Heart murmur in a child
Is it innocent or pathological?

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Heart murmurs in children are common and most are benign. A thorough investigation is required to determine if the murmur is innocent or a sign of pathological congenital heart disease.

Heart murmurs are common in childhood, occurring in one in every two children, whereas the prevalence of congenital heart disease has been estimated as one in every 100 children.[1,2] Despite this, a heart murmur in a child often provokes anxiety for both the parent and clinician. A common misconception is that a murmur is a diagnosis in itself indicating an organic heart problem. However, most murmurs are benign and the actual diagnosis is that of a normal heart and an innocent murmur. A systematic approach to investigating a heart murmur in a child includes taking a thorough history and performing an examination to determine if a murmur is innocent or a sign of a pathological congenital heart disease.

Clinical history

Risk factors

A thorough history should start with the antenatal period, as up to 20% of cases of congenital heart disease are the direct result of either genetic abnormalities or teratogens.[3] Therefore, antenatal history should focus on factors that are known to be associated with congenital heart disease, including a family history of it. Despite the prevalence of congenital heart disease being about 1% in the general population, this increases to up to 15% if there is a maternal history of such disease.[4]

Several commonly prescribed medications are teratogenic and can be associated with shunt lesions (e.g. atrial septal defect [ASD], ventricular septal defect [VSD], patent ductus arteriosus [PDA]) or obstructive lesions (e.g. pulmonary stenosis, aortic stenosis) in the fetus. Two such medications are sodium valproate (associated with ASD, VSD, aortic stenosis, pulmonary atresia with intact ventricular septum, and coarctation) and phenytoin (associated with pulmonary stenosis, aortic stenosis, coarctation and PDA). Other medications may be associated with potentially major heart defects, including Ebstein's

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anomaly with lithium use and conotruncal defects with use of retinoic acid. The use of illicit substances is also relevant as alcohol and amphetamines are both associated with the occurrence of shunt lesions.[4]

Finally, maternal medical conditions may predispose a child to particular heart diseases. Babies born to mothers with diabetes (including gestational diabetes) have a higher incidence of congenital heart disease, including transposition of the great arteries, whereas autoimmune conditions such as systemic lupus erythematosus may cause heart block in the child.

KEY POINTS

- A heart murmur is not a diagnosis.
- Murmurs are very common and may represent a normal heart or indicate pathological congenital heart disease.
- With a systematic approach, physicians may differentiate innocent from pathological murmurs with great sensitivity.
- All neonates with a murmur should be referred to a cardiologist for assessment.
- All patients with pathological murmurs, those with murmurs and syndromes associated with congenital heart disease and patients with undifferentiated murmurs should be referred to a cardiologist.

Symptoms

Symptoms arising from congenital heart disease vary with age and are much less specific than in adult populations. Infants may present with failure to thrive resulting from poor feeding. Infants with left-sided heart failure may be tachypnoeic, which is worsened with feeding. This causes infants to fatigue during feeding itself, leading to short, frequent feeds, increased work of breathing and diaphoresis. Infants may have poor weight gain and failure to thrive for a number of reasons; however, if there is no apparent organic cause and the patient has a heart murmur, they should be referred for specialist review.

Children and teenagers with heart murmurs are more likely to have a history of nonspecific lethargy and poor exercise tolerance when compared with their peers.[5] Common questions to ask these patients may include 'Do you keep up with friends at school?' and 'How many flights of stairs can you climb before tiring? How does this compare to your peers?' It is important to note that children with significant congenital heart disease may appear to be asymptomatic.

Clinical examination

Observation

An abundance of important information can be obtained from routine observations such as measurement of weight, oxygen saturations, blood pressure and heart and respiratory rates, as well as the evaluation of respiratory effort. A wide pulse pressure may be associated with a large PDA or significant aortic regurgitation, whereas an increased respiratory rate and effort suggests left-sided heart failure. Clues to the aetiology of a murmur include the presence of dysmorphic features, which could indicate a particular syndrome (Table 1).

Palpation

Palpation should be performed on the peripheral pulses (femoral, brachial and radial), the precordium, the abdomen for hepatomegaly and the suprasternal notch. Weak femoral pulses suggest

TABLE 1. SELECTED SYNDROMES AND ASSOCIATED CARDIAC LESIONS

Syndrome	Cardiac lesion
Fetal alcohol syndrome	Ventricular septal defect (VSD), atrial septal defect (ASD)
Trisomy 21	Atrioventricular septal defect, ASD, VSD
Turner syndrome	Coarctation of the aorta, bicuspid aortic valve
Noonan syndrome	Pulmonary stenosis, ASD, hypertrophic cardiomyopathy
Williams syndrome	Supravalvular aortic stenosis, peripheral pulmonary stenosis
Di George syndrome	Aortic arch and conotruncal abnormalities
Marfan syndrome	Mitral valve prolapse

the presence of coarctation, whereas bounding pulses may be seen in children with lesions causing low diastolic pressure, such as large PDAs and severe aortic regurgitation.

Next the apex beat may be palpated. The normal location of this differs with age, with the classic position of the fifth intercostal space midclavicular line from about age 7 years and before this in the fourth intercostal space just left of the midclavicular line.[4]

A heave is a precordial impulse resulting from volume loading. ASDs shunt left to right at the atrial level and therefore volume loading occurs in the right ventricle, producing a right ventricular heave palpable at the left sternal border. PDAs shunt left to right at the level of the pulmonary artery, completely avoiding the right ventricle, so volume loading occurs in the left side of the heart. VSDs shunt at the ventricular level, with blood flow preferentially directed into the pulmonary artery, and therefore, perhaps somewhat counterintuitively, volume loading occurs in the left side of the heart.

A thrill is the palpable vibration of a murmur and is evaluated by gently placing the finger tips over the sites of auscultation and in the suprasternal notch. Thrill location typically matches the site of auscultation (e.g. left upper sternal edge = pulmonary stenosis; right upper sternal edge = aortic stenosis; left lower sternal edge = VSD).

Auscultation

Auscultation in paediatrics can be challenging from both a technical and diagnostic perspective. The younger the child, the faster the resting heart and respiratory rates, which makes confirming the presence and characteristics of a murmur challenging.

Heart sounds

Evaluation of the murmur should be considered an 'active process' whereby it is systematically analysed. The first and second heart sounds should first be evaluated and then splitting of the second heart sound. The first heart sound corresponds to the closing of the atrioventricular valves (tricuspid and mitral valves) whereas the second heart sound is the result of closure of the aortic and pulmonary valves. Splitting of the first heart sound may be normal, but if it is widely split it may be the result of a bundle branch block or Ebstein's anomaly.[4]

Evaluating splitting of the second heart sound can be challenging because it varies with respiration. Physiological splitting is thought to result from a decreased intrathoracic pressure during inspiration causing increased venous return to the right side of the heart and subsequently delayed closure of the pulmonary valve. This explains the normal finding of a split second heart sound during inspiration with a single (or almost single) sound during expiration. If the child is compliant, fixed splitting can be evaluated by asking the patient to completely exhale. In a normal individual, there may be no splitting for a couple of beats, which then widens over the subsequent beats. Conversely, wide and fixed splitting seen in children with ASDs may be heard throughout expiration with little variation.[6]

If there is pulmonary hypertension, closure of the pulmonary valve (P2) may be loud and narrowly split from closure of the aortic valve (A2). The intensity increases due to the P2 being closed under higher pressure. (It closes earlier as the high baseline diastolic pressure in the pulmonary artery exceeds that of the right ventricle.) In cases of severe pulmonary hypertension, a single second heart sound may be heard.

To complete the assessment of heart sounds, a third or fourth heart sound may be audible. The third heart sound is a low-pitched sound heard best with the bell and at the cardiac apex. It occurs in early diastole during rapid filling and is commonly heard in hyperdynamic states (e.g. fevers) and in a volume-loaded left ventricle (e.g. left to right shunt lesions). The flow of heart sounds is similar to the cadence of the word 'Kentucky'. Although also low pitched, a fourth heart sound may occur in late diastole resulting from a stiff ventricle and is always pathological. This sound takes the pattern of the word 'Tennessee'.[7]

Murmur

The next step in the assessment of patients is evaluation of the heart murmur itself, which again should be performed in steps. First, the systolic component should be evaluated and then the diastolic component. A murmur is described in terms of its:

- quality – vibratory, musical, harsh, high or low pitched
- location/radiation
- timing – systolic, diastolic, continuous

- intensity – graded during systole from 1 to 6 and in diastole from 1 to 4 (Table 2).

Additional sounds can be difficult to appreciate but are very helpful in making a diagnosis. An ejection click occurs just after the first heart sound and is usually indicative of valvular pathology. Therefore, a click in the pulmonary region associated with an ejection systolic murmur is most likely valvular pulmonary stenosis, whereas the same at the apex is most likely aortic stenosis.

The timing of murmurs is important to help distinguish physiological murmurs from pathological ones. Any diastolic murmur should be considered pathological. If the murmur is loudest in the pulmonary or aortic valves, regurgitation is indicated. If it is loudest across atrioventricular valves, it indicates turbulent flow resulting from either an abnormal atrioventricular valve (e.g. mitral stenosis) or increased flow across that valve (the so-called diastolic rumble). All patients with diastolic murmurs should be referred for specialist review.

The most commonly heard murmurs are those in systole and may be divided into early, mid, late and pansystolic murmurs. Pansystolic murmurs are heard through the duration of the first and second heart sounds and is often difficult to hear. These murmurs are always pathological, with the underlying cause most commonly being a VSD. However, significant mitral and tricuspid regurgitation will also cause this type of murmur as they occur for the duration of ventricular systole. Early systolic murmurs share the same aetiology but in a different haemodynamic condition. Regurgitation of the mitral or tricuspid valve typically produces a pansystolic murmur, however this may also produce a short systolic murmur. The VSD producing an early systolic murmur results from a small muscular VSD that collapses on itself during systole and is often high pitched.

Midsystolic (more commonly ejection systolic) murmurs are always seen in patients with obstructive lesions such as aortic or pulmonary stenosis; however, these patients also have physiological flow murmurs or classic 'innocent' murmurs in high output states (e.g. fever or anaemia). Severe stenosis may sound more like a long systolic than ejection systolic murmur. The onset and offset of these murmurs are both between the first and second heart sounds so that both should be heard easily.

Finally, continuous murmurs are those that start in systole and continue through the second heart sound. The most common cause of this is a PDA, which has been described as a 'machine-like' murmur that is loudest in the left infraclavicular region or left sternal border.

Innocent murmurs

There are four classic types of innocent murmurs: Still's murmur, the pulmonary flow murmur, carotid bruit and a venous hum.

TABLE 2. GRADING OF MURMURS

Grade	Assessment
Systolic	
1/6	Barely audible
2/6	Soft murmur, easily audible
3/6	Loud, no thrill
4/6	Loud with a precordial thrill*
5/6	Audible with stethoscope half on the chest
6/6	Audible without stethoscope
Diastolic	
1/4	Barely audible
2/4	Soft but audible
3/4	Easily audible
4/4	Loud

* An isolated suprasternal or carotid thrill does not qualify for grade 4.

Still's murmur

The Still's murmur was described in 1909 by George Still and is the most common type of murmur in children. Classically, it occurs in children 3 to 6 years of age but may also be heard in neonates, infants and adolescents. It becomes louder during high-output periods (e.g. fever or tachycardia) and is commonly heard during assessment for viral illness. The murmur is midsystolic, has a vibratory quality and is loudest at the left sternal edge when the child is supine (and decreases when upright). It is grade 2 or below, with no diastolic component. The aetiology of this murmur is unclear and has been attributed to vibrations in the left ventricle mitral apparatus or a left ventricle false tendon.

Pulmonary flow murmur

The pulmonary flow murmur shares many of the same characteristics as the Still's murmur but its vibratory nature and location differ. It is loudest during high-output states, at the left upper sternal edge, and is grade 2 or less with no diastolic component and no clicks. It is typically louder in the supine position and has normal splitting of the second heart sound.

Carotid bruit

A carotid bruit is a high-pitched systolic murmur usually heard on the right side of the heart in the supraclavicular region resulting from flow in the aortic branches. It is grade 2 or less and may decrease in intensity with hyperextension of the shoulders.

MURMUR CHARACTERISTICS

Innocent	Pathological
• Grade 2 or less	• Grade 3 or above
• Systolic	• Pansystolic, diastolic or continuous (except venous hum)
• Musical/vibratory in nature	• Radiation
• Minimal radiation	• Harsh in nature
• Decrease in intensity when sitting	• Abnormal second heart sound
• Normal second heart sound	• Additional sounds (click, gallop rhythm)
• No added heart sounds (excluding no clicks or snaps)	• Heave/thrill
• Quiet precordium	

Venous hum

This innocent murmur is the exception to the rule of all innocent murmurs being systolic. A venous hum is a low-pitched continuous murmur heard over the anterior neck lateral to the sternocleidomastoids. Unlike its pathological counterpart, this murmur has a louder diastolic component (pathological continuous murmurs have a louder systolic component). It is heard most commonly in children aged from 3 to 8 years and is positional, decreasing when supine and disappearing with jugular compression.

A plan of action

With experience, the clinician may differentiate innocent from pathological murmurs with great sensitivity (Box).[8] Innovative methods are being developed to assist this, including a successful online program teaching Australian and Canadian medical students, and even using 'the Cloud' via the internet in remote areas.[9,10]

The decision then becomes which patients to investigate further, who to refer for echocardiogram and cardiology review, and in whom to do nothing. Investigating with routine chest x-rays is not recommended because it has a low sensitivity and positive predictive value.[11] The use of ECGs may assist in the diagnosis of a simple right-sided heart lesion (ASD or pulmonary stenosis); however, apart from large VSDs, the use of these may be limited.[12]

Neonates present a unique group in whom even paediatric cardiologists show low specificity in detecting a pathological murmur with auscultation.[13] Considering the potential for rapid and severe decline, it is recommended that all neonates with a murmur be investigated with special attention paid to weight gain, oxygen saturations and femoral pulses.

The need for referral is then based on the patient's history, results of the physical examination and the clinician's confidence in diagnosing innocent murmurs. Algorithms have been published to assist.[7] However, children with non-innocent or undifferentiated murmurs and all neonates should be referred to a specialist. In addition, murmurs in patients with syndromes known to have cardiac associations should be referred.

Conclusion

Heart murmurs in children can be a distressing finding for parents and provoke anxiety in physicians. Innocent murmurs and pathological murmurs have similar characteristics and they can be differentiated with experience. It is important to emphasise to parents that, although murmurs may be caused by heart abnormalities, a heart with an innocent murmur is still completely normal and as such the child should be treated normally. When there is doubt regarding the murmur, the child should be referred to a paediatric cardiologist without further investigations needed. Using this systematic approach provides the GP with a degree of confidence in the evaluation of a paediatric murmur and appropriateness of referral. MT

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