The cardiovascular system

3

This chapter presents a full account of the cardiovascular examination. However, in our experience, the candidate is usually requested only to examine a part of the system, e.g. 'feel the pulse', 'localize the apex beat' or 'listen to the heart'. You should therefore follow the examiners' instructions exactly in order to avoid antagonizing them.

INSPECTION

GENERAL

- General health, e.g. nutritional status, failure to thrive, tachypnoeic
- Dysmorphic features, e.g. Down's, Turner's, Noonan's or Williams syndrome.

FACE

Colour

Cyanosis Implies desaturated blood in the capillaries (>5 g/dL), giving the skin and mucous membranes a bluish discoloration, characteristic of right-to-left shunts within the heart or between the great arteries, or as a consequence of inadequate oxygenation of blood in the lungs. It corresponds to an arterial saturation of about 75% for a haemoglobin of 12–16 g/dL. Consequently, it can be present at a normal P_aO_2, if there is polycythaemia, or be difficult to detect if there is concomitant anaemia. It is clinically detected as follows:

- central (right-to-left shunts, cardiac or lung) – tongue
- peripheral (inadequate peripheral circulation) – nailbeds.

Pallor This is best detected in the oral mucosae, lips and conjunctivae. If associated with poor cardiac output, pulses may be weak and tissue perfusion poor.

Polycythaemia Often found in association with cyanotic congenital heart disease. These children have a high haematocrit and increased viscosity associated with an increased risk of cerebrovascular events.

Teeth

Comment on dental caries, indicating the importance of dental care as part of outpatient follow-up.

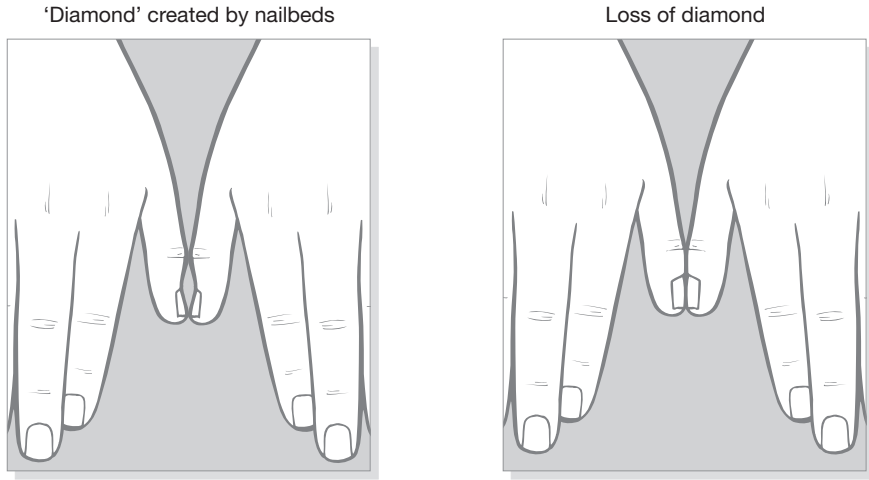


Fig. 3.1 Finger clubbing.

HANDS

Clubbing
Increased longitudinal and lateral curvature of the nails with loss of the acute angle between the proximal part of the nail and the skin, best seen at sites of largest surface area, such as thumbs and great toes (Fig. 3.1).

Bony abnormalities
- Absent radii – VACTERL syndrome
- Absent thumbs – Holt–Oram syndrome.

Rarities
- Splinter haemorrhages and Osler's nodes of infective endocarditis
- Tuberous and tendon xanthomata of familial hypercholesterolaemia. Feel over the elbows in a hypertensive child.

CHEST

Respiratory rate

Scars (Fig. 3.2)

Asymmetry
Look from the side in the same plane as the chest for:

- anterior bulge left chest – cardiomegaly
- left parasternal heave – right ventricular hypertrophy
- visible pulsations
- Harrison's sulci – in conditions with increased pulmonary blood flow or chronic asthma.

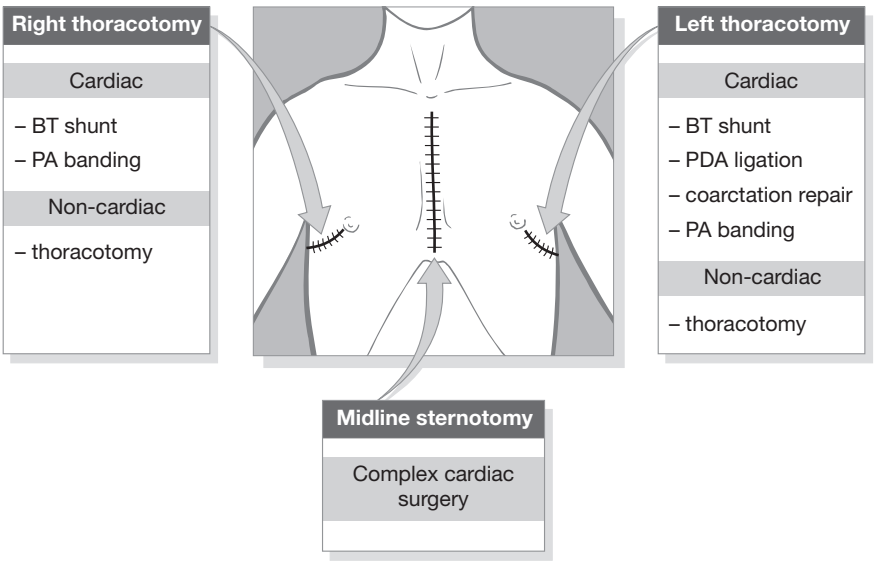


Fig. 3.2 Scars. BT, Blalock–Taussig; PA, pulmonary artery; PDA, patent ductus arteriosus.

PALPATION

PULSES

Brachial pulses

In young children the brachial pulse is the easiest to palpate. Ensure that both brachial pulses are present and equal in volume. If there is an *absent or reduced brachial pulse,* this is indicative of one of the following:

- classic left Blalock–Taussig shunt – absent left brachial pulse
- classic right Blalock–Taussig shunt – absent right brachial pulse
- left subclavian artery repair of coarctation – absent left brachial pulse
- flap aortoplasty repair of coarctation – reduced left brachial pulse
- previous cardiac catheterization – absent radial or brachial pulse
- cervical rib – either brachial pulse absent (especially on shoulder abduction)
- embolization
- congenital malformation – absent radial pulse.

Assess the following, using the right brachial pulse.

Rate Always count this over 10 seconds whilst deliberately looking at your watch. Never guess. Table 3.1 gives heart rates for healthy children. Abnormal rates could be:

- bradycardia (rare in exams)
  - — junior athletes!
  - — drugs (β-blockers and digoxin)
  - — complete heart block
- tachycardia – sinus tachy in anxious child.

Table 3.1 Heart rates in healthy children

Age (years)	Normal range (bpm)
0–2	80–140
2–6	75–120
>6	70–110

Rhythm

Regular
- Respiratory sinus arrhythmia (universal in young children).

Regularly irregular
- Pulsus bigeminous, coupled extrasystoles (digoxin toxicity).

Irregularly irregular
- Multiple extrasystoles – common in young children, they disappear on exertion
- Atrial fibrillation
  - — atrial septal defect (ASD)
  - — open heart surgery or atrial surgery
  - — Ebstein's anomaly of tricuspid valve
  - — rheumatic mitral stenosis (immigrant children only).

Check the apical rate by auscultation for the true heart rate as small pulses may not be transmitted.

Volume

Small volume
- Pump failure – heart failure
- Shock – circulatory failure due to hypovolaemia
- Outflow obstruction – aortic stenosis (AS) or pericardial effusion.

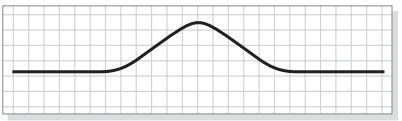
The first two are commoner in practice, but the third is commoner in exams.

Large volume
- Anaemia
- Carbon dioxide retention
- Thyrotoxicosis (very rare).

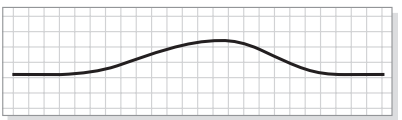
Varying volume
- Extrasystoles
- Atrial fibrillation
- Incomplete heart block.

Character The character of the pulse may be one of the following (see also Fig. 3.3):

- normal
- slow rising – moderate to severe aortic stenosis
- collapsing
  - — aortic incompetence (AI) (rare)
  - — patent ductus arteriosus (PDA) (large volume, rapid collapse – often a neonatal case)
- bisferiens – moderate aortic stenosis with severe aortic incompetence (very rare)

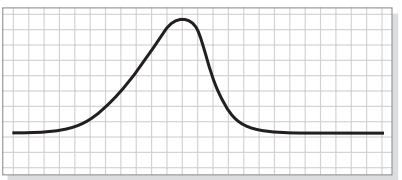


Normal



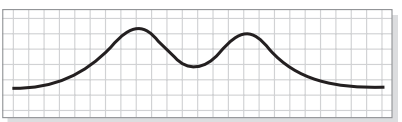
Slow rising

Moderate to severe aortic stenosis



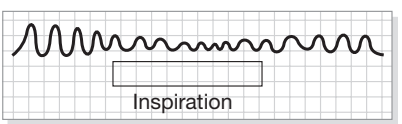
Collapsing pulse

Aortic incompetence (rare)
PDA (large volume, rapid collapse — often neonatal case)



Bisferiens

Moderate aortic stenosis with severe aortic incompetence (very rare)



Pulsus paradoxus

An exaggeration of a normal phenomenon, i.e. the fall in blood pressure on inspiration.

Fig. 3.3 Character of pulses.

- pulsus paradoxus – not a paradox at all but an exaggeration of a normal phenomenon, i.e. the fall in blood pressure on inspiration. If detected, offer to check by sphygmomanometry. A paradox of greater than 15 mmHg is abnormal. Causes include:
  — pericardial effusion
  — constrictive pericarditis
  — severe airways obstruction (asthma)
  (all are very unlikely in an exam).
- rapidly rising, ill-sustained, jerky – hypertrophic obstructive cardiomyopathy (HOCM).

Femoral pulses

Absence of femoral pulses is indicative of coarctation. This can be checked at the end of the examination. Radiofemoral delay is difficult to detect in children.

Suprasternal notch

Gentle palpation will detect a thrill in aortic stenosis. AS is a fairly common

exam case and is easily missed if you press too hard, so make sure you have seen at least one case *before* the exam.

The jugular venous pressure (JVP) is generally not an important part of the paediatric cardiovascular system. It can only be measured in older children and, whilst it is elevated in right heart failure, fluid overload and pericardial tamponade, none of these is likely in the exam.

BLOOD PRESSURE

Although you are rarely asked to do this in an exam, you must say you would do it and know how to do so if you are asked to measure it! The cuff must cover at least two-thirds of the upper arm, with a bladder that completely encircles the arm. In younger children, systolic blood pressure can be approximately determined by palpation of the brachial pulses as the cuff is deflated. In older children you must listen over the brachial pulse with a stethoscope. Record the blood pressure in the right arm and note whether the child is sitting, standing or supine. It is impossible to get accurate readings when a child is crying. Win cooperation by asking the child to 'see how strong you are' and by getting him/her to watch the mercury column rise and fall. Blood pressure varies with age but a rough guide is as follows:

- mean diastolic = 55 + age in years
- mean systolic = 90 + age in years.

The upper limits of normal are (mean + 20) mmHg for diastolic and (mean + 18) mmHg for systolic.

APEX BEAT

Position

This is described as the furthest lateral and inferior position at which the finger is lifted by the cardiac impulse, and is *normally the fourth intercostal space in the midclavicular line*. Always be seen to define the position by counting down from the second rib space which lies below the second rib (opposite the manubriosternal angle). Describe its position in relation to the midclavicular line, anterior and mid-axillary lines.

The beat may be:

- Displaced to the left
  - — cardiomegaly
  - — scoliosis
  - — pectus excavatum
- On the right side
  - — congenital dextrocardia: feel for the liver (Kartagener's syndrome)
  - — acquired dextroposition: heart pushed or pulled to the right
  - — left diaphragmatic hernia (rare in exams)
  - — collapsed lung on the right side (rare in exams).

Quality

- Sustained – with *pressure* overload in aortic stenosis
- Forceful – left ventricular hypertrophy
- Thrusting – with *volume* overload: an active large stroke volume ventricle in mitral or aortic incompetence, or left-to-right shunt

- Parasternal heave – right ventricular hypertrophy.

Thrills

The accompanying murmur is by definition at least 4/6 in intensity. Localize the site. For a *systolic thrill*:

- lower left sternal edge – ventricular septal defect (VSD)
- upper left sternal edge – pulmonary stenosis (PS).

Palpable heart sounds

A *second sound* reflects pulmonary hypertension.

NB. If you find an abnormality, think of possible causes before you listen, and what murmur you would expect to hear, e.g.:

- collapsing pulse – ?aortic incompetence
- suprasternal thrill – ?aortic stenosis.

AUSCULTATION

Listen over the four main areas of the heart whilst palpating the right brachial pulse with your left hand and in each area concentrate on:

- heart sounds
- added sounds
- murmurs.

Also present your findings in this manner, in order not to forget things.
The four main areas are (Fig. 3.4):

- apex (and axilla if there is a murmur)
- tricuspid area

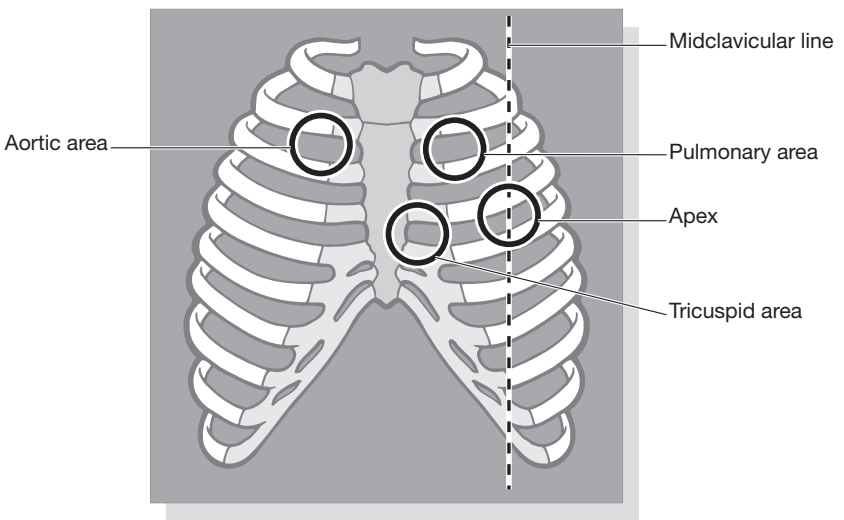


Fig. 3.4 Auscultation of praecordium.

- aortic area (and neck if there is a murmur)
- pulmonary area (listen over the back if there is a murmur).

Listen over the apex, first with the bell and then with the diaphragm of the stethoscope, and then continue with the diaphragm over the other areas. Always listen at the back – innocent murmurs do not radiate to the back. Murmurs of pulmonary stenosis radiate to the back. With an older, cooperative child, always listen again along the lower left sternal edge (LSE).

- Murmur loudest in expiration – left heart disease
- Murmur loudest in inspiration – right heart disease

HEART SOUNDS (Fig. 3.5)

Normal heart sounds

- First sound – sudden cessation of mitral and tricuspid flow due to valve closure
- Second sound – sudden cessation of aortic and pulmonary flow due to valve closure.

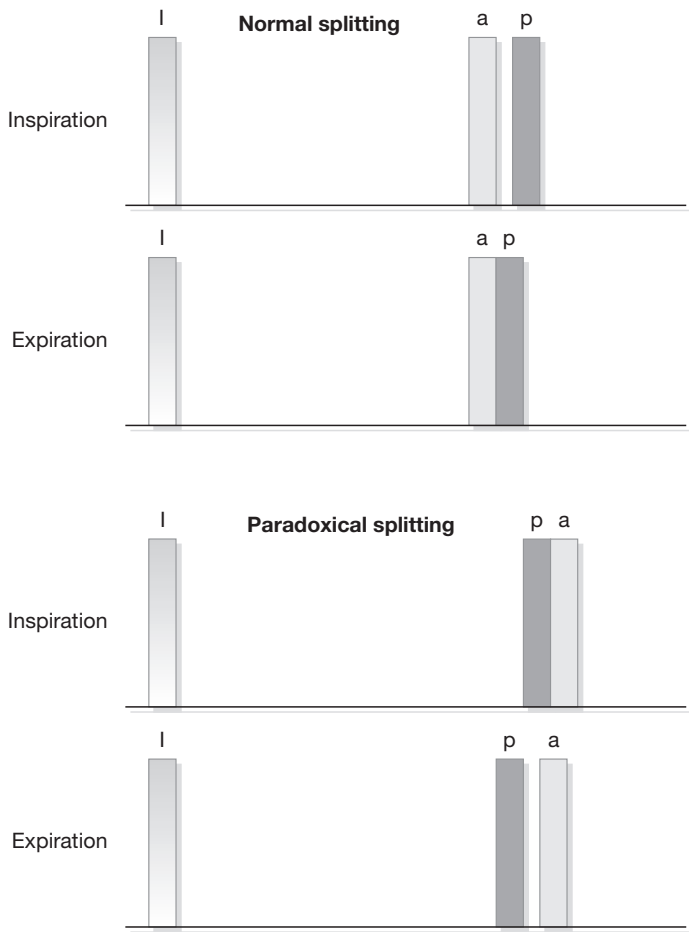


Fig. 3.5 Heart sounds. a, aortic valve; p, pulmonary valve.

Loud first sound
- ASD
- Mechanical prosthetic valve
- Mitral stenosis (MS) (very rare in paediatric exams).

Variable loudness of first sound
- Heart block
- Atrial fibrillation.

Loud second sound This is *very important* in paediatric cardiology. If it is of normal intensity and splits normally, many important conditions are excluded.

- Increased pulmonary flow – PDA, ASD, large VSD
- Pulmonary hypertension.

Split second sound
- Universal in healthy children and widens on inspiration. Aortic closure precedes pulmonary closure
- Fixed splitting (no change with respiration) – ASD
- Widely split – ASD, PS, right bundle branch block (RBBB)
- Reversed splitting (widens on expiration) – severe AS, left bundle branch block (LBBB).

Single second sound (inaudible pulmonary component)
- Tetralogy of Fallot
- Pulmonary stenosis.

ADDED SOUNDS

Third sound After the second sound, i.e. early diastole, low-pitched.

- Rapid ventricular filling, normal in healthy children
- Best heard with the bell over the apex
- May be confused with a split second sound or opening snap
- Heard in failure of either ventricle.

Fourth heart sound
- Never a normal finding
- Precedes first sound
- Failure of either ventricle
- Pulmonary hypertension.

Opening snap (Fig. 3.6)
- After second sound, high-pitched
- Mitral stenosis.

Ejection click (Fig. 3.6)
- After first sound, high-pitched, early systole
- Aortic or pulmonary stenosis.

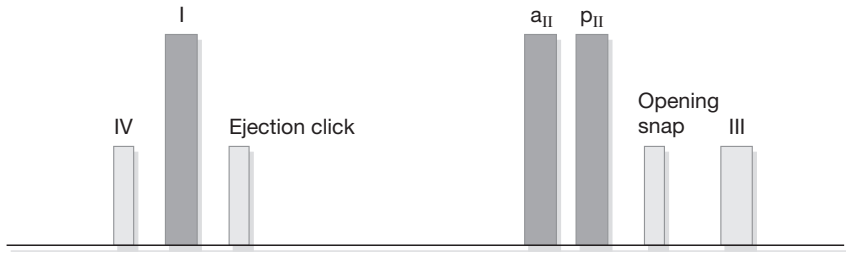


Fig. 3.6 Added heart sounds.

MURMURS

Try to define the following:

- Intensity (grades 1–6 if systolic, 1–4 if diastolic; grade 4 if thrill is palpable)
- Site where heard loudest
- Radiation
- Timing (systolic, diastolic or both)
- Duration (e.g. early diastolic or pansystolic)
- Pitch and quality (e.g. high or low, harsh or blowing)
- Changes with respiration or posture.

Remember to listen over the back (PDA, PS and coarctation).

Normal murmurs (previously called innocent or benign)
Cardiac murmurs are common in paediatrics. Accurate assessment of the child following the guidelines below will help to distinguish normal from pathological murmurs. Diagnosing a normal murmur positively rather than by exclusion will reduce unnecessary referrals and undue anxiety in the parents.

The '10 S' test of a normal murmur is as follows:

- **S**ymptom-free
- **S**ystolic
- **S**hort
- **S**oft
- **S**ite – heard over a small area only
- **S**plit second sound
- **S**itting/standing (i.e. varies with posture)
- **S**ternal depression (benign murmurs with pectus excavatum)
- **S**igns – no other abnormal signs, all pulses are normal
- **S**pecial tests (ECG and chest X-ray are normal).

There are basically five types of normal murmur which originate from increased flow velocity.

Still's murmur Early soft systolic murmur heard over the lower left sternal edge. Usually grade 2 in intensity but can be louder and often has a musical or buzzing quality to it. Murmur will decrease or disappear on hyperextension. Try it in an older, cooperative child.

Pulmonary flow murmur Soft ejection systolic murmur, usually ≤ grade 2, heard over the second left intercostal space. Rarely propagated posteriorly.

Can be confused with pulmonary flow murmur associated with ASD but there is no wide fixed splitting of second heart sound.

Venous hum Continuous murmur with diastolic accentuation heard below right clavicle and radiating to base. Often loud, grade 3, the intensity decreases when supine and can be obliterated by gentle neck compression. Still's murmur is often also present.

Supraclavicular or carotid bruit Best heard above the clavicles, although it transmits downwards.

Neonatal physiological peripheral artery stenosis murmur Maximal over upper left sternal edge and usually ≤ grade 2. Radiates throughout the thorax, to both axillae and to the back. Most disappear by 6 months of age and all have gone by 12 months.

Pathological murmurs

Using the following criteria – the seven cardinal signs – it is estimated that 95% of pathological murmurs would be identified:

- Pansystolic murmur
- Intensity ≥ grade 3
- Intensity maximal at upper left sternal edge
- Posterior propagation of murmur
- Harsh quality
- Early or mid-systolic click
- Abnormal second heart sound.

Classification of pathological murmurs

- Systolic (see Table 3.2)
- Diastolic (see Table 3.3)
- Continuous (see Table 3.4).

Table 3.2 Systolic murmurs

Cause	Type	Site
Innocent flow murmur	ESM	Left sternal edge or pulmonary area
Anaemia	ESM	Left sternal edge or aortic area
VSD	PSM	Left sternal edge, fourth intercostal space
PS	ESM	Pulmonary area, left second intercostal space
ASD	ESM	Pulmonary area, left second intercostal space
Aortic stenosis or bicuspid aortic valve	ESM	Aortic area, right second intercostal space to carotids in AS AS (rare), bicuspid valve (quite common) May radiate to carotids
Coarctation	PSM	Left sternal edge and between scapulae
HOCM	Late SM	Rare
Mitral regurgitation	PSM	Apex and left axilla
Mitral valve prolapse	Late SM	Apex

ESM, ejection systolic murmur; PSM, pansystolic murmur; HOCM, hypertrophic obstructive cardiomyopathy

Table 3.3 Diastolic murmurs

Cause	Site
ASD	Tricuspid flow murmur, low-pitched over sternal edge
VSD	Mid-diastolic mitral flow murmur with large defect
Mitral stenosis (rare)	Low-pitched at apex

Table 3.4 Continuous murmurs

Cause	Site
Innocent venous hum	Below either clavicle, may disappear when lying down or with legs elevated
PDA	Below left clavicle, radiates to back
Coarctation	Left sternal edge and between scapulae

Quality

- High frequency, blowing – mitral regurgitation (MR), aortic regurgitation (AR), pulmonary regurgitation (PR)
- Low frequency, harsh – AS, PS, VSD
- Lower frequency, rumbling – MS.

A summary of murmurs is presented in Figure 3.7.

ANYTHING ELSE?

- Feel for hepatomegaly
- Femoral pulses – do this at the end when examining babies as it is unpleasant and will make them cry
- Blood pressure – say you would like to do this if you have not already done so
- Height and weight – say you would like to plot these parameters on a growth chart appropriate for age and sex.

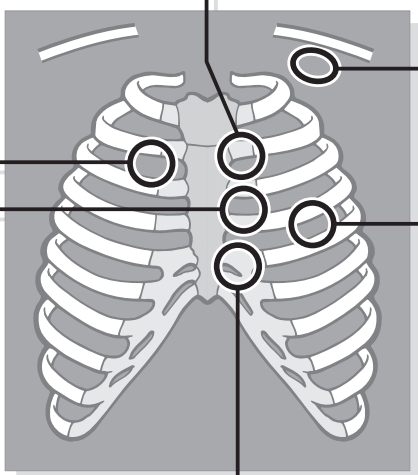
PRESENTATION OF HEART DISEASE

ASYMPTOMATIC MURMUR

- Neonatal check
- 6-week check
- Pre-school check
- Routine check with other illness.

Commonest causes

- VSD
- ASD
- PDA
- PS
- Coarctation
- AS.



URSE

Systolic
– aortic stenosis

Continuous
– right BT shunt

Venous hum

ULSE

Ejection systolic
– pulmonary stenosis
– aortic stenosis
– innocent

Left clavicle

Continuous
– patent ductus arteriosus

MLSE

Ejection systolic
– subpulmonary stenosis
– subaortic stenosis

Pulmonary stenosis
– ventricular septal defect
– Fallot's

Apex

Pulmonary stenosis
– mitral regurgitation
– ventricular septal defect

Late systolic
– mitral valve prolapse

Ejection systolic
– aortic stenosis

Mid-diastolic
– mitral stenosis

Back

Systolic
– coarctation
– peripheral pulmonary stenosis

Continuous
– patent ductus arteriosus
– ventricular septal defect

LLSE

Pulmonary stenosis
– ventricular septal defect
– tricuspid regurgitation

Diastolic
– tricuspid stenosis

Fig. 3.7 Summary of cardiac murmurs. URSE, upper right sternal edge; ULSE, upper left sternal edge; BT, Blalock–Taussig; MLSE, mid left sternal edge; LLSE, lower left sternal edge.

CYANOSIS

The child's age at presentation is important in determining the aetiology.

Presenting in first week of life – five Ts

- **T**ransposition of the great arteries – abnormal mixing
- **T**otal common mixing:
  - — total AV canal defect
  - — truncus arteriosus
- **T**otal pulmonary atresia – duct-dependent pulmonary circulation
- **T**ricuspid atresia – duct-dependent pulmonary circulation
- **T**ricuspid regurgitation and Ebstein's anomaly with right-to-left shunt via ASD.

Don't forget other causes of cyanosis in the neonatal period:

- respiratory
- persistant pulmonary hypertension of the newborn
- metabolic
- haematological
- sepsis.

Presenting after first week of life – two Ts

- **T**etralogy of Fallot (can present earlier if very severe)
- **T**otal anomalous pulmonary venous drainage (TAPVD).

HEART FAILURE

Less likely to be seen in an exam but you must know the causes, signs and symptoms.

Neonatal period (obstructed duct-dependent systemic circulation)

- Hypoplastic left heart syndrome
- Coarctation
- Critical aortic stenosis
- Tricuspid atresia
- Interrupted aortic arch.

Infancy

- VSD
- Atrioventricular septal defect (AVSD)
- Large PDA
- TAPVD.

Any age

- SVT
- Myocarditis
- Cardiomyopathy.

Signs

- Breathlessness
- Poor feeding
- Sweating
- Recurrent chest infections.

Symptoms
- Failure to thrive
- Tachypnoea
- Tachycardia
- Cardiomegaly
- Murmur/gallop rhythm
- Hepatomegaly
- Cool peripheries.

RARER PRESENTATIONS

Hypertension
Commonest causes are:

- cardiac – coarctation
- renal – reflux nephropathy secondary to urinary tract infection (UTI)
- catecholamine excess – neuroblastoma, phaeochromocytoma.

'Funny turns'

Cardiac arrhythmias

Presenting complaint
- Syncope – pallor
- Fits – blue.

Causes
- Supraventricular tachycardia (SVT)
- Prolonged PR interval – Lown–Ganong–Levene
- Prolonged QT syndrome
  - — Romano Ward (autosomal dominant (AD))
  - — Jervel–Lange–Nelson (autosomal recessive (AR) + deafness)

Cerebral events

Presenting complaint
- Fit
- Transient ischaemic attack (TIA)
- Stroke.

Causes
- Emboli – right-to-left shunt
- Thrombosis – polycythaemia
- Cerebral abscess.

Cyanotic spells
- Fallot's – infundibular spasm: 'spelling'.

Recurrent chest infections
Increased pulmonary blood flow/congestion.

- ASD
- VSD
- TAPVD.

Coincidental finding
- ECG – long QT interval
- Chest X-ray – cardiomegaly.

Subacute bacterial endocarditis
This is a very rare presentation.

COMMON LONG CASES

- Congenital heart disease
- Multisystem disorders.

CONGENITAL HEART DISEASE (CHD)

Certain congenital disorders are associated with heart disease.

Chromosomal abnormalities

- Down's syndrome (trisomy 21) – AVSD (30%), VSD, ASD
- Turner's syndrome (XO) – coarctation, aortic stenosis
- Cri-du-chat syndrome (5p–) – VSD
- Williams syndrome (microdeletion chr. 7) – supravalvular aortic stenosis, peripheral pulmonary stenosis
- Noonan's syndrome (AD, chr. 12) – pulmonary stenosis.

Intrauterine infection

- Rubella (esp. first trimester) – PDA, septal defects, peripheral pulmonary valve stenosis.

Maternal diseases

- Diabetes – increased incidence of all CHD, especially septal hypertrophy
- Systemic lupus erythematosus – congenital heart block.

Drugs in pregnancy

- Anticonvulsants – AS, PS, coarctation
- Excess alcohol – septal defects.

MULTISYSTEM DISORDERS

Some inherited causes of heart disease presenting in older children are listed below:

- Familial hypercholesterolaemia (AD) – hypertension, atherosclerosis, tendon xanthoma, corneal arcus
- Pompé's disease (type II glycogen storage disease, AD) – cardiomyopathy in infant/toddler
- Mucopolysaccharidoses (AR/X-linked) – storage material in valves may cause stenosis or regurgitation
- Marfan's syndrome (AD) – aortic regurgitation, mitral valve prolapse
- Ehlers–Danlos syndrome (AD) – aortic dissection
- Friedreich's ataxia (AR) – cardiomyopathy.

HISTORY (IMPORTANT POINTS)

Age of presentation

- Was CHD diagnosed antenatally?
- Congenital is more common than acquired

- Common associations with heart disease shown above
- 8% of CHD is associated with major chromosomal anomalies
- 10–15% is associated with non-cardiac anomalies
- If associated with a metabolic disorder, it will generally be picked up as part of routine screening once the underlying diagnosis is made. The child will usually have presented with other features of the underlying disorder.

How did it present?
- Symptomatic versus asymptomatic (see above).

Current symptoms
- Chronic limitation of exercise tolerance – quantify this
- How much school is missed?
- Headaches, 'funny turns', frequent chest infections, 'spelling'.

Family history
- Conditions associated with CHD
- Sudden/unexpected death at a young age
  - — hypertrophic obstructive cardiomyopathy
  - — arrythmias
  - — hypercholesterolaemia.

Treatment so far
- Cardiac catheterizations or surgery
- Admission for drug therapy (suggesting previous heart failure)
- Current medications?

Immunizations up to date?
- ***No vaccine is contraindicated*** in CHD per se
- Measles can be particularly serious in CHD.

EXAMINATION

This is as outlined above. *Don't forget to*:

- plot height and weight on growth chart
- measure blood pressure (upper right arm, and lower limb if there is coarctation)
- comment on dental caries.

INVESTIGATIONS

You must be able to discuss the logical sequence of investigations in a child with suspected heart disease.

Arterial blood gases (ABGs)
- Essential to confirm central cyanosis
- 'Hyperoxic/nitrogen washout' test – ABG is sampled from right radial artery to confirm central cyanosis; the child is then exposed to 100% oxygen for 10 minutes and the blood gas repeated (see Table 3.5).

Table 3.5 Cyanosis

Cause	Hyperoxic test
Lung disease (unless very severe)	P_aO_2 >15 kPa
Cardiac – transposition of the great arteries (TGA), truncus arteriosus (TA), pulmonary atresia (PA), large right-to-left shunt	P_aO_2 same
Common mixing (truncus arteriosus)	modest rise in P_aO_2

Chest X-ray
- Heart – size, shape, situs
- Valves – calcified/prosthetic
- Lungs – pulmonary oligaemia/plethora/vasculature
- Bony structures – rib notching (collaterals in coarctation).

Electrocardiogram
- Axis
- Conduction abnormalities
- P-wave abnormalities
- Ventricular hypertrophy.

Echocardiogram
- Detects most cyanotic conditions in the newborn
- Very useful for acyanotic conditions (septal defects, duct or valvular disease), particularly if accompanied by Doppler measurements of flow velocity.

Cardiac catheterization
- To measure pressure gradient across stenosed valve or outflow tract obstruction
- To quantify accurately the size of the shunt
- To determine the exact anatomy of complex lesions when surgery is considered
- For intervention by dilatation of valvular stenosis or coarctation.

TREATMENT

You would be expected to know how to manage the following.

THE BLUE BABY

- Oxygen – useless unless it has been demonstrated to improve P_aO_2.
- Prostaglandin
  - — Commenced if condition is 'duct-dependent', e.g. in:
    - — right ventricular outflow tract obstruction
    - — transposition of the great arteries (TGA)
    - — left ventricular outflow tract obstruction

  - Beware of side-effects:
    - hypotension
    - apnoea
    - fever
    - flushing
    - convulsions
- Correct acidosis
- Keep warm
- Prevent hypoglycaemia.

HEART FAILURE

Drugs

- Only if the child is symptomatic
- Diuretics – thiazide or loop diuretics are often used in combination with potassium-sparing to avoid the need for unpalatable potassium supplements
- ACE inhibitors – often used in conjunction with diuretics
- Digoxin – still widely prescribed although there is little evidence to support its use
- Dopamine – may be required if the child is hypotensive.

Feeding

- Passage of a nasogastric tube will reduce the work of breathing
- High-calorie feeds should be used with diuretics rather than fluid restriction as these babies often have high metabolic rates.

Monitoring

- Daily weights
- Assessment of liver size.

Ventilation May be required for severe heart failure and for apnoea secondary to prostaglandins.

Surgery Depends on the age of the child, but essentially there are two main reasons for performing surgery in the first year of life:

- severe heart failure with failure to thrive
- pulmonary hypertension with the potential to progress to pulmonary vascular disease.

ARRHYTHMIAS

Supraventricular tachycardia (SVT)

- The most common arrythmia in childhood
- Less than 25% have an underlying defect – commonly Wolff–Parkinson–White syndrome or Ebstein's anomaly
- If failure is evident, the arrythmia has been present for some time and treatment is urgent.

Treatment

- *Ventilation* – oxygen via mask or positive pressure
- *Circulatory support* – correction of acidosis
- *Vagal stimulation* – eyeball pressure, carotid sinus massage, submersion into ice-cold water should be tried although rarely successful in a very sick child; caution is required not to induce asystole, and thus it must always be done with a monitor attached
- *Adenosine* – this works by causing transient block of the AV node and may be of diagnostic and therapeutic benefit:
  - **diagnostic value:** if the dysrhythmia is atrial in origin, transient blockade of the AV node will slow the ventricular response to atrial tachycardia, atrial flutter and atrial fibrillation, which can then be diagnosed on the monitor. But use with caution, as there may be an accessory pathway in patients with atrial flutter or fibrillation and adenosine may increase conduction down anomalous pathways
  - **therapeutic value:** if the dysrhythmia is either AV nodal re-entry tachycardia or AV re-entry tachycardia, then adenosine may terminate the abnormal rhythm
  - **caution:** adenosine should NOT be used if the patient is on dipyrimadole, an anti-platelet drug, as dipyridamole will prolong the half-life of adenosine
- *Electrocardioversion with DC shock* – use in a severely ill child when the above has failed
- *Maintenance therapy* – digoxin or flecainide are effective.

ANTIBIOTIC PROPHYLAXIS OF ENDOCARDITIS

Prevention of endocarditis is necessary in patients with a heart-valve lesion, septal defect, patent ductus or prosthetic valve who are undergoing the following procedures:

- dental procedures, including local, general or no anaesthetic
- upper respiratory tract procedures
- genitourinary procedures
- gastrointestinal procedures
- (obstetric/gynaecological procedures).

A detailed description of antibacterial prophylaxis is given in the *British National Formulary*.

MANAGEMENT OF CONGENITAL HEART DISEASE

You need to be able to discuss the merits of medical versus surgical therapy and to be aware of current areas of debate, such as:

- How to manage VSD and when to close with surgery
- Management of ASD
- Medical or surgical treatment of PDA in the premature infant
- Management of TGA – anatomical correction versus balloon septostomy and Mustard procedure

- Total correction versus systemic to pulmonary shunt in Fallot's
- Correction of AV canal defects, especially in children with Down's syndrome.

COMMON CARDIOLOGY SHORT CASES

There is a plethora of complex congenital heart conditions but only nine common lesions, which can be categorized into acyanotic and cyanotic groups.

ACYANOTIC

Three 'holes' (left-to-right shunt)

- Ventricular septal defect (VSD)
- Atrial septal defect (ASD)
- Patent ductus arteriosus (PDA).

Three 'blocked pipes' (obstruction to flow)

- Pulmonary stenosis (PS)
- Coarctation of aorta
- Aortic stenosis (AS).

This group represents two-thirds of cases and the conditions are termed simple. The first three cases – left-to-right shunts – can lead to Eisenmenger's syndrome (pulmonary hypertension and reversal of the shunt, with consequent cyanosis). Any of the six can occur in combination but they usually occur in isolation, hence the term 'simple.'

CYANOTIC

Three 'blue babies'

- Transposition of the great arteries (TGA)
- Tetralogy of Fallot (TOF)
- Pulmonary atresia.

These account for the remaining one-third of the cases and the conditions are often complex lesions. By definition, there is a significant right-to-left shunt, or separate pulmonary and systemic circulation (transposition), but this is often complicated by other anomalies.

The algorithm for clinical examination and diagnosis (Fig. 3.8) may be helpful for the short cases as a guide to diagnosing the underlying heart disease. If you recognise the child as having an obvious syndrome then try to think of the likely underlying cardiac lesions and concentrate on the signs associated with that lesion during your examination.

Case histories of the common short cases can be found in more detail in *Paediatric Short Cases for Postgraduate Examinations* by A Thomson, H Wallace and T Stephenson (Churchill Livingstone, Edinburgh, 2003).

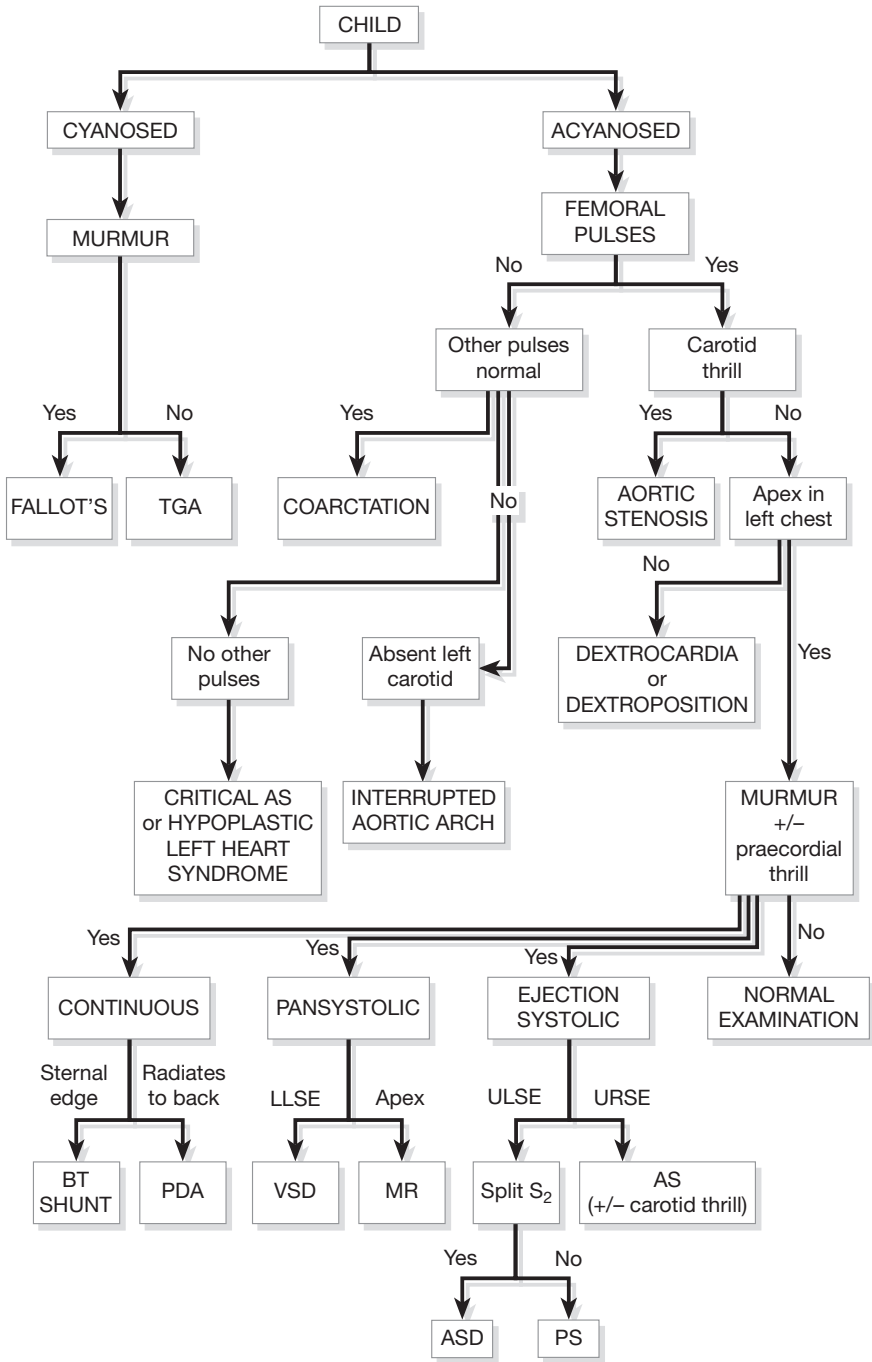


Fig 3.8 Algorithm for clinical examination.

SUMMARY OF CARDIOVASCULAR EXAMINATION

INSPECTION
Expose child appropriately and ideally position at 45°

Whole child
General health, nutritional status, dysmorphic features, sweating

Hands
Clubbing, peripheral cyanosis, xanthomas, splinter haemorrhages, absent thumbs, absent radii, abnormal palmar creases

Face
Plethoric, conjunctival injection, pallor, central cyanosis, teeth (conjunctival injection + gum hypertrophy = chronic cyanosis)

Chest
Respiratory rate, scars (thoracotomy = operations outside heart, sternotomy = intracardiac), symmetry – look from the side, deformity – Harrison's sulci, visible pulsation

PALPATION

Pulses
Both brachial and femoral (can do at the end), rate (count for 6 seconds then multiply by 10), quality, rhythm

BP
'I would like to measure the blood pressure at the end'

Apex
Locate apex beat (most lateral and inferior impulse) and count ribs to check position, normally fourth intercostal space in midclavicular line, nature of impulse; sustained in AS, forceful in LVH

Praecordium
Thrills or heaves, palpable P_2 in pulmonary hypertension
Suprasternal notch: thrill = aortic stenosis

AUSCULTATION

Heart sounds

- Loud S1: ASD, prosthetic valve,
- Loud S2: increased pulmonary blood flow (PDA, ASD, VSD), pulmonary hypertension
- Split S2: fixed split (ASD), wide split (ASD, PS, RBBB), reversed split (AS, LBBB)
- Single S2: tetralogy of Fallot, PS
- Extra HS: ejection click (AS/PS), mitral valve prolapse

Murmurs
Grade, timing, character, quality, position of maximum intensity, radiation (see later)

Back
Listen for murmurs and inspiratory crackles if in failure

ANYTHING ELSE?
Blood pressure
Femoral pulses
Feel for hepatomegaly
Plot height and weight on a growth chart appropriate for the patient's age and sex

INVESTIGATIONS
Saturation monitor, ABG, ECG, CXR, ECHO, cardiac catheterization