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Heart murmurs in children

See also the separate Heart auscultation article.

What is a heart murmur?

A heart murmur is an additional sound (other than the first or second heart sound), heard when auscultating the heart. Heart murmurs are common in asymptomatic and otherwise well children. Many murmurs are innocent and result from normal patterns of blood flow through heart and blood vessels. [1]

However, a murmur can be the sole manifestation of structural heart disease. More than 50% of newborns with congenital heart disease are unrecognised at birth. ^[2] Therefore, a careful evaluation is essential. If in doubt, referral to a paediatrician with expertise in cardiology (PEC) or a paediatric cardiologist is necessary.

Symptoms of heart murmur in children (presentation)

Does this child have heart disease? [3]

The key question which is raised on the detection of a heart murmur is: 'Does this child have a heart disease?' To answer this, three groups of factors need to be considered:

- 1. Are there any symptoms and signs of heart disease?
- 2. Are there any predisposing medical conditions?
- 3. What are the characteristics of the murmur?

Symptoms of heart disease

The symptoms vary with the age of the child and are more nonspecific in infants. One or more of the following may be present:

| Infant | Older Child |
|-------------------------|-------------------------|
| Breathlessness | Exercise intolerance |
| Poor feeding | Palpitations |
| Excessive sweating | Chest pain |
| Blue episodes | Syncope |
| Generally unwell | Pedal oedema |
| Not gaining weight | Positive family history |
| Positive family history | |

Signs of heart disease

| Infant | Older Child |
|---------------------------------|--|
| Tachypnoea | Tachypnoea |
| Tachycardia | Tachycardia |
| Hepatomegaly | Hepatomegaly |
| Poor peripheral pulses | Poor peripheral pulses |
| Low oxygen saturations | Elevated jugular venous pressure (JVP) |
| Faltering growth (growth chart) | Pedal oedema |
| | Basal lung crepitations |

Predisposing medical conditions

- Down's syndrome.
- Noonan's syndrome.
- Turner syndrome.
- Marfan's syndrome.
- Fetal alcohol syndrome.
- Fetal rubella infection.

Characteristics of the murmur

Each heart murmur must be analysed in terms of intensity (grades 1 to 6), timing (systolic or diastolic), location, transmission and quality (musical, vibratory, blowing, harsh, etc).

A grade 1 murmur is barely audible, grade 2 is soft but easily heard, grade 3 is loud but not accompanied by a thrill, while grade 4 is associated with a thrill. Grade 5 and 6 are very loud murmurs which may be audible with stethoscope partly or completely off the chest.

Most murmurs are systolic and location of a systolic murmur can point toward specific cardiac diagnosis as described in the table below:

| Upper left sternal border | Pulmonary stenosis (PS), atrial septal defect (ASD), innocent pulmonary flow murmur, tetralogy of Fallot (TOF), coarctation of the aorta (CoA), aortic stenosis (AS), patent ductus arteriosus (PDA) with pulmonary hypertension |
|----------------------------------|--|
| Upper right sternal border | AS, supravalvular AS, subaortic stenosis |
| Lower left sternal border | Ventricular septal defect (VSD), Still's murmur, hypertrophic obstructive cardiomyopathy (HOCM), tricuspid regurgitation (TR) |
| Apical area | Mitral regurgitation (MR), mitral valve prolapse (MVP), HOCM, vibratory innocent murmur |

Likewise timing of the murmur can help in identifying the cause of the murmur:

| Systolic | Diastolic | Continuous |
|----------|------------------------------|------------------------------------|
| VSD | Aortic regurgitation (AR) | PDA |
| TR | Pulmonary regurgitation (PR) | Venous hum |
| MR | Mitral stenosis (MS) | Arteriovenous malformations (AVMs) |
| PS | | |
| AS | | |
| ASD | | |
| PDA | | |

Pathological murmurs

Diastolic murmurs are pathological. Six cardinal signs are described which indicate that a systolic murmur is likely to be pathological, ie because of an underlying heart defect. These are:

- Holosystolic (pansystolic) murmur.
- Harsh murmur.
- Abnormal heart sounds.
- Early or mid-systolic click.
- Grade 3 murmur or greater.
- Heard over upper left sternal border.

Innocent murmurs^[4]

- Sensitive (changes with child's position or with respiration).
- Short duration (not holosystolic).
- Single (no associated clicks or gallops).
- Small (murmur limited to a small area and not radiating).
- Soft (low amplitude).
- Sweet (not harsh-sounding).
- Systolic (occurs and is limited to systole).

Note that, as innocent murmurs are flow murmurs, they may become louder (or only be audible during) states of relatively increased cardiac output, such as during exercise, with anxiety, during a fever, or with anaemia. [5]

Five types of innocent murmurs in childhood are described, all with diagnostic clinical features. However, the differential diagnosis always includes pathological murmurs because of various heart defects. These are summarised below:

| Innocent Murmurs | | |
|--|--|--------------|
| Name | Features | Differential |
| Still's murmur | Mid-left sternal border, mid- systolic, grades 2-3, twanging string, musical, vibratory sound | VSD |
| Pulmonary flow murmur | Upper left sternal border, mid- systolic, grades 1-3, grating | PS, ASD |
| Venous hum | Right and/or left infraclavicular, continuous, only heard in upright position, diastolic component louder than systolic | PDA |
| Carotid bruit (supraclavicular systolic murmur) | Supraclavicular area, ejection systolic, grades 2-3 | AS |
| Peripheral pulmonary stenosis (pulmonary flow murmur of newborn) | Upper left sternal border, grades 1-2, radiates to axillae and back, usually disappears by 6 months of age | PS |

Neonatal heart murmurs

Heart murmurs in neonates are much more likely to indicate structural heart disease and should prompt specialist assessment.

One study of 6,333 healthy newborn babies found heart murmurs in 87 (1.37%) neonates, of whom 42.5% had a structural cardiac malformation. Ventricular septal defect (62%) was the most common diagnosis, followed by atrial septal defect, pulmonary stenosis and patent ductus arteriosus. [6]

Even potentially life-threatening heart defects may have no other signs or symptoms in addition to the heart murmurs. A very thorough evaluation including detailed clinical examination, femoral pulse check along with pulse oximetry (pre- and post-ductal saturations) is mandatory. Those with clinical signs, difficult-to-palpate femoral pulses or low oxygen saturations require prompt echocardiographic assessment. Neonates with heart murmurs who are clinically asymptomatic should also be referred for a routine echocardiographic assessment.

Diagnosing heart murmurs in children (investigations)

ECG and CXR have limited use in the diagnosis of underlying pathology associated with pathological heart murmurs, with low sensitivity and specificity for identifying cardiac defects or anatomical abnormalities.

Echocardiography is the gold standard to diagnose congenital cardiac malformations definitively in paediatric patients. It is indicated in any child with an asymptomatic heart murmur which has attributes of a pathological murmur or when the examiner is not comfortable in making a clinical diagnosis of an innocent heart murmur. This can usually be done by a PEC in a district general hospital setting, minimising the need for referral to a tertiary cardiology service. ^[7]

Management of heart murmurs in children

Any child found to have a heart murmur should have a thorough clinical evaluation including pulse oximetry and palpation of femoral pulses. Clinically unwell children or those with red flags such as difficult-to-feel femorals or low oxygen saturations need urgent referral for specialist cardiac evaluation.

It is important to remember that absence of symptoms does not exclude important pathology. If in doubt, referral to a PEC or a paediatric cardiologist is essential.

Evaluation of murmurs [8]

A suggested framework for evaluating murmurs is given below.

Infants and children who are symptomatic with a heart murmur should be discussed with paediatric cardiology urgently, with a view to urgent inpatient echocardiography and further management. This applies also if:

- There are signs of heart failure or shock.
- Lower limb oxygen saturations are below 96%.
- There is >3% difference in pre- and post-ductal oxygen saturations.
- There are absent or weak femoral pulses.

In neonates and infants who are asymptomatic with a heart murmur, arrange echocardiography and/or a paediatric cardiology opinion as soon as possible (as an inpatient, or within a week), if:

- The murmur is loud (greater than 2 out of 6 intensity).
- The murmur is diastolic, pansystolic, or continuous.
- A heave is palpable.
- The location of the murmur is anywhere other than at the lower sternal edge.
- There is an abnormal ECG.
- The infant has dysmorphic features.

Congenital heart disease is unlikely if **all** of the following are present:

- The infant is otherwise well.
- There are no signs of heart failure.
- Normal pulses are palpable.
- Oxygen saturations are normal.
- The murmur is soft and midsystolic.
- The murmur is audible only at the lower sternal edge.

Neonates and infants meeting all of these criteria should be re-reviewed by, or referred to, paediatric services routinely, with a view to arranging an echocardiogram if the murmur persists on review.

In older children (beyond infancy), refer to paediatric cardiology for consideration of echocardiology if **any** of the following are present: [9]

- Loud murmur (greater than 2 out of 6 intensity).
- Diastolic murmur.
- Murmur associated with a click, S3 or S4, or fixed S2 split.
- Murmur becomes louder with standing or Valsalva manoeuvre.
- The murmur radiates to the carotids.
- The cardiovascular examination is otherwise abnormal (eg, abnormal pulse rhythm, asynchronous or variable upper and lower extremity pulses, or signs of heart failure).

If none of these features are present, and there are no risk factors for structural heart disease (personal or family history) an innocent murmur is likely, and reassurance and safety-netting advice can be given.

Patient and family education

Once a heart murmur is confirmed to be innocent, reassurance to the family regarding its benign nature is important. Although the murmur may never disappear and may persist into adulthood, the parent and child need to be specifically reassured that an innocent murmur is simply an additional noise audible to the clinician and not a disease or illness, and is therefore completely harmless.

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