

**Anaesthesia for non-cardiac surgery in children with congenital heart disease**

Based in part upon: Story E. Recognising cardiac disease in children *Anaesthesia Tutorial of the Week* 93 (2008)<sup>1</sup>

Isabeau Walker

Correspondence Email: [isabeauwalker@mac.com](mailto:isabeauwalker@mac.com)

**SUMMARY**

Children with congenital heart disease provide a challenge to the anaesthetist, but with careful planning, most can be anaesthetised safely. This article covers pathophysiology, recognition, and principles of anaesthesia for children with congenital heart disease, including anaesthesia for specific cardiac lesions.

**INTRODUCTION**

The risk of perioperative complications is higher in children with congenital heart disease (CHD) for both minor and major surgery, particularly in neonates and those with complex lesions.<sup>2</sup> Although CHD is uncommon, it is important for the anaesthetist to understand how to recognise a child with CHD, and the principles of anaesthetic management for children with both unrepaired and repaired lesions.

**INCIDENCE OF CONGENITAL HEART DISEASE**

The incidence of CHD is approximately 8:1000 live births. The frequency of different lesions varies between populations, with ventricular septal defect (VSD) being the most common in all populations (see Table 1).

Most CHD is untreated in low- or middle-income countries (LMIC). Babies with complex cardiac

disease are likely to die due to lack of access to corrective or palliative surgery, in particular those with duct dependent lesions. A child with untreated heart failure due to high pulmonary blood flow is more likely to die from pneumonia, which is the most common cause of mortality in the under 5 age group. If a child with high pulmonary blood flow remains untreated they may develop Eisenmenger's syndrome, which is associated with high mortality in childhood or early adult life.

However, as surgical systems improve, there are an increasing number of programmes to treat children with CHD in LMIC, many developed in partnership with international outreach teams, so that the outlook for children born with simple conditions such as VSD, atrial septal defect (ASD), patent ductus arteriosus (PDA), coarctation of the aorta or Tetralogy of Fallot is much more optimistic.

**Table 1.** The incidence of different types of congenital heart disease in children in the UK

Condition	Incidence
Ventricular septal defect (VSD)	32%
Patent arterial duct (PDA)	12%
Pulmonary stenosis (PS)	8%
Coarctation of the aorta (CoA)	6%
Atrial septal defect (ASD)	6%
Tetralogy of Fallot (TOF)	6%
Aortic stenosis (AS)	5%
Transposition of the great arteries (TGA)	5%
Hypoplastic left heart syndrome (HLHS)	3%
Hypoplastic right heart syndromes	2%
Atrioventricular septal defects (AVSD)	2%
Truncus arteriosus (common arterial trunk)	1%

**Isabeau Walker**

Consultant Paediatric  
Anaesthetist  
Great Ormond Street  
Hospital NHS Foundation  
Trust  
London  
WC1N 3JH

## CAUSE OF CONGENITAL HEART DISEASE AND ASSOCIATED ABNORMALITIES

The cause of most CHD is unknown, but may be related to chromosomal abnormalities, exposure of the mother to teratogens (e.g. alcohol, or drugs such as anti-epileptics or warfarin), or maternal disease (rubella, diabetes).

Recognisable chromosomal abnormalities are present in 25% of children with CHD. The diagnosis of a chromosomal abnormality should lead to assessment of the child for CHD. The most common chromosomal abnormality is Down syndrome (trisomy 21) – 40% of children with Down syndrome have CHD, most commonly atrioventricular septal defect (AVSD) or VSD. Chromosomal abnormalities associated with cardiac lesions are shown in Table 2.

## PATHOPHYSIOLOGY OF CONGENITAL HEART DISEASE

It is useful to classify CHD according to the pathophysiology of the major heart lesion, in particular, whether the child is acyanotic (i.e. the child is 'pink'), or the lesion is associated with cyanosis (i.e. the child is 'blue'). Most CHD is acyanotic. Measuring oxygen saturation by pulse oximetry shortly after birth is a useful screening test to identify cyanotic lesions such as tetralogy of Fallot, transposition of the great arteries (TGA), total anomalous pulmonary venous drainage (TAPVD) or duct dependent lesions such as hypoplastic left heart syndrome. However, the most common cause of cyanosis in the neonatal period is due to a respiratory problem. A description of common lesions is shown in Table 3.

## Left to right shunting lesions: ASD, VSD, AVSD, PDA

The child with acyanotic heart disease presents with signs and symptoms of cardiac failure. The severity of the symptoms depends on the volume of the shunt:

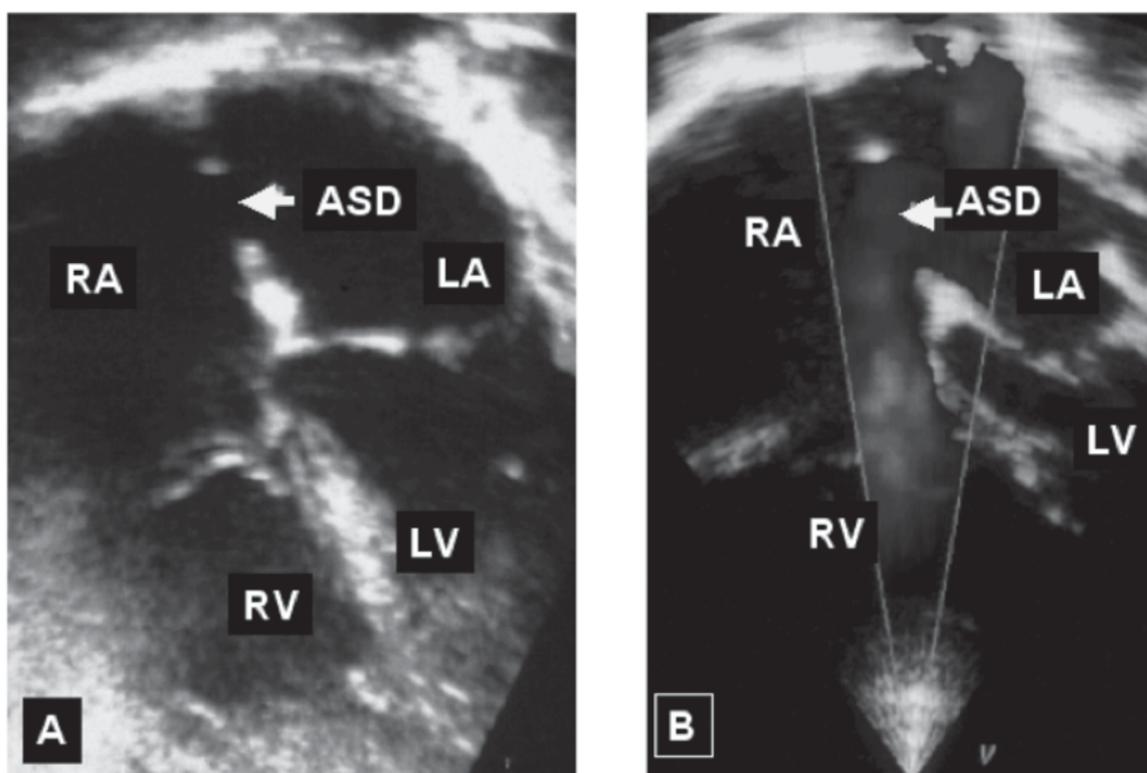
- *Atrial septal defect, ASD.* (See Figure 1). A secundum ASD is associated with a low volume left to right shunt with right heart overload. The child may present with recurrent chest infections, failure to thrive, or may be asymptomatic.
- *Ventricular septal defect, VSD.* A 'restrictive VSD' is small, and causes a low volume shunt with few symptoms. An 'unrestrictive VSD' is large, associated with a high volume left to right shunt and symptoms of severe cardiac failure in infancy
- *Atrioventricular septal defect, AVSD.* (See Figure 2). There is an abnormal atrioventricular valve, an atrial septal defect ('primum ASD'), +/- a ventricular septal defect. This results in a high volume shunt and signs of severe cardiac failure in infancy leading to early pulmonary hypertension. Untreated, children with AVSD develop Eisenmenger's syndrome (see later) by the age of 1-2 years, with 4% survival beyond 5 years.
- *Patient ductus arteriosus, PDA.* (See Figure 3). If the duct is restrictive the child may present with relatively few symptoms initially, or signs and symptoms of cardiac failure. Premature neonates with a large unrestrictive PDA may have severe cardiac failure and be ventilator dependent.

**Table 2.** Chromosomal abnormalities associated with cardiac lesions

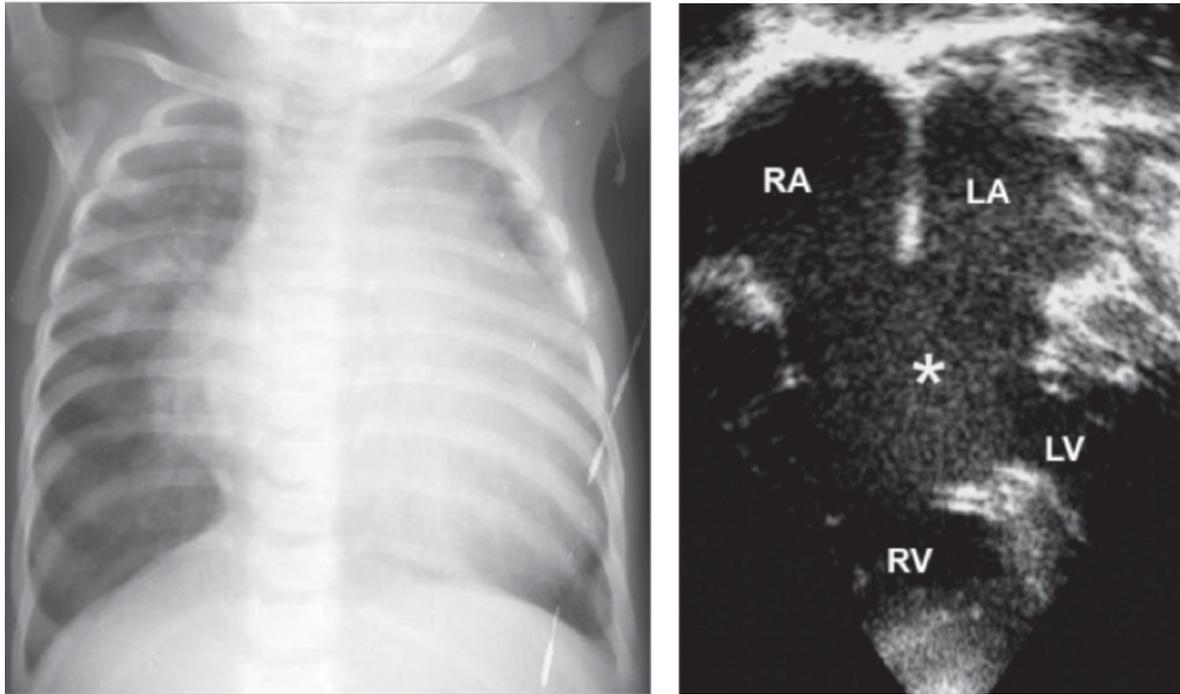
Abnormality	Clinical features and associated conditions	Typical cardiac lesion
Down syndrome Trisomy 21	Typical facial appearance, single palmar crease, short stature, learning difficulties, lax joints (including cervical spine instability), hypothyroidism, obstructive sleep apnoea, leukaemia, duodenal atresia.	AVSD, VSD
DiGeorge syndrome ('Catch 22') 22q11 deletion	Learning difficulties, cleft palate, hypocalcaemia, absent thymus (frequent respiratory infections), typical facial appearance	Aortic arch abnormalities, VSD
Marfan's syndrome	Abnormally tall stature, long fingers, scoliosis, abnormal shaped chest, high arched palate, retinal detachment, inguinal hernia, spontaneous pneumothorax	Aortic root dilatation and dissection
Apert syndrome	Craniosynostosis (premature fusion of cranial sutures), syndactyly (fused fingers), deafness	PS, VSD
Charge association	Abnormal iris (coloboma), choanal atresia (abnormal nasal passageway), developmental delay, abnormal genitalia, ear deformity	Variety, including VSD, AVSD
VATER	Vertebral abnormalities, anal atresia, tracheo-oesophageal fistula, renal abnormalities	VSD, TOF
Goldenhar syndrome	Hemifacial microsomia (poorly developed maxilla/mandible), difficult intubation, ear abnormalities, cleft palate	VSD, TOF

**Table 3.** Classification of common congenital heart lesions

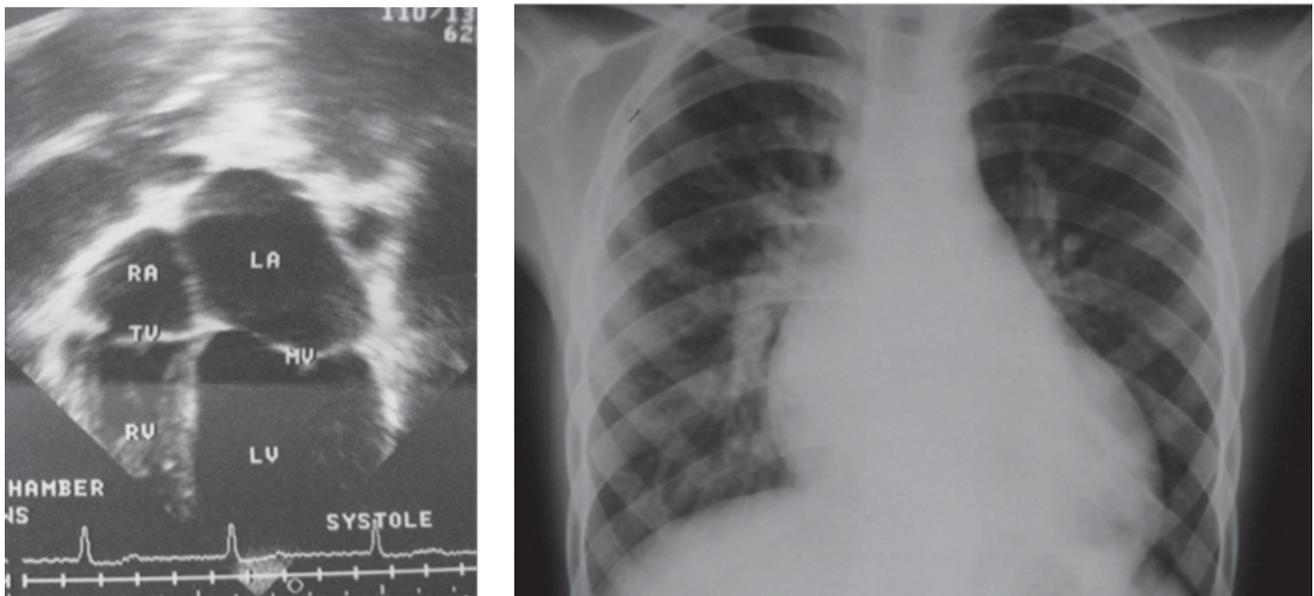
Acyanotic congenital heart disease – ‘pink’ babies		
	Common examples	Comment
<b>Left-to-right shunt</b> ‘Restrictive’ lesions  ‘Non-restrictive’ lesions	Small ASD, VSD, PDA  Large VSD, PDA, AVSD, common arterial trunk	Large (‘non-restrictive’) lesions are associated with severe congestive cardiac failure in infancy.  If unrepaired, may lead to pulmonary hypertension and reversal of shunt (Eisenmenger’s syndrome).
<b>Obstructive lesions</b>	Aortic stenosis, coarctation of the aorta, pulmonary stenosis	Severity of lesion determines age at presentation – neonates with severe obstruction may be critically ill with a duct dependent circulation
Cyanotic lesions – ‘blue’ babies		
<b>Right-to-left shunt</b>	Tetralogy of Fallot: • VSD with aortic override • Right ventricular outflow tract obstruction • Right ventricular hypertrophy,	May present with severe cyanosis and hypercyanotic ‘spells’; or if unrepaired in an older child, with cyanosis, fatigue and a history of ‘squatting’.
<b>Transposition of the Great Arteries (TGA)</b>	TGA may be associated with ASD, VSD, PDA	Long term survival requires intervention in early infancy (arterial switch operation)
<b>Single ventricle physiology</b>	Hypoplastic left heart syndrome (HLHS), hypoplastic right heart syndrome, tricuspid atresia.	Duct dependent circulation; survival requires intervention in the neonatal period
<b>Common mixing</b>	Total anomalous pulmonary venous drainage (TAPVD).	Present with heart failure and/or cyanosis – survival requires intervention in the neonatal period



**Figure 1.** Echocardiographic appearance of secundum ASD with left-to-right flow



**Figure 2.** CXR and echocardiographic appearance in a child with heart failure secondary to AVSD



**Figure 3.** Echocardiographic appearance and CXR in a child with a PDA, showing left heart volume overload and prominent pulmonary vasculature

### Tetralogy of Fallot

Tetralogy of Fallot (TOF) is associated with a right to left shunt. The abnormality in TOF is due to:

- VSD with aortic override.
- Right ventricular outflow tract obstruction (RVOTO) (muscle bundles below the pulmonary valve; valvar or supra-valvar obstruction).
- Right ventricular hypertrophy due to the RVOTO.

The child presents with cyanosis from birth and may develop cyanotic 'spells' in infancy, due to spasm of muscle bundles in the right ventricular outflow tract in response to adrenergic stimulation, for instance when the child is upset or cries. An older child who has not had corrective surgery may 'squat' when tired to increase pulmonary blood flow (by increasing systemic vascular resistance), and will have clubbing. (See Figure 4).

A modified Blalock-Taussig (mBT) shunt may be placed as a palliative procedure in the first few weeks of life to provide a secure



**Figure 4.** Finger clubbing in a child with Tetralogy of Fallot

blood supply to the lungs before corrective surgery is performed. This is an arterial shunt between the innominate artery and the right pulmonary artery. The shunt is 'restrictive' so that the pulmonary blood supply is adequate, but not too high; the ideal SpO<sub>2</sub> is around 85%. If the shunt is too large (unrestrictive), the child will have too high a pulmonary blood flow, causing signs of cardiac failure, with low systemic pressure due to excessive 'runoff' from the innominate artery to the lungs.

Surgery for TOF should ideally be at a few months of age. Complete correction involves closure of the VSD and relief of the obstruction at the right ventricular outflow tract. The surgical course may be complicated, but the saturation will be normal after surgery and the long-term outlook is usually good. Some patients may develop pulmonary regurgitation, right heart dilation and arrhythmias if a patch has to be placed across the pulmonary annulus.

### Duct dependent circulation

In utero, the placenta is the main site for gas exchange for the developing foetus and the blood flow to the foetal lung is minimal. Blood from the right ventricle bypasses the lungs and passes directly to the aorta via a foetal vessel called the arterial duct. After birth, a number of changes occur in transition from the foetal to the newborn circulation, one of which is closure of the arterial duct so that blood now perfuses the lungs. (See Basic Science article, this edition of *Update*, page 4)

There are certain severe congenital cardiac lesions that are only compatible with life if the arterial duct remains open – the circulation is said to be 'duct dependent'. If undiagnosed at birth, these babies typically present with acute collapse as the duct closes within the first 5 days of life (the differential diagnosis is septic shock).

#### Examples of duct dependent circulations:

- Critical coarctation of the aorta (duct dependent systemic circulation). There is extreme narrowing of the aorta where the arterial duct joins the aorta, and blood supply to the lower half of the body is only possible if blood passes from

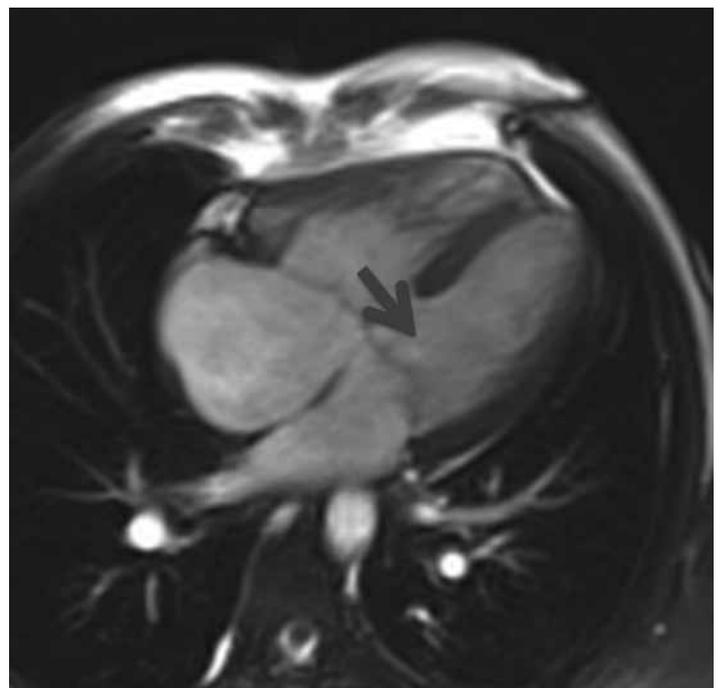
the pulmonary artery to the descending aorta via the duct. The blood in the pulmonary artery is deoxygenated, so the oxygen saturation in the feet ('post-ductal') will be lower than the oxygen saturation in the right hand ('pre-ductal')

- Pulmonary atresia (duct dependent pulmonary circulation). In pulmonary atresia, the only blood supply to the lungs is that which passes from the aorta to the pulmonary artery via the duct.

Continued survival of these babies requires infusion of prostaglandin E1 to keep the duct open until urgent cardiac surgery can be performed.

### Eisenmenger's syndrome

A large ('unrestricted') VSD allows blood to pass from the left ventricle to the right ventricle ('left-to-right shunt'), which results in high pulmonary blood flow and congestive cardiac failure. The normal physiological response to high pulmonary blood flow is for the resistance in the pulmonary vessels ('pulmonary vascular resistance', PVR) to increase. With time, the PVR rises until eventually it exceeds the systemic vascular resistance, and the flow across the VSD reverses - this is known as Eisenmenger's syndrome. Clinically, this is associated with an initial improvement in symptoms of cardiac failure as pulmonary blood flow reduces, followed by increasing cyanosis as the shunt reverses to become right-to-left. Surgical closure of the shunt is not possible at this stage as the resistance to flow through the pulmonary circulation is too high and right ventricular failure will occur. Individuals with Eisenmenger's syndrome are deeply cyanosed with clubbing of fingers; they may develop haemoptysis, endocarditis or cerebral abscess, and will eventually die from cardiac failure.



**Figure 5.** Cardiac MRI of patient with AVSD and right-to-left shunt in Eisenmenger's syndrome

Corrective cardiac surgery must be undertaken before the onset of pulmonary vascular disease to avoid Eisenmenger's syndrome. The ideal age for surgery is critically dependent on the underlying lesion. Eisenmenger's syndrome occurs before the age of 1 year in children with very high pulmonary blood flow, such as unrepaired AVSD, but may occur at 40-50 years in adults with unrepaired ASD who have had moderately increased pulmonary blood flow over many years. (See Figure 5).

## RECOGNISING CONGENITAL HEART DISEASE IN CHILDREN

It is important to recognise the child with CHD so that they can be prepared for surgery:

- Treat cardiac failure (e.g. VSD, AVSD).
- Recognise a condition that may be associated with acute decompensation during surgery (hypercyanotic 'spell' in Tetralogy of Fallot; loss of cardiac output in aortic stenosis, coarctation; cardiomyopathy; pulmonary hypertensive crisis in Eisenmenger's syndrome).
- Ensure antibiotic prophylaxis is given to children at risk of endocarditis.

Children rarely present with the symptoms classically associated with heart disease in adults (i.e. chest pain, shortness of breath, swollen ankles); they usually present with symptoms such as failure to thrive, frequent chest infections, or unexplained 'funny turns'. A careful history and examination is key, as are special investigations such as CXR, ECG, SpO<sub>2</sub>, echocardiogram, or occasionally cardiac catheterisation. Cardiac MRI is increasingly used.

### History

Ask about the following:

*Pregnancy:* Maternal disease, drug and alcohol intake

*Birth history:* A history of prematurity is associated with PDA. Birth asphyxia is associated with persistent foetal circulation (persistent pulmonary hypertension of the newborn, PPHN)

### Cardiac symptoms

- Cyanosis - central cyanosis is an important cardiac symptom that is difficult to detect and may often be missed by parents. Central cyanosis is seen as blue discoloration of the tongue and lips and may be confirmed using pulse oximetry.
- Hypercyanotic 'spell'. This is a classic symptom of Tetralogy of Fallot (see Table 3). The child may become deeply cyanosed when upset or crying, and may become limp and unresponsive; this is a sign of acute reduction in pulmonary blood flow associated with sudden increase in the dynamic obstruction to the right ventricular outflow tract. Older children with uncorrected TOF may 'squat' to increase pulmonary blood flow – this position increases the systemic vascular resistance and reduces the right-to-left shunt across the VSD. Cyanotic spells are uncommon

in the newborn. Older babies may be placed in the knee to chest position in response to a cyanotic spell. The differential diagnosis for episodic cyanosis is fits or respiratory problems such as croup or asthma.

*Respiratory symptoms.* Breathlessness due to increased pulmonary blood flow is a common respiratory symptom in children with cardiac failure, for instance due to a large VSD or AVSD. In babies, this presents as slow feeding, breathlessness, cold clammy sweatiness and poor weight gain. An older child may have limited exercise tolerance and not keep up with their peers. Frequent respiratory tract infections and poor weight gain are common in older children with an ASD, although there may not be overt breathlessness.

*Funny turns and chest pain* – funny turns are an unusual presentation for cardiac disease in children, more commonly associated with simple faints, or neurological disease such as epilepsy. Sudden collapse may be due to arrhythmias, and collapse with exercise is a very worrying sign in a child with significant left ventricular outflow tract obstruction such as aortic stenosis. Most chest pain in children is due to musculoskeletal problems, especially in older children. Coronary artery abnormalities, and hence chest pain due to angina, is rare. A young infant with angina due to Kawasaki disease or Anomalous origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) may present with 'quiet' episodes associated with reduced activity and sweatiness – more commonly with breathlessness and listlessness due to left ventricular failure.

*Poor weight gain* – this is common in conditions causing heart failure or associated with increased pulmonary blood flow such as VSD. Older children with acquired heart disease such as endocarditis or cardiomyopathy may have anorexia and weight loss. Conditions associated with cyanosis and reduced pulmonary blood flow such as TOF are not usually associated with poor weight gain.

*General enquiry* – this may reveal other symptoms suggestive of a complex congenital disorder such as Down's syndrome, a family history of cardiac disease, or symptoms suggestive of acquired heart disease such as rheumatic fever or endocarditis.

### Examination

Examine the child using the standard routine of inspection, palpation, percussion and auscultation:

- Look for dysmorphic features, for instance suggestive of Down's syndrome, or the presence of an associated congenital abnormality such as cleft palate or spinal deformity.
- Signs of poor weight gain and failure to thrive should be sought (weigh the child and compare to standard growth charts).
- Signs of breathlessness will be apparent as increased respiratory rate, and in the infant, intercostal and sub-costal recession, nasal flaring and grunting. In an older child, chest deformity may be associated with longstanding ventricular enlargement.

- Peripheral cyanosis is common in children at any age and is not important. Central cyanosis is always important but may be missed in a child with severe anaemia or children with pigmented skin. Look at the colour of the tongue – if blue, it suggests the SpO<sub>2</sub> is <85%. Saturation must be confirmed using a pulse oximeter. Long standing cyanosis may be associated with ‘clubbing’ of the nails of the hands and feet. Children who have long standing cyanosis develop compensatory polycythaemia and possibly complications such as cerebral thromboembolism.
- The jugular venous pulse is very difficult to see in children <5 years who have chubby necks and who move around a great deal – the liver size gives a much better estimate of venous pressure.
- Palpate the pulses to assess rate, rhythm, volume and character. Radio-femoral delay or absent femoral pulse is seen in coarctation; differential right and left radial pressures are seen in aortic arch interruption.
- A suprasternal ‘thrill’ may be felt in aortic stenosis, occasionally in pulmonary stenosis or other causes of aortic arch anomaly leading to a ‘palpable’ murmur. A palpable ‘heave’ indicates ventricular hypertrophy.
- The normal position of the cardiac apex is the 4th intercostal space inside the nipple line in a child <5 years, the 5th intercostal space at the nipple line in a child >5 years.
- Estimate the liver size by palpation. The normal neonate may have 1cm of liver palpable, an older child may have a liver edge palpable – anything more may indicate increased right atrial pressure, usually due to heart failure, or a non-cardiac cause of hepatomegaly.
- Dependent peripheral oedema is a late sign of cardiac failure in children, and may be felt by palpation over the sacrum.
- Percussion may be useful to estimate liver size and the presence of ascites (rarely due to cardiac failure in children).

## Heart murmurs

### Innocent cardiac murmurs

The most common murmur in children is a functional, innocent or physiological heart murmur, which is heard in 10% of normal children. The classical innocent murmur in children is known as the ‘Still’s’ murmur. Innocent murmurs may also be due to flow murmurs associated with increased cardiac output, heard in children with a fever or anaemia. The heart is structurally normal in all children with an innocent murmur. A murmur in a child may be classified as innocent if the child has no other signs or symptoms of cardiac disease, and the murmur has certain characteristic features:

- Soft (no thrill)
- Systolic and short (never pansystolic)
- Asymptomatic

- Best heard at the left sternal edge, no radiation
- Changes with posture – softer when standing.

### Pathological cardiac murmurs

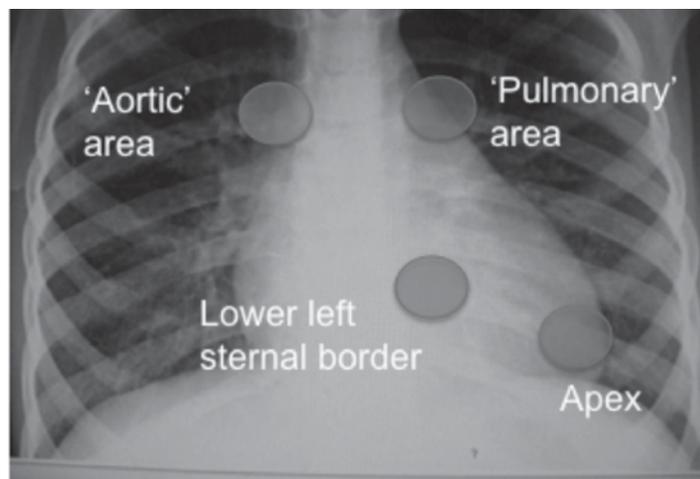
Cardiac murmurs associated with a left to right shunt such as a VSD may not be very obvious in the newborn period when the pulmonary vascular resistance is high. The pulmonary vascular resistance falls in the first few days of life - the loud systolic murmur due to a VSD will become apparent and the child may develop signs of heart failure as the flow increases across the shunt. A murmur is the result of turbulent flow and is graded as soft, moderate, or loud; a very loud murmur may also be felt (‘thrill’). See Table 4 and Figure 6 for description and location of pathological cardiac murmurs.

### Investigations

Special investigations include the Chest Xray, ECG, echocardiography and cardiac catheterisation.

**Chest Xray.** Consider the age of the patient and if the film was taken in the sitting or lying position. Evaluate the chest Xray systematically:

- A - Adequacy and alignment. The film should be sufficiently penetrated to just visualise the disc spaces of the lower thoracic vertebrae through the heart shadow. At least 5 anterior rib ends should be seen above the diaphragm on the right hand side. Alignment can be assessed by ensuring that the medial ends of both clavicles are equally spaced about the spinous processes of the upper thoracic vertebrae.
- B – Bones. Check the ribs, clavicles and vertebrae. (Rib notching is sometimes seen in severe coarctation of the aorta).
- C - Cartilage and soft tissues.
- Lungs - Compare side-to-side and upper, middle and lower thirds of the chest. Look for pleural effusions, pneumothorax, vascular markings that are increased or decreased (plethoric or oligoemic), fluid in the fissures, white lung areas which could be consolidation or haemorrhage.



**Figure 6.** Best location to hear cardiac murmurs

**Table 4. Pathological murmurs in children**

Murmur	Location murmur heard best
<b>Systolic murmurs</b>	
'Soft' = grade 2/6; 'Moderate' = grade 3/6; 'Loud with thrill' = grade 4/6	
<b>Ejection systolic</b>	<i>Aortic stenosis</i> : Upper right sternal edge +/- carotid thrill <i>Pulmonary stenosis or ASD</i> : Upper left sternal edge, no thrill <i>TOF</i> : Long harsh murmur (in a child with cyanosis)
<b>Pansystolic</b>	<i>VSD</i> : Lower left sternal edge +/- thrill <i>Mitral regurgitation (NB. rheumatic fever)</i> : Apex
<b>Diastolic murmurs (unusual in children)</b>	
'Soft' = grade 2/4; 'Moderate' = grade 3/4; 'Loud' = grade 4/4	
	<i>Aortic regurgitation (NB. endocarditis)</i> : Lower left sternal edge, sitting forward, collapsing pulses
<b>Continuous murmurs</b>	
	<i>Patent ductus arteriosus (PDA)</i> : Left infraclavicular region <i>Arteriovenous malformation</i> : Any site (head, shoulder, lungs).

- Heart: Look at the size of the heart; is it enlarged? Is the shape unusual? (See below). In normal infants the heart is up to 60% of the thoracic diameter, 50% thereafter. (See Figure 7). Remember that a normal cardiac shadow does not rule out cardiac disease.
- The upper mediastinum: In children under the age of 18 months, the normal thymus may simulate superior mediastinal widening (above the level of the carina) (See Figure 7).
- D – Diaphragms. The border between the heart and the diaphragm and between the diaphragm and the ribs (cardiophrenic and costophrenic angles) should be clear on both sides. Loss of definition of the left diaphragm behind the heart suggests left lower lobe collapse, an abnormal hump suggests diaphragmatic rupture, a hazy diaphragm suggests effusion or collapse in the bordering lung segment, and an elevated diaphragm suggests phrenic nerve palsy

There are some classical appearances of the chest Xray in children:

- 'Egg-on-side' = Transposition of great arteries in a neonate
- Boot shaped heart = Tetralogy of Fallot (right ventricular hypertrophy and reduced pulmonary markings (See Figure 8))
- 'Snowman in a snow storm' = Obstructed total anomalous pulmonary venous connection in a neonate
- Globular heart. Usually associated with pericardial effusions, may be secondary to pericarditis or dilated cardiomyopathy

- Situs. In the normal situation (situs solitus) the heart is on the left with the gastric bubble on the left and the liver on the right. In situs inversus these relations are reversed
- Oligaemic lung fields are seen in conditions associated with reduced pulmonary blood flow such as TOF and pulmonary atresia
- Plethoric lung fields are seen in children with left to right shunts, especially VSD and AVSD.

#### *The electrocardiogram (ECG)*

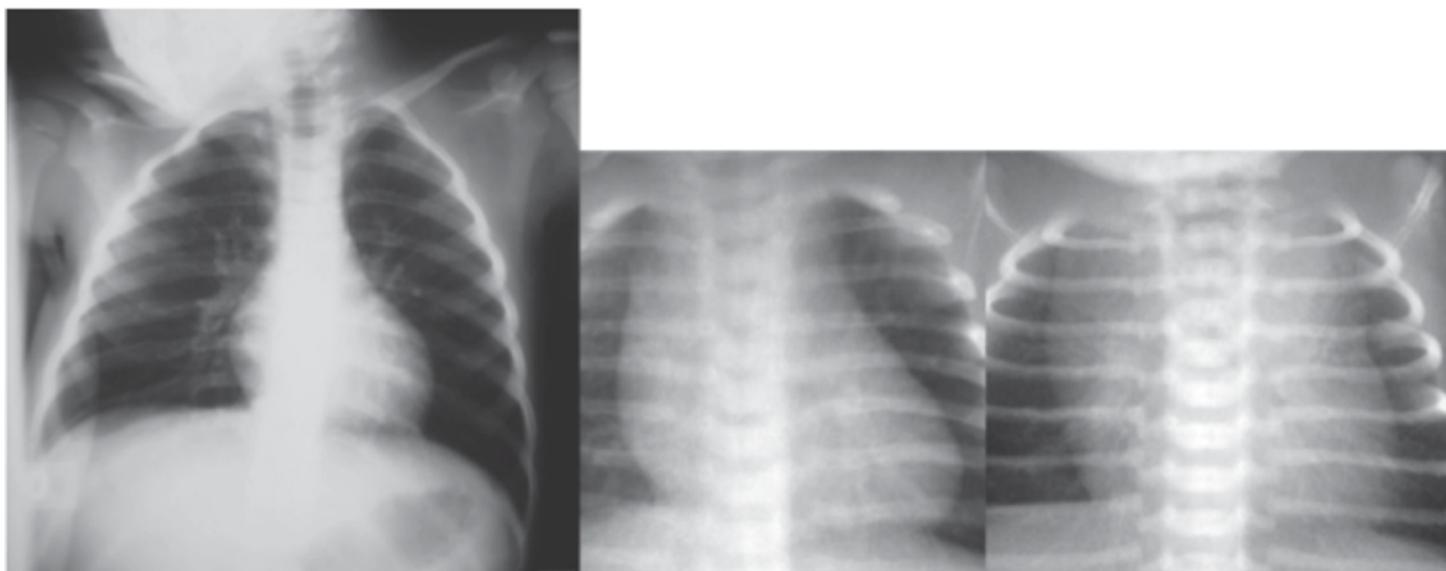
The ECG may be useful to investigate rhythm and conduction abnormalities, as well as assessing chamber hypertrophy and strain. Interpretation of the paediatric ECG is complex and must take the child's age into account, with comparison to tables of normal values.

#### *Echocardiography*

Echocardiography is a form of cardiac imaging that uses reflection of ultrasound pulses from interfaces between tissue planes. It can be used to generate detailed real time images of the cardiac anatomy. It has become the standard investigation for all patients with valvular heart disease, congenital heart disease, myocardial and pericardial disease, and to assess myocardial function.

The heart is examined in a systematic way, deciding first the orientation of the heart within the body (normal = solitus); the connections between the atria and the ventricles (normal = concordant), ventricles and major vessels (normal = concordant); and the side of the aortic arch (normal = left). The normal position and connections is often described in short hand as 'SCCL'. The echocardiologist then describes the ventricular function, the valves, the shunts, and the size of the major vessels.

**Figure 7.** Normal chest Xrays in children



**Normal 3 year-old**

**Normal newborn**

Doppler ultrasound may be used to estimate pressure gradients across valves and shunts using the following formula:

$$\text{Pressure} = 4 v^2, \text{ where } v \text{ is the velocity measured by Doppler m.sec}^{-1}$$

The 'z score' is used to describe the heart compared to normal values for the age of the child; if the aortic valve has a z score of -2, this suggests the valve is 2 standard deviations smaller than the normal average size.

#### Cardiac catheterisation

Cardiac catheterisation is used to answer specific diagnostic questions in children with congenital heart disease. A catheter can be passed into the heart chambers under Xray control to measure intracardiac pressures and oxygen saturations, or for radiological imaging by injection of contrast media. Interventional cardiology is increasingly providing definitive treatment for a number of conditions, for instance closure of ASD or PDA by insertion of an occlusion device, balloon dilatation of pulmonary stenosis, or diathermy ablation of abnormal conduction pathways.

### PRINCIPLES OF ANAESTHESIA IN CHILDREN WITH CONGENITAL CARDIAC DISEASE

Unnecessary surgery should be avoided due to the increased risk associated with anaesthesia, particularly in neonates, and children with complex (cyanotic) disease. It is important to optimise the condition of the child preoperatively, to understand the physiology of the lesion, and know how to avoid acute decompensation during surgery.

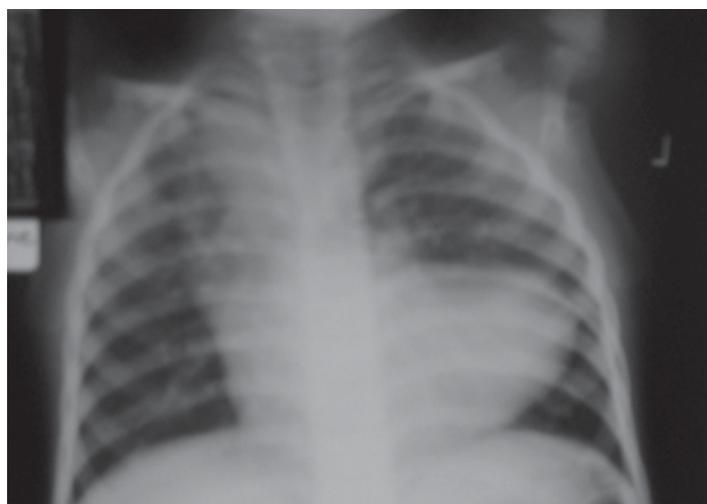
The general principles of anaesthesia are as follows:

- Avoid deep (halothane) anaesthesia
- Support ventilation where possible

- Avoid air emboli, particularly for cyanotic children with right-to-left shunts; make sure there are no air bubbles in the drugs or fluids given
- Consider endocarditis prophylaxis
- Consider the effect of anaesthesia on the pulmonary blood flow, and the balance between systemic and pulmonary blood flow.

#### Pulmonary blood flow

Pulmonary blood flow may be low due to increased pulmonary vascular resistance (PVR), which may be due to excessive muscularisation of the pulmonary arterioles secondary to long-standing high pulmonary blood flow. Pulmonary vascular resistance may be increased acutely, particularly in neonates, due to:



**Figure 8.** Chest Xray of a child with tetralogy of Fallot and right-sided aortic arch showing a 'boot shaped' heart

- Hypoxia
- Acidosis
- Over/under inflation of the lungs.

The most powerful pulmonary vasodilator available therapeutically is oxygen. The effect of oxygen is particularly evident in the first weeks of life when the pulmonary vasculature remains very reactive. It is sometimes important to avoid giving too much oxygen. For instance, if a patient with high pulmonary blood flow is given 100% oxygen to breathe (e.g. a baby with a large unrepaired AVSD), this will cause the pulmonary vascular resistance to fall, pulmonary blood flow will increase and the symptoms of cardiac failure, such as breathlessness, will get worse. A saturation of 94% is adequate in children with unrepaired AVSD, and excessive oxygen should be avoided.

Other pulmonary vasodilators include:

- Nitric oxide, an inhaled pulmonary vasodilator, used for the treatment of pulmonary hypertension in neonates
- Sildenafil, a cGMP-specific phosphodiesterase type 5 inhibitor
- Bosentan, an endothelin receptor antagonist.

If a patient has impaired pulmonary blood flow due to pulmonary hypertension, for instance, a child with sepsis and persistent pulmonary hypertension of the newborn, they should be treated with oxygen, nitric oxide +/- sildenafil if available.

### **ANAESTHESIA FOR SPECIFIC CARDIAC LESIONS**

Children who have had corrective surgery e.g. for ASD, VSD or PDA, may be treated the same as any other child. Children with uncorrected cardiac lesions may be more challenging.

#### **Uncorrected left-to-right shunt; ASD, VSD, AVSD, PDA**

##### *Problems*

- Increased pulmonary blood flow
- Cardiac failure.

##### *Anaesthesia for uncorrected left-to-right shunt*

- Careful preoperative assessment; if possible, treat cardiac failure with diuretics (furosemide, spironolactone PO). In older children, exclude signs of Eisenmenger's syndrome and shunt reversal
- Avoid deep halothane anaesthesia
- Avoid excessive oxygen; keep  $FiO_2$  at a minimum, accept  $SpO_2$  approx. 94% (but see below for Eisenmenger's syndrome)
- Support the ventilation to reduce the work of breathing (increased pulmonary blood flow causes lung compliance to fall).

#### **Eisenmenger's syndrome**

Irreversible pulmonary hypertension has developed as a consequence of long-standing or significantly raised pulmonary blood flow. There is now right-to-left shunting or bidirectional shunting and the

patient is cyanosed. The mortality from Eisenmenger's syndrome is 75% at 30 years of age. (See figure 5).

##### *Problems*

- Shunt reversal
- Cyanosis

##### *Anaesthesia for Eisenmenger's syndrome*

The patient is severely unwell, and may present with the following signs and symptoms:

- Limited exercise tolerance
- Polycythaemia, hyperviscosity syndrome (fatigue, headache, blurred vision, cerebral thrombosis, cerebral abscess)
- Coagulopathy
- Thromboembolic events
- Endocarditis.

This is very high-risk surgery, with an estimated perioperative mortality of 10-14% even for minor surgery. The risks and benefits of proceeding with surgery have to be considered carefully. The following is recommended:

- Polycythaemia is associated with hyperviscosity. Consider venesection if the haematocrit is >60. Do not let the patient become dehydrated; start IV fluids preoperatively.
- Give high inspired oxygen at all times.
- Use a careful balanced general anaesthetic and close monitoring. Induce anaesthesia with small incremental doses of midazolam and fentanyl IV, and a small dose of induction agent (ketamine, etomidate).
- Support ventilation to reduce the work of breathing, but do not over/under inflate the lungs (increases PVR).
- Avoid increases in pulmonary vascular resistance, as this will make the cyanosis worse.
- Do NOT use a spinal, as this will cause a fall in systemic vascular resistance, and will encourage the right to left shunt.
- Maintain oxygen carrying capacity; do not allow the patient to become anaemic, and maintain haemoglobin 12-14g.dl<sup>-1</sup> (Hct 36-42).
- Consider a pulmonary vasodilator if possible – sildenafil or inhaled NO

##### *Management of a pulmonary hypertensive crisis*

In the event of a pulmonary hypertensive crisis with falling  $SpO_2$  and blood pressure, you must intervene to reduce the PVR and increase cardiac output:

- Give 100% oxygen and hyperventilate the patient
- Give analgesia
- Treat metabolic acidosis

- Support the circulation – adrenaline infusion
- Start a pulmonary vasodilator if available (inhaled NO, sildenafil via a nasogastric tube).

The patient must be monitored in a high dependency area or ICU after surgery.

## Tetralogy of Fallot

### Problems

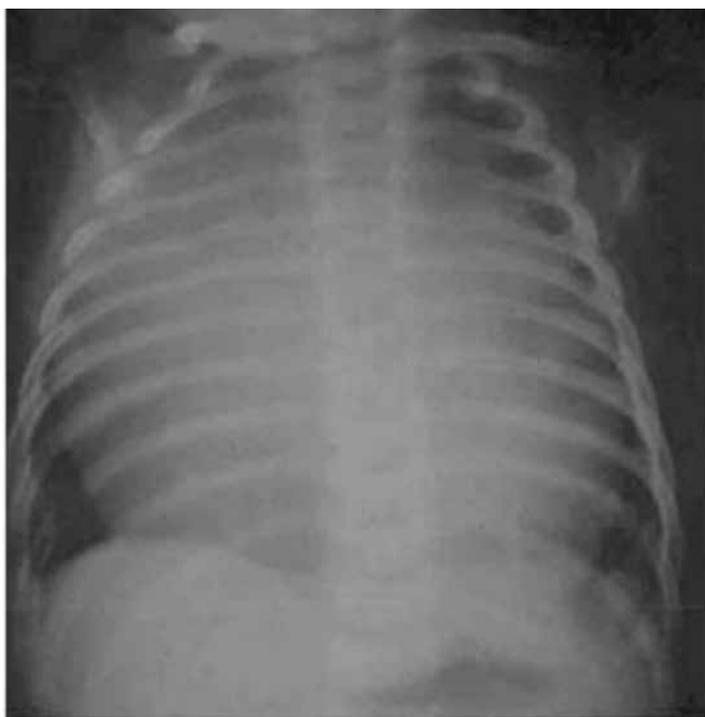
- Reduced pulmonary blood flow due to obstruction at the right ventricular outflow tract (NB PVR is LOW)
- Hyperviscosity syndrome and polycythaemia due to long-standing cyanosis.

### Anaesthesia for children with unrepaired TOF

- Sedative premedication; keep the child calm
- Balanced anaesthesia with good analgesia; avoid dehydration, keep well-filled, avoid sympathetic stimulation to avoid hypercyanotic spells.

### Management of a hypercyanotic 'spell' during surgery in unrepaired TOF

- Increase the FiO<sub>2</sub>.
- Deepen anaesthesia.
- Give a bolus of Ringer's 20ml.kg<sup>-1</sup> to increase cardiac output and blood flow to the lungs.
- Increase the systemic vascular resistance to reduce the right-to-left shunt; phenylephrine bolus 1mcg.kg<sup>-1</sup> IV, titrated



**Figure 9.** Large pericardial effusion in a child

to effect. Do NOT use adrenaline as this will worsen the right ventricular outflow tract obstruction ( $\beta$ 1-receptor agonist effect).

- Consider a  $\beta$ -blocker – esmolol or propranolol IV.

### Anaesthesia for children with a modified BT shunt (mBT shunt)

- Check the shunt is patent before the start of surgery – SpO<sub>2</sub> should be around 85%, and a shunt murmur should be audible (continuous murmur in the second intercostal space, mid-clavicular region on the right).
- Avoid dehydration as this may result in the shunt becoming blocked.
- Keep the blood pressure up; if the blood pressure falls, pulmonary blood flow will also fall and the oxygen saturation will drop.

## High-risk lesions

There are a number of high-risk lesions that should ring alarm bells; these 'red flag' patients must be assessed and optimised before surgery, and the risk/benefits of non-cardiac surgery considered very carefully.

The following are high-risk lesions:

- Eisenmenger's syndrome. (See above)
- Aortic stenosis. Avoid a fall in SVR – do not use spinal anaesthesia
- Cardiomyopathy. May be dilated or restrictive, and the child will have limited reserve. Ketamine is useful for induction, and the child should be monitored carefully postoperatively
- Pericardial effusion. Must be drained by the cardiologist before surgery (See Figure 9)
- Endomyocardial fibrosis. Manage as for restrictive cardiomyopathy.

## ENDOCARDITIS PROPHYLAXIS

Children who have had bacterial endocarditis, who have a prosthetic valve, and who have congenital heart disease are at high risk of bacterial endocarditis. The current advice from the American Heart Association is to give prophylactic antibiotics in the following situations:

- High risk dental surgery e.g. involving suturing or dental infection
- Give antibiotics as indicated for the surgical procedure.

## ANAESTHESIA FOR A CHILD WITH AN UNDIAGNOSED CARDIAC LESION

There may be situations in LMIC where the anaesthetist is presented with a child with a murmur and an undiagnosed cardiac condition. The following approach is advised:

## Elective surgery

Refer for investigation if possible.

## Emergency surgery:

- 'Well' child, pink, breathless with cardiomegaly. Likely to be a condition associated with left to right shunt such as an ASD, VSD or PDA. The child may have heart failure with increased work of breathing. Support ventilation during surgery to reduce the work of breathing.
- Young child, 'well', but has always been blue. Likely to be tetralogy of Fallot. The problem is reduced pulmonary blood flow. Give high-inspired oxygen and avoid sympathetic stimulation.
- Any neonate, or older child with poor function, funny turns +/- cyanosis. These cases are high risk and only life-saving surgery should be contemplated. Use a balanced anaesthetic technique with positive pressure ventilation. Ketamine is a good agent to use in these high-risk cases.

All children with undiagnosed cardiac disease should be referred to a cardiologist as soon as possible for investigation.

## CONCLUSION

Children with CHD may provide a challenge to the anaesthetist, but with careful planning, most can be anaesthetised safely. It is important to understand the physiology of the particular cardiac lesion, to prepare in advance, and to do the simple things well.

## ACKNOWLEDGEMENT

I would like to thank Dr Jan Marek who provided the echocardiographic images for this article.

## REFERENCES

1. Story E. Recognising cardiac disease in children. *Anaesthesia Tutorial of the Week* 93 (2008). Available at: [http://www.wfsahq.org/components/com\\_virtual\\_library/media/4bd1c43b45969453c47a9a1c96723d46-64f3a3069773de361efae976bb23f931-93-Recognising-Cardiac-Disease-in-Children.pdf](http://www.wfsahq.org/components/com_virtual_library/media/4bd1c43b45969453c47a9a1c96723d46-64f3a3069773de361efae976bb23f931-93-Recognising-Cardiac-Disease-in-Children.pdf)
2. Baum VC, Barton DM, Gutgesell HP. Influence of congenital heart disease on mortality after non-cardiac surgery in hospitalized children. *Pediatrics* 2000; **105**: 332-5.
3. Paediatric cardiology: an introduction. Archer N, Burch M. Chapman Hall Medical. Philadelphia 1998.