



# Evaluation of asymptomatic heart murmurs

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## KEYWORDS

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**Summary** Cardiac murmurs are a common finding in asymptomatic paediatric patients. The most common causes are innocent murmurs and minor structural heart disease. Beyond the age of 6 weeks patients are unlikely to have major underlying heart disease. Presentation of asymptomatic murmurs may be variable and assessment strategies depend on the age of the patient and should include clinical examination with measurement of blood pressure, evaluation for presence of cyanosis, electrocardiogram and echocardiography in selected patients.

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## Practice points

- Sole clinical evaluation by non-specialists is inadequate in differentiating innocent from pathological murmurs
- Clinical examination is not sufficiently sensitive to identify cyanosis in children
- Simple clinical examination combined with pulse oximetry and an electrocardiogram (ECG) will allow effective prioritization of referrals between non-specialists and paediatric cardiologists
- The chest X-ray (CXR) is of no value in the routine evaluation of asymptomatic murmurs

- It is unlikely that a major or complex congenital cardiac lesion will present for the first time after 6 weeks of age

## Introduction

Heart murmurs are a common finding in children, their incidence depending partly on the effort with which they are sought.<sup>1</sup> The 'diagnosis' of a heart murmur induces a high degree of anxiety in parents, who usually associate a suggestion of possible heart disease with poor prognosis and terminal illness, as not infrequently witnessed in elderly relatives. They are unprepared for the possibility of heart disease in their child, and usually lack knowledge and understanding of basic paediatric cardiac entities such as 'innocent murmurs'.

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However, there are some simple rules that enable practitioners to make an educated guess about the nature and significance of heart murmurs. In his 1960 review Fogel<sup>1</sup> emphasizes that the cardiac cycle is not silent. The flow of blood around the heart and great vessels always produces some sound, although frequently at a threshold below that of the human ear aided by a stethoscope. It is therefore not unusual for a murmur to be audible in patients without heart disease, even though the incidence of murmurs in routine practice does not seem to reach the 50–60% quoted by some authors.

Innocent murmurs are most commonly of crescendo–decrescendo mid-systolic type, of lower intensity (grade I–II) being audible at the (lower) left sternal border, non-radiating, and increasing in intensity in supine position. They usually have associated normal, variable splitting of the second heart sound, but are not associated with clicks or snaps. In contrast to systolic murmurs it is axiomatic that diastolic murmurs, whilst unusual, are always pathological with the exception of venous hums that occasionally extend into diastole when the patient is sitting up. Functional murmurs are more readily audible in high-output cardiac states, for example, with fever or excitement.

Apart from their specific character, innocent murmurs are defined through the normality of all other findings.<sup>2</sup> However, the lack of identifiable abnormality does not necessarily guarantee cardiac normality. Significant lesions do not always have classical associated findings, and even if they do, the average clinician may find it difficult to identify subtleties such as fixed splitting of the second heart sound, clicks or opening snaps. Frequently these findings are only confirmed retrospectively after echocardiography has demonstrated a specific cardiac abnormality.

## Age considerations

### The neonate and young infant

A lower threshold for referring asymptomatic infants with 'routine' murmur for a more detailed cardiac evaluation, will result in a higher probability of identifying structural heart disease.<sup>3–6</sup> Physiological changes that occur during the transition from in utero to ex utero life may mask even significant congenital cardiac lesions, which should be considered during the initial assessment. A good example is a large ventricular septal defect (VSD) in which a left-to-right shunt may be minimal soon after birth due to the high pulmonary vascular

resistance, which only gradually declines over the first few weeks of life. Neonates with large VSDs are therefore commonly asymptomatic early in life, and the classical finding of an audible murmur is often not present during this period. Similarly, infants with duct-dependent lesions such as pulmonary atresia, may have only very subtle symptoms or signs early on, but are likely to become acutely unwell if a detailed evaluation is delayed and the duct constricts a few days after birth.

However, even though there is a relatively high incidence of structural heart disease in neonates presenting with, for the most part, asymptomatic cardiac murmurs (50–75%),<sup>5,6</sup> it is clear that even in this population serious lesions generally present symptomatically, with a murmur being identified subsequently.

However, not all infants require immediate specialist assessment once a murmur has been identified and the key to an appropriate management strategy is thorough clinical assessment and prioritization.

### The older child

In contrast to neonates and infants, an asymptomatic murmur identified in an older child is much more likely to be 'innocent' in nature, and it is frequently an incidental finding during an intercurrent illness. Cardiac abnormalities that can present with an asymptomatic murmur in the older child are either acquired cardiac lesions such as rheumatic heart disease, or more commonly congenital cardiac abnormalities that are either of minor severity or that have slowly progressed over time (Table 1). Acquired cardiac abnormalities are less likely to present with asymptomatic murmurs, and instead frequently present with symptoms of the primary pathological process before the development of an audible murmur.

In the absence of a characteristic history of a progressive cardiac lesion there is no immediate need to refer older children with asymptomatic murmurs for specialist cardiac assessment. In fact the re-evaluation of an incidental murmur noted during an acute illness, once the child has recovered, will allow the practitioner to reassure parents in those cases where the murmur has spontaneously disappeared.

### Murmur variability: now you hear it now you don't

There are several possible reasons for a murmur being audible on some occasions and not on others.

**Table 1** Congenital and acquired lesions that may present in the older child with an asymptomatic murmur.

Congenital lesions that may present in the older child with an asymptomatic murmur:

- Atrial septal defect (ASD)
- Aortic coarctation
- Aortic or pulmonary valve stenosis
- Small ventricular septal defects with minor left–right shunt
- Small persistent arterial duct (PDA) with minor left–right shunt
- Mitral regurgitation (MR)

Acquired lesions that may present in the older child with an asymptomatic murmur:

- Rheumatic valvar heart disease (e.g., MR)
- Cardiomyopathies (e.g., hypertrophic obstructive cardiomyopathy)
- Cardiac manifestations of systemic disease (e.g., muco-polysaccharidoses)
- Endocarditic valve lesions

This is obviously a feature of an innocent murmur that may appear with high output states such as fever. This is particularly the case with pulmonary flow murmur.

However, this variability is not only limited to innocent murmurs. The physiological changes of the neonatal period, for example, characterized by a slow drop in pulmonary vascular resistance, explain why VSD and persistent arterial duct (PDA) murmurs may be inaudible in the first few days of life.

Murmurs secondary to congenital regurgitation of the atrio-ventricular (AV) valves (tricuspid or mitral) are likely to be apparent early in life, but they may become more audible if progressive ventricular dilatation leads to a higher degree of AV valve regurgitation.

In contrast, murmurs that result from congenital stenosis of a semi-lunar valve (aortic or pulmonary) are usually present from birth unless the stenosis is acquired or progressive. As in most cases flow across these valves is 'obligatory' there is little opportunity for change; though absolute intensity will vary with flow and gradient. It is important to emphasize that any progressive congenital or acquired cardiac lesion can be associated with change in appearance and intensity of associated cardiac murmurs.

## Cardiac lesions and asymptomatic murmurs

### Minor lesions

Haemodynamically insignificant lesions such as mild pulmonary stenosis, mild aortic stenosis and small

ventricular septal defects usually give characteristic cardiac murmurs of ejection systolic or pansystolic type. The intensity of these murmurs should be grade 1–3 and there is usually no associated thrill. Unless a murmur is very loud and 'omnipresent', its localization and radiation is expected to match standard clinical descriptions.

Minor lesions are unlikely to be associated with findings other than the murmur itself. Heaves and thrills are extremely rare, although not impossible, especially in slim children with, for example, mild–moderate pulmonary valve stenosis.

Generally most minor lesions do not require any immediate intervention at presentation, and can be followed with a 'watch-and-wait' approach. Thus there is no need for immediate referral for definitive cardiac assessment in those patients in whom a minor lesion is suspected.

### Significant 'asymptomatic' lesions

There is a small, but significant, group of patients with congenital heart disease, who do not follow the classical rules of clinical presentation. Remarkably severe lesions can balance themselves so as to minimize clinical symptoms. For example in our own institution we have seen patients with tetralogy of Fallot presenting as late as 50 years of age with clinical symptoms related to atrial arrhythmias, having lived a virtually normal life up to this stage.

Sometimes these conditions become evident when a secondary illness, such as a viral respiratory tract infection, disturbs the cardiovascular balance to a point where decompensation occurs. However, sometimes an apparently minor murmur in an otherwise asymptomatic patient may be the first

hint of a significant cardiac problem. In this context one should bear in mind that the intensity of a murmur is a product of both the flow and pressure gradient, and may not necessarily reflect the severity of the underlying cardiac disease.

Progressive pulmonary vascular disease as a result of a moderate or large VSD, for example, may lead to a reduced intensity of the associated cardiac murmur. A high pulmonary vascular resistance will also mask heart failure as it will lead to a reduced left-to-right shunt. In a few children, these changes can occur quite quickly, in others, the pulmonary vascular resistance may never fall significantly after birth. These children may consider themselves unlucky in that their unusual physiology has prevented them from developing symptoms related to the underlying disease, which would otherwise ring alarm bells.

Asymptomatic cyanosis—an oxymoron some might say—is not common but is physiologically quite easy to comprehend. The body does not require full saturation of haemoglobin per se as long as oxygen delivery to the tissue is adequate. As oxygen delivery is the product of cardiac output and oxygen carriage it follows that a mild degree of desaturation may well be compensated through a slight increase in cardiac output, without any noticeable symptoms. However, it is unlikely that careful assessment with our scheme would be normal in such a child. At the very least they will have abnormal saturations or electrocardiogram (ECG) patterns.

## Evaluation of asymptomatic murmurs

The aim of any evaluation strategy of asymptomatic cardiac murmurs should be to limit unnecessary investigations and referrals as much as possible, whilst at the same time providing an appropriate safety net to diagnose severe underlying heart disease at an early stage before any adverse events occur. Any such strategy has to be adapted further to the needs and level of anxiety of the individual patient and family.

Around 80% of all congenital heart disease that will eventually present in the first year of life will have had neonatal screening check without symptoms.<sup>3</sup> Although 50% of these patients will pass the neonatal screening examination, the out-of-hospital mortality of undiagnosed congenital heart disease in the first 6 weeks of life is about 1.6%.<sup>3</sup> However, the mortality of undiagnosed congenital heart disease in older infants that have correctly or incorrectly passed the 6-weeks screening examina-

tion has been reported to be extremely low, if not close to 0%.<sup>3</sup> These figures illustrate very well the importance of differentiating between infants and older children when applying an evaluation strategy for asymptomatic cardiac murmurs (Fig. 1).

## Clinical examination

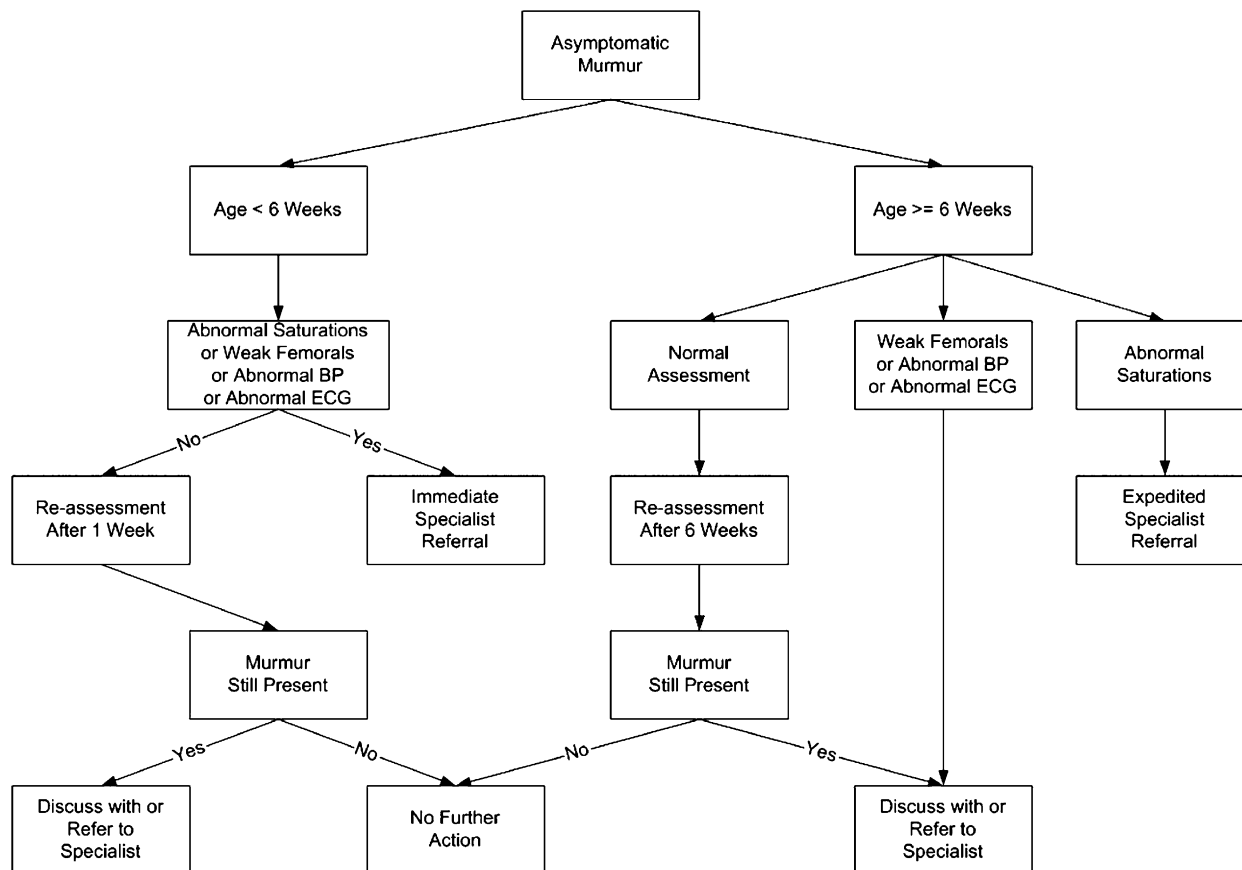
Any child with a heart murmur deserves a full examination of the cardiovascular system, at least incorporating an assessment of peripheral pulses, evaluation for possible cyanosis and documentation of a non-invasive blood pressure (BP) recording. As by definition this article deals with asymptomatic murmurs we would not expect to find any evidence of heart failure or respiratory compromise on clinical examination (and therefore will not discuss the clinical signs of decompensated cardiac disease).

### Peripheral pulses

Both brachial (infant) or radial (older child) and both femoral pulses should be routinely examined and compared. Weak femoral pulses, especially if discrepant to the quality of brachial or radial pulses or associated with radio-femoral delay, are suggestive of aortic coarctation or aortic arch obstruction, and should, therefore, lead to further more detailed investigations. Weak pulses all round can be an incidental finding, but could also be the result of significant left ventricular outflow tract obstruction or aortic stenosis, as well as poor ventricular function, although the latter is unlikely in the absence of symptoms.

### Cyanosis and transcutaneous oxygen saturations

Though the identification of cyanosis is considered a standard clinical skill there is remarkably little evidence regarding the sensitivity and specificity of clinical examination for varying degrees of cyanosis. Surprisingly cyanosed children can be missed by medical services at a number of levels. It is important to emphasize that a cyanosed child is not necessarily 'blue' but may instead present with a subtle degree of paleness, especially in the context of coexisting anaemia, such as physiological anaemia of the newborn. One should also remember to differentiate between central and peripheral cyanosis during clinical examination. Circumoral pallor is a particularly common finding in children and does not have a significant association with cardiac disease. In one study of emergency referrals of children with respiratory difficulties, in whom cyanosis might be more easily recognized because of a higher index of suspicion, a



**Figure. 1** Flowchart suggesting a strategy for evaluation of asymptomatic cardiac murmurs by the general paediatrician or practitioner.

clinical examination was found to be only 33% sensitive at a threshold of 92%.<sup>7</sup>

The value and feasibility of routine screening pulse oximetry in identifying congenital cardiovascular malformations is questionable when used non-selectively in neonates,<sup>8,9</sup> but it may serve as a helpful technique when evaluating infants in whom congenital heart disease is suspected. When using pulse oximetry to screen for congenital cardiovascular malformations the sensitivity can be further increased through comparison of right arm and lower limb saturations.<sup>9</sup> Even though it is probably not feasible to use these devices in primary care environments it seems foolhardy not to use such a technology in secondary and tertiary care.

#### BP

The standard oscillometric method of obtaining BP recordings in the neonate should be routinely applied in all infants upon first clinical encounter. However, the value of four limb BP measurements in the neonate is questionable with differences of as high as 20 mmHg having been identified to be

'more likely to be due to random variability in measurement than to coarctation of the aorta'.<sup>10</sup> If the evaluation of the femoral pulses suggests possible coarctation in a neonate, then echocardiography is clearly indicated. Straightforward upper limb BP measurements may be more valuable in the older child where these measurements are less fraught and more reproducible. For coarctation in the older child absolute levels of upper limb BP are much more important, representing as they do the risk of end-organ damage.

#### ECG and chest X-ray

The value of chest X-ray in the diagnosis of congenital heart disease is limited. Several studies have shown that chest X-rays have an extremely low sensitivity and specificity for identifying cardiac lesions.<sup>11-14</sup> Because chest X-rays also expose the child to ionizing radiation, their routine use cannot be considered appropriate in the asymptomatic child.

The ECG also has a low sensitivity in identifying congenital cardiac lesions in asymptomatic children with cardiac murmurs. As most of these murmurs

will either be innocent or the underlying cardiac abnormalities will have no physiological effect, ECG changes are not seen. In addition, the value of the ECG to the general paediatrician is also limited by confidence in its interpretation, not least related to the wide variation of normal during infancy and childhood. However, being able to identify the QRS and *p*-wave axis, bundle branch block patterns and the presence of ventricular hypertrophy goes a long way in making best use of this diagnostic modality in paediatric patients. Identifying ECG abnormalities may on occasions suggest a more specific cardiac abnormality, such as left superior axis being associated with atrio-ventricular septal defect, but more importantly it will provide a useful indicator for the need for further echocardiographic evaluation. The low prevalence of significant pathology in asymptomatic patients with cardiac murmurs makes the negative predictive value high despite low sensitivity. However, the ECG is non-invasive and easily transmitted for expert review, and should, therefore, be considered as part of the routine initial assessment.

### Echocardiography

Echocardiography remains the gold standard for diagnosing congenital cardiac lesions. However, this method is not always readily available to the general paediatrician, and is used mostly as an assessment tool for the paediatric cardiologist.

The role of echocardiography in the evaluation of asymptomatic murmurs has changed significantly over the last 10 years. Though sensitivity and specificity of clinical assessment by a paediatric cardiologist are quite well validated<sup>15-18</sup> the ease and lack of contra-indication to echocardiography make it a difficult investigation to refuse, not least on the background of increasing medical litigation, as well as patient and parent expectation.

Even though it is quite unlikely that a significant pathology would be missed in older children when innocent murmurs are diagnosed solely on clinical examination, minor lesions may masquerade under the impression of innocence. Danford and colleagues identified 16 patients with cardiac abnormalities on echocardiography, out of 187 who were clinically thought to have innocent murmurs, four of whom required invasive interventions.<sup>19</sup>

One of the major concerns using the strategy of echocardiographic evaluation in all patients with cardiac murmurs is the fact that it is associated with a significant pick-up of otherwise trivial conditions, such as silent and small arterial ducts, patent foramen ovale and small atrial septal

defects, which may cause unnecessary anxiety in patients and relatives.

Whether or not general paediatricians and practitioners should diagnose innocent murmurs is a debate best summed up by Pelech<sup>20</sup> who uses the term 'comfort factor' to refer to the degree of uncertainty which an individual clinician is willing to tolerate. Studies of the sensitivity and specificity of non-specialist assessment are mixed, but would not generally support an adequate degree of diagnostic precision<sup>21,22</sup> On the background of increasing medical litigation it is probably wise for the non-specialist to have a low threshold for referring patients with asymptomatic murmurs that have been noted on multiple clinical encounters for further specialist assessment.

### Conclusion

Asymptomatic murmurs are a common finding in infants and older children, with innocent murmurs, as well as minor structural heart disease, accounting for the majority of these cases. Assessment strategies employed by the general paediatrician and practitioner need to be customized to the age of the patient. If referral for specialist review by a paediatric cardiologist is considered the following information should be supplied, which will allow the cardiologist to appropriately prioritize the patient:

- Age and clinical status of the child including family history and associated extra-cardiac anomalies
- Description of the murmur (volume, character, position, radiation)
- Peripheral pulse assessment, as well as BP recording
- If possible, differential upper and lower limb saturations
- ECG evaluation

If all the above findings are normal the parents may be reassured that, even in the possible presence of structural heart disease, the possibility of a clinically significant lesion is low.

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