Should I do this case? – the paediatric murmur

How do you deal with anaesthesia in a child with a murmur?

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Johan Diedericks has a clinical interest in paediatric anaesthesia, particularly paediatric cardiothoracic anaesthesia, and a research interest in cardiac function in anaesthesia.

In a review on anaesthesia-related cardiac arrest in children, Linda Mason states that murmurs should be characterised before anaesthesia, especially in infants.[1] It is accepted that auscultation skills are on the wane as new diagnostic modalities emerge.[2] However, it is impossible to refer every murmur heard for special investigation, since about 50 - 72% of paediatric murmurs are normal or innocent.[3,4] With a wide description of incidence of innocent murmurs (depending on the experience of the examiner and examining conditions) and a very low incidence (0.5%) of cardiac disease in children, it is difficult to confidently make a decision.[5] Some important considerations are given in Table I.

Table I. Considerations of a murmur discovered at preoperative visit

- Is it dangerous? May it cause haemodynamic instability intraoperatively?
- In some lesions (e.g. ASD or VSD) paradoxical embolism and a reversion to a fetal-type circulation may occur during anaesthesia (transitional circulation)*
- Is antibiotic prophylaxis indicated; what drug and what dose?
- Should the case be postponed and the murmur investigated (expensive)?
- Should surgery continue with later follow up?
- Could it be ignored?
- Patient and parent information and reassurance and continuity of follow-up

*Hypoxia, hypercarbia, acidosis, hypotension, hypothermia, painful stimulation are precipitants. Pulmonary hypertension increases this risk.
ASD = atrial septal defect.
VSD = ventricular septal defect.

Innocent and pathological murmurs

The characteristics of innocent and pathological murmurs are given in Tables II and III. The difficulty is that there is overlap in the findings with an innocent or pathological murmur. Coleman *et al.*[6] studied a cohort of 444 children with innocent murmurs (21% of the total number investigated). Innocent murmurs were defined as those murmurs with no cardiac lesion (83%), or with a minor lesion (17%). The minor lesions were atrial septal defect (ASD), small ventricular septal defect (VSD), mild pulmonary stenosis or regurgitation, and patent ductus arteriosus (PDA). Coleman found that a clinical diagnosis of VSD was as often disproved as confirmed and that the clinical diagnosis of ASD was seldom confirmed. Mild pulmonary stenosis was rarely clinically diagnosed but frequently discovered on cardiac catheterisation. Two of their patients developed bacterial endocarditis (with a small VSD and insignificant pulmonary stenosis). It is therefore clear that although

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Table II. Characteristics of innocent murmurs[5-7]

- Early systolic or continuous
- Soft (3/6 or less)
- Crescendo-decrescendo
- Examples:
  - Still's murmur – most common, 'musical or vibratory, groaning, squeaking, creaking, rasping', 1 - 3/6, no thrill, inside apex or low parasternal, but may be over whole precordium, softer or disappear on standing, reappears on squatting
  - Pulmonary ejection murmur – systolic, high-pitched 'blowing', second left interspace, may be heard at the apex, left sternal border, aortic area and neck, normal split-second heart sound during inspiration (not expiration)
  - Supraclavicular arterial bruit (rare)
  - Late systolic cardiorespiratory murmur (rare)
  - Continuous murmur (venous hum) – systolic and diastolic hum, loudest in sitting during inspiration, disappears or diminishes in the supine position or with pressure over the supraclavicular area due to reduced jugular venous flow

lesions may be haemodynamically insignificant, there are other consequences. The intensity of small VSD murmurs may vary from 2/6 to 6/6 and can be early systolic rather than pansystolic.[8]

Any murmur detected preoperatively should be evaluated to determine whether it is innocuous. In the vast majority of cases this should be determined by clinical means (although it is not foolproof). This evaluation can be done by a general practitioner or a specialist anaesthetist. Referrals could be to a paediatrician or a paediatric cardiologist.

Clinical evaluation of murmurs in children[7,9]

The clinical evaluation should start with historical features that may suggest that the murmur is pathological. For example prematurity,

Table III. Characteristics of pathological murmurs[5-7]

- Diastolic, pansystolic or late systolic
- Usually loud (3/6 or more)
- Associated with a thrill
- Symptoms or signs of cardiac disease
- Continuous
- S_1 inaudible or not single
- Most do not change significantly on standing (the systolic murmur of the rare, but dangerous, hypertophic obstructive cardiomyopathy increases on standing)[7]
- Examples:
  All murmurs caused by cardiac lesions. Some people classify ASD, small VSD, mild pulmonary stenosis or regurgitation, and PDA as innocent murmurs. Although these lesions may be haemodynamically innocent during anaesthesia, children may develop bacterial endocarditis[6]

Table IV. Questions to determine the clinical effect of a murmur[9]

Children

- Does he/she run? Like peers?
- Is he/she calmer or slower than peers?

Cyanosis

- Does he/she turn blue? During feeding/when crying?
- Does he/she lose consciousness?
- Does he/she stop playing and squat?

Infant

- Is feeding prolonged?
- Does he/she sweat during normal care?
- Does he/she have swollen eyes in the morning?

other congenital malformations, feeding intolerance, failure to thrive, respiratory symptoms, particularly repeated infections, cyanosis, chest pain, syncope, or a family history of sudden death are all worrying. Table IV shows the questions to ask. Examination should be in a quiet room, the patient preferably lying down. Sitting in the mother's lap can help pacify the child.

If palpation indicates increased precordial activity, an ASD, moderate to large VSD or significant PDA may be present. A thrill may be felt at the lower left sternal border (VSD), left upper sternal border (pulmonary valve stenosis) or suprasternal notch (aortic stenosis). Both brachial and femoral pulses bilaterally should be equal in timing and intensity and blood pressure in the right arm normal, to exclude aortic coarctation.

The first heart sound (S_1) is normally a single sound caused by closure of the mitral (MV) and tricuspid (TV) valves. If S_1 is inaudible, some other sound is obscuring it (think VSD, AV regurgitation, PDA, severe pulmonary stenosis). The murmurs that cause this effect are often called holosystolic. If S_1 appears split it is either caused by a click or by asynchronous closure of the MV and TV. Pulmonary valve (PV) ejection clicks begin shortly after AV valve closure, vary with respiration and are best heard at the upper left sternal border. Aortic valve clicks begin shortly after S_1 and are loudest at the apex. Mitral valve clicks are best heard at the apex when standing.

The second heart sound (S_2) is caused by closure of the aortic and pulmonary valve, and has two components, the aortic second sound (A_2) and the pulmonary second sound (P_2). The sound splits during inspiration as more blood is drawn into the right ventricle and subsequently the PV closes later. A loud, single S_2 indicates pulmonary hypertension (with RV overload) or congenital heart disease involving the semilunar valves. Murmurs are graded 1 - 6/6, and are timed as early, middle or late systolic. The 'character' of the murmur may help with diagnosis. A 'harsh' murmur occurs when blood flows at high velocity from a high- to a low-pressure chamber. Examples are VSD and semilunar valve stenosis. 'Whooping' or 'blowing' sounds occur at the apex with mitral regurgitation, and a crescendo-decrescendo 'flow murmur' describes the innocuous functional murmur. However, similar murmurs may also be heard with ASD, mild semilunar valve stenosis, subaortic obstruction, aortic coarctation, and very large VSD.

Most pathological murmurs do not change in intensity during position changes, the most important exception being the murmur of hypertrophic cardiomyopathy (HOCM).

Most pathological murmurs do not change in intensity during position changes, the most important exception being the murmur of hypertrophic cardiomyopathy (HOCM) (Table III). The venous hum is heard all over the anterior chest and is present if the child is upright and disappears when he lies down or with pressure over the jugular vein. The position of highest intensity of murmurs varies with lesions (Table V).

Table V. Areas of highest intensity for common paediatric murmurs[7]

Area	Murmur
Upper right sternal border	Aortic stenosis, venous hum
Upper left sternal border	Pulmonary stenosis, pulmonary flow murmurs, ASD, PDA
Lower left sternal border	Still's murmur, VSD, tricuspid regurgitation, HOCM, subaortic stenosis
Apex	Mitral regurgitation

Every murmur must be thoroughly evaluated clinically and if there is uncertainty about the innocuous nature of the murmurs according to the suggested algorithm, the surgery should be postponed.

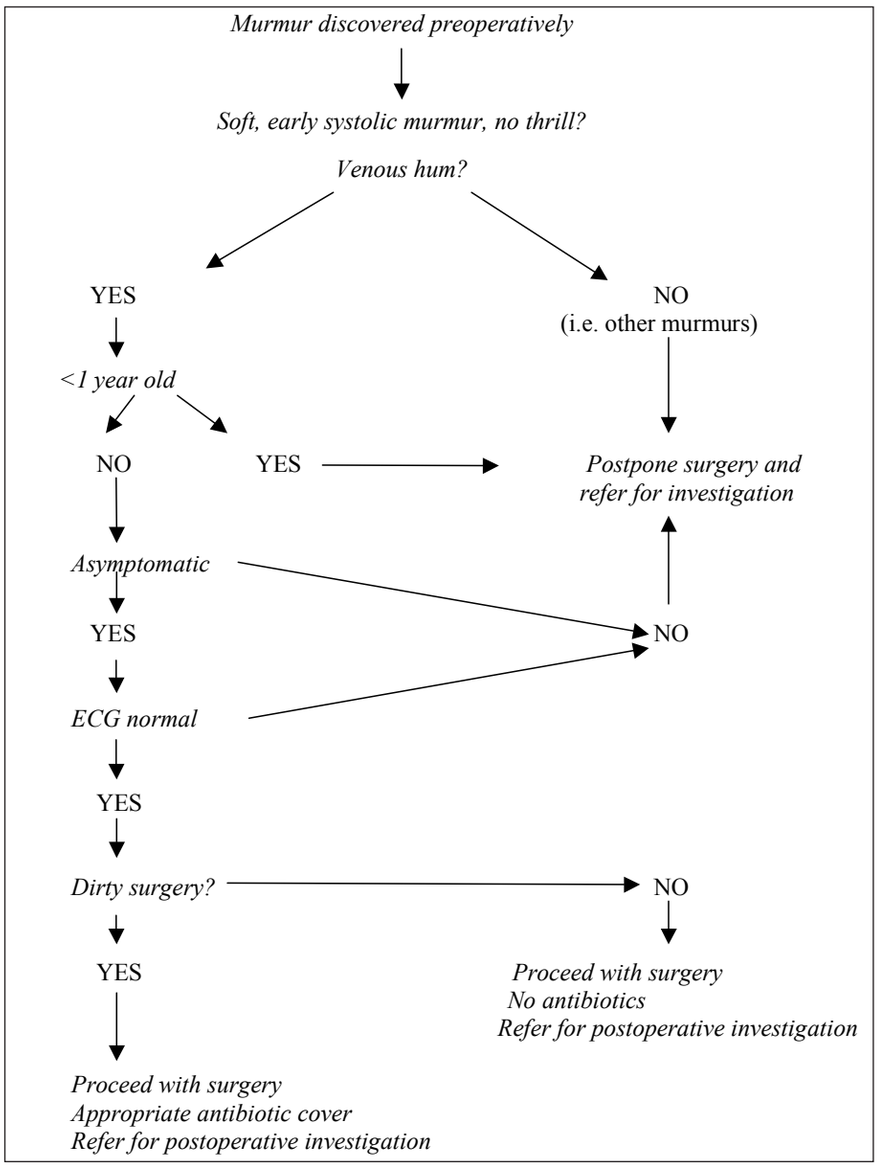

Fig. 1. An assessment algorithm.

Table VI. Differentiation between an ASD and innocent murmur [7]

Physical sign	Innocent murmur	ASD
Precordial activity	Normal	Increased
S_1	Normal	Normal
S_2	Splits, moves with respiration	Fixed widely split
Systolic murmur (supine)	Crescendo-decrescendo Possible vibration lower left sternal border	Crescendo-decrescendo 'Flow' at upper sternal border
Systolic murmur (standing)	Decrease in intensity	Does not change
Diastolic murmur	Venous hum	Inflow 'rumble' across tricuspid valve area

The ASD is most frequently incorrectly diagnosed. A differentiation between a functional murmur and ASD is given in Table VI.

Assessment algorithm

Which children with murmurs can then be anaesthetised, and which would have to be further investigated? McEwan *et al.* suggested a logical sequence that it is useful to follow.[5] A modified version of this is given in Fig. 1. Most murmurs will be innocent and one can proceed to anaesthesia. Without corroborating history, the most dangerous lesions that must be excluded are HOCM and aortic stenosis. Both lesions may be asymptomatic with only the murmur, but may cause fatal haemodynamic derangement during anaesthesia. In both conditions the ECG usually shows left ventricular hypertrophy and left axis deviation. Be wary of any child with an R wave in V5 or V6 that is greater than 40mV. Refer such ECGs for evaluation. Other dangerous lesions such as significant pulmonary stenosis, tetralogy of Fallot or coarctation of the aorta will usually have typical symptoms, signs and ECG abnormalities.[5]

Although 'insignificant' lesions such as ASD, small VSD or pulmonary stenosis may be of no haemodynamic importance, there is the possibility of endocarditis. Surgery that may cause a bacterial surge must be covered by antibiotics (dentectomy, oral surgery, upper respiratory tract surgery, genitourinary instrumentation or surgery, and gastrointestinal procedures). The perioperative guidelines for antimicrobial therapy were recently upgraded and a summary is shown in Table VII.[10] The risk of dental procedures has been shown to be smaller than previously thought, and prophylaxis is only necessary when there is manipulation of gingival tissue or the periapical region of teeth or perforation of the oral mucosa. Prophylaxis is also only necessary in certain 'at-risk' cardiac conditions (Table VII).[10]

In summary

Most children with heart murmurs will have an insignificant murmur that is unlikely to cause haemodynamic problems during anaesthesia. However, every murmur must be thoroughly evaluated clinically and if there is uncertainty about the innocuous nature of the murmurs according to the suggested algorithm, the surgery should be postponed. Otherwise anaesthesia can be administered with appropriate antibiotic prophylaxis. The child should be referred postoperatively for evaluation and follow-up since some seemingly insignificant murmurs may over years be unmasked as specific pathology.

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Table VII. Cardiac lesions at risk of endocarditis and the surgical procedures that require prophylaxis. Regimens according to the American Heart Association[9,10]

At-risk cardiac lesions	Operative procedures requiring prophylaxis	Bacterial prophylaxis suggested for children

• Prosthetic valves
• Previous infective endocarditis
• Congenital heart disease (CHD):
  • Unrepaired cyanotic CHD, including palliative shunts and conduits
  • Repaired congenital heart defects with prosthetic material within 6 months of surgery (not yet epithelialised)
  • Repaired CHD with residual defects
• Cardiac transplants with valvular disease
	Prophylaxis for dental procedures that involve:
	• Manipulation of gingival tissue
	• Manipulation of the periapical region of teeth
	• Perforation of the oral mucosa
	No prophylaxis for:
	• Injections of local anaesthesia in non-infected tissue
	• Dental radiographs
	• Placement or adjustment of removable prosthodontic or orthodontic appliances
	• Placement of orthodontic brackets
	• Shedding of deciduous teeth
	• Bleeding from trauma to lips or oral mucosa
		Single dose 30 - 60 minutes before procedure:
		Oral or unable to take oral medication:
		• Amoxillin 50 mg/kg po
		• Ampicillin 50 mg/kg or cefazolin or ceftriaxone IV or IM
		Allergic to penicillins or ampicillin – oral:
		• Cephalexin 50 mg/kg PO or
		• Clindamycin 20 mg/kg PO or
		• Azithromycin or clarithromycin 15 mg/kg PO
		Allergic as above, unable to take oral medication:
		• Cefazolin or ceftriaxone 50 mg/kg IM or IV
		• Clindamycin 20 mg/kg IM or IV

	Respiratory tract-invasive procedures that incise the mucosa: • Tonsillectomy • Adenoidectomy • Bronchoscopic biopsy (but not for bronchoscopy that does not break mucosa) • Procedures to treat established infection and drainage of abscess or empyema	As above Choose agent active against viridans group of streptococci. Consider vancomycin with beta lactam-sensitive patients or methicillin resistance
	Gastrointestinal and genitourinal tract (GIT & GUT): • No prophylaxis for procedures • Only prophylaxis when there is established GIT or GUT infection	As above
	Infected skin, skin structure or musculoskeletal tissue	As above

In a nutshell

- Cardiac murmurs in children may have serious haemodynamic implications during anaesthesia and surgery.
- Innocuous and pathological murmurs have characteristics that differ, but there is unfortunately also overlap.
- Most murmurs (>70%) are innocuous and anaesthesia can be safely administered.
- It is essential to investigate any murmur detected preoperatively to exclude dangerous murmurs.
- An appropriate history and thorough clinical examination will enable diagnosis of most innocuous murmurs.
- For some cardiac lesions perioperative antimicrobial prophylaxis is necessary for some procedures.
- Patients should be referred for postoperative evaluation and follow-up.
- Patients and parents should be informed of the implications and need for prophylaxis and follow-up.