

Anaesthetist's evaluation of a child with a heart murmur

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Abstract

Although between 50% and 85% of children may have a heart murmur, most heart murmurs are innocent. Murmurs can occur because of children's high cardiac output and small vessels, branching at acute angles, which promote turbulent flow. Innocent murmurs can be described according to seven characteristics, but not all murmurs adhere to these rules. There are seven different types of innocent murmurs. An innocent murmur can often be diagnosed by taking a history and conducting an examination. An electrocardiogram is helpful in assessing left ventricular mass if aortic stenosis (AS) or hypertrophic cardiomyopathy (HOCM) is anticipated. If uncertainty exists, the differential diagnosis should guide decision-making. If a high-risk lesion, such as an AS or HOCM is anticipated, it should be assessed first, while a more benign lesion, such as an atrial septal defect, could be evaluated by a cardiologist after surgery. The type of surgery is also a deciding factor. Any child under one year of age has a much higher risk of having pathology which may be severe, and will need to be referred before surgery. Endocarditis prophylaxis and the risk of air emboli should always be considered.

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Introduction

Between 50% and 85% of children can have a heart murmur at some stage.^{1,2} As the incidence of congenital heart disease is only 0.8%,³ most of these murmurs are likely to be innocent. Therefore, the anaesthetist is often required to evaluate a patient with a murmur and has to decide whether to defer or proceed with surgery.

Physiology

Murmurs result from turbulent flow.⁴ Usually, blood flow is laminar. Turbulent flow is more likely if Reynold's number is over 3 000:

$$Re = \rho DV / \eta$$

(ρ = density, D = diameter, V = flow velocity, η = viscosity).⁵

This explains why anaemic patients (low η) develop flow murmurs. High-flow velocity often causes turbulent flow. The flow acceleration of blood passing through a constriction, e.g. stenosis, causes turbulence, resulting in a murmur.

Children have a high cardiac output (V), and small vessels (D) which branch at acute angles, promoting turbulent flow. Sound intensity diminishes by the square of the distance as it travels away from its source.² Since in children there is a short distance from their skin to the heart and vessels, it is more likely that a clinician will hear a murmur. Typically, innocent murmurs will change with alterations in flow, because of respiration or altered position.

Evaluation

Innocent murmurs are described according to seven characteristics¹ (Table I).

Table I: Characteristics of innocent murmurs

- Systolic
- Soft (< grade 3)
- Short duration (not pansystolic)
- Sweet
- Small (a limited area and not radiating)
- Sensitive (changes with position or respiration)
- Single (no clicks or gallops)

Not all innocent murmurs adhere to these rules, and adolescents may have an innocent isolated S_3 .⁶ The S_3 may disappear when standing upright. Certain red flags (Table II) increase the likelihood of underlying pathology.

Table II: Pathological features of a heart murmur

- Pansystolic murmur.
- Diastolic murmur.
- Maximal over pulmonary area (*upper left sternal border*).
- Grade 3 or higher.
- Harsh quality.
- Changes typical of hypertrophic cardiomyopathy, where systolic anterior motion of the mitral valve (systolic anterior motion) increases in the following situations: increased intensity on standing, increased intensity with Valsalva, a decrease or no change on passive leg raise and a decrease or no change when changing from standing to squatting.
- Abnormal S_2 .
- Ejection systolic click.
- Active precordium.

Assessment of a murmur starts with a routine evaluation (taking a history and conducting an examination). Special examinations, such as a chest X-ray and an electrocardiogram (ECG), do not add much value,¹ but if aortic stenosis (AS) or HOCM need to be eliminated, assessment of the left ventricular muscle mass on ECG is important.

History

A family history of cardiac disease, sudden death or sudden infant death should be sought. If a first-degree relative has congenital heart disease (CHD), the risk of cardiac disease increases 3-10 times.¹ HOCM is an autosomal dominant trait and ventriculoseptal defects (VSD) and mitral valve prolapse have a high degree of penetrance.

Important in the pre- and perinatal history are exposure to toxins (alcohol and lithium), infections (rubella), maternal diabetes and prematurity [patent ductus arteriosus (PDA)]. Associated syndromes, chromosomal disorders, connective tissue disorders and metabolic disorders should be excluded.

A history of the following cardiac symptoms is relevant (Table III).

Table III: Symptoms of cardiac disease

- *Constitutional symptoms:* Constitutional symptoms include poor feeding, fatigue, diaphoresis, exercise intolerance, failure to thrive and developmental delay (syndromes).
- *Cardiovascular symptoms:* Cardiovascular symptoms include cyanosis, chest pain (aortic stenosis and hypertrophic cardiomyopathy), dizziness, palpitations and near-syncope, and syncope (aortic stenosis and hypertrophic cardiomyopathy).
- *Respiratory symptoms:* Respiratory symptoms include frequent infections, "asthma", a chronic cough and exertional dyspnoea.

Babies develop puffy eyes (rather than swollen feet), especially after sleeping. Patients who present to the emergency room with acute heart failure generally experience dyspnoea, nausea and vomiting, fatigue and coughing.

It is not often that chest pain and syncope are symptoms of cardiac disease in children.⁶ Chest pain is common, but less than 1% of children with chest pain have cardiac disease. Up to 15% of children experience syncope before age 21, but most of them have no cardiac pathology. Although rarely pathological, these symptoms may be associated with life-threatening conditions (HOCM and AS), and patients should be evaluated carefully.

Examination

Up to 25% of patients with abnormalities of other organ systems may have cardiac defects.¹ Defective segmentation of the sternum may be associated with cardiac conditions and situs inversus can reveal more heart defects.

A crying child cannot be assessed properly so the heart should be examined early, and in particular, before blood is taken. Similarly, initially it is important to try to focus on auscultation of the heart sounds and extra clicks, and to ignore the murmur, and then to listen to the murmur specifically.

An active precordium may indicate an AS, moderate to large VSD, or a significant PDA.⁷ Blood pressure should be measured in all four limbs to exclude coarctation. Hepatomegaly is a useful sign since children tend to develop biventricular failure.⁸

Types of innocent murmurs

Various innocent murmurs exist, of which the most common was described by George Still in 1909.²

Still's

Still's murmur is a musical, vibratory, low-pitched murmur (like a tuning fork) which is heard at the left lower sternal border, which may extend to the apex. It is early systolic, often gets softer when standing, and typically occurs between the ages of 2 and 6 years. The murmur may originate from relative left ventricular outflow tract narrowing, fibromuscular bands in the left ventricle, or from the right ventricular outflow tract.

Aortic systolic

Aortic systolic murmur is an ejection systolic murmur, heard at the upper right sternal border, and is more common in adolescents and adults. HOCM must be excluded and referral may be warranted.²

Pulmonary flow

Authors do not always differentiate pulmonary flow murmur from the peripheral pulmonary arterial stenosis murmur (see below). The pulmonary flow murmur is heard at the upper left sternal border (second or third intercostal space), but may also be heard over the apex or the lower neck.⁴ It peaks later (mid-systole), is harsh and becomes softer when the patient stands. Typically, it occurs during adolescence and in patients with pectus excavatum,⁶ kyphoscoliosis or in those with a straight back.² Echocardiography⁴ has shown that it probably originates from a relatively big stroke volume over a relatively small left ventricular outflow tract.

Pulmonary stenosis (PS) is differentiated by an ejection click, thrill, a higher pitch and longer duration, and a widely split S_2 . The pulmonary flow murmur of an AS is associated with a fixed split S_2 , active precordium and a diastolic flow rumble.²

Peripheral pulmonary arterial stenosis

Peripheral pulmonary arterial stenosis is heard in the axillae and back, is soft (grade 1-2), low-pitched and early to mid-systolic. It occurs in infants because of the relative discrepancy in the size of the main and branch pulmonary arteries. During foetal life, most of the cardiac output is diverted via the duct, leaving little flow in the small pulmonary arteries. The acute angle at which the branch pulmonary arteries come off increases the chance of turbulent flow.⁶ It may be impossible to distinguish this murmur from that of pathological peripheral PS. Pulmonary valvular or arterial stenosis and an AS must be excluded.

Supraclavicular systolic

Supraclavicular systolic murmur is a brief, low-pitched, early systolic, crescendo-decrescendo murmur, heard above the clavicles of children and young adults. It may also be heard in the upper chest and can extend up into the neck. It diminishes with sudden hyperextension of the shoulders. The murmur probably originates from the

brachiocephalic vessels as they come off the aorta,² and must be differentiated from AS, PS and coarctation.¹

Innocent murmurs that are heard during diastole have a systolic component and can be regarded as continuous murmurs.

Venous hum

Venous hum occurs between 3 and 8 years of age. It is best heard over the lower neck, lateral to the sternocleidomastoid, and resolves when lying down or turning the head away.¹ It is grade 1-6, has a whirring quality and is continuous, but accentuated during diastole. Arteriovenous malformation (AVM) and PDA are pathological continuous murmurs which should be excluded.

Mammary soufflé

Mammary soufflé is heard during late pregnancy or lactation and sometimes during adolescence. It is caused by blood flow in the mammary vessels and is heard over the breast. There is a distinct gap between S_1 and the onset of the murmur because of delayed arrival of blood flow, and it may extend into diastole. Again, PDA or AVM must be excluded.²

Conclusion

If a child with an unexpected murmur presents for anaesthesia, the following must be considered:⁷

- How the underlying pathology increases the risk of an anaesthetic and surgery. If the differential diagnosis includes HOCM or AS, anaesthesia could pose a much higher risk than if a small VSD is suspected.
- Whether endocarditis prophylaxis is necessary.
- The risk of paradoxical embolism.
- What the haemodynamic aims of anaesthesia for each specific lesion would be to optimise cardiorespiratory function, and which complications would need to be avoided, e.g. pulmonary oedema, cyanosis, systolic anterior motion, increased shunt or decreased cardiac output.

Table IV: Features of pathological murmurs

Lesion	Location	Qualities
Ventriculoseptal defect	Left lower sternal border	Pansystolic; bigger lesion is softer murmur, split S_2
Atrioseptal defect	Upper left sternal border	Increased pulmonary flow: ejection systolic; fixed split S_2
Patent ductus arteriosus	Upper left sternal border	Continuous, crescendo-decrescendo; may conceal S_2
Tetralogy of Fallot	Upper left sternal border	Outflow tract murmur, ejection systolic; cyanosis
Pulmonary stenosis	Upper left sternal border	Ejection systolic; radiates to axillae, back, infraclavicular area, click
Coarctation	Interscapular	Ejection systolic; radio-femoral delay, differential blood pressure
Aortic stenosis	Upper right sternal border	Ejection systolic; thrill, radiates to carotids

The following is suggested:

- If the anaesthetist is convinced that the murmur is innocent, surgery should proceed. An innocent murmur is likely if the typical features of an innocent murmur are present, the rest of the physical examination is normal, the child is asymptomatic and nothing in the history indicates an increased risk of heart disease.¹
- Children who are younger than one year of age may be asymptomatic, despite having severe abnormality. They should be referred. Newborn babies have a 1% incidence of heart murmurs, but 31-86% of newborns with a murmur have a heart defect.¹
- Any findings that are consistent with cardiac conditions with high anaesthetic risk should prompt referral. These include signs of HOCM or AS, left ventricular hypertrophy on ECG, cardiomyopathy, the presence of extracardiac abnormalities, and complications of CHD such as cyanosis, cardiac failure, arrhythmias and pulmonary hypertension.³
- If there is uncertainty about the murmur, but the child is asymptomatic, it is likely to be an uncomplicated lesion (AS, small VSD, PDA or mild PS),⁷ and it is reasonable

to proceed with surgery, depending on the nature of the procedure. Referral to a cardiologist should be performed later. The need for antibiotic prophylaxis according to guidelines should be considered and meticulous care given to ensuring that air embolism is avoided.

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