When to Call the Cardiologist: Treatment Approaches to Neonatal Heart Murmur

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Moderate and severe forms of congenital heart disease (CHD) occur in about 6 per 1,000 live births. If tiny muscular ventricular septal defect (VSD) and bicuspid aortic valve as well as other lesions of a less serious nature are included, it is estimated that the true incidence of congenital heart disease might be as high as 75 per 1,000 live births.[1] CHD is a leading cause of infant mortality in United States.[2] Early detection and prompt intervention is critical in reducing infant mortality associated with CHD.

Careful clinical examination is extremely important and is always the first step in ruling out CHD. It is not infrequent to find a heart murmur while examining a baby in the newborn nursery. The clinician is then faced with a deci-

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sion whether the murmur is physiologic or pathologic. The focus of this article is to help with such a decision.

HEART MURMUR IN NEWBORNS

Soon after birth, the newborn baby is adapting to an extra-uterine life with dramatically new circulation in which oxygen exchange occurs in the lungs, and thermoregulation becomes more challenging.[3] To adapt to this new hemodynamic and metabolic state, there are three critically important changes that occur in newborn circulation: 1) Pulmonary blood flow increases dramatically to about 20 times that of a fetus; 2) Central shunts through the ductus venosus, foramen ovale and ductus arteriosus are abolished; and 3) Combined ventricular output increases dramatically to meet the demands of respiratory work and thermoregulation.[4] Such dramatic changes in circulation can lead to transient murmurs and rapidly changing clinical examination of newborns in the first few hours of life.

Experience and literature suggest that a 'heart murmur' is probably the most common reason for cardiology consultation in most hospitals.[5] It has been reported that "murmur in a baby" causes significant parental anxiety, which might be relieved by cardiology consultation.[6,7]

Detection of a murmur depends on how soon after birth the baby is examined, the condition under which the examination is performed, the expertise of the listener and the frequency of the examination. Hence, it is not surprising that the reported prevalence of heart murmurs in newborns is extremely variable from as low as 0.6% to as high as 77.4%.[8-11] Murmurs heard in the first few hours of life might reflect a closing ductus or transient tricuspid regurgitation from perinatal stress or a peripheral pulmonic flow murmur due to turbulence resulting from sudden increase in pulmonary blood flow. The prevalence of heart murmurs decreases significantly over the first few days of life.[12]

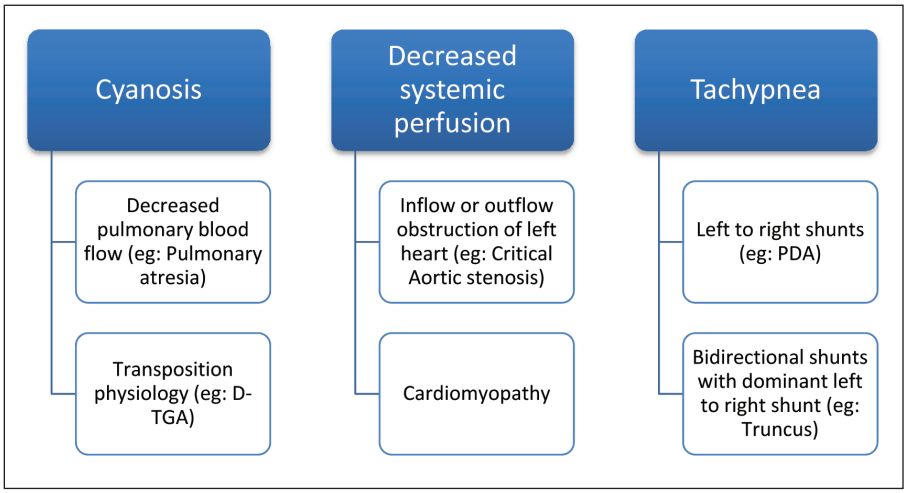

Figure courtesy of Deepak Kamat, MD, PhD, FAAP.

Figure 1. Clinical manifestations of critical congenital heart diseases. D-TGA = D-transposition of the great vessels ; PDA = patent ductus arteriosus.

Several studies have evaluated the predictive value of heart murmurs as a sole finding in the diagnosis of CHD. Ainsworth et al studied 7,000 newborns and concluded that presence of a murmur had a sensitivity of 44% and a positive predictive value of 54% in predicting structural heart disease.[8] In another series in which echocardiography was performed to evaluate for possible heart disease based on suspicious physical examination, fewer than 15% of subjects were found to have significant CHD.[13] Singh et al[14] evaluated 205 echocardiograms done in neonatal nursery because of a murmur: major structural lesions were identified in 2% of them. Half were coarctation, 26% of the echoes were normal, 38% had defects like small VSD or atrial septal defect (ASD). Transient circulatory changes like patent ductus arteriosus, tricuspid regurgitation accounted for the other 34%.[14] Similar data have been reported by others.[15]

Clinical experience plays a significant role in the ability to recognize the murmur as pathological or otherwise. Studies have shown that neonatologists and pediatric cardiologists are better than pediatricians and pediatric trainees in diagnosing pathological murmurs with confidence.[16,17] Interestingly, one study found that pediatric cardiologists could identify neonates with CHD with a sensitivity of 80.5% and a specificity of 90.9%.[18]

CLINICAL PRESENTATION OF NEWBORNS WITH CHD

Congenital heart disease in the newborn period can be classified into two broad categories: Critical CHD and non-critical CHD. Critical CHD includes those lesions that can lead to death or severe morbidity if unrecognized in early infancy.[4,19] Examples of such lesions include coarctation of aorta, hypoplastic left heart syndrome, and transposition of great arteries. Non-critical CHD include ASD, tiny muscular VSD, bicuspid aortic valve, and mild pulmonary stenosis.

Critical CHD have three cardinal signs in the newborn period (see Figure 1): 1) Cyanosis; 2) Decreased systemic perfusion; and 3) Tachypnea.

The vast majority of critical congenital heart diseases are ductal dependent for either systemic or pulmonary blood flow. Prompt recognition and initiation of prostaglandin E (PGE) is critical to stabilize the baby. Some patients may need emergent interventions like balloon atrial septostomy or, rarely, surgery.

It is important to remember that many newborns with critical CHD such

as transposition of great arteries, valve atresia, and total anomalous pulmonary venous return may not have a murmur.[19] Newborns with ductal-dependent lesions like pulmonary atresia usually have a large unrestrictive ductus and can develop a murmur later when the pulmonary vascular resistance falls or when the ductus starts to constrict.

On the other hand, newborns with non-critical CHD may be completely asymptomatic or have murmur as their sole clinical manifestation.

TREATMENT APPROACHES TO NEWBORNS WITH HEART MURMUR

History

The value of a careful history is often underestimated. Feeding difficulty, such as not being able to feed for long time, taking breaks during feeding, as well as tachypnea, fussiness, color change either pallor or cyanosis, and sweating during feeding may suggest the presence of CHD. A family history of CHD, known heritable syndromes, maternal history during pregnancy such as poorly controlled diabetes, medications, drug abuse, infections, and presence of known genetic syndromes in a baby are clues towards to specific heart defects.

General Examination

General assessment of the baby is very important as to rule out critical CHD. Is the baby cyanotic? Is there poor perfusion? Is there respiratory distress or tachypnea? Does the baby have congenital anomalies of a known genetic syndrome?

Acro-cyanosis is common in newborns and should not be confused with true cyanosis. Cyanosis can be due to CHD, respiratory failure, or pulmonary hypertension of newborn. Cyanosis might not be apparent till oxygen saturation drops below 85%; saturation should be measured when cyanosis is difficult to be assessed clinically. Pulse oximetry has been suggested recently as a good screening test to rule out critical CHD and is endorsed by the American Heart Association and American Academy of Pediatrics in a recently published scientific statement.[19] The statement suggests using saturation < 95% in lower extremity after 24 hours of life as an indication for further evaluation. Saturation should ideally be measured in the right ear lobe and in the leg. Right ear lobe is the safest place to get a pre-ductal saturation as the right arm might not be pre-ductal in the presence of an aberrant right subclavian artery. Differential saturation with higher saturation in the lower limbs suggests transposition or supra-cardiac total anomalous pulmonary venous return. Similarly, differential saturation with lower saturation in the lower extremities might suggest coarctation of aorta, pulmonary hypertension, critical left-sided obstructive lesions or infra-diaphragmatic total anomalous pulmonary venous return.

Decreased systemic perfusion as assessed by cool extremities, poor capillary refill, weak pulses and low blood pressure can be due to sepsis, metabolic disorders, anemia or endocrine disorders as well as due to cardiac lesion. Respiratory distress or tachypnea in a newborn is more likely to be due to respiratory pathology, but careful clinical examination, as well as chest X-ray and blood-gas analysis can be helpful in suggesting that the symptoms are due to cardiac pathology. Presence of hepatomegaly

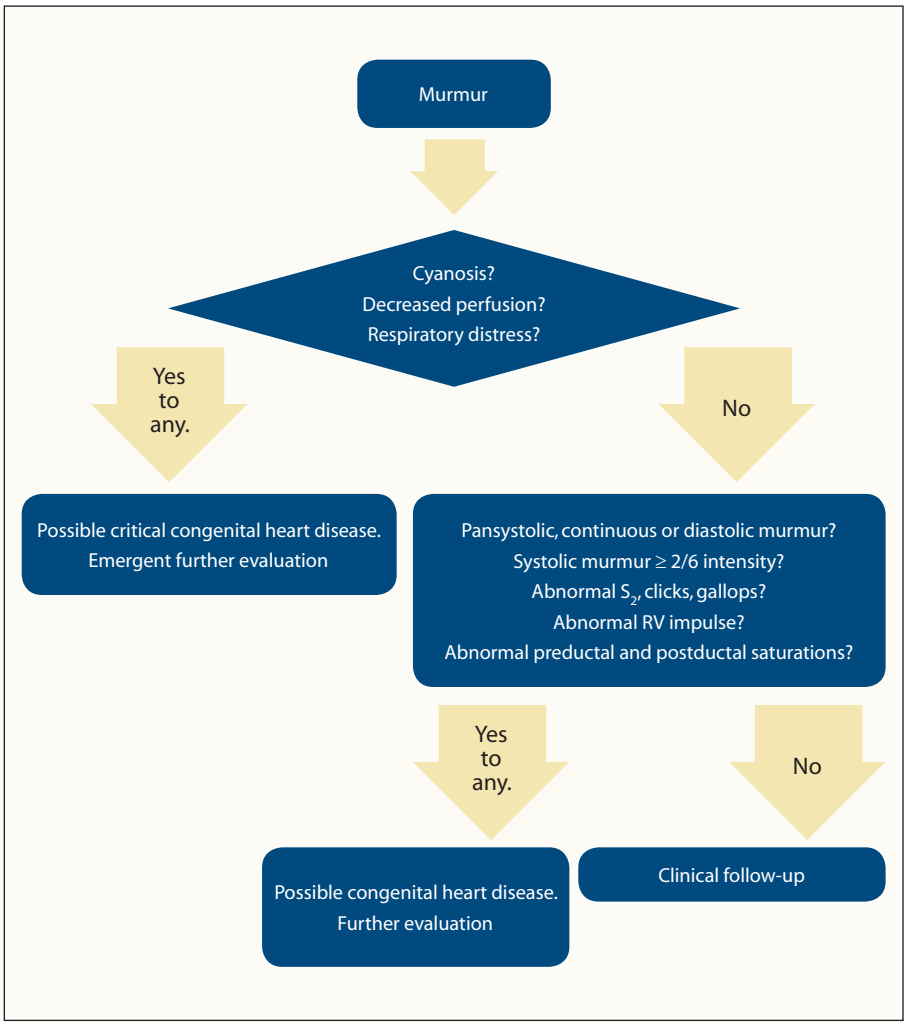

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Figure 2. Flow diagram for evaluation of a newborn with heart murmur. RV = right ventricle.

may be due to elevated right atrial filling pressures.

Cardiac Examination

The cardiac examination should begin with palpation of the precordium. Normal newborns have a parasternal or sub-xiphoid impulse because of a dominant right ventricle (RV). Increased RV impulse might suggest transposition or RV outflow tract obstruction. An absent RV impulse may in turn suggest inflow obstruction to the RV. One should feel for any thrills, which are rare in newborns. A palpable thrill in a cyanotic newborn almost always suggests tricuspid atresia with VSD, as this is the only lesion where the VSD jet points anteriorly. Careful attention should be paid to the first and second heart sounds. A single S_2 might be a result of transposed aorta or single outflow tract. A widely split S2 may suggest total anomalous pulmonary venous return. Presence of clicks might suggest truncus arteriosis or Ebstein anomaly. A gallop may be heard in presence of heart failure.

A murmur should be evaluated to define its timing (early systolic, late systolic, pan systolic, diastolic), quality, location, intensity (graded 1-6), frequency and radiation.[20] The point where highest frequencies are heard is more reliable for determining the location rather than loudness because the highest frequencies do not travel far from the lesion and the frequency of the murmur correlates with the pressure gradient across the lesion. Diastolic murmurs are rare in newborns and are almost always related to structural heart disease. Continuous murmurs may be heard in the presence of large patent ductus. A to-and-fro murmur in a baby with severe respiratory distress suggests absent pulmonary valve syndrome.

An outline for evaluation of a newborn baby with a murmur has been suggested in Figure 2. If the newborn baby with heart murmur is symptomatic with signs of poor perfusion, cyanosis or respiratory distress, he/she should be evaluated by a cardiologist immediately. In an asymptomatic neonate with heart murmur, a careful examination is important. In the absence of any other abnormality on cardiac examination, a newborn baby with ejection systolic murmur of ≤ 2/6 in intensity (considered to be low frequency) can be observed safely with serial clinical examination and outpatient follow-up.

SUMMARY

Heart murmurs in newborns are common but are not always a result of significant CHD. It is important to identify those with critical CHD, as early diagnosis leads to better outcomes. A careful history, general physical and cardiac examination should help the clinician in determining if the child needs further evaluation. ■

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