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Review Article

Cardiac Murmurs in The Newborn – When to Worry?

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Abstract

Congenital Heart Disease (CHD) contributes to a large proportion of mortality among infants and young children. Newborns (birth to 1 month of age) are at higher risk of having a serious lesion requiring early intervention, than older infants and children. Detecting a murmur in a newborn on physical exam can provide a clue to the presence of CHD, but its utility is limited by provider expertise and neonatal factors, such as rapid heart rate and respiratory symptoms. Furthermore, not all murmurs are pathological. Health care providers including primary care physicians, pediatricians or nurse practitioners often face difficulties when determining whether a murmur warrants further investigation. We aim to describe key differences between innocent and pathological heart murmurs in the newborn. Further, we describe current screening protocols for Critical Congenital Heart Defects (CCHD) that may assist primary care physicians in deciding when to refer for further evaluation.

Keywords: Congenital Heart Disease, Murmur, Pulse Oximetry Screening

Abbreviations: AAP: American Academy of Pediatrics; AS: Aortic Stenosis; AVSD: Atrioventricular Septal Defects; CCHD: Critical Congenital Heart Defects; CHD: Congenital Heart Disease; CPS: Canadian Paediatric Society; PDA: Patent Ductus Arteriosus; PFO: Patent Foramen Ovale; POS: Pulse Oximetry Screening; TGA: Transposition of The Great Arteries; VSD: Ventricular Septal Defect

Evaluating A Newborn with A Heart Murmur

The incidence of Congenital Heart Disease (CHD) in the general population is less than 1% [1-6]. Heart murmurs are a common finding in infants and in older children. Less than 1 percent of newborn infants have an audible murmur in the first few days of life [4,5]. In fact, a murmur may be the only clue to a severe and potentially life-threatening CHD, known as a Critical Congenital Heart Defect (CCHD) [7]. More than half of newborns with a heart murmur have a structural heart lesion when confirmed on echocardiogram [7-9]. However, not every murmur is pathological; the remainder are referred to as innocent, or physiological murmurs that originate from normal flow patterns in the absence of anatomic abnormalities of the heart. Cardiac murmurs in the newborn may also be the result of a transitional circulation, which is usually related to the Patency of the Ductus Arteriosus (PDA), or its closure. The Patent Foramen Ovale (PFO) is another part of the transitional circulation that is not usually clinically detectable [10].

Determining the etiology of a murmur in the newborn presents a challenge for health care providers. Because of this, they remain the most common reason for pediatric cardiologist consultation in neonatal intensive care units and nurseries [11,12]. A thorough evaluation including a complete history and physical examination is a crucial first step to identifying those newborns

with pathological murmurs at increased risk for CHD. Ul Haq, et al., showed that 14% of patients with a positive family history of CHD had an anatomical heart defect themselves [13]. Infants of diabetic mothers are at increased risk for Ventricular Septal Defects (VSD), Transposition of The Great Arteries (TGA) and Aortic Stenosis (AS) [13,14]. Intrauterine infections such as maternal rubella predispose the infant to a Patent Ductus Arteriosus (PDA) and peripheral pulmonary stenosis [4]. There is also an association with maternal alcohol consumption and VSD, as well as maternal exposure to Lithium with Ebstein's anomaly [15]. Newborns with genetic syndromes such as Trisomy 21, 22q11.2 deletion or Turner's syndrome are also at increased risk for Atrioventricular Septal Defects (AVSD), conotruncal abnormalities and coarctation of the aorta, respectively [13]. Certain red flags on history including failure to thrive, feeding accompanied by excessive sweating, pallor, tachypnea or increased work of breathing should raise suspicion of heart failure in the context of CHD [15].

Physical examination findings of dysmorphic features or characteristics of genetic syndromes may lead the clinician to investigate the presence of a cardiac anomaly [4,15]. Other findings including cyanosis indicate a right-to-left intracardiac shunt or a single ventricle physiology, whereas findings of respiratory distress, tachycardia, tachypnea and hepatomegaly raise suspicion for heart failure. Palpation for femoral pulses along with a 4-limb blood pressure measurement are important to evaluate for an aortic coarctation [5,15]. On cardiac auscultation, the characteristics of the murmur may further help to differentiate between a pathological and innocent murmur. Loud (greater than grade III/VI) murmurs, diastolic and pansystolic murmurs, those that are harsh with unusual radiation or have abnormal second heart sounds, are more concerning for an underlying structural heart lesion [4,5,15].

When red flags are present in the history or physical examination, in conjunction with a neonatal heart murmur, referral to a pediatric cardiologist should be considered. A summary of the history and physical examination findings that should prompt consideration of cardiology referral are presented in Tables 1 & 2.

Table 1: Historical Findings Suggestive of Structural CHD Prompting Pediatric Cardiology Referral

Historical Finding	Significance
Family History	
CHD	CHD more common in children with a first-degree relative who has CHD, high penetrance with ventricular septal defect and mitral valve prolapse [4]
Sudden cardiac death	Increased risk of hypertrophic cardiomyopathy, inherited in an autosomal dominant pattern
Sudden infant death syndrome	May be secondary to undiagnosed CHD
Prenatal and Perinatal History	
In utero exposure to non-pharmacological toxins	Maternal alcohol consumption is associated with an increased risk of atrial and ventricular septal defects, and Tetralogy of Fallot
In utero exposure to teratogenic pharmacological agents	Some studies have demonstrated that SSRI exposure has been associated with a small, but statistically significant increased risk of ventricular septal defects and bicuspid aortic valve [4] Lithium exposure is associated with Ebstein's anomaly Valproate is associated with coarctation of the aorta and hypoplastic left heart syndrome
Intrauterine infection	Maternal rubella infection is associated with patent ductus arteriosus and peripheral pulmonary stenosis
Maternal diabetes mellitus	Increased risk of ventricular septal defect, transposition of the great arteries, and aortic stenosis
Preterm delivery	50% of newborns weighing less than 1500 g at birth have CHD, most commonly patent ductus arteriosus [4]

Alarm Symptoms	
Failure to thrive Feeding intolerance as exhibited by excessive sweating, pallor, tachypnea, or increased work of breathing	May indicate CHF in the context of CHD; poor weight gain commonly reflects decreased cardiac output or left-to-right shunting of blood with resultant pulmonary hypertension

Table 1: Historical Findings Suggestive of Structural CHD Prompting Pediatric Cardiology Referral.

Table 2: Physical Examination Findings in Newborns with Heart Murmurs Prompting Pediatric Cardiology Referral	
Finding	Significance
Abnormal vital signs	Arrhythmia, tachycardia, hypoxia and tachypnea may indicate underlying structural heart disease Blood pressure discrepancy between upper and lower limbs is suggestive of coarctation of the aorta (pressure gradient of >20 mmHg between upper and lower extremities)
Adventitious breath sounds	Wheezing may be associated with cardiac asthma Crackles may be associated with pulmonary congestion due to congestive heart failure
Dysmorphic features	Newborns with genetic syndromes such as Trisomy 21, 22q11.2 deletion, or Turner's syndrome are at increased risk for Atrioventricular Septal Defects (AVSD), conotruncal abnormalities and coarctation of the aorta, respectively [13]
Abnormal cardiac S_2	Wide split fixed S_2 in atrial septal defect
Capillary refill	Normal capillary refill is less than 2 to 3 seconds; a delayed capillary refill may indicate poor perfusion secondary to reduced cardiac output
Systolic ejection click	Semilunar valve stenosis
Weak or absent femoral pulses	Coarctation of the aorta
Ascites and hepatomegaly	Congestive heart failure

Table 2: Physical Examination Findings in Newborns with Heart Murmurs Prompting Pediatric Cardiology Referral.

Pulse Oximetry Screening

Although some pediatricians and neonatologists may exclude heart disease based on clinical exam alone, the clinical exam is less reliable in younger children, and up to half of babies with CHD may be missed and can only be ruled out more definitively through echocardiogram [15,16]. The ability to accurately identify pathologic murmurs in the newborn by clinical examination alone varies even among subspecialists, due to differences in clinical expertise, with sensitivities ranging from 80.5 to 94.9 percent and specificities ranging from 25 to 92 percent [17,18]. Moreover, CHD may occur even in the absence of a heart murmur or other clinical findings [2,4]. One study found that by examination alone, detection rates for pathological heart murmurs in the newborn were approximately 50 percent when confirmed by echocardiogram [5]. Because of these challenges, other tools have therefore arisen to help physicians screen for CHD in the newborn.

The introduction of Pulse Oximetry Screening (POS) has been shown to be an accurate, cost-effective and noninvasive screening method for the detection of CCHD with high specificity (99.9%) and moderate sensitivity (76.5%) [1,2,19-21], with sensitivity increasing (82-92%) when combined with physical examination. CCHD lesions include duct-dependent cardiac abnormalities (hypoplastic left heart syndrome, pulmonary atresia, transposition of the great arteries and tricuspid atresia) and cyanotic heart lesions including tetralogy of Fallot, total anomalous pulmonary venous return and truncus arteriosus (Table 3) [3,6,20].

Cyanotic	May Be Cyanotic
Hypoplastic left heart syndrome Pulmonary atresia with intact ventricular septum Total anomalous pulmonary venous return Tetralogy of Fallot Transposition of the great arteries Tricuspid atresia Truncus arteriosus	Coarctation of the aorta Double outlet right ventricle Ebstein's anomaly Interrupted aortic arch Defects with single ventricle physiology

Table 3: Structural heart lesions detectable using pulse oximetry screening [6].

Early detection of CCHD is critical to ensure rapid intervention and surgical management. The Canadian Paediatric Society (CPS) and American Academy of Pediatrics (AAP) recommend the implementation of routine POS for all newborns [3,20]. A positive screen is indicated by saturations below 90% on either the right hand (preductal saturation) or any foot (postductal saturation), below 95% in both extremities after three measurements, or a difference of more than 3% between preductal and postductal saturations after three measurements (Figure 1) [3,20,21]. Infants with a positive screen should have further evaluation, including assessment by a pediatric cardiologist and echocardiogram. Though uncommon, false positive screens may result from other conditions causing hypoxemia, such as hemoglobinopathies or persistent pulmonary hypertension of the newborn. Routine POS should be performed between 24-36 hours of age, as screening before 24 hours increases this rate of false positives [2].

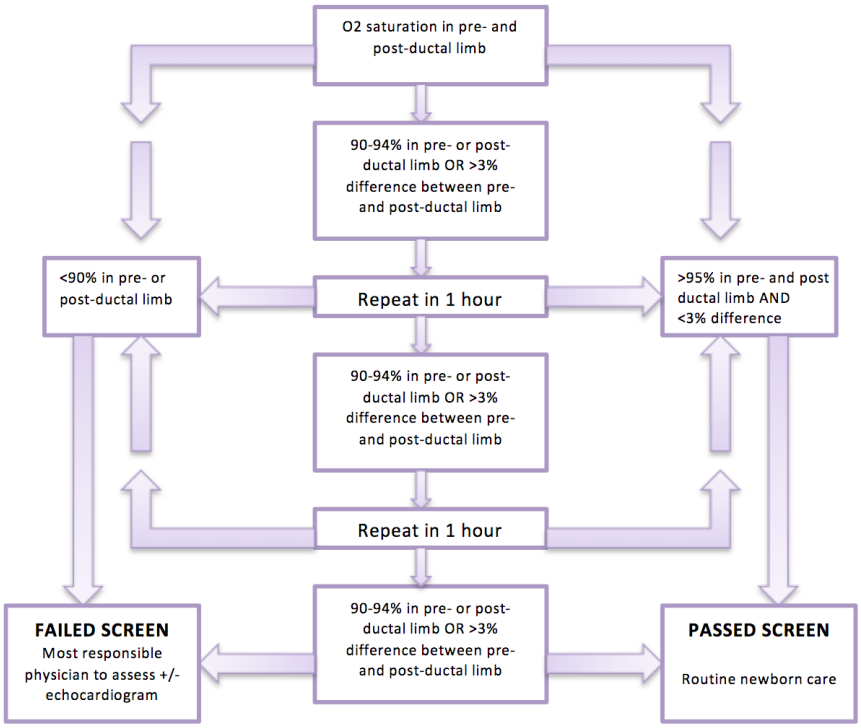

Figure 1: Pulse oximetry screening guidelines for the detection of critical congenital heart disease; adapted from the Canadian Pediatrics Society [3].

Determining When to Refer in The Setting of CHD

In the setting of CHD, a thorough history, physical examination including auscultation for neonatal murmur, and pulse oximetry screening must be carefully combined in order to determine when to refer to pediatric cardiology. The utility of POS is in differentiating newborns at risk for CCHD who require urgent pediatric cardiology referral. We recommend that POS should be implemented universally prior to a newborn being discharged from hospital. Newborns with a cardiac murmur who pass the POS but have red flags on history or physical examination as indicated in (Tables 1 and 2) should also be referred for further assessment. Finally, a newborn with a murmur and negative POS with no red flags should be followed closely by their primary physician to monitor for any symptoms of cyanosis, heart failure, failure to thrive or feeding intolerance. If red flags arise on reassessment, or the murmur characteristics become concerning, the newborn warrants further evaluation. It should be noted that our final recommendation may vary by clinical experience, geographic location and availability of follow-up.

When referring to pediatric cardiology, we recommend that no further diagnostic testing be conducted by the primary physician. Chest x-rays and electrocardiograms rarely assist in the diagnosis of a heart murmur and are therefore not cost-effective. In a study performed by Mackie, et al., the addition of electrocardiography did not improve the sensitivity in the detection of CHD [17].

In geographically remote areas, where pediatric cardiologists may be unavailable for urgent referral, phonocardiography may be considered. Phonocardiography involves digital heart sound recordings which can be reviewed by a pediatric cardiologist remotely, to distinguish between innocent murmurs and murmurs that are likely to be pathologic. Newborns with murmurs that are determined likely to be pathologic should be transported to a clinical centre which can accommodate echocardiogram and examination by a pediatric cardiologist [4].

Conclusion

Neonatal heart murmurs continue to pose a challenge to healthcare providers. Based on their audible characteristics alone, there may be insufficient information to confidently rule out a structural heart lesion. However, complete history and physical examination remain crucial to identify the critical features that may alert the clinician to the presence of critical congenital heart disease (CCHD). Although Pulse Oximetry Screening (POS) has been shown to improve the detection of CCHD, any newborn with a cardiac murmur should be seen by a health care provider after discharge, as vigilant follow-up of all newborns is recommended, as it is possible a newborn with CCHD may escape early screening. For newborns who have clinical findings suggestive of a cardiac condition (ie, Failure to thrive/poor feeding, cyanosis, decreased pulses, respiratory distress) or have a pathologic cardiac examination, an urgent referral to a pediatric cardiologist for immediate assessment is warranted.

What's New

1. Congenital heart disease must be considered in the workup for a newborn with a heart murmur
2. Pulse oximetry screening should be done routinely for every newborn 24-36 hours after birth
3. Pulse oximetry screening can help identify critical congenital heart lesions that warrant urgent cardiology follow-up and further workup
4. Close follow-up of all newborns with a murmur is warranted

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