RECOGNISING CARDIAC DISEASE IN CHILDREN

ANAESTHESIA TUTORIAL OF THE WEEK 93

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Consider the following 2 cases and discuss the questions below before reading the tutorial.

Case A

A 3 month old baby presents with poor feeding and failure to thrive. She was born at term after a normal pregnancy. Mum describes that the baby always breathes very fast, that she takes a long time to feed and gets very sweaty. On examination the baby is pink, malnourished and breathless. She has a respiratory rate of 50 and has marked intercostal and sub-costal recession. She has a loud pan-systolic murmur and her liver is palpable at 2cm. A CXR shows increased pulmonary vascular markings and a large heart (>60% of the thoracic diameter).

Case B

A 9 year old girl presents with fever and chest pain. Her mother informs you that she had a sore throat a few weeks ago for which she did not receive any treatment. On examination you notice that her right knee and left elbow are swollen and painful. On auscultation of her heart you hear a loud pansystolic murmur.

Questions:

1. What is meant by "cyanotic" or "acyanotic" cardiac lesions? List example of each.
2. What is Eisenmengers syndrome?
3. What is meant by duct dependence?
4. How can you recognise cardiac disease in children?
5. What is the likely diagnosis for case A and where does it fit into your classification?
6. List as many different cardiac lesions as you can think of and describe the type, and location of the murmur associated with it eg aortic stenosis, ejection systolic murmur, upper right sternal edge.
7. Describe how you would systematically look at a paediatric CXR and some of the features you may be looking for in cardiac disease.
8. What investigations, if available, could help you to identify cardiac disease?

9. What is the diagnosis for case B and what is the likely cause of the heart murmur? How many children worldwide do you think are affected with this condition?
10. What are the features of an 'innocent' heart murmur?

Tutorial:

Heart disease in children may be **congenital** or **acquired.** This tutorial will consider the different types of heart disease in children and the recognition and investigation of children with heart disease.

1. Congenital heart disease

i. Structural heart defects

The incidence of congenital heart disease (CHD) varies between 5 and 8/1000 live births. The cause of most congenital heart disease is unknown, but is likely to be related to genetic defects, teratogens (such as maternal alcohol or drugs, including anti-epileptics or warfarin), or maternal disease (rubella, diabetes).

The incidence of different types of structural heart disease varies between populations, but ventricular septal defect (VSD) is the most common lesion in all populations. The incidence of different types of CHD is shown in table 1 below.

Table 1. The incidence of types of CHD in the UK

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

ii. Genetic defects

Recognisable chromosomal abnormalities are present in 25% of children with CHD. The diagnosis of a chromosomal abnormality should lead to active assessment for CHD. The most common chromosomal abnormality is Down's syndrome (trisomy 21) – 40% of children with Down's syndrome have CHD, most commonly atrioventricular septal defect (AVSD) or VSD. Genetic defects associated with cardiac lesions are shown in table 2 below:

Table 2. Genetic defects associated with cardiac lesions

Abnormality	Clinical features and associated conditions	Typical cardiac lesion
Down's 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

iii. Classification of structural defects

It is useful to classify congenital heart disease according to the pathophysiology of the major heart lesion, in particular, whether the lesion is associated with cyanosis ('blue') or is acyanotic ('pink'), also whether the lesion is associated with abnormal flow between the cardiac chambers (abnormal 'shunt'), obstruction to flow, abnormal connections of the major vessels or abnormal mixing.

Table 3. Classification of common congenital heart lesions

	Common examples	Comment
Acyanotic lesions - pink		
Left to right shunt i. 'Restrictive' ii. 'Non-restrictive'	 Small ASD, VSD, PDA Large VSD, PDA, AVSD	Large ('non-restrictive') defects are associated with severe congestive cardiac failure in infancy. If unrepaired, may lead to pulmonary hypertension and reversal of shunt (Eisenmenger's syndrome, see later). The age at which this occurs depends on the magnitude of the shunt (occurs in infancy for non-restrictive shunt, may occur in adulthood for small, restrictive shunts)
Obstructive	Aortic stenosis, coarctation of the aorta, pulmonary stenosis	Severity of lesion determines age at presentation – neonates with severe obstruction may be critically ill
Cyanotic lesions - blue		
Right to left shunt	Tetralogy of Fallot (Right ventricular outflow tract obstruction, right ventricular hypertrophy, VSD with aortic override)	May present with severe cyanosis and hypercyanotic 'spells'
Transposition of the Great Arteries (TGA)	TGA may be associated with ASD, VSD, PDA	Long term survival requires intervention in early infancy (arterial switch operation)
Common mixing	Anomalous pulmonary venous drainage with ASD, common arterial trunk, single ventricle	Present with heart failure and cyanosis – survival requires intervention in the neonatal period

iv. 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 from the pulmonary artery 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 including expansion of the lungs (which reduces

pulmonary vascular resistance) and closure of the arterial duct, so that blood now perfuses the lungs. 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'. 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 include critical coarctation of the aorta (duct dependent systemic circulation) or pulmonary atresia (duct dependent pulmonary circulation). In critical coarctation there is extreme narrowing of the aorta just where the arterial duct joins the aorta, and blood supply to the lower half of the body is only possible if it passes from the pulmonary artery to the descending aorta via the duct. 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 prostglandin E1 (prostin) to keep the duct open until urgent cardiac surgery is possible. NB prostin also causes pyrexia and apnoeas and these babies may require ventilation while on prostin infusions.

v. Eisenmenger's syndrome

Lesions associated with left to right shunt, such as AVSD, cause high pulmonary blood flow and congestive cardiac failure. The normal physiological response to high pulmonary blood flow is for the pulmonary vascular resistance to increase. With time, the pulmonary vascular resistance will exceed the systemic vascular resistance, and the flow across the shunt will reverse (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, may develop haemoptysis, endocarditis or cerebral abscess, and will eventually die from cardiac failure.

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 the age of 40-50 years in adults with unrepaired ASD who have had moderately increased pulmonary blood flow over many years. Corrective cardiac surgery to avoid Eisenmenger's must be undertaken before the onset of pulmonary vascular disease, the ideal age dependent on the severity of the underlying lesion.

vi. Further details and diagrams of the normal heart and common congenital heart lesions.

See the following web links for further details of congenital heart lesions:
http://www.med.yale.edu/intmed/cardio/chd/
http://www.nlm.nih.gov/medlineplus/ency/article/001114.htm

vii. Arrhythmias in children

Another form of congenital heart disease in children may be the presence of abnormal conduction pathways that lead to the development of cardiac arrhythmias, particularly supraventricular tachycardia.

Supraventricular tachycardia may be caused by abnormal conduction pathways such as the Wolf-Parkinson-White syndrome, characterised on the ECG by a short P-R interval and an abnormal delta wave on the upstroke of the QRS complex. The child may develop sudden episodes of tachycardia associated with heart rates of >240beats/min, up to 300beats/min. These appear on ECG as narrow complex rhythms. The child may feel faint and uncomfortable during an arrhythmia. The arrhythmia may terminate spontaneously or be terminated by vagal manoeuvres such as brief immersion of the face in ice cold water, carotid sinus massage (massage to the carotid artery in the neck), or in older children, a Valsalva manoeuvre. Specific treatment is with rapid bolus of intravenous adenosine, or if this fails, synchronised cardioversion. Beta blockers may be used to prevent further episodes of tachyarrhythmia – many children will grow out of these episodes with time. Transcatheter radiofrequency ablation of the accessory pathway is undertaken in specialist centres for older children whose symptoms are not controlled by drugs.

Supraventricular tachycardia must be differentiated from **ventricular tachycardia** (VT). In VT the rate is often slower (<200 beats/min), with wide complexes on the ECG with no association between the QRS complex and the P wave. The child is often symptomatic, presenting with acute collapse. Ventricular tachyarrhythmias are often associated with underlying heart disease such as cardiomyopathy. **Torsades de Pointes** is a particular form of sinusoidal ventricular arrhythmia that occurs in the rare congenital long QT syndrome. If a child with ventricular tachycardia is acutely symptomatic they will require electrical cardioversion; if not, then treatment with an antiarrhythmic drug such as flecainide or amiodarone, or in the case of Torsades de Pointe, intravenous magnesium.

Bradycardia. Heart rate less than 90/minute. May be congenital (complete heart block) or acquired following infection (myocarditis, rheumatic fever) or cardiac surgery (damage to the conduction pathways).

2. Acquired Heart Disease

Acquired heart disease is much more common in adults than in children, but a there are number of diseases affecting the heart that may be acquired during childhood.

i. Rheumatic heart disease

Rheumatic fever is uncommon in the developed world, but still occurs with an incidence of 1-2/1000 in developing countries. Rheumatic fever is due to an abnormal immune response to the Lancefield Group A streptococcus (*Streptococcus pyogenes*), which results in a pancarditis that may lead to permanent damage to the heart and heart valves. The left sided valves are most commonly affected, the mitral valve more commonly than the aortic valve. Mitral regurgitation is seen initially; mitral stenosis

may develop over time. Approximately 8 million children are affected by rheumatic heart disease worldwide.

The typical picture is of a child 5-15 years of age who suffers from an untreated streptococcal throat infection; 1-5 weeks later they present with a fever, swollen joints and signs of cardiac disease (breathlessness due to heart failure). There is no specific test for rheumatic fever, but the diagnosis is based on the presence of two major criteria, or one major and two minor, in a child with evidence of a recent streptococcal throat infection.

Table 4. Criteria for diagnosis of rheumatic fever.

Major criteria

- Arthritis
- Carditis
- Sydenham's chorea
- Erythema marginatum (rash with clear centre)
- Subcutaneous nodules

Minor criteria

- Fever
- swollen joints
- raised inflammatory markers (ESR, CRP)
- first degree heart block
- previous history of rheumatic fever

Treatment of acute rheumatic fever

The most important treatment of acute rheumatic fever is prevention, associated with improvements in socioeconomic conditions.

Once acute rheumatic fever has occurred, treatment includes penicillin to eradicate the streptococcal infection, high dose aspirin as an anti-inflammatory drug, or steroids if the cardiac disease is severe. Heart failure is treated with diuretics, and urgent heart valve replacement may occasionally be required. Long-term antibiotics throughout childhood are essential to prevent recurrences and to reduce the risk of subsequent valvular heart disease. Penicillin is commonly given as a monthly intramuscular injection. Heart surgery may be required at a later stage to correct long-term damage to the heart valves.

ii. Infective endocarditis

Children who have congenital heart defects or who have rheumatic heart disease may develop infective endocarditis in which the endocardial tissue is infected by organisms such as *Streptococcus viridans*, *Staphylococcus aureus*, or *Staphylococcus epidermidus*. The child presents with general malaise, arthralgia, fever, splenomegaly and the signs of their underlying heart disease. The classical signs of splinter haemorrhages, petechiae, haemorrhagic lesions (Janeway lesions), tender Osler's

nodes, or Roth's spots (retinal haemorrhage) are not always seen in children. Microscopic haematuria is common, as is anaemia and a raised ESR. Serial blood cultures may reveal the causative organism.

Infective endocarditis may affect the heart valves and cause severe heart failure and death. Large lumps of infected material (vegetations) may embolise or cause abscesses in the aortic root, brain, lung, kidney or spleen. Treatment of endocarditis is at least 6 weeks of antibiotics to clear the infection. Surgery may occasionally be required to remove large clumps of infected material or treat the effects of severe valve damage.

It is important to prevent the development of endocarditis in susceptible individuals by administering prophylactic antibiotics, for instance when undergoing dental or genitourinary procedures that may be associated with a bacteraemia.

iii. Kawasaki disease

Kawasaki disease is a rare disease that primarily occurs in children under the age of 5 years. It is an inflammatory condition associated with skin rashes, fever, cracked lips and peeling of the skin of the hands and feet. It causes inflammation of blood vessels including the coronary arteries that may result in coronary aneurysms and myocardial infarction. Treatment is symptomatic with anti-inflammatory agents (e.g. high dose aspirin)

iv. Other causes of acquired heart disease in children

Infective pericarditis may occur secondary to septicaemia or pulmonary infection. Viral infection may result in **cardiomyopathy**. **AIDS** related **myocarditis** is seen in end stage disease.

Heart failure may also be a late manifestation of **severe anaemia** or **severe malnutrition**

3. Recognising Cardiac Disease in Children

Although rare in children, is essential that the anaesthetist recognises the presence of heart disease prior to surgery:

- to treat cardiac failure prior to surgery (VSD, AVSD)
- to recognise a condition that may be associated with acute decompensation during surgery (hypercyanotic episode in Tetralogy of Fallot, loss of cardiac output in aortic stenosis, coarctation, cardiomyopathy)
- to minimise the risk of air emboli through abnormal shunts (TOF)
- to ensure antibiotic prophylaxis is given to children at risk of endocarditis

Children rarely present with the symptoms classically associated with heart disease in adults (chest pain, shortness of breath, swollen ankles) – rather they present with a

variety of 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 and pulse oximetry. An echocardiogram is the gold standard investigation required to confirm the diagnosis. Cardiac catheterisation is used as a diagnostic tool to answer specific questions, for instance to measure the pulmonary vascular resistance, or investigate the anatomy of the pulmonary vessels.

i. History

There are important features to identify in the history including pregnancy and birth history, any cardiac symptoms, and a general enquiry:

Pregnancy – maternal disease, drug and alcohol intake

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

Cardiac symptoms

- **Cyanosis** - central cyanosis for instance due to Tetralogy of Fallot (TOF) 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 non-invasively using pulse oximetry.

- **Hypercyanotic 'spell'** – is a classic symptom of Tetralogy of Fallot (right ventricular outflow tract obstruction with VSD, right ventricular hypertrophy, aortic override of the VSD). After an episode of crying the child may become deeply cyanosed and may even 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 learn to 'squat' in response to a hypercyanotic spell – this position increases the systemic vascular resistance and reduces the right-to-left shunt across the VSD. Cyanotic spells are uncommon in the newborn, but 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 ASDs, although there may not be overt breathlessness.

- **Funny turns and chest pain** – funny turns are an unusual presentation for cardiac disease in children, much 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

ii. Examination

The child should be systematically examined and in particular for features of cardiac disease following the standard routine of inspection, palpation, percussion and auscultation:

Inspection

- The child should be inspected for **dysmorphic features**, for instance suggestive of Down's syndrome, or the presence of an associated congenial 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 long standing 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. It is detected by looking at the colour of the tongue – blue colouration suggests a saturation of less than 85%. Central cyanosis is not improved by breathing 100% oxygen and its presence should be confirmed by 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 < 5years who often have fat necks and who move around a great deal – the liver size gives a much better estimate of venous pressure.

Palpation

- The pulses should be palpated to assess rate, rhythm, volume and character. **Radio-femoral delay** or absent femoral pulse is seen in coarctation, differential right and left radial pressures 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 < 5years, the 5th intercostal space at the nipple line in a child > 5 years.

- The **liver size** should be estimated by palpation. The normal neonate may have 1cm of liver palpable, an older just 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 in children, and may be felt by palpation over the sacrum.

Percussion

- Percussion may be useful to estimate liver size and the presence of ascites (rarely due to cardiac failure in children)

Auscultation

- **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 increasing heart failure as the flow increases across the shunt. The loudness of cardiac murmurs may be graded.

Table 5. Cardiac murmurs in children

Systolic murmurs Soft = grade 2/6, Moderate = grade 3/6 Loud with thrill = grade 4/6
Ejection systolic murmur Upper right sternal edge +/- carotid thrill = Aortic stenosis Upper left sternal edge, no thrill = Pulmonary stenosis or ASD Long harsh murmur with cyanosis = TOF
Pansystolic murmur Lower left sternal edge +/- thrill =VSD Apex = Mitral regurgitation (rheumatic fever)
Diastolic murmur Soft = grade 2/4 Moderate =grade 3/4 Loud = grade 4/4
Unusual in children Lower left sternal edge, sitting forward, collapsing pulses = Aortic regurgitation (endocarditis)
Continuous murmur
Left infraclavicular region = Patent ductus arteriosus Any site (head, shoulder, lungs) = Arteriovenous malformation

Innocent cardiac murmurs.
The commonest murmur heard in children is the functional, innocent or physiological heart murmur, which is heard in 10% of normal children. The classical innocent murmur in children is the 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

iii. Investigations

Special investigations include the CXR, ECG, echocardiography and cardiac catheterisation.

Chest X-Ray. In looking at the CXR in a child, always consider the age of the patient and if the film was taken in the sitting or lying position. The CXR should be evaluated 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 third of the chest. Look for pleural effusions, pneumothorax, vascular markings that are increased or decreased (plethoric or oligaemic), fluid in the fissures, white lung areas which could be consolidation or haemorrhage.

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. 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).

D – Diaphragms
The boarder between the heart and the diaphragm and 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 CXR 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
- **'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. Doppler ultrasound may be used to estimate pressure gradients across valves and VSDs. Echocardiography has become the standard investigation for all patients with valvular heart disease, congenital heart disease, myocardial and pericardial disease, and in assessing myocardial function.

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 X-ray control to measure intracardiac pressures and oxygen saturations, or for radiological imaging by injection of contrast media. Interventional cardiology is a growing speciality that provides definitive treatment for a growing number of conditions, for instance closure of ASD or PDA by insertion of occlusion devices, balloon dilatation of pulmonary stenosis, or diathermy ablation of abnormal conduction pathways.

Further reading

Paediatric cardiology: an introduction. Archer N, Burch M. Chapman Hall Medical. Philadelphia 1998

Answers to Questions.

Most answers are covered in the tutorial text.

1. A lesion is “cyanotic” if it allows deoxygenated blood to bypass the lungs and mix with oxygenated blood in the systemic circulation. This makes the patient cyanosed with low saturations. A lesion where there is either no mixing of oxygenated and deoxygenated blood, or the lesion allows oxygenated blood to pass from the left side of the heart to the right is termed “acyanotic”. The patients’ saturations are normal.
2. See text.
3. See text.
4. See text.
5. The diagnosis in Case A could be any of the acyanotic lesions. She has a history starting at birth so it is unlikely to be due to an acquired infective illness.
6. See table 5.
7. See text.
8. See text.
9. Case B: Rheumatic heart disease with mitral regurgitation. About 8 million children are affected worldwide.
10. See text.