

# Management of asymptomatic heart murmurs in infants and children

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## Abstract

Murmurs heard in neonates and older children may be innocent or signify heart disease. The neonatologist or general paediatrician must decide which infants and children need specialist referral, and if so, the urgency of referral. Neonates with clinical features suggesting heart disease should be referred urgently. Neonates with murmurs and no other pathological features must be reviewed regularly during the first few weeks of life if they are not referred to a cardiologist. Older children with asymptomatic murmurs should be referred routinely to a cardiologist if the paediatrician or parents are concerned about the possible significance of the murmur.

**Keywords** children; congenital heart disease; murmur; neonate

## Introduction

In the past 25 years, echocardiography has revolutionised the management of heart murmurs in infants and children. Prior to this, the only resources available to the general paediatrician to aid management were his or her clinical acumen, chest x-ray and electrocardiography, and referral to a paediatric cardiologist. The latter were few in number and apart from greater clinical experience and expertise the only added resource they had to offer was cardiac catheterisation. Since subjecting asymptomatic children with murmurs to the latter often seemed disproportionate, paediatricians undertook responsibility for clinical management of many such children. This often involved periodic review, not only of children with innocent murmurs, but also of some with a clinical diagnosis of asymptomatic and uncomplicated heart disease, such as ventricular septal defect.

This practice demanded of paediatricians that they maintained their clinical skills, offered continuity of family care, and

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provided gate-keeping access to scarce paediatric cardiology resources. The practice carried several disadvantages, however. Difficulty in categorically excluding heart disease led to children with innocent murmurs being subjected to unnecessary outpatient review, inappropriate advice concerning antibiotic prophylaxis and possibly even more harmful constraints. In contrast, diagnosis of significant heart disease may have been delayed in some children.

The advent of echocardiography appeared, at first glance, to solve these problems. It permits prompt diagnosis of most types of congenital heart disease. It can help to exclude heart disease in children with innocent murmurs, thus obviating the need for inappropriate outpatient review and unnecessary precautions. Echocardiography is not a panacea, however. It demands considerable training and ongoing practice to maintain skills. Undue reliance on echocardiography may supplant the clinical skills that older paediatricians had to maintain. A normal scan does not always provide the expected reassurance to parents. Finally, incidental findings, in particular persistent patent foramen ovale, may not only induce parental anxiety but also mandate further review, thus defeating the purpose of conducting a 'therapeutic' scan. Paediatricians, therefore, must continue to exercise clinical judgement in determining which children and families need specialist referral, and when. This paper is written from the perspective of the general paediatrician or neonatologist, working in a climate of possibly increasing pressure to refer all infants and children with murmurs to a specialist.

## Objectives in the assessment and management of murmurs

The paediatrician must consider several issues when faced with an infant or child with an asymptomatic murmur. The first priority is the prompt recognition of structural heart disease. The second is the reasonably confident exclusion of such disease, and reassurance of the child and family. These objectives must be achieved by the judicious use of specialist resources, that is, paediatric cardiology and echocardiography. Since resources do not currently permit specialist referral of all such children, these objectives may conflict with one another. Thus, in some children, the pronouncement that a murmur is innocent is made on the balance of probabilities, and parents and clinicians may have to accept some uncertainty. The probability of identifying significant heart disease is greater in neonates than in older children and uncertainty is less acceptable. We will, therefore, outline the management of murmurs in neonates separately from their management in older children.

## Asymptomatic murmurs heard in the neonatal period

### Prevalence

The reported prevalence of murmurs in the neonatal period varies from 0.6 to 1.9 per 1000 infants.<sup>1-4</sup> This variation may depend on the timing of examination, whether preterm infants and infants with other cardiovascular symptoms and signs are included, and on the size of the population studied. In general, there is an inverse association between population size and the prevalence of murmurs.<sup>1</sup>

### Pathological associations

Many types of congenital heart disease can be associated with an asymptomatic murmur. The most common association is ventricular septal defect (VSD); and patent ductus arteriosus (PDA) if preterm infants are included.<sup>5</sup> Many VSDs identified in the neonatal period are small and close spontaneously.<sup>3</sup> Murmurs can also, however, be associated with obstructive or cyanotic congenital heart disease, in which early identification and treatment are essential.<sup>2,6,7</sup>

### Innocent (functional) murmurs

Non-pathological murmurs in the neonate are generally related to blood flow through the ductus arteriosus or to turbulent flow in the pulmonary arteries.<sup>1</sup> Arlettaz *et al.* conducted serial clinical and echocardiographic review of 50 neonates with 'innocent murmurs' and 50 controls.<sup>8</sup> All infants with murmurs had a patent foramen ovale (PFO) compared with 82% of controls, 50% with a murmur had pulmonary branch stenosis (PBS) compared with less than 12% of controls and 60% of infants with a murmur had a PDA, compared with 6% of control infants. In all cases the duct had closed by 6 weeks, and the PBS had resolved by 6 months. Patency of the foramen ovale persisted in 66% of infants.

### Investigation and management: referral or review?

Careful clinical examination for other cardiovascular findings is mandatory, and should be repeated daily if the infant remains in hospital. Measurement of blood pressure in all limbs is often done, but its variability in normal neonates casts doubts upon its reliability in identifying or excluding duct-dependent heart disease.<sup>9</sup> Chest x-ray is unlikely to be of value as it may be normal in asymptomatic infants with heart disease. Electrocardiography may be of value, as an abnormal axis or changes suggesting ventricular hypertrophy raise the possibility of heart disease. Pulse oximetry is receiving increasing attention as a possible screening tool for congenital heart disease.<sup>10</sup> It should be conducted in all neonates found to have a murmur. It may detect subtle or evolving hypoxaemia, and differences between upper and lower limb oxygen saturations may point to a duct-dependent lesion.<sup>11</sup>

The challenge facing the clinician is to decide whether to refer a neonate with a murmur for urgent cardiological assessment and echocardiography. In infants who have any other cardiac symptoms or signs, who have dysmorphic features or congenital abnormalities, who have a harsh or a loud murmur, or who show hypoxaemia on pulse oximetry this decision is straightforward. Whether to refer infants who are otherwise asymptomatic and have normal oxygen saturation is more problematic. Proponents of early referral dispute the frequency of innocent murmurs in neonates and cite the challenges posed by physiological changes in the circulation after birth and early discharge of neonates.<sup>1,2,7</sup> They argue that routine neonatal examination is insensitive at detecting congenital heart disease, and that identification of a murmur at least gives an opportunity for targeted investigation of infants at particularly high risk. In support of this case is the extensive experience of infants presenting critically ill, or dying, with obstructive left heart malformations, before diagnosis and timely treatment.<sup>1,7</sup>

The number of units with a paediatrician or neonatologist with a special interest in cardiology is increasing and in these

units early echocardiography of all infants with a murmur may be feasible. There are issues relating to quality control, accumulating experience and maintaining skill. Also, the findings on echocardiography conducted by a non-specialist may need to be confirmed later by a cardiologist. Some lesions, such as anomalous pulmonary venous drainage, can be very difficult to identify, even by experienced practitioners.

Many units do not have this facility and the general paediatrician or neonatologist must still decide which infants to refer and which may safely be reviewed. For an otherwise well infant, with no symptoms or signs other than a medium to low intensity murmur, recent experience suggests that careful and timely clinical review is safe and effective, and may reduce the need for cardiological referral of all such infants. Patton and Hey reported their experience in a maternity unit where routine neonatal examination was conducted by advanced neonatal nurse practitioners.<sup>4</sup> About 10% of all neonates were found to have a murmur. They were discharged if otherwise clinically well and if pulse oximetry was normal. The parents were given verbal and written advice concerning murmurs and significant symptoms. The infants were reviewed during the first and second weeks of life. If the murmur was still present at 2 weeks of age, the infant was referred to a paediatric cardiologist. One-third of the latter were found to have a structural heart defect. During the 8-year period of audit, the number of infants with congenital heart disease diagnosed after discharge declined. In the past 4 years, only 6% of cases were identified after discharge, and none were life-threatening.

A similar recent study reported the performance of senior house officers in evaluating murmurs heard on routine examination.<sup>3</sup> During a 2-year period, 112 such infants were identified. Infants with significant clinical features were referred for immediate senior assessment. The rest were reviewed in a dedicated clinic run by a consultant neonatologist with experience and training in echocardiography. All 12 infants referred for immediate cardiological assessment because of clinical concern had significant pathology – 11 had structural heart disease and one had pulmonary hypoplasia. The remaining 100 infants were referred to the 'murmur clinic'. Of the 78 who attended, only 16 infants still had a murmur. Three of these had a normal echocardiogram, seven had pulmonary branch stenosis and six had minor heart lesions (VSD, PDA and pulmonary valve stenosis (PS)). Pulmonary branch stenosis resolved eventually in all infants. Of the 22 who defaulted, none were known to have presented to the hospital, or elsewhere, with heart disease.

These studies were relatively small and the findings cannot be used to guarantee that clinical review is always as safe as urgent cardiological referral. They do suggest, however, that a carefully structured system, with appropriate training, provision of appropriate information to parents, facility for review of infants during the first weeks of life and audit of outcomes is likely to be safe for the many infants who have either an innocent murmur or non-life-threatening heart disease. It also provides substantial, although not absolute, safeguards that may identify infants with evolving symptoms or signs of serious heart disease.

Depending on local facilities and arrangements, regular review of neonates with a murmur in the first few weeks may be no less draining on resources than early cardiological referral. In such instances, the latter approach may be preferable.

## Management of asymptomatic murmurs beyond the neonatal period

In evaluating the older infant or child found to have a murmur, the same general principles should inform the paediatrician's practice – appropriate referral if the clinical features suggest pathology and reassurance if they do not. There are, however, differences in emphasis compared with the evaluation of neonatal murmurs. First, identification of life-threatening heart disease is less likely in an older child presenting with a murmur. Second, compared with neonates, murmurs first heard in older infants and children are more likely to be innocent. In contrast, the prevalence of asymptomatic murmurs (up to 70% of children) is greater in older children than in neonates, so the obligation to 'gate-keep' remains. Finally, echocardiography does not always provide definitive reassurance in the evaluation of asymptomatic murmurs. McCrindle reported that, even after children with murmurs were reported to have a normal heart following echocardiography, 10% of parents still had anxieties and misconceptions, often still conflating a murmur with heart disease.<sup>12</sup> Liberal use of echocardiography does not replace the need for careful clinical management and communication with parents.

### Pathological associations

Pathological murmurs may be associated with a jet effect due to blood flowing through a stenotic valve (e.g. aortic stenosis) or through a left to right shunt (e.g. VSD), or to increased pulmonary blood flow (e.g. atrial septal defect (ASD)). Table 1 lists the common types of congenital heart disease that may be associated with an asymptomatic murmur, along with possible associated clinical features. Less commonly, a murmur may point to cardiomyopathy,

### Common types of congenital heart disease that may present with an asymptomatic murmur beyond the neonatal period

Type of congenital heart disease	Clinical features
Atrial septal defect	Wide, fixed splitting of second heart sound
Ventricular septal defect	Discrete blowing or harsh murmur Thrill
Aortic stenosis	Harsh murmur that may radiate to neck Ejection click Thrill
Pulmonary stenosis	Ejection click
Patent ductus arteriosus	Murmur heard over upper precordium or back Diastolic murmur in older children Prominent pulses
Aortic coarctation	Low volume or delayed femoral pulses Hypertension

Table 1

or rheumatic valvular disease. Some of the associated clinical features, such as ejection clicks and wide splitting of the second heart sound, may be difficult for the non-specialist to identify or exclude, and their absence does not exclude pathology.

### Innocent murmurs

Four types of innocent murmur are generally recognised in children.<sup>13</sup> Table 2 lists these types and summarises their characteristic features. McCrindle's review offers a comprehensive account of the clinical features of innocent murmurs for interested readers.

### Clinical evaluation and investigation

With the exception of the venous hum, innocent murmurs are exclusively systolic in timing, with an ejection quality such that the heart sounds are heard easily. They are typically of low to medium intensity. Some innocent murmurs vary in intensity with respiration, posture and physical manoeuvres such as Valsalva or compression.

Obviously, the presence of any cardiovascular symptoms or signs points to the likelihood of a murmur being pathological. Other clinical features suggesting pathology include high intensity, harsh quality, and diastolic or pansystolic timing. Some of these characteristics, such as intensity and quality, are subjective, and evaluation of their significance depends on the experience and skill of the clinician. Also, many heart lesions, such as ASD, mild PS and a small PDA, may be associated with a low-intensity, ejection systolic murmur and no other clinical signs. How can the clinician distinguish such lesions from an innocent murmur?

In the pre-echocardiography era, paediatricians frequently relied on chest x-ray and electrocardiography in this situation. Extensive evidence, however, suggests that these investigations are unhelpful in distinguishing innocent from pathological murmurs.<sup>14,15</sup> They may, indeed be misleading and yield false-positive

### Common innocent murmurs in children

Innocent murmurs	Characteristic features
Carotid bruit	Ejection systolic Heard best above clavicles and over upper precordium
Venous hum	Heard best above clavicles Systolic and diastolic components Accentuated by sitting forward Diminished by pressure, supine posture
Still's murmur	Vibratory ejection systolic Localised to left sternal edge Accentuated by fever, activity
Pulmonary outflow murmur	Ejection systolic Heard best at upper left sternal edge Peak incidence in late childhood

Table 2

results. Right bundle branch block, for example, which is commonly seen on the electrocardiograph of healthy children, may be interpreted as suggesting an ASD.<sup>14</sup> Also, non-specialists may misinterpret chest x-ray findings as suggesting increased pulmonary vascularity or cardiomegaly.<sup>14</sup>

If further investigation is considered necessary, therefore, echocardiography is the investigation of choice. At present in the UK, referral for this entails referral to a paediatric cardiologist. Indications for such referral include the presence of clinical features suggesting pathology as outlined above, parental anxiety, and lack of confidence on the part of the clinician that a murmur is benign.<sup>16</sup> Other indications, which may overlap with the latter two, include the finding of an apparently new murmur in an older child, a family history of heart disease such as cardiomyopathy, or the finding of a murmur in a child undertaking strenuous physical activity. Some of these indications are not clearly defined, and much depends on the clinician's and the family's tolerance of uncertainty. Echocardiography should not, however, be assumed to be a panacea, a reassurance tool.<sup>12</sup> It may not alleviate disproportionate parental anxiety, which should be investigated and managed holistically. The authors have anecdotal experience of several children who were referred repeatedly from primary care with parental 'cardiac neurosis' that was not alleviated by normal findings on echocardiography. Also, incidental findings may engender even further anxiety, in particular the finding of a persistent PFO. There is increasing concern that this may be associated with an increased risk of stroke in adult life.<sup>17</sup> Its identification in young infants now often generates the need for repeat scanning to confirm closure. What if it is identified in an older child who is being scanned largely to reassure the family or the referring clinician? Surely the cardiologist must inform the child or family, and give some account of its possible significance? Clearly, avoiding referral in the first place does not solve this problem, but the point is that liberal use of echocardiography for reassurance may in some cases be counterproductive.

If the clinician is confident, based on clinical assessment, that a murmur is innocent, he or she should say so, explain the grounds for this opinion to the parents and discharge the child. With the availability of echocardiography, there is now no place for the old practice of bringing such children back for indefinite clinical review and advising antibiotic prophylaxis and other inappropriate constraints. If the clinician cannot be so confident, the child should be referred.

### Conclusions and future developments

Paediatric cardiology and echocardiography are finite resources that at present cannot extend to the evaluation of all infants and children with asymptomatic murmurs. The responsibility of the paediatrician is to decide which infants and children need referral, and which can be managed safely and appropriately in secondary care. Neonates with murmurs and other symptoms or signs, or those in whom the murmur sounds pathological, should be referred urgently. Neonates with no such accompanying features may be managed locally provided carers are given comprehensive relevant information and advice, and provided the infant is reviewed regularly by senior clinicians during the first few weeks of life. If the murmur persists beyond this time, or if other symptoms or signs develop, they should be referred.

Older infants and children with asymptomatic murmurs can be managed at greater leisure. If the clinician and parents are happy with the clinical diagnosis of an innocent murmur, no further investigation or follow-up are needed. This practice is necessary in order to 'gate-keep' paediatric cardiology resources, but it cannot guarantee the exclusion of a minor asymptomatic heart lesion.<sup>18</sup> If the clinician or parents are not confident that a murmur is innocent, the child should be referred. Again, a normal echocardiogram does not always provide full reassurance and incidental findings, such as a PFO, may engender further anxiety.

If further evaluation of screening pulse oximetry in the newborn confirms its promise, its introduction may complement the practice of careful clinical management of young infants with clinically innocent murmurs. This practice demands a clear pathway for such management, including audit of the unit's experience. The increasing introduction of paediatricians with a special interest in cardiology will hopefully support safe local management of children with innocent murmurs or minor heart disease, and enhance efficient utilisation of tertiary paediatric cardiology services.<sup>19</sup> ◆

### REFERENCES

- 1 Richmond S, Wren C. Early diagnosis of congenital heart disease. *Semin Neonatol* 2001; **6**: 27–35.
- 2 Ainsworth SB, Wyllie JP, Wren C. Prevalence and clinical significance of heart murmurs in neonates. *Arch Dis Child Fetal Neonatal Ed* 1999; **80**: F43–F45.
- 3 Farrer KF, Rennie JM. Neonatal murmurs: are senior house officers good enough? *Arch Dis Child Fetal Neonatal Ed* 2003; **88**: F147–F151.
- 4 Patton C, Hey E. How effectively can clinical examination pick up congenital heart disease at birth? *Arch Dis Child Fetal Neonatal Ed* 2006; **91**: F263–F267.
- 5 Du Z-D, Roguin N, Barak M. Clinical and echocardiographic evaluation of neonates with heart murmurs. *Acta Paediatr* 1997; **86**: 752–756.
- 6 Mellander M, Sunnegardh J. Failure to diagnose critical heart malformations in newborns before discharge – an increasing problem? *Acta Paediatr* 2006; **95**: 407–413.
- 7 Wren C, Reinhardt Z, Khawaja K. Twenty-year trends in diagnosis of life-threatening neonatal cardiovascular malformations. *Arch Dis Child Fetal Neonatal Ed* 2008; **93**: F33–F35.
- 8 Arlettaz R, Archer N, Wilkinson AR. Natural history of innocent heart murmurs in newborn babies: controlled echocardiographic study. *Arch Dis Child Fetal Neonatal Ed* 1998; **78**: F166–F170.
- 9 Crossland DS, Furness JC, Abu-Harb M, et al. Variability of four limb blood pressure in normal neonates. *Arch Dis Child Fetal Neonatal Ed* 2004; **89**: F325–F327.
- 10 Thangaratinam S, Daniels J, Ewer AK, et al. Accuracy of pulse oximetry in screening for congenital heart disease in asymptomatic newborns: a systematic review. *Arch Dis Child Fetal Neonatal Ed* 2007; **92**: F176–F180.
- 11 Granelli A, Mellander M, Sunnegardh J, et al. Screening for duct-dependent congenital heart disease with pulse oximetry: a critical evaluation of strategies to maximise sensitivity. *Acta Paediatr* 2005; **94**: 1590–1596.

- 12** McCrindle BW, Shaffer KM, Kan JS, et al. An evaluation of parental concerns and misconceptions about heart murmurs. *Clin Pediatr (Phila)* 1995; **34**: 25–31.
- 13** McCrindle BW. The outpatient evaluation of heart murmurs. *Balliere's Clinical Paediatrics* 1996; **4**: 1–15.
- 14** Birkebaek NH, Hansen LK, Oxhoj H. Diagnostic value of chest radiography and electrocardiography in the evaluation of asymptomatic children with a cardiac murmur. *Acta Paediatr* 1995; **84**: 1379–1381.
- 15** Rajakumar K, Weisse M, Rosas A, et al. Comparative study of clinical evaluation of heart murmurs by general pediatricians and pediatric cardiologists. *Clin Pediatr (Phila)* 1999; **38**: 511–518.
- 16** McCrindle BW, Shaffer KM, Kan JS, et al. Factors prompting referral for cardiology evaluation of heart murmurs in children. *Arch Ped Adolesc Med* 1995; **149**: 1277–1279.
- 17** Kenny D, Turner M, Martin R. When to close a patent foramen ovale. *Arch Dis Child* 2008; **93**: 255–259.
- 18** Danford DA, Martin AB, Fletcher SE, Gumbiner CH. Echocardiographic yield in children when innocent murmur seems likely but doubts linger. *Pediatr Cardiol* 2002; **23**: 410–414.
- 19** Katumba-Lunanya JL. Neonatal/infant echocardiography by the non-cardiologist: a personal practice, past, present and future. *Arch Dis Child Fetal Neonatal Ed* 2002; **86**: F55–F57.

### Practice points

- Neonates with significant-sounding heart murmurs, symptoms or associated congenital abnormalities should be referred urgently for cardiological assessment
- Neonates with innocent-sounding murmurs and no symptoms should be reviewed regularly during the first few weeks of life
- Older infants and children should be referred routinely to a cardiologist if the paediatrician or parents are not confident that the murmur is innocent
- Echocardiography may aggravate parental anxiety if it identifies incidental, possibly significant, findings such as persistent patent foramen ovale