5.4 Cardiac disorders

5.4.A Congenital and rheumatic heart disease

**BOX 5.4.A.1 Minimum standards**
- Electrocardiograph.
- Chest X-ray.
- Prostaglandin E.
- Oxygen.
- Beta-blockers.

Introduction

Congenital heart disease occurs in 5–8 in 1000 live births.

Every country should have immediate access to a hospital that can surgically correct the easily curable acquired or congenital heart defects. The reality is very different with more than 90% of countries without access to such facilities.

Investigations or treatments, which are likely to be unavailable or irrelevant in the absence of a specialist cardiac centre are highlighted.

**TABLE 5.4.A.1 Percentage frequency of common congenital heart defects in the UK**

<table>
<thead>
<tr>
<th>Heart Defect</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect (VSD)</td>
<td>32%</td>
</tr>
<tr>
<td>Persistent ductus arteriosus (PDA)</td>
<td>12%</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>8%</td>
</tr>
<tr>
<td>Atrial septal defect (ASD)</td>
<td>6%</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>6%</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>6%</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>5%</td>
</tr>
<tr>
<td>Transposition of the great arteries</td>
<td>5%</td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>3%</td>
</tr>
<tr>
<td>Atrioventricular septal defect (AVSD)</td>
<td>2%</td>
</tr>
</tbody>
</table>

Cardiac defects may present as any of the following:
- cyanosis in the newborn period
- cyanosis in the older infant
- cardiovascular collapse in the newborn period
- cardiac failure in infancy
- an asymptomatic murmur.

This section explains how to recognise the presence of congenital heart disease in each of these clinical scenarios, and provides enough information to make a working diagnosis. Management decisions can then be made when modern imaging techniques are not immediately available.

**The cyanotic newborn**

**Is there a cardiac problem?**

When a child is referred as a ‘blue baby’, first check to see whether there is genuine central cyanosis. Examine the lips and tongue for blue discoloration, and confirm the clinical impression by measuring the oxygen saturation (less than 94% is abnormal). If there is central cyanosis this may have a cardiac or respiratory cause.

**TABLE 5.4.A.2 Features that distinguish cardiac from respiratory cyanosis in the neonate**

<table>
<thead>
<tr>
<th>Cardiac cyanosis</th>
<th>Respiratory cyanosis*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild tachypnoea but no respiratory distress</td>
<td>Respiratory distress</td>
</tr>
<tr>
<td>May have cardiac signs on examination</td>
<td>Chest X-ray: abnormal lung fields</td>
</tr>
<tr>
<td>Arterial blood gas: pO2 ↓, pCO2 ↓ or normal</td>
<td>Arterial blood gas: pO2 ↓, pCO2 ↑ or normal</td>
</tr>
<tr>
<td>Fails hyperoxia test</td>
<td>Passes hyperoxia test</td>
</tr>
</tbody>
</table>

*A respiratory cause for cyanosis is more likely in infants born preterm.

The hyperoxia test is performed as follows:
1. Ensure that there is good IV access.
2. Monitor oxygen saturations continuously.
3. Give 100% oxygen for 10 minutes.
4. Take an arterial blood gas sample in the right arm (preductal).
   - If pO2 is lower than 20 kPa (150 mmHg), a cardiac cause of cyanosis is likely (the test is ‘failed’).
   - If pO2 is higher than 20 kPa, a respiratory cause of cyanosis is likely (the test is ‘passed’).
   - Although pulse oximetry cannot reliably be used in place of an arterial blood gas, a resting saturation of less than 80% and a saturation of less than 90% after 10 minutes in 100% oxygen suggests cyanotic heart disease requiring early intervention.

**Note:** Oxygen administration could cause closure of the arterial duct, precipitating profound hypoxaemia in some types of cyanotic congenital heart disease.

- Prostaglandin E (which opens the duct) should therefore be available at the time of the test and should be given if oxygenation deteriorates.

Persistent pulmonary hypertension of the newborn (PPHN) may often mimic cyanotic heart disease using these clinical criteria. However, PPHN is usually distinguished by a history of fetal distress, resuscitation is often needed at birth, and there may be neurological signs. Improvements
in oxygenation may be possible after intubation and ventilation, and saturations in the right arm may be significantly higher than those in the feet, suggesting right-to-left shunting across the arterial duct.

What type of cardiac defect is present?
Cyanotic cardiac defects can be divided into three broad categories, as described below.

Once cyanotic congenital heart disease is suspected, attempt to place the defect in one of the three categories. This may be done using Table 5.4.A.3, which describes the typical findings in each physiological category. These guidelines assist the clinician but are not infallible, and the nature of the defect is sometimes only clear after echocardiography.

Cyanotic heart diseases
Low pulmonary blood flow
In defects where there is low pulmonary blood flow the physiology is the same regardless of the precise anatomy of the defect. Deoxygenated blood returning from the systemic veins cannot flow through the right side of the heart to the lungs. The pulmonary blood supply is therefore via the arterial duct. The deoxygenated blood from the right side of the heart shunts to the left side of the heart (via either an atrial or a ventricular septal defect), and the left ventricle receives both deoxygenated blood from the right heart and oxygenated blood from the pulmonary venous return. Blood entering the aorta is therefore not fully oxygenated, and the child appears cyanosed. If the duct closes, the infant becomes profoundly cyanosed and is unlikely to survive unless pulmonary blood flow is rapidly restored. This is duct-dependent pulmonary circulation, an example of which is shown in Figure 5.4.A.1.

Complete transposition of the great arteries (TGA)
Here the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle (see Figure 5.4.A.2). Systemic venous return enters the right side of the heart and is recirculated to the systemic arteries. Pulmonary venous return enters the left side of the heart and is recirculated to the lungs. Oxygenated blood and deoxygenated blood are therefore separated in two parallel circuits. Oxygenated blood enters the systemic circulation only when there is mixing between the two circuits. Mixing occurs at atrial level (across the foramen ovale) and at ductal level (while

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**Figure 5.4.A.1** The circulation in pulmonary atresia with intact ventricular septum. PDA, patent ductus arteriosus; Ao, aorta; PA, pulmonary artery; LA, left atrium; LV, left ventricle; RV, right ventricle; RA, right atrium.

**Figure 5.4.A.2** Transposition of the great arteries. For explanation of abbreviations, see legend to Figure 5.4.A.1.

**Figure 5.4.A.3** The circulation in double-outlet left ventricle. For explanation of abbreviations, see legend to Figure 5.4.A.1.
the duct remains open). Systemic oxygen saturation reflects the amount of mixing (which in turn depends on the size of these communications). If the atrial communication is small, oxygenation may therefore be duct dependent.

Common mixing lesions
In common mixing lesions, oxygenated pulmonary venous blood and deoxygenated systemic venous blood mix in one of the cardiac chambers. An example is shown in Figure 5.4.A.3. The systemic output is therefore only partly oxygenated. The relative amounts of pulmonary and systemic blood in the mixture determine the oxygen saturation and the mode of presentation. If pulmonary blood flow is high, cyanosis is minimal, and the child usually presents at about 2 months of age in heart failure. If pulmonary blood flow is low (the complex lesion may coexist with pulmonary stenosis), cyanosis is severe and is often detected early.

Once the defect has been placed in one of these categories, immediate management decisions can be made. Although it is not imperative to reach a more specific diagnosis, an anatomical diagnosis can sometimes be made using clinical information and simple investigations.

### TABLE 5.4.A.3 Features to help distinguish the three types of cyanotic congenital heart defect

<table>
<thead>
<tr>
<th>Condition</th>
<th>Low pulmonary blood flow</th>
<th>Complete TGA</th>
<th>Common mixing lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>pO₂ at rest</td>
<td>Often ≤ 35 mmHg</td>
<td>Often ≤ 35 mmHg</td>
<td>Often ≥ 45 mmHg</td>
</tr>
<tr>
<td>SaO₂ at rest</td>
<td>&lt; 80%</td>
<td>&lt; 80%</td>
<td>80–90%</td>
</tr>
<tr>
<td>pO₂ hyperoxia test</td>
<td>Often ≤ 50 mmHg</td>
<td>Often ≤ 50 mmHg</td>
<td>75–200 mmHg</td>
</tr>
<tr>
<td>SaO₂ hyperoxia</td>
<td>&lt; 90%</td>
<td>&lt; 90%</td>
<td>90–100%</td>
</tr>
<tr>
<td>Chest X-ray</td>
<td>Reduced pulmonary vascular markings</td>
<td>Normal or increased pulmonary vascular markings with or without narrow mediastinum</td>
<td>Normal or increased pulmonary vascular markings</td>
</tr>
</tbody>
</table>

### TABLE 5.4.A.4 Conditions with low pulmonary blood flow

<table>
<thead>
<tr>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Critical pulmonary stenosis</td>
</tr>
<tr>
<td>Pulmonary atresia with intact ventricular septum</td>
</tr>
<tr>
<td>Tetralogy of Fallot (with severe right ventricular outflow tract obstruction)</td>
</tr>
<tr>
<td>Pulmonary atresia with ventricular septal defect</td>
</tr>
<tr>
<td>Absent right atrioventricular connection</td>
</tr>
</tbody>
</table>

### Pulmonary atresia with intact ventricular septum and critical pulmonary stenosis

These two conditions are similar pathologies with either complete or almost complete closure of the pulmonary valve. Both are often associated with hypoplasia of the right ventricle. There is either no murmur or a soft murmur at the lower left sternal border (tricuspid regurgitation). The chest X-ray usually shows a normal heart size. The precordial leads on the ECG usually show decreased right ventricular voltages (small R waves in leads V1 and V2) and dominant left ventricular voltages (prominent S waves in leads V1 and V2 and prominent R waves in leads V5 and V6).

### Tetralogy of Fallot and pulmonary atresia with ventricular septal defect

These two pathologies are also similar, but in Fallot’s tetralogy the right ventricular outflow tract is patent, albeit narrow, generating a high-pitched ejection systolic murmur at the upper left sternal border. In both defects, the cardiac silhouette on the chest X-ray has a concavity on the left heart border where there is usually a convexity produced by the right ventricular outflow tract and pulmonary artery. The ECG shows dominant right ventricular voltages (normal neonatal RS progression).

### Absent right atrioventricular connection (also known as tricuspid atresia)

There is often a long harsh systolic murmur (this may arise from a restrictive ventricular septal defect or pulmonary stenosis). The precordial leads on the ECG show decreased right ventricular voltages and dominant left ventricular voltages. The QRS axis is characteristically directed to the left and superiorly between 0 and –90 degrees.

### Complete transposition of the great arteries

There is usually no murmur. The ECG shows dominant right ventricular voltages (normal neonatal RS progression). Therefore, if a newborn is severely cyanosed and otherwise appears clinically normal, actively look for a narrow mediastinum on the chest X-ray to help to make the diagnosis.

### Management of defects with low pulmonary blood flow or complete TGA

- Do not give oxygen after the hyperoxia test, as it may precipitate ductal closure.
- Start IV prostaglandin E (PGE) to maintain ductal patency. There are two formulations, namely prostaglandin E1 (PGE1) and prostaglandin E2 (PGE2). Commence PGE1 at 25 nanograms/kg/minute or PGE2 at 5 nanograms/kg/minute.
- PGE infusion is made up by adding 30 micrograms/kg of prostaglandin to 50 mL of 5% dextrose (if the pump runs at 1 mL/hour this is equivalent to 10 nanograms/kg/minute).
- If saturations are initially very low and fail to improve 10 minutes after starting PGE, intubate and ventilate the baby in air and increase the PGE dose to 50 nanograms/kg/minute. The dose can be increased further to a maximum of 100 nanograms/kg/minute if there is still no response. Prostaglandin sometimes causes vasodilatation, so fluid boluses may be required at high doses of PGE in order to maintain blood pressure.
- PGE often causes hypoventilation and apnoea (particularly at doses of PGE2 above 10 nanograms/kg/minute). If oxygen saturations initially improve with PGE and then start to fall, assess respiratory effort. If respiration is shallow or slow, intubate and ventilate in air.
- If oxygen saturations start to fall after PGE is started, and respiratory effort appears adequate, increase the
Cardiac failure. Few weeks of life, resulting in progressively worsening pulmonary vascular resistance decreases over the first few weeks of life, resulting in cyanosis. With time, the right ventricular outflow tract obstruction limits blood flow to the pulmonary arteries, causing deoxygenated blood to shunt right to left across the VSD, resulting in cyanosis. In tetralogy of Fallot, there is right ventricular outflow tract obstruction (usually a convexity produced by the right ventricular outflow tract and pulmonary artery). The right ventricular outflow tract obstruction limits blood flow to the pulmonary arteries, causing deoxygenated blood to shunt right to left across the VSD, resulting in cyanosis. With time, the right ventricular outflow tract obstruction usually becomes more severe, causing further reductions in pulmonary blood flow, more right to left shunting, and increasing cyanosis.

Cyanotic defects with high pulmonary blood flow

In cyanotic defects with high pulmonary blood flow (mostly common mixing defects), pulmonary blood flow increases as pulmonary vascular resistance decreases over the first few weeks of life, resulting in progressively worsening cardiac failure.

Management of common mixing lesions

- Monitor the child on the neonatal unit.
- Arrange for an echocardiogram as soon as possible to define the anatomy.
- If oxygen saturations fall progressively to less than 70%, commence PGE and arrange for an urgent paediatric cardiology review.
- Once the anatomy is defined it may be possible to discharge the baby without further treatment (after paediatric cardiology advice has been obtained).

The older infant with cyanosis

Is there a cardiac problem?

When an older infant presents with cyanosis, cardiac pathology is likely if:

- Respiratory distress is not severe.
- There is no carbon dioxide retention.
- Respiratory pathology is not evident on the chest X-ray.
- The cardiovascular examination is abnormal (see below).

What type of cardiac defect is present?

The cyanotic defects that commonly present after the neonatal period are tetralogy of Fallot and cyanotic defects with high pulmonary blood flow. They may escape detection at birth because cyanosis is initially only mild. In tetralogy of Fallot, there is right ventricular outflow tract obstruction and a large ventricular septal defect (VSD) (right ventricular hypertrophy and aortic override are the other components of the tetralogy). The right ventricular outflow tract obstruction limits blood flow to the pulmonary arteries, causing deoxygenated blood to shunt right to left across the VSD, resulting in cyanosis. With time, the right ventricular outflow tract obstruction usually becomes more severe, causing further reductions in pulmonary blood flow, more right to left shunting, and increasing cyanosis.

Cyanotic defects with high pulmonary blood flow

In cyanotic defects with high pulmonary blood flow (mostly common mixing defects), pulmonary blood flow increases as pulmonary vascular resistance decreases over the first few weeks of life, resulting in progressively worsening cardiac failure.

<table>
<thead>
<tr>
<th>TABLE 5.A.5 Cyanotic defects with high pulmonary blood flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>Truncus arteriosus</td>
</tr>
<tr>
<td>Total anomalous pulmonary venous connection</td>
</tr>
<tr>
<td>Double-outlet left ventricle</td>
</tr>
<tr>
<td>Absent right atrioventricular connection with large ventricular septal defect (tricuspid atresia)</td>
</tr>
<tr>
<td>Pulmonary atresia with large or multiple aorto-pulmonary collateral arteries</td>
</tr>
<tr>
<td>TGA with large ventricular septal defect</td>
</tr>
</tbody>
</table>

Findings in defects with high pulmonary blood flow

- May present with cyanosis at 2–6 weeks of age.
- Active praecordium.
- Murmur usually present (may be systolic, diastolic or continuous).
- Increased pulmonary vascular markings on chest X-ray.

Management of cyanotic defects with high pulmonary blood flow

Define the anatomy by echocardiography. Manage cardiac failure medically (see Section 5.4.B). Surgical correction or pulmonary artery banding will be necessary in most cases.

Findings in tetralogy of Fallot

- May present with increasing cyanosis.
- May present with an ejection systolic murmur at the upper left sternal border.
- Reduced pulmonary vascular markings on chest X-ray, and concavity on the left heart border where there is usually a convexity produced by the right ventricular outflow tract and pulmonary artery.
- Children are often asymptomatic, but there may be sudden periods of increased cyanosis known as hypercyanotic spells.

Characteristics of hypercyanotic spells

- Spells often occur on waking from sleep or after feeding.
- The infant becomes restless and agitated.
- There is increased cyanosis and pallor.
- Respiration is often rapid and shallow.
- In severe spells, crying is followed by limping or loss of consciousness.
- Spells usually last 1–5 minutes, but may last longer when severe.
- The ejection systolic murmur shortens or becomes inaudible.

Management of tetralogy of Fallot

The anatomy should be confirmed by echocardiography, preferably within a few weeks of presentation, and surgical correction should be carried out between 6 and 12 months of age (although it can be carried out later).

- Hypercyanotic spells may be life-threatening. If a child starts to have such spells, discuss this with a paediatric cardiologist immediately, as it is an indication for urgent surgery.

If hypercyanotic spells are more than a few minutes in duration, treat them urgently as follows:

- Knee–chest position.
- Give oxygen by face mask.
- Give an IV bolus of Ringer-lactate or Hartmann’s solution.
Neonatal cardiovascular collapse

Is there a cardiac problem?
When a child presents in shock in the first month of life, the working diagnosis is often dehydration or sepsis. The following features help to distinguish cardiac causes of poor systemic output from non-cardiac causes:

-Collapse in the first 2 weeks of life
-Poor feeding, lethargy and tachypnoea prior to collapse
-Dependent oedema and cardiomegaly on chest X-ray
-Lack of response to intravascular volume expansion.

What type of cardiac defect is present?
Left heart obstruction is the most likely cardiac cause of cardiovascular collapse with low systemic output in the first 2 weeks of life:

**TABLE 5.4.A.6 Left heart obstruction**

<table>
<thead>
<tr>
<th>Critical aortic stenosis</th>
<th>Hypoplastic left heart syndrome (HLHS)</th>
<th>Coarctation of the aorta</th>
<th>Interrupted aortic arch</th>
</tr>
</thead>
</table>

**FIGURE 5.4.A.4 Hypoplastic left heart.** For explanation of abbreviations, see legend to Figure 5.4.A.1.

**Hypoplastic left heart syndrome**

In hypoplastic left heart syndrome all of the left heart structures are small (see Figure 5.4.A.4). There is insufficient forward flow through the left ventricle and the aortic valve to support the systemic circulation. Pulmonary venous return cannot pass through the left heart, so it crosses the atrial septum and enters the right atrium, mixing with systemic venous return. Mixed pulmonary and systemic venous blood enters the right ventricle and is pumped to the pulmonary arteries and also across the arterial duct to supply the systemic circulation. Ductal flow passes to the descending aorta and retrogradely around the aortic arch to supply the head and neck vessels and the coronary arteries. Ductal flow is not fully oxygenated, so there is a degree of central cyanosis. When the duct closes, the cardiac output falls precipitously, the infant becomes shocked, and cardiac failure develops. This is duct-dependent systemic circulation. The haemodynamics are the same in critical aortic stenosis.

**Coarctation of the aorta**

Coarctation of the aorta consists of a narrowing in the descending aorta close to the aortic end of the arterial duct. Contractile tissue may extend from the duct into the aorta so that when the duct closes it draws in the adjacent section of aorta, causing obstruction. Flow to the head and neck vessels is maintained, but flow to the lower body distal to the coarctation site is dramatically reduced. The infant becomes shocked and acidotic. Cardiac failure develops secondary to high systemic afterload. This is also an example of the systemic circulation depending on ductal patency (although systemic blood flow may not directly depend on a right-to-left shunt through the duct). In interrupted aortic arch, perfusion to the lower part of the body depends on right-to-left ductal flow and presentation is similar to that with coarctation.

The following features help to distinguish between the lesions:

- If all of the pulses are weak or absent, consider HLHS or critical aortic stenosis.
- If the right arm pulses are palpable and the femoral pulses are weak or absent, consider coarctation or interrupted aortic arch (note, however, that all pulses may initially be impalpable if the cardiac output is poor).
- If four limb blood pressures demonstrate significantly lower blood pressures in the legs than in the right arm (a gradient of more than 20mmHg), consider coarctation or interrupted aortic arch.
- Coarctation often presents towards the beginning of the second week of life.
- HLHS often presents in the first 2 days of life.
- In HLHS, the ECG shows reduced left ventricular voltages (small R waves in leads V5 and V6).

Other cardiac causes of cardiovascular collapse in the first few weeks of life are supraventricular tachycardia (SVT) (see Section 5.4.C) and cyanotic congenital heart disease with duct-dependent pulmonary blood flow (when the duct closes, the ensuing profound hypoxaemia causes acidosis and cardiovascular collapse). SVT should be evident on the ECG and cyanotic heart disease should be suspected when the oxygen saturation remains low after instituting the management described below for left heart obstruction.
Emergency management of low systemic output secondary to left heart obstruction

1. Check the ECG (to exclude SVT as a cause of collapse).
2. Obtain peripheral IV access if not already established (if IV access is difficult, intra-osseous access should be obtained).
3. Give a fluid bolus of 10 mL/kg Ringer-lactate or Hartmann’s solution if not already given.
4. Intubate and ventilate if there is significant respiratory distress (high PEEP, 8–10cmH2O).
5. Once ventilated, commence prostaglandin E1 or E2 at 100 nanograms/kg/minute (give for 30 minutes, then reduce to 25 nanograms/kg/minute, reducing again to 10 nanograms/kg/minute when stabilised). If the initial clinical condition is not poor, commence PGE2 at a lower dose of 10 nanograms/kg/minute, which should avoid PGE-related apnoea and the need for ventilation.
6. Admit the child to the paediatric ICU.
7. Check blood sugar levels, full blood count, urea and electrolytes, coagulation levels and magnesium levels, and correct abnormalities.
8. Take blood cultures and treat with IV antibiotics, as sepsis cannot be excluded.
9. Check arterial blood gas (using the right arm if possible).
10. Give IV furosemide 1 mg/kg if the chest X-ray shows pulmonary oedema.
11. Insert central venous access and an arterial line.
12. Reassess whether further intravascular volume is needed (give if the central venous pressure is low).
13. Give dopamine 5–10 micrograms/kg/minute if perfusion remains poor or the blood pressure remains low.
14. Give adrenaline 0.1–0.2 micrograms/kg/minute if perfusion remains poor or the blood pressure remains low (by central venous access only).
15. If acidosis is profound and not improving with other measures, give IV sodium bicarbonate 4.2%.
16. Ask for an urgent paediatric cardiology review and advice.

Asymptomatic murmurs

When a child presents with an asymptomatic murmur, first examine them for cyanosis and measure the oxygen saturation. If there is desaturation, refer the child for an echocardiogram, as cyanotic congenital heart disease requires a detailed anatomical assessment. Tetralogy of Fallot is the most likely diagnosis. If cyanosis is excluded, the child may have an innocent cardiac murmur or one of the following defects.

<table>
<thead>
<tr>
<th>TABLE 5.4.A.7 Initially asymptomatic heart lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Left-to-right shunts</strong></td>
</tr>
<tr>
<td>Small to moderate-sized VSD</td>
</tr>
<tr>
<td>Small to moderate-sized PDA</td>
</tr>
<tr>
<td>Atrial septal defect (ASD)</td>
</tr>
<tr>
<td>Partial AVSD</td>
</tr>
<tr>
<td><strong>Left or right heart obstruction</strong></td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
</tr>
<tr>
<td>Aortic stenosis</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
</tr>
</tbody>
</table>

Innocent murmurs are characterised as follows:
- The Still’s murmur is a vibratory short systolic murmur heard at the lower left sternal border or apex.
- The venous hum is a soft continuous murmur heard best below the clavicles, and is abolished by pressure over the jugular vein or lying down with the neck flexed.
- The pulmonary flow murmur is a soft ejection systolic murmur at the upper left sternal border, and may be confused with an ASD or mild pulmonary stenosis.
- The neck bruit is an ejection systolic murmur that is maximal above the clavicle and may be confused with aortic stenosis.

The cardiac defects are characterised as follows:
- In coarctation, the right arm blood pressure is often elevated, the femoral pulses are weak or impalpable, and there is brachiofemoral delay.
- The patent ductus arteriosus (PDA) has a continuous murmur that is loudest in the left infraclavicular region.
- The ventricular septal defect (VSD) has a harsh pansystolic murmur that is loudest at the lower left sternal border radiating to the lower right sternal border.
- Aortic stenosis, pulmonary stenosis, atrial septal defect (ASD) and partial atrioventricular septal defect (AVSD) all have an ejection systolic murmur at the upper left sternal border.
  - In aortic stenosis the ejection systolic murmur is harsh and may be heard at the upper right and left sternal border. The murmur radiates to the carotid arteries and there is often a carotid thrill. There may be an ejection click at the apex if the stenosis is at valvar level.
  - In pulmonary stenosis, the ejection systolic murmur is harsh and radiates to the back. There may be an ejection click along the left sternal border if the stenosis is at valvar level.
  - In an atrial septal defect (ASD) there is a soft ejection systolic murmur at the upper left sternal border from increased flow across the pulmonary valve. There is sometimes a fixed widely split second heart sound, and there may be a mid-diastolic murmur at the lower left sternal border (from increased flow across the tricuspid valve) when the left-to-right shunt is large.
  - In partial atroventricular septal defect (AVSD) there is an abnormal atrioventricular valve and a defect in the atrial septum. There may be a blowing pansystolic murmur at the lower left sternal border or apex from atrioventricular valve regurgitation. The ejection systolic murmur may mimic an ASD, but the defect is distinguished by a superior QRS axis on the ECG.

Unless the murmur is clearly innocent, perform an ECG and chest X-ray.

Right ventricular hypertrophy (RVH) is indicated by an R wave in lead V1 > 98th centile for age (£ 20mm is always abnormal), a neonatal RS progression beyond the neonatal period (dominant R waves in lead V1 and dominant S waves in lead V6) or an upright T wave in lead V1 before the teenage years.

Left ventricular hypertrophy (LVH) is indicated by T inversion in leads V5 and V6, loss of the Q wave in lead V6 or the amplitude of the R wave in lead V6 plus S wave in lead V1 > 98th centile for age (£ 50mm is always abnormal). RVH may indicate significant right heart obstruction or high pulmonary artery pressure (secondary to a large left-to-right shunt or pulmonary vascular disease). LVH may indicate significant left heart obstruction. Cardiomegal and
increased pulmonary vascular markings on the chest X-ray may indicate a large left-to-right shunt.

Any child who is thought to have an anatomical defect on the basis of the clinical examination, or any child with an abnormal ECG or chest X-ray, should if possible be referred to a paediatric cardiologist for an echocardiogram and opinion. If there is evidence of a significant left-to-right shunt (see Section 5.4.B) in a VSD or PDA, the referral should be as soon as possible, as there is still a risk of pulmonary vascular disease even when the child does not present in heart failure.

Rheumatic fever
For the diagnosis and treatment of rheumatic fever, see Section 5.13.

Long-term consequences of rheumatic fever
After an attack of acute rheumatic fever there may be permanent valve damage. Rheumatic heart disease occurs when acute valve inflammation is followed by scarring and fibrosis, resulting in various degrees of shortening, thickening, rigidity, deformity, retraction and fusion of the valve cusps. The commonest valve lesions are mitral regurgitation, mitral stenosis and aortic regurgitation.

Rheumatic heart disease is most severe and progressive in (1) children who initially have severe carditis in (2) children who have recurrent attacks of acute rheumatic fever. The prognosis is more favourable if recurrences are prevented (residual cardiac disease may disappear or improve and valve damage only worsens in a few cases). It is therefore crucial to maintain continuous antibiotic prophylaxis to prevent further valve damage, particularly as children are prone to develop a recurrence after the initial attack (below).

Mitral regurgitation
Mitral regurgitation is the commonest valve lesion in children with rheumatic heart disease. Patients are often asymptomatic during childhood as symptoms are caused by left ventricular failure which may take as long as two decades to develop. However, cases may present before adolescence and mitral regurgitation may be rapidly progressive in regions where the incidence of rheumatic fever is high and recurrent rheumatic fever is common. Mitral regurgitation may be diagnosed by the presence of a blowing apical pansystolic murmur radiating to the left axilla. There may also be a third heart sound and a short low frequency mid-systolic murmur from increased transmural flow.

Features of severe mitral regurgitation:
- Easy fatigue (caused by low cardiac output)
- Shortness of breath on exertion (caused by pulmonary oedema)
- Hyperdynamic apical impulse and pansystolic murmur
- Apical impulse displaced laterally and inferiorly
- The chest X ray demonstrates cardiomegaly and left atrial enlargement (a double density on the right heart border and elevation of the left main bronchus)
- The ECG demonstrates left atrial enlargement (broad bifi d P waves in lead II and a prominent negative component to the P in V1) and left ventricular hypertrophy
- Signs of pulmonary hypertension (see below).

If there are features of severe mitral regurgitation, the child should be urgently referred for a paediatric cardiology opinion as surgery is likely to be necessary. Ideally all children with mitral regurgitation should be evaluated by echocardiography annually, as progressive left heart dilation may result in irreversible left ventricular dysfunction if referral is delayed until symptoms develop. Medical treatment should be given for heart failure (captopril is particularly useful) but children who are unwell enough to require this often need either a mitral valve repair or a mitral valve replacement with a mechanical valve or bioprosthesis.

Mitrall stenosis
If there is effective antibiotic prophylaxis, mitral stenosis usually develops slowly over 5–10 years and is often not sufficiently severe to cause symptoms in childhood. The reality in countries where there is inadequate prophylaxis and recurrent attacks of rheumatic fever are common is that mitral stenosis may progress much more rapidly and symptoms may be evident 6 months to 3 years after the first attack. Mild stenosis does not cause symptoms, moderate stenosis causes shortness of breath on exertion and severe stenosis causes easy fatigue, shortness of breath at rest, orthopnoea, paroxysmal nocturnal dyspnoea and haemoptysis. Mitral stenosis may be diagnosed by the presence of a low frequency mid-diastolic murmur maximal at the apex. The murmur may be accentuated by exercise and is often accompanied by a loud first heart sound and a diastolic opening snap. The murmur becomes longer as the severity of the stenosis increases. In severe cases there are also signs of pulmonary hypertension.

Signs of pulmonary hypertension:
- Left parasternal heave
- Loud second heart sound
- Early diastolic murmur of pulmonary regurgitation at the upper left sternal border
- Elevated JVP and hepatomegaly if there is right heart failure.

The chest X ray and ECG often show left atrial enlargement when there is moderate mitral stenosis. Radiographic signs of pulmonary oedema may be evident when stenosis is severe. ECG changes of right ventricular hypertrophy and right axis deviation are present when there is pulmonary hypertension. Symptoms should be treated with diuretics and a low-sodium diet. Digoxin is only indicated in rare cases where there is atrial fibrillation secondary to left atrial enlargement. Symptomatic children and children with signs of pulmonary hypertension should be referred for paediatric cardiology review as surgery is often necessary. The options for treatment are open or closed mitral commissurotomy, mitral valve replacement and percutaneous catheter balloon mitral commissurotomy.

Aortic regurgitation
Aortic regurgitation is less common than mitral regurgitation and frequently occurs in combination with mitral valve disease. Affected children usually remain asymptomatic for many years as symptoms only become evident when left ventricular dysfunction develops secondary to chronic left ventricular volume overload. Severe symptomatic aortic regurgitation may however become established within 1–2 years of the initial attack of rheumatic fever if recurrence is not prevented. Once symptoms appear deterioration is often rapid. Symptoms include exercise intolerance, shortness of breath on exertion and chest pain in a few severely affected cases. Examination reveals a blowing decrescendo
early diastolic murmur maximal at the mid to lower left sternal border. The murmur is loudest sitting forward with the breath held in expiration.

Signs of moderate to severe aortic regurgitation:
- The murmur lengthens and may be throughout diastole
- Hyperdynamic apex
- Apical impulse displaced laterally and inferiorly
- Wide pulse pressure
- Collapsing pulses
- Visible pulsations in the suprasternal notch and neck vessels
- Systolic murmur at the upper right sternal border (from increased aortic valve flow).

If patients are symptomatic or have signs of severe aortic regurgitation they should be referred for paediatric cardiology assessment as surgery may be necessary. Marked cardiomegaly on the chest X ray or multiple ventricular ectopics on the ECG should also prompt referral. Ideally all children with aortic regurgitation should have an echocardiogram at least annually as it is important to assess left ventricular dilation and function to ensure that surgery is carried out before irreversible left ventricular dysfunction develops. Exercise tolerance may be improved by captopril treatment and medical treatment for heart failure may be necessary in severe cases. Surgical options include aortic valve reconstruction, aortic valve replacement with an aortic homograft or mechanical valve and transferring the patient’s own pulmonary valve to the aortic position (Ross procedure).

5.4.B The child with heart failure and cardiomyopathy

**BOX 5.4.B.1 Minimum standards**
- Electrocardiograph.
- Chest X-ray.
- Furosemide.
- Spironolactone.
- Anticoagulant.

**Introduction**
Heart failure occurs when the heart is unable to pump enough blood to meet the metabolic needs of the body. The term is often used to indicate the clinical changes that occur when the cardiac pump cannot meet the workload it is presented with. This may occur either because the pump is weak (due to a primary abnormality of the cardiac muscle) or because the workload imposed on the heart is higher than normal. The latter is the case in congenital heart disease, where heart failure occurs because the heart is pumping against a high resistance (in the case of obstructive lesions) or because it is volume loaded (commonly in left-to-right shunting cardiac lesions).

Left-to-right shunting cardiac defects are the commonest cause of heart failure in infancy identified in well-resourced countries.

In resource-poor countries most heart failure is related either to severe anaemia or to fluid overload when treating infections or severe malnutrition, particularly with IV fluids or during blood transfusion (see below).

**The physiology of left-to-right shunts**
A large defect between the ventricles or great arteries allows free communication between the left and right sides of the heart. Left and right heart pressures therefore equalise, and pulmonary artery pressure is maintained at systemic level. The pulmonary vascular resistance then determines the pulmonary blood flow. In the newborn period the pulmonary vascular resistance is high, which limits the pulmonary blood flow and therefore the left-to-right shunt across the defect. Over the first 6 weeks of life, the pulmonary vascular resistance gradually falls, allowing pulmonary blood flow and the left-to-right shunt to increase. This gives rise to heart failure, which usually appears after 4 weeks of age. If the pulmonary arteries are exposed to high pressure and flow for a prolonged period, pulmonary vascular disease develops. This normally becomes significant between 12 and 18 months of age. High pulmonary vascular resistance secondary to pulmonary vascular disease reduces the left-to-right shunt, and symptoms of heart failure gradually resolve. Eventually, pulmonary resistance becomes so high that flow across the defect becomes right to left, and cyanosis develops (Eisenmenger’s syndrome). The pulmonary artery pressure remains high throughout, and it is only the amount of flow through the lungs that changes.

**Is heart failure present?**

**TABLE 5.4.B.1 Diagnosis of heart failure secondary to congenital heart disease in infancy**

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Easily tired</td>
<td>Failure to thrive</td>
</tr>
<tr>
<td>Poor feeding</td>
<td>Tachypnoea</td>
</tr>
<tr>
<td>Breathlessness (particu-</td>
<td>Increased respiratory effort</td>
</tr>
<tr>
<td>larly during feeds)</td>
<td>Tachycardia &gt; 160 bpm</td>
</tr>
<tr>
<td>Sweaty (particularly</td>
<td>Sweating</td>
</tr>
<tr>
<td>during feeds)</td>
<td>Pallor</td>
</tr>
<tr>
<td></td>
<td>Hepatomegaly</td>
</tr>
<tr>
<td></td>
<td>Gallop rhythm</td>
</tr>
</tbody>
</table>

**What type of cardiac defect is present?**
Heart failure in the first few weeks of life is a medical emergency.

The following causes should be considered:
- supraventricular tachycardia
- complete atrioventricular block
- high-output cardiac failure
- left heart obstruction.

Perform an ECG to detect supraventricular tachycardia and heart block. Check the haemoglobin level, as severe anaemia may cause high-output cardiac failure. Also examine the baby for cranial and hepatic bruits, as cranial and hepatic arteriovenous malformations are a potential (although very rare) cause of high-output cardiac failure.
If these tests are negative, refer the child to a paediatric cardiologist, as a left heart obstructive lesion is likely and there may be duct-dependent systemic circulation. Consider the use of prostaglandin to keep the ductus arteriosus open until the referral can be achieved (see Section 5.4.A).

Heart failure in infancy presenting after the first few weeks of life may be caused by any of the following:
- The left-to-right shunting lesions listed in Table 5.4.B.2
- Cyanotic congenital heart defects with high pulmonary blood flow
- The same causes that present in the first few weeks of life
- Myocarditis or cardiomyopathy.

### TABLE 5.4.B.2 Common left-to-right shunting lesions that cause heart failure

| Large ventricular septal defect (VSD) |
| Atrioventricular septal defect with large ventricular component (AVSD) |
| Large persistent ductus arteriosus (POA) |

Examine the child for cyanosis and measure the oxygen saturation. It should be possible to detect those children with cyanotic defects immediately (note, however, that children with AVSD are sometimes mildly desaturated). Next, attempt to detect the children with left-to-right shunts, looking for the following features which are present in significant shunts:
- Hyperdynamic precordial impulse
- Apical impulse displaced laterally and inferiorly
- Apical mid-diastolic murmur (from increased flow across the mitral valve)
- Loud second heart sound (from increased pulmonary artery diastolic pressure)
- Cardiomegaly and increased pulmonary vascular markings on the chest X-ray
- Signs of heart failure and pulmonary oedema on the chest X-ray in severe cases.

If these examination findings are not present and there is no evidence of SVT or a hyperdynamic circulation (see above), a left heart obstructive lesion should be considered. Some of these are eminently treatable conditions and if they are suspected the child should be referred for paediatric cardiology review without delay.

If there is evidence of a large left-to-right shunt, refer the child to a paediatric cardiologist within a few weeks. These signs must not be missed, as a remediable cardiac defect is rendered inoperable by delay. Although it is not imperative to make a more specific diagnosis, the following clinical features discriminate between the three most common left-to-right shunts:
- The persistent arterial duct has a continuous murmur that is maximal in the left infraclavicular area.
- A large ventricular septal defect has a quiet pansystolic murmur at the lower left sternal border. The ECG shows a characteristic superior QRS axis (between −30 and −180 degrees).

### Heart failure in later infancy and childhood

In addition to the symptoms seen in early infancy (easily tired, poor feeding, breathlessness particularly during feeds, excess sweating particularly during feeds), older children may have decreased exercise tolerance, shortness of breath on exertion and when lying flat.

The signs of heart failure are cyanosis or SaO₂ < 94%, basal lung crepitations, failure to thrive, tachypnoea > 50 breaths/minute for children aged 2–12 months, tachypnoea > 40 breaths/minute for children aged 12 months to 5 years and the cut-offs for tachycardia are > 120 bpm aged 1–5 years and > 100 bpm after age 5 years.

There is usually increased respiratory effort, sweating, pallor and hepatomegaly.

In older children the hepatomegaly may be tender, a gallop rhythm may be heard and a raised jugular venous pressure may be observed.

In addition to the congenital heart defects described in Section 5.4.A the following causes of heart failure should be considered:
- Severe anaemia
- Severe malnutrition
- Excessive intravenous fluids
- Rheumatic fever
- Myocarditis
- Cardiomyopathy
- Infective endocarditis
- Constrictive pericarditis (rare and most often caused by tuberculosis) (see Section 6.1.N).

Anaemia is a common and often severe problem in poorly resourced settings (see Section 5.11.A). When the haemoglobin falls below 7 g/dl cardiac output must increase to maintain oxygen delivery and heart failure frequently develops with a haemoglobin < 5 g/dl. The treatment is careful blood transfusion, but the increase in intravascular volume may precipitate worsening heart failure. Blood must therefore be infused slowly in small boluses and an exchange transfusion may be needed if there is clinical deterioration. Furosemide 1 mg/kg IV may be given during transfusion (see Section 5.11.A).

Protein-calorie malnutrition is also an important cause of cardiac failure in disadvantaged countries (see Section 5.10.B) with specific contributions from certain vitamin deficiencies (see Section 5.10.A). Although cardiac failure is unusual at presentation, it may occur after several days of refeeding. Rapid refeeding can cause a hypermetabolic state, demanding an increase in cardiac output which cannot be met by the malnourished heart which has a decreased cardiac reserve. The problem is exacerbated by coexisting anaemia, blood transfusion, inappropriate intravenous fluid administration and high sodium diets.

The other common causes of cardiac failure are dealt with individually in the sections below.

### Management of heart failure

Monitor heart and respiratory rates, respiratory distress and oxygenation regularly during treatment of acute heart failure. It is necessary to both control the symptoms of failure and to determine and treat the underlying cause.
Cardiomyopathy and myocarditis

Myocarditis and dilated cardiomyopathy both cause impairment of myocardial contractility. This results in a dilated poorly functioning heart. Children present with heart failure, sometimes in association with shock. They may also more rarely present with ventricular arrhythmias.

The unexpected onset of heart failure in a previously well child should suggest the diagnosis. However, in the first 3 months of life, heart failure associated with cardiomalgaly is more likely to be caused by congenital heart disease than by heart muscle disease. Echocardiography is therefore particularly important in this age group, to discriminate between the two potential causes of heart failure.

In addition to the features of heart failure listed earlier in Table 5.4.B.1, signs may include lateral displacement of the apex beat and an apical pansystolic murmur from mitral regurgitation. The chest X-ray often demonstrates cardiomalgaly. It is not essential to identify whether the child has cardiomyopathy or myocarditis, as the management of both conditions is the same. However, the latter may be suggested by a preceding viral illness or evidence of acute myocardial damage with elevated blood levels of creatinine kinase or troponin. Myocarditis may be confirmed by identifying enterovirus by polymerase chain reaction (PCR) or serology.

In most cases, the cause of the cardiomyopathy remains unknown. However, it is important to perform a 12-lead ECG in all cases of cardiomyopathy, as it may reveal two particular conditions that are reversible causes of poor heart function:

- Incessant tachyarrhythmias may cause cardiomyopathy. In cardiomyopathy there is often sinus tachycardia, which appears on the ECG as a heart rate faster than that expected for the child's age, with each QRS complex being preceded by a P wave that is positive in both lead I and lead aVF. If the QRS complexes are not preceded by P waves, or P-wave morphology is unusual, the rhythm is not sinus rhythm and a tachyarrhythmia must be suspected. Sometimes the tachyarrhythmia heart rate is only marginally higher than that expected for the child's age, but many months of mild tachycardia have resulted in poor function. If the arrhythmia is successfully controlled with anti-arrhythmic drugs or radiofrequency ablation, the heart function should normalise.

- Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) presents with severely impaired cardiac function at around 3 months of age. The ECG will show transmural anterolateral myocardial infarction in most cases. If the coronary artery is reimplanted in the aortic root the function will usually recover.

Post-intervention management is aimed at supporting the heart while function spontaneously recovers. It includes the following:

- Furosemide and spironolactone (see above, under heart failure)
- Captopril (see above, under heart failure)
- Digoxin:
  - 5 micrograms/kg orally twice a day (in children under 5 years old).
  - 3 micrograms/kg orally twice a day (in children over 5 years old).
  - Plasma levels should be in the range 0.8–2.0 micrograms/litre (check the level after 5 days, and at least 6 hours after giving a dose).
- Aspirin 3–5 micrograms/kg once a day if function is poor, to prevent thromboembolism.
- Anticoagulation if cardiac function is very poor:
  - Heparin IV, initially 20 U/kg/hour, titrated to APTT 2–3 times normal.
  - Warfarin if anticoagulation is needed long term.
- Intubation and ventilation if pulmonary oedema is severe.
- Inotropic support (dobutamine 5–10 micrograms/kg/minute, dopamine 5–10 micrograms/kg/minute, milrinone maximum dose of 0.7 micrograms/kg/minute). Ventilation and inotropic drugs should be a last resort if the child is deteriorating despite other measures, as it can be difficult to wean them off intensive care support once these steps are taken.
- Once the child is stabilised, introduce carvedilol at a dose of 50 micrograms/kg (maximum dose 3.125 mg) twice a day, doubling the dose at intervals of at least 2 weeks up to an upper limit of 350 micrograms/kg (maximum 25 mg) twice a day. Use echocardiography to check that cardiac function has not deteriorated before each dose increment, and monitor blood pressure for 4 hours after every dose increment. Carvedilol promotes myocardial remodelling.

Bacterial endocarditis

Endocarditis should always be suspected in a child with
a cardiac defect when there is a fever without a focus. Infection develops on injured areas of endothelium or on abnormal or damaged heart valves. In some cases the onset may be sudden with obvious signs of sepsis and cardiac fail-ure (secondary to valve damage). However, in most cases the onset is insidious and the diagnosis is unclear. There may be fever, malaise, fatigue, arthralgia, anorexia and weight loss. It may occur in a child previously thought to have a normal heart but with an undiagnosed congenital heart defect or undiagnosed episode of rheumatic fever.

Signs of endocarditis:
- Pyrexia
- Microscopic haematuria
- Splenomegaly
- Changing heart murmur
- Petechiae
- Neurological abnormalities (caused by cerebral abscess or infarction)
- Splinter haemorrhages, Janeway lesions, Osler’s nodes and Roth’s spots (characteristic but rare).

The diagnosis is made by isolating bacteria from the blood. At least three sets of blood cultures must be obtained from different puncture sites. If possible, antibiotics should be withheld until multiple blood cultures have been obtained and should only be started when the diagnosis is clear or there is a pressing clinical urgency. Blood cultures will be negative in 10–15% of cases. Echocardiography helps to make the diagnosis if vegetations are seen, but a negative echocardiogram does not exclude the diagnosis.

Organisms most commonly isolated in endocarditis:
- *Streptococcus viridans* (commonest overall)
- *Staphylococcus aureus* (most cases of fulminant endocarditis)
- Coagulase-negative staphylococci (if the patient has a central venous line or is immunocompromised e.g. with HIV).

If the organism is *Streptococcus viridans*, IV benzylpenicillin 25 mg/kg 6 hourly and gentamicin 7.5 mg/kg once daily are given for two weeks, followed by a further two weeks of oral amoxicillin. If the organism is *Staphylococcus aureus*, IV flucloxacillin 25 mg/kg 6 hourly is given for 4 weeks, coupled with IV gentamicin 7.5 mg/kg once daily (or sodium fucidate) 6–7 mg/kg 8 hourly for the first two weeks. Vancomycin 10 mg/kg 6 hourly is used in place of flucloxacillin if the organism is a coagulase negative *Staphylococcus* or the patient is allergic to penicillin.

The effectiveness of treatment is monitored by symptoms and inflammatory markers (WBC, ESR and CRP). Surgery is necessary when the organism cannot be eradicated, when there is evidence of embolisation, where there is a large mobile vegetation at risk of embolisation or when there is severe cardiac failure from valve damage.

### 5.4.C The child with a cardiac arrhythmia

**80X 5.4.C.1 Minimum standards**
- Electrocardiograph.
- Defibrillator.
- Adenosine.
- Beta-blockers.
- Flecaïnide.
- Amiodarone.
- Atropine.

**Supraventricular tachycardia**

Supraventricular tachycardia (SVT) is the commonest tachyarrhythmia in childhood.

It may present with poor systemic output and heart failure in infancy, or palpitations and dizziness in later childhood. SVT can be distinguished from sinus tachycardia because the rate is usually more rapid (200–300 beats/minute) than can be explained by the child’s level of activity, fever, agitation or pain. The ECG in most cases shows narrow QRS complexes without a preceding P wave. The commonest cause of SVT in childhood is an accessory pathway, which is an abnormal bundle of muscle fibres bridging from the atrium to the ventricle. In accessory pathway-mediated tachycardia, depolarisation passes down from the atrium to the ventricle through the atrioventricular node, and then returns back up to the atrium using the accessory pathway. If the electrical wavefront then passes down the atrioventricular node again and once again returns up to the atrium via the accessory pathway, SVT has initiated. Some, but not all, accessory pathways are evident on the resting ECG because forward conduction across the accessory pathway in sinus rhythm causes a slurred stroke just before the QRS complex, known as a delta wave. The condition is often known as the Wolff–Parkinson–White (WPW) syndrome. The best method of treating this condition is by radiofrequency ablation of the abnormal pathway by means of a catheter passed into the atria, but this is a skilled technique that is only available in specialist centres.

Some patients have a different type of SVT, where the electrical wavefront loops back on itself to form a ‘short circuit’ entirely within the atrioventricular node. This is less often seen in early childhood, but becomes more common towards adolescence. A totally different mechanism of tachycardia occurs when there is an abnormally rapid atrial discharge (atrial flutter or atrial ectopic tachycardia). This is relatively rare in childhood.

**Management of SVT**

1. Record a 12-lead ECG in tachycardia,
2. While attempts are made to terminate the tachycardia, record a rhythm strip (this can often be easily run off a standard defibrillator),
3. In the infant, try to terminate the SVT by facial immersion in ice-cold water.
4. To terminate symptomatic or prolonged attacks of SVT in the older child, try vagal manoeuvres such as ice-cold packs on the face, the Valsalva manoeuvre or carotid sinus massage.
5. If tachycardia persists, obtain IV access (via a large antecubital vein if possible) and give a rapid bolus of
Section 5.4

Adenosine 100 micrograms/kg followed by a rapid crystalloid flush.

6 If the SVT is not terminated, give larger doses of adenosine, doubling the dose until a maximum dose of 400 micrograms/kg is reached.

7 If adenosine terminates the tachycardia transiently, and then SVT re-initiates, anti-arrhythmic drug treatment needs to be initiated straight away to prevent constant recurrence of the arrhythmia.

8 If adenosine successfully terminates the tachycardia, it is not compulsory to initiate anti-arrhythmic treatment. As SVT is not dangerous beyond infancy, anti-arrhythmic drugs are only given if the child wants to avoid further attacks (this decision is usually influenced by the frequency and duration of attacks). In infancy, SVT may cause serious haemodynamic compromise. In view of this, all infants who present with SVT should be started on anti-arrhythmic medication, which should be continued until the child's first birthday.

9 If adenosine does not terminate the tachycardia at all, carry out synchronised DC cardioversion, after anaesthesia, intubation and ventilation with 0.5 joules/kg, rising to 2 joules/kg in steps if the first shocks are unsuccessful.

10 In rarer cases, adenosine causes only transient block of the atrioventricular node, revealing rapid atrial activity in the form of P waves or sawtooth flutter waves. When atrioventricular nodal conduction returns after a few seconds, the tachycardia is re-initiated. These tachycardias require either anti-arrhythmic drug treatment to be initiated straight away or DC cardioversion.

Ventricular tachycardia (VT) is normally diagnosed when there is a broad complex tachycardia. However, the latter is more often seen in childhood when there is SVT with bundle branch block. In view of this, adenosine should be given if a child presents with regular broad complex tachycardia. If the adenosine terminates the tachycardia, this proves that SVT was the correct diagnosis. If there is irregular broad complex tachycardia, do not give adenosine (this pattern indicates a dangerous arrhythmia). If the tachycardia persists even after high-dose adenosine, ventricular tachycardia is likely. Not all childhood ventricular tachycardia is dangerous. If the child is haemodynamically stable, attempts can be made to terminate the tachycardia with anti-arrhythmic drugs (see below). If there is haemodynamic compromise, immediate DC cardioversion is required.

**Direct current (DC) cardioversion**

Sedate or anaesthetise the child unless they are in extremis. Use paediatric paddles if the child weighs less than 10 kg. Place one paddle over the apex of the heart in the mid-axillary line and the other immediately below the clavicle just to the right of the sternum. If there are only adult paddles and the child weighs less than 10 kg, place one on the back and one over the lower chest anteriorly. The first shock should be 0.5 joules/kg, and subsequent shocks should be increased stepwise to a maximum of 2 joules/kg.

**Anti-arrhythmic drugs**

This is only a guide, and other types of drug within a class can be given.

**First choice**

**Beta-blockers** (oral doses given)

- **Infants**: propranolol 1 mg/kg/dose three times a day.
- **Children** (when cannot swallow tablets): atenolol 1–2 mg/kg once a day.
- **Older children** (when can swallow tablets): bisoprolol 0.2–0.4 mg/kg once a day (tablets come as 2.5 mg, so use multiples of this amount).

**Second choice**

**Flecainide** (oral doses given)

- **Under 12 years of age**: initially 2 mg/kg/dose twice a day. It is possible to increase to 3 mg/kg/dose if tachycardias persist (maximum of 8 mg/kg/day).
- **Over 12 years of age**: 50–100 mg twice a day (maximum of 300 mg a day).

It is preferable to measure the flecainide level after 5 days just before the next dose is due to be administered, to check that the plasma level has not exceeded 800 micrograms/litre.

Avoid feeds for 30 minutes before or after giving oral flecainide, as the absorption of the drug is significantly affected by milk and dairy products.

**Third choice**

**Beta-blocker and flecainide together**

If the tachycardia does not respond to the above drugs in the acute setting, or the child's haemodynamic status is borderline, IV amiodarone is the safest option.

**IV amiodarone**

Give a loading dose of 5 mg/kg over 2 hours (dilute with 5% dextrose only). Then continue infusion at a rate of...
5–20 micrograms/kg/minute (maximum of 1.2 grams in 24 hours).

Consider stopping the infusion 4–8 hours after the SVT has resolved.

If tachycardia recurs after stopping amiodarone, give a further loading dose and recommence infusion, continuing for at least 1 day after tachycardia resolution.

As amiodarone has a large number of side effects, consider switching to either a beta-blocker or flecaïnide once the tachycardia has been controlled and the child is haemodynamically stable.

Make up the amiodarone infusion as follows:
15 mg/kg in 50 mL of 5% dextrose (1 mL/hour = 5 micrograms/kg/hour: such a slow infusion will need an electrically driven syringe pump).

Amiodarone is incompatible with sodium chloride. Therefore do not make up with and do not flush lines with this solution.

Amiodarone can be given through a peripheral line, but serious tissue damage may be caused by the drug if extravasation occurs, so central access is preferred. If peripheral access is used, dilute the infusion to a concentration between 600 micrograms/mL and 2 mg/mL. This dilution will be more appropriate in situations where electrically driven syringe pumps are not available, but the infusion needs close monitoring.

### Congenital complete heart block
- Consider this in any newborn who has a consistent bradycardia without apparent cause, such as terminal respiratory failure or very severe shock.
- P waves are dissociated from QRS complexes on the 12-lead ECG.
- Perform an echocardiogram to exclude structural heart disease.
- Check for anti-Ro and anti-La antibodies in the child’s mother (the underlying cause in the majority of cases).
- Monitor the heart rate for 24–48 hours.
- Assess perfusion and blood pressure, and examine for signs of heart failure.
- Arrange for a permanent pacemaker if there is inadequate cardiac output, heart failure, structural heart disease or the heart rate is < 50 beats/minute.
- Atropine 20 micrograms/kg or isoprenaline infusion 0.02–0.2 micrograms/kg/minute can be used for emergency treatment of severe bradycardia with inadequate cardiac output.

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### 5.5 Shock

#### 5.5.1 Shock

**Introduction**

‘Shock’ occurs when the circulatory system fails to deliver adequate amounts of primarily oxygen, but also nutrients, to the tissues, and fails to remove unwanted metabolites from the tissues for excretion.

**Pathology at cell level**

At a cellular level, the end result of shock is anaerobic metabolism (oxygen-depleted metabolism). This is an inefficient mechanism and requires much more energy than aerobic metabolism (the normal oxygen-dependent system). In addition, anaerobic metabolism builds up excess toxic acid products in the cells which cannot be removed by the failed circulation. Cellular function deteriorates and there is a downward spiral of increasing loss of homeostasis, the onset of disseminated intravascular coagulation, and after a short while so much cell death occurs in vital organs that recovery is impossible and the patient dies.

In the early stages of shock the body has mechanisms to try to combat this process. The circulatory system is under the control of the sympathetic nervous system. This system regulates the flow of blood in health and in disease to all organs so as to respond to demands on different organs. In health, more blood is sent to muscles if a person is exercising, more to the digestive system if they are eating, and more to the skin if their body is too warm.

In shock, the sympathetic nervous system attempts to protect the vital organs by diverting blood away from muscle, skin and the digestive system and directing it to the heart, brain and kidneys. This gives rise to some of the earlier signs of shock, such as cold peripheries, increased capillary refill time, cerebral anxiety or agitation, tachycardia to increase cardiac output, and reduced urine output as the kidneys conserve fluid.

Later signs such as depressed consciousness, weak pulses, falling blood pressure and acidotic breathing show that the body’s compensation mechanisms are failing. It can be seen that it is vital to recognise and treat shock in the patient as soon as possible, as this will give the best chance of patient recovery.

**Clinical diagnosis of shock**

The signs of shock are listed below, although not all of them are present in all types of shock.
- **Tachycardia** (best measured with a stethoscope).