctional or low atrial in origin, and is usually a normal variant. In cases of concern, consultation with a specialist could be considered.

Premature Beats

The most common cause of an irregular rhythm in a term neonate is the presence of premature atrial contractions (PAC). Most often they are benign and will resolve in a matter of days.\(^{20}\) The ECG shows an early beat preceded by a normal or inverted P wave and a pause following the premature beat. If the interval between the sinus beat and PAC shortens, the premature electrical activity from the atrium finds the ventricles in their relative refractory period and the beat is conducted with aberration, making the beat look like a premature ventricular contraction (PVC) but the complex is preceded by a P wave. If the interval shortens even further, the ventricular response may be dropped entirely, leaving a long pause (Fig. 4). The variation in QRS morphology may lead one to think that there is a combination

Fig. 4. PACs with varying coupling intervals. The tracings (A, B) are simultaneous. Note PACs (gray arrows) followed by a long pause. The PAC occurs during the refractory period of the ventricles and is not conducted. If the PAC occurs a bit later, it finds the ventricles in their relative refractory period and the PAC is conducted with aberration, looking like a PVC in strip (B) (black and white arrow), but the beat is clearly preceded by a P wave. When the PAC occurs even later, it is conducted normally (solid black arrow). (Adapted from Scagilotti D, Deal BJ. Benign cardiac arrhythmias in the newborn. In: Emmanouilides GC, Riemenschneider, TA, Allen HD, et al, editors. Moss and Adams heart disease in infants, children, and adolescents. 5th edition. Williams and Wilkins; 1995. p. 629; with permission.)
of PACs and PVCs, but that is not the case. All are isolated PACs and are entirely benign.

PVCs are less common unless the patient is on cardiac drugs, postoperative from cardiac surgery, or has significant hyperkalemia. Isolated PVCs are usually also benign and will resolve within a few days. However, obtaining serum electrolytes and an echocardiogram could be considered to rule out pathologic causes.

**Congenital Third-Degree Heart Block**

With congenital third-degree heart block, the atria beat at their inherent rate, 110 to 150 beats per minute, and the ventricles beat at their rate of 60 to 70 beats per minute with no relationship between the two (Fig. 5). Congenital third-degree heart block is frequently seen in babies born to women with systemic lupus erythematosus; if not already known, the diagnosis should be suspected when the dysrhythmia is discovered. Because this condition can be present for quite some time in utero, the infant’s cardiovascular system can compensate for the low rate by increasing stroke volume to maintain cardiac output. It is uncommon for a neonate to be symptomatic and need pacing, but consultation with a specialist is advised.

**Supraventricular Tachycardia**

SVT is not uncommon in the neonate and must be distinguished from sinus tachycardia (Fig. 6). Most times it occurs in the absence of structural heart disease but can be associated with lesions that produce a large right atrium, such as PS/PA with massive tricuspid regurgitation. SVT in utero is also a common cause of nonimmune hydrops fetalis, caused by CHF, which may resolve when the SVT breaks.

SVT is also frequent in instances of the various pre-excitation syndromes (ie, Wolf-Parkinson-White). Heart rates are in the range of 210 to 220 beats per minute and do not change with activity of the infant. SVT in the presence of a normal heart can be tolerated for several hours; unless the baby is symptomatic, with signs of CHF and metabolic acidosis, there is usually sufficient time to diagnose and treat the infant safely. Although maneuvers that produce vagal stimulation, ice to the forehead, and

![Fig. 5. Complete heart block (third-degree heart block). Atrial rate is 145 beats per minute (P-P interval 0.42 seconds) and regular. Ventricular rate is 60 beats per minute (R-R interval 1.0 second) and regular. P waves and QRS complexes are independent of each other. (From Artman M, Mahony L, Teitel DF. Neonatal cardiology. New York: McGraw-Hill; 2002. p. 165; with permission.)](Image)
painful stimuli can break the SVT, their benefit is questionable because SVT frequently recurs until maintenance medication is administered. When used in excess, these interventions have their own inherent risks. Consultation with a specialist is usually indicated.

**PULSE OXIMETRY SCREENING**

Universal newborn screening is the process by which newborns are tested shortly after birth for conditions that can cause severe illness, disability, or death. Through early

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**Fig. 6.** SVT (A) and sinus tachycardia (B). SVT (A) with rate of 315 beats per minute. QRS complexes are normal, but no P waves are seen. With sinus tachycardia (B) rate is 230 beats per minute. P waves (arrows) are visible preceding a normal QRS. Running the paper at 2\times speed helps to uncover and identify the P waves. (*From* Artman M, Mahony L, Teitel DF. Neonatal cardiology. New York: McGraw-Hill; 2002. p. 166, 171; with permission.)
identification and treatment, newborn screening provides an opportunity to reduce morbidity and mortality. Babies with CCHD are at significant risk of disability or even death if their condition is not timely diagnosed and treated. With the vast improvements in fetal cardiac ultrasonography and the addition of POS, great strides are being made in our diagnostic abilities for detecting CCHD. However, even when prenatal ultrasounds are performed by those with specific training in CHD, fewer than 50% of cases of proven CHD are identified. Further, it has been estimated that up to 30% of infants with unrecognized CCHD may be discharged each year from newborn nurseries in the United States. The Centers for Disease Control and Prevention (CDC) estimates that each year about 1200 more newborns with CCHD could be identified at birth hospitals using POS.

Universal POS was added to the federally recommended uniform screening panel through endorsement by the Secretary of Health and Human Services (HHS) in September 2011. In December 2011, the American Academy of Pediatrics (AAP) published their endorsement of the HHS recommendations for POS for CCHD in the January issue of the journal Pediatrics. POS for CCHD has now become a national standard of care and is part of most but not all state newborn screening panels. Data regarding US state participation in POS for CCHD can be found online on the AAP Web site. The method of using POS for CCHD as a compliment to existing practices of care has been shown to be cost-effective and associated with improved detection and outcomes for babies. Estimated costs run from about $5 to $14 per newborn screened.

In a 2012 meta-analysis of 13 studies with data for 229,421 newborn infants, the overall sensitivity of pulse oximetry for detection of CCHD was 76.5% (95% confidence interval [CI] 67.7–83.5) and specificity was 99.9% (95% CI 99.7–99.9).

A recommended standard protocol for POS of well newborns and best-practice advice regarding implementation are now readily available to newborn health care providers. Utilization of the AAP-endorsed CCHD POS algorithm is recommended (Fig. 7).

Newborn POS is most successful in identifying 7 primary and 5 secondary cardiac lesions (Table 3). Obtaining both preductal and postductal pulse oximetry measurements is essential because defects with right-to-left shunting of desaturated blood through the ductus arteriosus will not be detected with only preductal readings. It is recommended that the probes be placed on both the right hand and one foot. Screening at least 24 hours after delivery substantially reduces the false-positive rate (0.05% after 24 hours vs 0.5% before 24 hours).

<table>
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<tr>
<th>Table 3</th>
<th>Screening targets for pulse oximetry</th>
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<tr>
<td><strong>Primary Screening Targets</strong></td>
<td><strong>Secondary Screening Targets</strong></td>
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<tr>
<td>Hypoplastic left heart syndrome</td>
<td>Coarctation of the aorta</td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td>Interrupted aortic arch</td>
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<tr>
<td>Tetralogy of Fallot</td>
<td>Ebstein anomaly of the tricuspid valve</td>
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<td>Total anomalous pulmonary venous return</td>
<td>Double outlet right ventricle</td>
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<td>Transposition of the great vessels</td>
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<tr>
<td>Tricuspid atresia</td>
<td></td>
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<tr>
<td>Persistent truncus arteriosus</td>
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</table>

It is important to remember that the current POS protocol will not detect all forms of CHD whether they are critical lesions or not.

Data from Refs.
Fig. 7. Algorithm for POS. Effect of altitude: It is important to note that the oxygen saturation thresholds for a positive screening result may vary at high altitude. Appropriate studies need to be performed at higher altitudes to establish reliable thresholds. (Adapted from Kemper AR, Mahle WT, Martin GR, et al. Strategies for implementing screening for critical congenital heart disease. Pediatrics 2011;128(5):e1267; with permission.)
**Pulse Oximetry Clinical Assessment**

Babies with saturation less than 90% in the right hand or foot should be *immediately referred for clinical assessment* (see Fig. 7). Babies with 3 failed readings (pulse oximetry <95% in the right hand and foot OR >3% difference between the right hand and foot) should receive

- Clinical assessment (infectious and pulmonary pathology should be excluded)
- Echocardiogram
- Referral to pediatric cardiology, immediately if symptomatic, expeditiously if asymptomatic

1. Passed Screens
   - A screen is considered passed if:
     - The oxygen saturation is 95% or greater in the right hand and foot with less than 4% difference between the two readings; screening would then be complete.

2. Failed Screens
   - A screen is considered failed if:
     - Any oxygen saturation measure is less than 90% (in the initial screen or in repeat screens).
     - Oxygen saturation is less than 95% in the right hand and foot following 3 measurements, each separated by 1 hour.
     - A greater than 3% absolute difference exists in oxygen saturation between the right hand and foot on 3 measurements, each separated by 1 hour.

Any infant who fails the screen should have a diagnostic echocardiogram performed and be referred to a pediatric cardiologist for further management.

*It is important to remember that it is possible for a baby to have a normal POS and still have a congenital heart defect.*

**EVALUATION AND STABILIZATION WHEN CRITICAL CONGENITAL HEART DISEASE IS SUSPECTED**

After careful review of the history and physical examination, the physician must decide about the need for further intervention. If the term neonate seems well and has passed POS but has a benign dysrhythmia or has a grade i to ii mid- to high-frequency murmur that is localized, a follow-up examination in 24 hours should be sufficient to decide on the need for further referral. However, if the baby does not pass POS, obtaining both an echocardiogram and consulting with a pediatric cardiologist would be prudent.

When the infant seems ill, general interventions should be initiated. Obtain a serum glucose to screen for hypoglycemia. Check 4 extremity blood pressures. Screening for sepsis and the initiation of antibiotics should also be considered. A chest radiograph can be obtained and may show cardiomegaly or abnormal pulmonary vascularity. However, in many instances, the chest radiography will be normal even in the presence of a CCHD lesion. The real value of the chest film is not to support or dismiss the diagnosis of CHD but rather to identify other causes of distress in the newborn, such as a pneumothorax or primary pulmonary disease. Unless a dysrhythmia is present, an ECG is usually not helpful.

If the infant is extremely ill, early control of the airway and placement of umbilical artery and venous catheters would be strongly advised. When the index of suspicion of CCHD is high, consideration should be given to initiating an infusion of prostaglandin E₂ (PGE₂).

PGE₂ stabilizes a PDA and will usually reopen a constricted or closed ductus, providing a reliable means of PBF in patients with CCHD and improvement in
CHF. PGE$_2$ is infused at 0.02 to 0.1 µg/kg/min. The infusion is usually begun at 0.05 µg/kg/min and can be titrated depending on the changes in oxygenation, increase in blood pressure, and decrease in acidosis. Ideally PGE$_2$ is infused into a reliable peripheral intravenous line; however, administration through an umbilical venous catheter or an umbilical artery catheter will suffice at least temporarily. Apnea can occur when initiating therapy, especially at higher doses. Therefore, it is important to be ready to establish a stable airway when initiating therapy.

Taking these measures should aid in stabilizing the newborn allowing for subsequent transport to an appropriate critical care unit while awaiting further interventions.

REFERENCES