There are a number of commonly encountered acute cardiac presentations in older infants and children that are essential to recognise early in order to institute appropriate emergency care. Many of these will be the first clinical presentation of a child; therefore the attending doctor or nurse, with no specific diagnosis, will need to know the basic physiology of the cardiovascular system and the common derangements that may occur. For children with a known diagnosis, such as the above-mentioned one, attending practitioners must be able to manage life-threatening complications, as these patients will often be seen in a non-tertiary setting.

This article highlights the clinical presentation and immediate management of common cardiac emergencies in children beyond the neonatal period, illustrated with typical clinical scenarios, ECG recordings or X-ray/echocardiographic images. These are grouped according to their mode of presentation as follows:

- cyanosis/hypercyanotic spells
- acute congestive heart failure
- shock
- tachyarrhythmias/bradyarrhythmias
- pericardial effusion/cardiac tamponade.

Cyanosis/hypercyanotic spells

The child with cyanotic congenital heart disease often goes undiagnosed and can present for the first time in a hypercyanotic spell ...

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A 2-year-old boy with known tetralogy of Fallot and a right modified Blalock-Taussig shunt is referred to you in the emergency room with severe cyanosis, having been ill for the past two days with a runny nose, cough and poor appetite. He is irritable but alert with deep sighing and rapid respiration. You can't hear a shunt murmur but a soft ejection murmur is present. What has happened and how will you manage him (see below)?

Clinical features

The clinical features are central cyanosis (confirmed with a pulse oximeter) (Fig. 1), irritability (due to acute cerebral hypoxia), hyperpnoea secondary to metabolic acidosis (confirmed with arterial or venous blood gas), a softer ejection systolic murmur (ESM) than at baseline (due to acutely reduced pulmonary blood flow), and signs of the trigger (e.g. dehydration from gastroenteritis, pyrexia from a respiratory tract infection). A chest X-ray usually shows oligaeic lungs and a 'boot' heart. In the patient described above, it is likely that his shunt has acutely blocked – he needs to be managed as a ‘spelling tet’.

Management

- Keep the child calm (the mother should be with the child!) – avoid painful procedures without sedating the patient
- Sedation – chloral hydrate 50 mg/kg orally or morphine 0.1 - 0.2 mg/kg IM/IV/SC or ketamine (2 - 4 mg/kg IM or 1 - 2 mg/kg IV) – repeat as necessary
- Posture – knee-chest/head down
- Blood volume expansion – 10 ml/kg 0.9% saline IV bolus and repeat as necessary until the liver is palpable (do not be afraid of giving IV fluids – minimal risk of heart failure in a tet!)
- Sodium bicarbonate 1 - 2 mmol/kg IV as a 4.2% solution – repeat according to blood gas (acidosis perpetuates a vicious cycle of worsening hypoxia)
- Beta-blocker – propranolol 0.5 - 1 mg/kg orally or, better, esmolol IV bolus and infusion (if available)
- Facemask oxygen (especially if the trigger is a chest infection)
- Discuss with paediatric cardiologist and refer to tertiary centre as soon as possible.

Many congenital and acquired heart diseases may present with heart failure.
Increasing intensity of the ESM, in addition to rising oxygen saturations, provides reassuring signs of response to treatment. Failure to respond to these measures necessitates intubation/ventilation with sedation/paralysis and urgent transfer to a tertiary (preferably cardiac) facility where, in an ICU setting with invasive monitoring, alpha-agonists such as phenylephrine or noradrenaline are useful to stabilise the child pending an emergent surgical systemic-to-pulmonary shunt or definitive corrective procedure.

Acute congestive heart failure

An 8-year-old boy presents to the medical emergency department with respiratory distress, unable to lie flat. He has a 10 cm liver, displaced apex beat with a tachycardia and a loud apical pansystolic murmur. His mother says he has recently been tiring easily and was treated for tonsillitis in the previous month. You then notice his swollen left knee. What is the diagnosis?

Many congenital and acquired heart diseases may present with heart failure (HF). The main acquired causes present acutely, and include myocarditis/dilated cardiomyopathy, valvular dysfunction from infective endocarditis or acute rheumatic fever, acute glomerulonephritis, prolonged tachyarrhythmias and heart block. In contrast, HF secondary to congenital heart disease is usually a more insidious process and can occur at any age but most often presents in infancy; e.g. at 4 - 6 weeks (or earlier in premature infants) for large L to R shunts.

Clinical features

The clinical features are tachycardia/tachypnoea, difficult feeding (infants) (tiring, dyspnoea, sweating) and poor weight gain/tachypnoea, difficult feeding (infants) (tiring, dyspnoea, sweating) and poor weight gain. Clinical features are tachycardia and a loud apical pansystolic murmur. He is also tachypnoeic, with occasional crackles in the lung fields. His abdomen is soft and nontender. The clinical features are tachycardia/tachypnoea, difficult feeding (infants) (tiring, dyspnoea, sweating) and poor weight gain.

Management

- **Non-pharmacological measures**, e.g. modest fluid restriction (usually temporary until established on diuretics and tolerating feeds well), posture (head up), supplemental oxygen if desaturated, correct conditions increasing oxygen/metabolic demand (e.g. pyrexia, anaemia, sepsis), attention to fluid and electrolyte balance, and optimisation of nutrition.
- **Diuretics**, usually furosemide 1 mg/kg 6 - 12-hourly IV or orally, with spironolactone 1 mg/kg 12-hourly orally (for potassium-sparing effect, may also slow deleterious ventricular remodelling – stop all other K+ supplements).
- **ACE inhibitors**, e.g. captopril, starting with a test dose of 0.1 mg/kg (monitoring blood pressure to detect hypotension), increasing slowly over a few days to 3 - 4 mg/kg/day in 3 divided doses as tolerated – ensure normal renal function and K+, contraindicated in neonates (except under specialist supervision).
- **Digoxin** is still used in HF caused by myocardial dysfunction or supraventricular tachycardia, but its role in HF secondary to volume overload conditions, such as large L to R shunts, is doubtful (avoid in suspected acute viral myocarditis and ensure normal renal function and K+ to prevent toxicity) – dose 5 μg/kg 12-hourly per mouth (tds loading is not required).
- **Beta-blocker therapy** (e.g. carvedilol) may be beneficial in some children with chronic mild to moderate HF (given under specialist supervision only).
- **Refer to tertiary paediatric cardiac centre** (preferably on the same admission) to establish a definitive diagnosis, as many causes of HF are amenable to specific therapy, e.g. surgical correction of congenital or acquired heart defects (in this patient's case mitral valve surgery may be indicated).

Shock

A 2-month-old boy is referred with severe respiratory distress and a diagnosis of acute bronchiolitis. You request an immediate bronchiodilator ‘neb’ but then notice marked cardiomegaly on his chest X-ray. Further examination reveals that he is peripherally ‘shut down’, with a ‘galloping’ heart and no murmurs. He appears to have acidic breathing. What is a more likely differential diagnosis?

The causes of cardiogenic shock in children are similar to those implicated in HF. However, the majority of congenital causes present in the neonatal period and are therefore discussed elsewhere in this issue (see article ‘Neonatal cardiac emergencies’). It can be difficult to differentiate cardiogenic from septic shock but an enlarged liver and heart are clues to a cardiac aetiology. If in doubt, antibiotics (usually cefotaxime or ceftriaxone) should be given early (but after a blood culture), as septicemia remains the commonest cause of shock in children.

Clinical features

The clinical features are poor peripheral perfusion (cool extremities with prolonged capillary filling time), reduced volume pulses, tachycardia/tachypnoea, often hepatomegaly, clinical and radiological cardiomegaly with/without pulmonary oedema (may cause wheezing), occasional ‘gallop rhythm’ and murmurs (e.g. mitral regurgitation), oliguria/anuria, lethargy, hypotension (late sign) and metabolic acidosis/high lactate on blood gases. You suspect this patient to be in cardiogenic shock, possibly due to acute myocarditis, and correctly proceed as follows:

It can be difficult to differentiate cardiogenic from septic shock but an enlarged liver and heart are clues to a cardiac aetiology.

Management

- Admission to a paediatric ICU as soon as possible, but do not delay initiating treatment – have a resuscitation trolley at hand (Fig. 2).
- Respiratory/ventilatory support is usually indicated (e.g. CPAP or mechanical ventilation with sedation and paralysis) – intubation to be done by most senior practitioner available using cardio-stable drugs, e.g. etomidate and vecuronium (high risk of cardiac arrest at intubation).
- Inotropes, e.g. dobutamine 5 - 15 μg/
kg/min IV infusion, via central or peripheral line; failure to respond to this may require adrenaline 0.05 - 0.2 μg/kg/min via central line. An ideal drug is milrinone, which is both an inotrope and a peripheral vasodilator, but its availability is limited.

- Fluid resuscitation may be needed initially as the child may be intravascularly volume depleted (cautious 5 - 10 ml/kg aliquots assessing response after each bolus).
- Diuretics only once the patient is no longer in shock (usually furosemide 1 mg/kg - 12-hourly IV initially with spironolactone 1 mg/kg 12-hourly orally).
- Ideally, there should be CVP and invasive arterial pressure monitoring to guide the above-mentioned treatment.
- Attention must be given to fluid, electrolyte and glucose balance.
- Refer to tertiary centre as soon as stabilised to establish a definitive diagnosis that may be amenable to specific therapy. In our example scenario, the patient is found on echo to have an anomalous left coronary artery from the pulmonary artery (ALCAPA) – an uncommon but curable cause of dilated cardiomyopathy – and has successful corrective surgery 2 days later.

Tachyarrhythmias

These can be divided into narrow QRS complex (<90 msec) and wide QRS complex (>90 msec) tachycardias. The majority of children presenting with narrow complex tachycardias have an AV re-entrant tachycardia via an accessory pathway – also loosely called an ’SVT’ (Fig. 3) – typically with a rate of 240 +/- 40/min (or >2 x normal rate for age); other causes include ectopic and multifocal atrial tachycardias, atrial flutter, and nodal/AV junctional ectopic tachycardia. Sinus tachycardia in infants is usually less than 220/min (or <2 x normal rate for age) and has a definite ’warm up’ and ’cool down’ phase with an obvious trigger, e.g. fever, anaemia.

An important rule of thumb is that wide complex tachycardia in children is ventricular tachycardia until proved otherwise ...

**Management**

- Try to ’capture’ the tachycardia on a 12-lead ECG recording (preferably with a rhythm strip) to enable a specific diagnosis to be made (this can be faxed or e-mailed to a cardiologist).
- However, do not delay if haemodynamically unstable – provide immediate treatment.
- Remember to treat any reversible causes of arrhythmias, e.g. 4 Hs and 4 Ts according to the Advanced Paediatric Life Support (APLS) guidelines (hypoxaemia, hypovolaemia, hypo-/hyperthermia, hyper-/hypokalaemia, as well as Ca, Mg and blood glucose derangements and tamponade, tension pneumothorax, toxins/poisons/drugs, thromboembolism).
- Wide complex tachycardia (probable VT): immediate synchronised DC cardioversion 1 J/kg increasing to 2 J/kg if no response (sedate if conscious without delaying cardioversion as VT can quickly degenerate into pulseless VT or VF).
- If no response to cardioversion and still shocked, consider amiodarone 5 mg/kg IV over a few minutes (over 20 - 30 min if not in shock).
- For torsades de pointes give magnesium sulphate 25 - 50 mg/kg IV over a few minutes.

- Note that pulseless VT should be managed as for VF (according to APLS guidelines).
- Narrow complex tachycardia: if in shock, initial attempt at a vagal manoeuvre (e.g. diving reflex, carotid sinus massage or a Valsalva in an older child), but if unsuccessful progress rapidly to synchronised DC cardioversion as for VT or immediate adenosine (see below), whichever is quicker to administer.
- If haemodynamically stable, attempt vagal manoeuvres – if no response, adenosine 100 μg/kg rapid IV bolus increasing by 100 μg/kg every 2 minutes up to a maximum of 500 μg/kg (300 μg/kg for neonates) or 12 mg total dose (Fig. 5).

Bradyarrhythmias

Excluding sinus bradycardia, the main cause of bradycardia in children is complete heart block (CHB), which is either:

- congenital – presenting often in the neonatal period (see accompanying article 'Neonatal cardiac emergencies'). The ventricular rate is usually 45 - 80/min with a narrow QRS complex escape; or
- acquired – mainly post-cardiac surgery, rarely from severe myocarditis or poisoning/drug overdoses. This form may be temporary or permanent. The ventricular rate tends to be slower (40 - 60/min), typically with a wide QRS complex escape (Fig. 6).

Clinical presentation

As for tachycardias, the clinical presentation ranges from asymptomatic to cardiogenic shock or SCD. CHB may cause heart failure, especially if associated with congenital heart disease. Dizziness or syncope are worrying
symptoms and are more likely to occur at a heart rate below 40 - 45/min, especially with exercise (risk of SCD).

**Management**
- Atropine (bolus 20 μg/kg, minimum dose 100 μg, maximum 600 μg), adrenaline (10 μg/kg bolus) followed by infusions or an isoprenaline infusion may be attempted pending urgent transfer to a tertiary paediatric cardiac centre for definitive ventricular pacing.
- Bolus of 10 - 20 ml/kg IV crystalloid if the patient is hypotensive/shocked.
- If there is an inadequate response to the above, transcutaneous or transoesophageal temporary pacing should be promptly instituted if available (discuss with paediatric cardiologist) – this requires patient sedation +/- ventilation for comfort.
- The definitive treatment is permanent pacing, which is usually achieved via surgically placed epicardial wires in young children.
- Transvenous pacing may be used as a temporary pacing measure (via an RV pacing lead) or for permanent pacing in older children and adults.

**Pericardial effusion/cardiac tamponade**
The main causes of pericardial effusion in infants and children are viral and tuberculous pericarditis. Less common causes include acute rheumatic fever, bacterial (purulent) pericarditis, post-cardiac surgery and connective tissue or malignant disease, e.g. lymphoma.

**Clinical features**
The clinical features depend not only on the amount of fluid but, more importantly, on the speed of fluid accumulation – the more rapid, the more likely to cause cardiogenic shock, i.e. cardiac tamponade. Suggestive features include precordial pain, palpable apex beat, muffled heart sounds, pericardial friction rub or occasionally a ‘pericardial knock’ (a sign of constrictive pericarditis) and the characteristic globular cardiomegaly on chest X-ray (Fig. 7). Signs of tamponade include tachycardia with pulsus paradoxus, hepatomegaly/distended neck veins together with poor perfusion and hypotension.

**Management**
- If haemodynamically stable without any features of tamponade (usually in children with a chronic history), refer to tertiary centre as soon as possible for definitive diagnosis (by echocardiography (Fig. 8), and pericardiocentesis or surgical drainage).
- If signs of cardiac tamponade are present, urgent decompression of the pericardial space is mandatory and lifesaving as deterioration to cardiac arrest is inevitable. This can be achieved by aspirating even a small amount of fluid (e.g. 10 - 20 ml) from a sub-xiphoid approach (14 - 18 G IV cannula aiming at the left shoulder at 30 - 45° to skin – stop advancing the needle as soon as fluid has been obtained and aspirate as much as possible before removing cannula). Note that fluid may be haemorrhagic. This is often enough to improve cardiac filling and output – at least temporarily.
- Blood volume expansion with 20 ml/kg crystalloid IV bolus, repeated as necessary.
- Once stabilised, refer urgently to tertiary facility.

**Conclusion**
The child with a cardiac emergency may be challenging to many health practitioners. However, with application of a sound systematic clinical approach and knowledge of the common presentations, as outlined above, there is much that can and should be done as immediate primary care to prevent death and major morbidity – the ‘golden hour’ of care applies just as in other resuscitation situations. This emergency care also allows for stabilisation of the child, enabling a safe transfer to a tertiary paediatric cardiac centre for more definitive diagnosis and management.

**Bibliography/suggested further reading available at www.cmej.org.za**

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**IN A NUTSHELL**
- There are a number of congenital and acquired heart diseases in older infants and children that may present as life-threatening emergencies.
- These may be undiagnosed and present for the first time as emergencies, or they may present with complications in a child with a known diagnosis.
- Every health practitioner caring for children should be aware of the common diagnoses, expected complications and their immediate management.
- The most commonly encountered emergencies are cyanosis/hypercyanotic spells, acute congestive heart failure, cardiogenic shock, tachyarrhythmias/bradyarrhythmias and pericardial effusion/cardiac tamponade.
- The emphasis should be on early recognition of haemodynamic derangements and immediate appropriate intervention in order to prevent death and significant morbidity.
- Early consultation with a paediatric cardiologist is desirable especially if there is doubt as to the diagnosis or poor response to initial treatment.
- Once a child is stabilised, prompt referral to a tertiary paediatric cardiac centre is usually required for definitive diagnosis and management.