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   Fallot’s tetralogy
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Surgery for congenital heart disease
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8.1 INTRODUCTION

Preparation

Cardiology short cases are a hard part of the exam. In this chapter we have tried to give useful information and to highlight some of the important areas and more common conditions seen. It is essential to have a good system of cardiovascular examination and some background knowledge (read through paediatric cardiology in one of the standard texts) to help with the interpretation of physical signs. The best places to see children with signs are on the cardiology ward (pre-elective operation) and in the cardiology clinic. These children will be most representative of the cases you are likely to see in your exam. You should ask a cardiology specialist trainee or consultant to critique your examination technique and point out to you how they would best elicit the relevant signs. There are many important physical signs that you may never have seen outside of a specialist clinic/ward. They are often surprisingly easy to elicit and once seen they are often not forgotten.

During the examination

It is important to adapt your examination, if appropriate, and to be responsive to whatever the examiner asks. Clearly, if the examiner asks you to do cardiovascular examination then the full examination should be done. If you are asked to examine the heart then it is less clear. There are different strategies to deal with this although no consensus view. It is, however, in our experience often best to do the full cardiovascular examination unless the examiner specifically tells you to take short cuts. If the examiner asks you just to listen to the heart then that may be all he/she expects in that instance. If you are unclear what is expected of you, then it is perfectly reasonable to check with the examiner directly.

It is not unreasonable in the case of a small child to move straight on to auscultation at the very beginning of the examination if the patient is quiet, in order to listen to heart sounds and murmurs before the child becomes upset. It is probably best to explain yourself to the examiner if you perform the examination this way. Remember to complete the examination.

Taking blood pressure and checking jugular venous pressure (JVP; see Section 8.4) are part of a complete examination and you should
include them in all your practice cases so that you get used to doing them quickly under pressure for children at a variety of ages. Because they can often take up a disproportionate amount of time and are so often normal it may be acceptable to leave them to the end, but you should make every effort to let the examiner know that you haven’t overlooked them and give the examiner the prerogative to stop you from attempting them if he or she does not think they are relevant. If in doubt – do it!

Particularly with the cardiovascular system, there are many signs that have the potential to give very accurate diagnostic information, but require years of experience to be fully appreciated, such as split second heart sounds and abnormal JVP waveforms. As a general rule, if a sign is not obvious it is probably not there or not worth mentioning. Don’t spend so long on these aspects of the examination that you neglect the bigger picture.

In most short case scenarios it is sensible to talk your way through the examination of the case. In cardiovascular system examination however, particularly if the diagnosis is not obvious, then it is wise to keep your own counsel; although clearly if you see something obvious, eg Down syndrome, cyanosis or chest wall scarring, it is appropriate to mention it. Many of the signs you discover will be much easier to interpret once you have gathered all the information, particularly when it comes to interpreting murmurs. You would be wise to not volunteer any information about heart sounds and murmurs until you have listened as fully as you intend to, including checking for radiation and response to manoeuvres. This will give you the maximum time to assimilate all the information and put it together into a cohesive, reasonable sounding interpretation.

The keys to success in this station are to have a well-practised fluent technique, to be able to detect the relevant signs with confidence, to be able to formulate them into a sensible differential diagnosis and to have enough background knowledge of the more common conditions to be able to answer some questions about the significance of your findings and further management.
8.2 CLINICAL EXAMINATION

Suggested approach to cardiovascular examination

Examine this child’s heart

This boy had a cardiac procedure as an infant, please examine his cardiovascular system

Initial procedure

- Introduce yourself to the parents and the child

General observations

- Age
- Growth: more than just worth mentioning, growth is crucial to determining management in congenital heart disease
- Colour: pink, pale or blue (or jaundice as in Alagille’s)
- Dysmorphic features (could give clue to underlying cardiac lesion)
- Monitoring (may have oxygen saturation probe)
- Supplemental oxygen
- Increased work of breathing (recession, tachypnoea)

Hands to arms

- Perfusion: cool or warm hands
- Clubbing: if you are unsure whether it is present, don’t spend a long time looking for it, as it may not be significant enough to comment on. Best to look for ‘loss of diamond’ when comparing index fingers (first sign of clubbing). Clubbing is present after long-standing arterial desaturation (over 6 months). Could be present in pink, well-saturated children if they have recently had corrective cardiac surgery (or if it has a ‘non-cardiac’ cause)
- Evidence of endocarditis: see notes below
- Radial pulse (less relevant in babies) and brachial pulse: compare both sides
  - Make a show of counting pulse rate
  - Comment on rate (and whether this is normal for this age group; see Box), rhythm, character
Hyperconvex/hypoplastic nails | Turner syndrome
Short fourth and fifth metacarpals | Turner syndrome
Single palmar crease | Down syndrome
Long tapering fingers | DiGeorge syndrome
Arachnodactyly | Marfan syndrome
Radial abnormalities causing thumb/wrist dysmorphism | VACTERL syndrome
Overlapping fingers | Trisomy 18
Polydactyly | Trisomy 13

Table 8.1: Syndromes with hand abnormalities and heart defects

<table>
<thead>
<tr>
<th>Normal ranges for heart rate (resting) in beats per minute</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neonates</td>
</tr>
<tr>
<td>2 years</td>
</tr>
<tr>
<td>4 years</td>
</tr>
<tr>
<td>6 years and over</td>
</tr>
</tbody>
</table>

- **Brachial/carpotid pulse characters**
  - **Collapsing**: run-off lesions, classically aortic regurgitation, could also be seen in patent ductus arteriosus, large arteriovenous fistula
  - **Thready**: congestive cardiac failure, circulatory shock
  - **Asymmetrical**: post-coarctation repair (using L subclavian flap)
  - **Slow rising**: flattened pulse amplitude, implies impaired ejection from left ventricle (eg aortic stenosis)
  - **Pulsus paradoxus**: cardiac tamponade/pericarditis, severe asthma/high intrathoracic pressures

**Tip**
Don’t suggest the pulse character is abnormal unless it is very obviously abnormal. If you are unsure, it is less likely to be significant and you should move on
Blood pressure

You should make a judgement about how quick and easy it would be to check the BP manually at this point. In older children this can be done rapidly and you should do it. In younger, less co-operative children it can take a while to do this but it is still an essential part of a complete, structured examination. If you plan to come back to BP at the end then make sure the examiners are aware of this or it may appear that you have simply forgotten. You will need to practise doing this on children of various ages so that you don’t take too long over it in the exam. Your approach should be:

- Explanation
- Position child as you want them to be
- Select correct cuff size
- Inflate the cuff whilst palpating the brachial/radial pulse until the pulse disappears
- Inflate the cuff a further 10 mmHg
- Place the stethoscope over the brachial artery
- Deflate the cuff slowly until regular sounds (the Korotkov’s sounds) can just be heard through the stethoscope
- Note the systolic pressure as accurately as possible
- Continue to deflate the cuff slowly; the sounds get louder then suddenly become muffled and disappear
- Record the point at which the sounds become muffled as the diastolic pressure
- Interpret the results

All children need an explanation of what is about to happen. This should start with ‘have you had your blood pressure checked before?’ Young children need very careful reassurance and this should be well practised. It is likely that if the examiner has indicated that you should take the blood pressure of a young child then there will be a lot riding on your sensitive, paediatric approach.

Avoid cuffs that are too small as they may cause a falsely high BP. Cuff width should be approximately two-thirds of the length of the upper arm (or 50% of the circumference of the arm), and the bladder should encircle the limb almost completely.

Technically you should take at least two readings before you say what the blood pressure is. This is probably only necessary if you have been specifically asked to check the BP. You should let the examiner know what the result was from the first reading and then say ‘I would normally confirm this with a second blood pressure reading’

In reality BP is often measured by a Doppler machine or Dinamap®. If you are not familiar with how to use these you should ask one of the nursing
staff to teach you. Don’t be caught out by not being able to use a simple piece of equipment like a blood pressure monitoring machine!

Interpreting whether BP readings are normal is difficult. There are published normal ranges. As a rough guide a systolic BP should be no more than 90 plus double the child’s age. If you find a BP that is outside this range you should comment on the fact that it is not a normal reading and that you would check the value against a table of normal ranges. Table 8.2 is a rough guide. Ideally you should use a table that provides information about maximum normal blood pressures at different ages and height centiles. Remember particularly if the reading is abnormal to check a second one.

### Table 8.2: Approximate values for blood pressure in children

<table>
<thead>
<tr>
<th>Age</th>
<th>Mean BP should be equal to or above gestational age (e.g. mean of &gt;40 mmHg for term infant)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newborn</td>
<td></td>
</tr>
<tr>
<td>Infancy</td>
<td>80–95 mmHg systolic</td>
</tr>
<tr>
<td>Preschool</td>
<td>80–100 mmHg systolic</td>
</tr>
<tr>
<td>School age</td>
<td>90–110 mmHg systolic</td>
</tr>
<tr>
<td>&gt;12</td>
<td>100–120 mmHg systolic</td>
</tr>
</tbody>
</table>

Face and neck

- Conjunctival pallor (approach with care)
- Central cyanosis – becomes clinically apparent when saturations are below 85%. This is not necessarily true if the patient is anaemic or dark skinned. You should say that the patient is not clinically cyanosed unless they are very obviously blue. In practice, for the purposes of the exam, if you are not sure and the patient is not clubbed, do not spend a long time looking.
- Dental caries if present
- Carotid pulse character
- Jugular venous pressure – in older children (see notes below)

Chest

Inspection

- Expose fully wherever possible
- Shape (chest wall deformities)
- Comment on any visible impulses (could indicate hyperdynamic apex beat)
- Scars (see Box)
### Causes of chest wall scars

**Right lateral thoracotomy**
- Modified BT shunt
- Lung causes (lobectomy)
- Tracheo-oesophageal fistula repair

**Left lateral thoracotomy**
- Modified BT shunt
- Coarctation repair
- Patent ductus arteriosus ligation
- Pulmonary artery banding
- Lung causes (lobectomy)

**Median sternotomy**
- Any bypass surgery (most corrective surgery of intracardiac abnormalities requires bypass)

**Chest drain scars**
** Mediastinal drain scars**
**Chest wall pacemakers** (often in left pectoral region)

### Palpation
- Apex beat (right or left): the first and most important thing is to look for dextrocardia at this point; if you miss it now it will be difficult not to look foolish when you eventually realise. Place a hand on each side of the chest and remove the hand that hasn't got the strongest impulse underneath it (if you detect dextrocardia you must remember to look for associated abnormalities/Kartagener's syndrome).
- You can then proceed to determine:
  - Site: normal (4th or 5th intercostal space, midclavicular line) or displaced
  - Character: hyperactive characterises lesions with volume overload (large left to right shunt/valvular regurgitation)
  - Right ventricular heave: a palpable right ventricular impulse indicative of high right-sided pressures (for example, right ventricular outflow tract obstruction)
- Thrills
  - Upper left sternal border – pulmonary stenosis (rarely patent ductus arteriosus)
  - Upper right sternal border – aortic stenosis
- Lower left sternal border – ventricular septal defect
- Suprasternal notch – aortic stenosis (occasionally pulmonary stenosis, patent ductus arteriosus, coarctation of the aorta)
- Carotid arteries – aortic stenosis (occasionally coarctation of the aorta)

Auscultation

Listen in four areas:

1. Mitral (apex)
2. Tricuspid (lower left sternal edge)
3. Aortic (right second intercostal space)
4. Pulmonary (left second intercostal space)

Several candidates have been asked to point out the areas to the examiner.

*Remember this is on the assumption that the heart is structurally normal and dextrocardia is not present.*

Heart sounds

Take note of these before assessing the nature of any murmur

- Are they both present?
- First heart sound – loud when cardiac output high, eg when nervous or excited, anaemia and pyrexia. Soft when cardiac output is low or there is poor left ventricular function
- Is the second heart sound normal?
- If the second heart sound is louder than the first when listening over the apex, then this could indicate pulmonary hypertension

The following are classically heard by consultant cardiologists and medical students but very rarely by the rest of us. Once again, if it is not obvious don’t spend a long time looking for it, as it is unlikely to be significant for the purposes of the exam. You should however know the significance and implications of the following and how to detect them if they are there:

1. Splitting of second heart sound
   - Pulmonary valve closes later than aortic valve in the normal cardiac cycle
   - Best listened for at the upper left sternal border
   - Increased separation between sounds in inspiration and decreased separation (becoming single second heart sound) in expiration are normal
   - Widely split: classically occurs in atrial septal defect (where the right ventricle is overfilled and pulmonary valve closure is delayed), mild pulmonary stenosis and right bundle branch block
8: Cardiovascular

- Fixed splitting of the second sound (atrial septal defect): when there is no variation with respiration, the two atria are functioning as one and respiration has the same effect on both the pulmonary and systemic circulations.
- Single S2 occurs when there is pulmonary hypertension (pulmonary valve closes quickly), transposition of the great arteries or abnormal aortic or pulmonary valves (atresia or severe stenosis).
- Paradoxical splitting (aortic valve closes last) – left bundle branch block, severe aortic stenosis.

2. Third heart sound ('Tennessee')
   - Often heard in normal children.
   - Can indicate reduced ventricular compliance.
   - Best heard with bell over apex/lower left sternal border.

3. Fourth heart sound ('Kentucky')
   - This occurs shortly before the first heart sound.
   - Almost always pathological but rare in children.
   - Indicates reduced ventricular compliance.

4. Gallop rhythm
   - Implies loud S3 or S4 and tachycardia.
   - Always pathological.

5. Ejection clicks
   - Classically occur in aortic valve and pulmonary valve stenosis, but may also be heard when there is a large dilated aorta in tetralogy of Fallot, coarctation of the aorta, patent ductus arteriosus or when there is a dilated pulmonary artery in pulmonary hypertension. The sound is made by the forceful opening of the valve.
   - Aortic click – lower left sternal edge and apex.
   - Pulmonary click – second left intercostal space.

Murmurs

Make sure you have fully assessed a murmur before you comment on it. A full assessment consists of the following six components:

1. Timing in relation to cardiac cycle: patterns of murmur are given in Table 8.3.
### Patterns of Murmur

<table>
<thead>
<tr>
<th>Systolic</th>
<th>Diastolic</th>
<th>Continuous</th>
</tr>
</thead>
</table>
| Ejection systolic (crescendo/decrescendo)  
• Could be innocent  
• Aortic stenosis  
• Pulmonary stenosis  
Panystolic  
• VSD  
• Mitral regurgitation  
• Tricuspid regurgitation  
Systolic click (occurs midsystole) with late systolic murmur  
• Mitral valve prolapse  
Ejection click (occurs early in systole)  
• Aortic valve disease | Early diastolic murmur  
• Aortic regurgitation (best heard at LLSB, with bell, in expiration, sitting forward)  
Mid diastolic  
• Mitral/tricuspid stenosis  
Opening snap/diastolic rumble  
• Mitral stenosis | • PDA  
• ASD  
• Blalock–Taussig shunt  
• AV malformation  
• Aneurysm  
• Collateral vessels  
• Venous hum  
• Peripheral pulmonary stenosis  
• Aortopulmonary window |

### Sites of Maximal Intensity

**Table 8.3:** Patterns of murmur. ASD, atrial septal defect; AV, arteriovenous; LLSB, lower left sternal border; PDA, patent ductus arteriosus; VSD, ventricular septal defect

2. Site of maximal intensity (see Table 8.4)  
*Remember this is on the assumption that the heart is structurally normal and dextrocardia is not present.*

<table>
<thead>
<tr>
<th>Upper left sternal edge = pulmonary area</th>
<th>Upper right sternal edge = aortic area</th>
<th>Lower left sternal edge</th>
<th>Apex</th>
</tr>
</thead>
</table>
| • Pulmonary stenosis  
• Pulmonary flow murmur  
• Pulmonary artery stenosis  
• PDA  
• RVOT obstruction (e.g. Fallot’s) | • Aortic valve stenosis  
• Subaortic stenosis  
• Supravalvular aortic stenosis | • Aortic regurgitation (loudest over 3rd or 4th intercostal space)  
• VSD  
• AVSD  
• Vibratory innocent murmur (Still’s)  
• HOCM  
• Tricuspid regurgitation | • MR  
• Mitral stenotic murmurs  
• Mitral valve prolapse |

**Table 8.4:** Sites of maximal intensity. AVSD, atrioventricular septal defect; HOCM, hypertrophic obstructive cardiomyopathy; MR, mitral regurgitation; PDA, patent ductus arteriosus; RVOT, right ventricular outflow tract; VSD, ventricular septal defect
3. Character: high or low pitched/musical/vibratory/blowing

4. Murmur-enhancing moves:
   - Left lateral position – brings out S3 and mitral murmurs
   - Sitting forward – brings out aortic murmurs
   - Change with inspiration/expiration – expiration brings out left-sided murmurs
   - Valsalva – for hypertrophic obstructive cardiomyopathy

5. Radiation (neck, axilla and back)
   - To neck – aortic in origin
   - To back/sides of chest – pulmonary in origin (eg pulmonary stenosis, branch pulmonary artery stenosis, right ventricular outflow tract obstruction/infundibular stenosis as in Fallot’s)
   - To axilla – pulmonary valve murmurs or mitral regurgitation
   - Look for murmur over the back at this stage (for coarctation – just below tip of left scapula)

6. Grade (I–VI)

### Grading of Murmurs

<table>
<thead>
<tr>
<th></th>
<th>Barely audible</th>
</tr>
</thead>
<tbody>
<tr>
<td>III</td>
<td>Medium intensity</td>
</tr>
<tr>
<td>IV</td>
<td>Loud but no thrill</td>
</tr>
<tr>
<td>V</td>
<td>Loud with a thrill</td>
</tr>
<tr>
<td>VI</td>
<td>Very loud but still requires stethoscope to be on the chest</td>
</tr>
<tr>
<td></td>
<td>So loud, can be heard with stethoscope off the chest</td>
</tr>
</tbody>
</table>

Auscultate basal lung fields before moving on.

**Abdomen**

- Hepatomegaly
- Situs solitus (midline liver) or situs inversus: associated with atrial isomerism; association with malrotation so may have evidence of Ladd’s procedure scar
- Abdominal pacemaker: epigastric scar; may not be able to feel pacemaker box
- Scars from postoperative peritoneal dialysis (sometimes necessary for fluid/ascites management post bypass surgery)
- Ascites
Evidence of protein-losing enteropathy (can occur after bidirectional Glenn procedure)

- Splenomegaly (occurs in infective endocarditis)

Groin

Announce that you are about to do this and remember to preserve modesty in older children

- Scars from cardiac catheterisation/central lines
- Femoral pulses: compare character with upper limb pulses and look for radio-femoral delay (a sign that collateral circulation has developed to bypass an aortic coarctation)

Don’t forget to mention

- Blood pressure if not already done
- Jugular venous pressure if not already done
- Femoral pulses if not already done
- Check growth chart
- Peripheral or sacral oedema
- Fundoscopy/dipping urine for evidence of infective endocarditis
### 8.3 Cases

The following are representative of some of the cases that you may see in the exam.

**Case one: Complex cyanotic congenital heart disease**

*Patient: 8-month-old girl. She appears small for her age although I would like to plot her on a centile chart. She is tachypnoeic at rest. She is centrally cyanosed. She has no dysmorphic features. She is clubbed and has peripheral cyanosis. She has an absent right brachial and radial pulse, there is no radio-femoral delay. She has a right-sided thoracotomy scar. On auscultation she has a normal first heart sound and a single second heart sound. She has a continuous murmur over the right side of her chest. (At the end of this examination you should suggest palpating the abdomen for a liver and taking the blood pressure.)*

This set of clinical signs would fit with a child who has a complex cyanotic congenital heart lesion and has had a shunt procedure.

**Notes**

Possible diagnoses include:

- Pulmonary atresia without a ventricular septal defect
- Double outlet right ventricle
- Univentricular heart with pulmonary atresia

You would not be expected to come up with that diagnosis, but carefully eliciting the clinical signs will allow a best guess. In this case, with the shunt and the single second heart sound, a diagnosis of pulmonary atresia would be the first best guess. Fallot’s is less likely because there is no systolic murmur of pulmonary stenosis and in pulmonary stenosis there should be two components of the second sound.
Case two: Trisomy 21, with AVSD and Eisenmenger’s syndrome

**Patient:** 13-year-old girl. She has facial features of Down syndrome. She is deeply cyanosed at rest. She is clubbed and peripherally cyanosed. Her pulses are normal in character and the rate is 95 beats per minute. There is no radio-femoral delay. There are no scars visible on the thorax. Her apex beat is not displaced. She has a right ventricular heave. On auscultation she has a normal first heart sound, a loud second heart sound and a pansystolic murmur, maximal at the left sternal edge but heard all over the chest. I would like to go on to palpate her abdomen for a liver.

This young girl with Down syndrome is most likely to have atrioventricular septal defect with shunt reversal resulting in Eisenmenger’s syndrome.

**Notes**

Down syndrome children who have no scars and are cyanosed at the age of 13 are likely to have a defect causing Eisenmenger’s – be prepared to talk about Eisenmenger’s syndrome and its prognosis. This child could just as well be seen in the History Taking/Management Planning Station or the Communication Station.
Case three: Cavopulmonary shunt

Patient: 20-month-old girl. She appears to be thriving although I would like to plot her on a centile chart. She has no dysmorphic features and appears to be comfortable at rest. She is mildly cyanosed centrally. Her pulses are normal throughout. She has no radio-femoral delay. She has a median sternotomy scar but no thoracotomy scars. Her apex beat is localised to the 5th intercostal space on the right; there are no heaves or thrills. She has normal heart sounds on the right side and appears to have dextrocardia. There is a continuous murmur.

This set of clinical signs would fit with a child who has a complex congenital cardiac defect and who is likely to have had a central shunt procedure, eg cavopulmonary anastomosis.

Notes

If the heart sounds are difficult to hear, as a matter of routine listen to the right side of the chest to exclude dextrocardia. Mild cyanosis in a child with a median sternotomy scar suggests a palliative shunt procedure for those children for whom definitive surgery is not possible. The child described had a complex cardiac defect involving right atrial isomerism, dextrocardia, a complete AV canal defect and a single ventricle with pulmonary atresia. A precise diagnosis would not be expected.
Case four: Innocent murmur

Patient: Jack, aged 4. He looks well and is thriving. There is no clubbing. His pulse and blood pressure are normal. There are no scars. His apex beat is normal and in the 5th left intercostal space. There are no heaves or thrills. There is a 2/6 ejection systolic murmur, maximal in the pulmonary area, which does not radiate. The murmur is quieter when the child sits forward. The heart sounds are normal. Chest X-ray and ECG are normal.

This is likely to be a pulmonary flow murmur.
Case five: Complex congenital cardiac disease

Patient: 3-year-old boy. He looks well and is not tachypnoeic at rest. His right radial pulse is absent. He has a scar from cardiac catheterisation. He has a right thoracotomy scar and midline sternotomy scar. I presume he has complex congenital heart disease, now repaired, and I would like to proceed to listen to the heart …

It is not possible to achieve a specific diagnosis in this case.