Neonatal cardiac emergencies

The neonatal period is one that fills many generalists with fear – this article will help to dispel these concerns.

George A Comitis, MB ChB, DCH (SA), DA (SA), FCPaed (SA), Cert Cardiology (SA) Paed

Consultant, Paediatric Cardiology Service of the Western Cape, Red Cross War Memorial Children's Hospital, Cape Town, and Division of Critical Care and Children's Heart Diseases, School of Child and Adolescent Health, University of Cape Town

George Comitis is the young blood of the paediatric cardiology team at Red Cross Children's Hospital. He was lucky enough to be trained in the same unit after spending two wet years at the Royal Brompton Hospital as a Fellow in Paediatric Cardiac Intensive Care, during which he became tired of not being able to talk properly to his patients.

Correspondence to: G Comitis (gam.comitis@uct.ac.za)

A day 2 term neonate, delivered uneventfully following prolonged rupture of membranes, is referred to you with respiratory distress since the previous night in the postnatal ward. He has obvious central cyanosis and on auscultation you hear no murmurs. His arterial blood gas shows severe hypoxaemia with a metabolic acidosis. You initially suspect congenital pneumonia but then notice that his chest radiograph shows clear 'black' lung fields and a boot-shaped heart. What is a more likely diagnosis? Why was he apparently well for the first day?

Many congenital heart diseases (CHDs) present in the neonatal period. This article focuses on critical congenital heart defects with an incidence of 3 - 3.5/1 000 live births[1] (almost half of all CHDs). These have been defined as lesions that are 'duct dependent' (i.e. dependent on a patent ductus arteriosus (PDA) for pulmonary or systemic blood flow) or require intervention (surgical or catheter) within the first month in order to sustain life or prevent major morbidity.[2] In addition, common neonatal arrhythmias are discussed as these can also present as emergencies.

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A clear appreciation of the pathophysiology of each particular CHD requires a sound understanding of the fetal, transitional and neonatal circulations, i.e. the normal physiological changes that occur after birth.

As detailed antenatal anomaly scanning is not routinely available in South Africa, the majority of neonates with critical CHD present clinically without a known diagnosis. Even when available, detection rates for CHD are modest, currently ranging from 57% to 67%.[3,4] Despite the numerous different cardiac lesions, there is only a limited spectrum of clinical presentations that the practitioner is likely to encounter, i.e.:

- cyanosis
- shock
- tachyarrhythmias/bradyarrhythmias
- acute congestive heart failure.

This article discusses these presentations and provides practical acute management guidelines using clinical scenarios. Where overlap exists with conditions occurring in older infants and children, the reader is referred to the companion article 'Common paediatric cardiac emergencies' in this journal for further details.

Cyanosis

A detailed description of cyanotic CHD is beyond the scope of this article. Rather, a few pointers are given to assist in distinguishing between respiratory and cardiac causes of cyanosis in the neonate (Table I), as the former represents the main differential diagnosis, with mention of the more common lesions. Thereafter essential emergency care is discussed.

Table I. Distinction between respiratory and cardiac cyanosis in the neonate

	Respiratory	Cardiac
History	Risk factors, e.g. meconium-stained liquor, prematurity, CNS depression	Suggestive antenatal scan, infant of diabetic mother, syndromes, previous children with CHD, etc.
Onset	At birth	Hours to days after birth
Examination	Respiratory distress – tachypnoea, grunting, recession	Comfortable unless duct-dependent pulmonary circulation with closing PDA
Auscultation	Lung crackles/wheezes, reduced air entry	Clear lungs, normal air entry, murmur not always present
Blood gas	High pCO_2, low pO_2	Normal/low pCO_2, low pO_2
Hyperoxia test*	'Passed'	'Failed'
Chest X-ray	Lung infiltrates, normal heart size	Clear lung fields, oligaemia or plethora, heart normal or enlarged, 'typical' cardiac silhouette†
ECG	Normal	May be abnormal and provide clue to diagnosis

*Hyperoxia test – measure right radial or brachial arterial pO_2 in 100% O_2 (closed headbox or rebreather mask for 10 -15 min): pO_2 <150 mmHg (20 kPa) → cardiac, pO_2 >150 mmHg → respiratory (note: there may be a few exceptions to this rule).

†'Typical' X-ray patterns are only occasionally seen – 'egg on the side' (narrow mediastinum) for transposition, boot heart for tetralogy/pulmonary (and sometimes tricuspid) atresia, 'snowman' sign for supracardiac total anomalous pulmonary venous drainage (TAPVD), cardiomegaly with plethora for truncus arteriosus.

It is useful to remember the most common cyanotic CHDs as the 5 Ts: in order of frequency – tetralogy of Fallot (and pulmonary atresia), transposition of great arteries, tricuspid atresia, TAPVD, and truncus arteriosus. In addition, an important differential is persistent pulmonary hypertension of the newborn (PPHN) – suggestive features would be a history of risk factors for PPHN (such as perinatal asphyxia or meconium aspiration) and differential cyanosis, i.e. oxygen saturation/pO_2 in the feet (postductal) lower than the right hand (preductal) due to R to L shunting across the PDA. NB: This is also a feature of critical aortic arch obstructions (see below).

In the introductory scenario, the baby probably has an RV outflow tract obstructive lesion (extreme tetralogy, critical pulmonary stenosis or atresia), i.e. duct-dependent pulmonary circulation, and was relatively well until spontaneous closure of the PDA on day 2 caused acute hypoxaemia and acidosis. The patient requires resuscitation as follows:

Management

- Open the PDA ASAP! Mainstay therapy for reopening a PDA and keeping it open is prostaglandin E_1 (alprostadil) IV infusion (starting dose 50 - 100 ng/kg/min, if poor response (saturations and acidosis not improving) increase to max 300 - 400 ng/kg/min, maintenance dose 10 - 25 ng/kg/min) or prostaglandin E_2 (dinoprostone) orally/via nasogastric tube (dose 25 µg/kg or quarter tablet (500 µg strength) every ½ - 1 hour) – the latter should be available at all hospitals as an emergency drug, the former usually only at referral centres. Oral PGE_2 is very safe (baby unlikely to come to any harm if diagnosis is not cyanotic CHD), but IV PGE_1 can cause apnoea/hypoventilation and hypotension; hence used more in ICU or high-care settings. Prostaglandins may be used to attempt reopening of a PDA in a neonate up to 30 days of age.
- Blood volume expansion – 10 ml/kg 0.9% saline IV bolus, repeat as needed until liver edge palpable – to improve preload and hence encourage further opening of the PDA and pulmonary blood flow through the duct.
- Sodium bicarbonate empirically (as acidosis aggravates hypoxic pulmonary vasoconstriction) – 1 mmol/kg IV as a 4.2% solution, repeat according to blood gas.
- Correct other derangements, e.g. hypoglycaemia, hypocalcaemia, concomitant sepsis.
- Oxygen administration – aiming for saturations 75 - 85% (depending on exact diagnosis).
- Inotropes/vasopressors may be indicated to maintain adequate systemic perfusion and encourage pulmonary perfusion (by increasing systemic vascular resistance).
- If poor response to above, intubation/ventilation with sedation +/- paralysis to minimise metabolic demands and improve oxygenation.
- Consult with paediatric cardiologist and, once stabilised, transfer to tertiary centre for definitive diagnosis (usually with echocardiography) and management – this may entail cardiac catheterisation and often palliative surgery (e.g. modified Blalock-Taussig shunt).

Shock

A 1-week-old term neonate who was well at birth and discharged on day 2 presents to you in medical emergency with respiratory distress, vomiting and inability to feed for 1 day. He is centrally pink, alert and bright-looking but irritable with tachycardia and poor peripheral perfusion. His brachial pulses are easily felt but lower limb pulses are absent. What is the likely differential diagnosis?

The practitioner faced with a shocked neonate needs to have a high index of suspicion for CHD as the presentation can often be mistaken for neonatal sepsis. The usual lesions implicated are the critical left-sided outflow tract obstructions such as critical aortic stenosis (AS), hypoplastic left heart syndrome (HLHS), critical coarctation and interrupted aortic arch, which all represent duct-dependent systemic circulations. These are also the most commonly missed critical CHD diagnoses.[5] In critical AS and HLHS the R to L PDA flow provides all the systemic perfusion and in the arch obstructions provides flow to the lower body causing differential cyanosis (pink arms, blue feet). However, this sign is less prominent or absent if there is intracardiac mixing of systemic and pulmonary venous blood (e.g. coexistent large ASD or VSD).

Important, and often missed (suggesting an arch obstruction), are discrepant pulses (as in the above scenario) and blood pressure (lower limb blood pressure less than that of upper limb, or even undetectable). Radio-femoral delay is impossible to detect at the fast heart rates of neonates – rather examine for diminished volume or absent pulses. The above neonate likely has a critical aortic arch obstruction with acute loss of lower body perfusion causing shock and metabolic acidosis after spontaneous ductal closure on day 5 - 6 (preserved brain perfusion, hence alert). He needs resuscitation as follows:

Management

- Arrange urgent admission to ICU and do not delay treatment.
- Again – open the PDA ASAP! Prostaglandin therapy as above for duct-dependent pulmonary circulation. Note that oral PGE_2 may be less reliable as gut absorption may be compromised in this setting. However, oral PGE_2 must be started if IV PGE_1 is not available.
- Blood volume expansion – 0.9% saline in 5 - 10 ml/kg aliquots, monitoring liver size, to assist opening the PDA and improving systemic perfusion (baby also likely to be hypovolaemic with poor feeding and vomiting).
- The rest of the resuscitation and stabilisation would be similar to the above approach for cyanosis, e.g. inotropes (especially if coexistent LV dysfunction), oxygen to achieve preductal saturations (>85% in this case), and often mechanical ventilation.
- It is important to check for and manage end-organ damage such as renal failure, liver dysfunction and gut ischaemia/necrotising enterocolitis as per routine ICU protocols.
- Transfer to tertiary centre for definitive management – confirmation of diagnosis by echocardiography and, usually, primary surgical repair of the arch obstruction.

Tachyarrhythmias

A 1-day-old neonate is referred to you soon after birth with a heart rate of 280/min. He was delivered by caesarean section for fetal tachycardia but had good Apgar scores and there were no risk factors for congenital sepsis. He is alert, comfortable in room air and perfusing well without signs of cardiac failure. What is the differential diagnosis?

The most common neonatal tachyarrhythmias (excluding sinus tachycardia which usually does not exceed 220/min) are atrioventricular re-entry tachycardia (AVRT) (associated with an accessory AV pathway) followed by atrial flutter. Less likely causes include AV node re-entry tachycardia, ectopic and multifocal atrial tachycardia and

junctional tachycardias. Note that these are all narrow QRS complex tachycardias. Wide complex tachycardias are less commonly seen and, as for older children, merit urgent consultation with a paediatric cardiologist. They are best regarded and treated as ventricular tachycardia rather than SVTs with aberrant conduction.

You decide to administer adenosine. This has only a very transient effect slowing the rate to about 130/min – after a few seconds the rate is again 280/min. Fig. 1 shows the rhythm strip you recorded during adenosine administration. What is the likely diagnosis and how will you treat it?

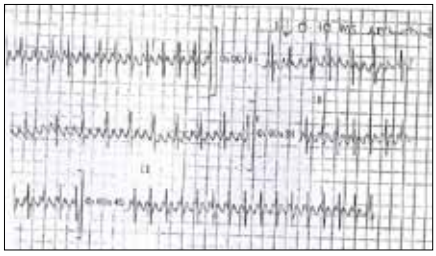

Fig. 1. Neonate with tachycardia: continuous rhythm strip (lead II) with adenosine.

Fig. 1 shows the typical 'saw tooth' baseline of atrial flutter, more manifest after adenosine, which slows the ventricular rate.

Management

- Atrial flutter: adenosine is not effective. The treatment of choice is synchronised DC cardioversion – as little as 0.5 J/kg is often enough to cardiovert to sinus rhythm. Ensure that paediatric paddles are used with adequate electrode jelly and the correct application front and back of the chest for a neonate. Remember to sedate the baby for comfort.
- AVRT: similar approach as for older children – if shocked, initial attempt at vagal manoeuvre and, if unsuccessful, progress immediately to synchronised DC cardioversion 1 J/kg, then 2 J/kg if no response *or* adenosine, whichever is quicker to administer. The starting dose of adenosine for neonates is 150 µg/kg rapid IV bolus; if no response, increase by 50 µg/kg every 2 minutes to a maximum single dose of 300 µg/kg.
- Wide complex tachycardia (probable VT): synchronised DC cardioversion 1 J/kg, then 2 J/kg if no response (sedate if conscious without delaying cardioversion).
- Treat any aggravating and reversible factors, e.g. hypoglycaemia, electrolyte imbalance, acidosis.
- It is good practice to record the tachyarrythmia on a 12-lead ECG as well as a rhythm strip before and during any therapy (cardioversion or adenosine as in the above case) so that the rhythm can be analysed by a cardiologist.
- If still no response, consult a paediatric cardiologist and refer to a tertiary centre (options include amiodarone, digoxin or propranolol, depending on the diagnosis).

Bradyarrhythmias

A baby is delivered at term by caesarean section for presumed 'fetal distress' (fetal bradycardia noted on cardiotocogram (CTG). You attend the delivery and note that the baby is vigorous at birth but remains bradycardic at 48/min. Her ECG is shown in Fig. 2. What is the diagnosis?

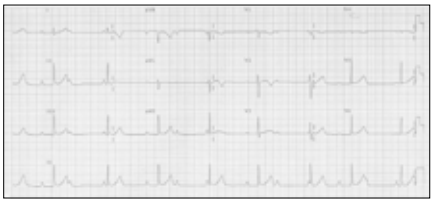

Fig. 2. Neonate with bradycardia.

The commonest cause of bradycardia in the neonate (defined as a heart rate <70/min) is congenital complete heart block (CHB). Fig. 2 is a typical example showing atrioventricular dissociation and a narrow QRS escape. It is often associated with maternal lupus erythematosus or other connective tissue diseases (which are often asymptomatic in the mother). Less commonly, cases may be isolated or associated with CHD.

Acute management

- The majority of neonates (with heart rates above 50 - 55 bpm) will be asymptomatic with good cardiac output and hence will not require immediate treatment – they may be referred, on the same admission, to a paediatric cardiologist for exclusion of structural heart disease and ongoing observation.
- If the neonate is symptomatic (signs of heart failure or cardiogenic shock), treat as for the older child – positive chronotropes (atropine, adrenaline or isoprenaline), blood volume expansion and temporary pacing under sedation (transcutaneous or transoesophageal) – see accompanying article 'Common paediatric cardiac emergencies' in this issue.
- Eventually all patients will require permanent cardiac pacing but this can often be delayed for years in the stable patient.

Acute congestive heart failure

A 2-week-old neonate presents with respiratory distress and was unable to feed for 3 days. He is tachycardic with bounding pulses, the apex is displaced and the liver is enlarged. There is a loud ejection systolic murmur and single S2. What is the differential diagnosis?

Many of the critical CHDs as well as the arrhythmias discussed in this article can present with acute heart failure (HF). The important point is to differentiate HF from respiratory distress due to lung disease or neonatal sepsis – as in older children, useful clues for HF are cardiomegaly and hepatomegaly.

Management

- Non-pharmacological measures, e.g. fluid restriction, nutritional support, oxygen and ventilatory support as indicated, correct aggravating factors such as anaemia or sepsis.
- Diuretics, usually furosemide 0.5 - 1 mg/kg 8 - 12-hourly IV or orally with spironolactone 1 mg/kg 12-hourly per mouth.
- ACE inhibitors are generally contraindicated in neonates (high risk of severe hypotension and renal failure).
- Inotropes/vasodilators such as dobutamine may be indicated in severe HF.
- Refer to tertiary paediatric cardiac centre (on the same admission) for definitive management. The above-described neonate is found to have truncus arteriosus and proceeds to full surgical repair after 5 days of stabilisation on anti-failure treatment as above.

'Typical' X-ray patterns are only occasionally seen ...

Conclusion

The practitioner faced with an acutely ill neonate needs to have a high index of suspicion for a cardiac aetiology. Obvious

clues would include all of the above presentations. A thorough cardiovascular examination should always be undertaken – without delaying emergency care – including a hyperoxia test in the cyanosed neonate, a check for discrepant pulses and blood pressures, and differential cyanosis. A chest X-ray and 12-lead ECG are essential first-line investigations to narrow the differential diagnosis. Immediate stabilisation as outlined above, followed by prompt referral to a tertiary centre, provides the best opportunity to minimise morbidity and mortality.

References and bibliography/suggested further reading available at www.cmej.org.za

IN A NUTSHELL

- By definition, all neonates with critical CHD will require catheter or surgical intervention in the first month of life.
- Despite numerous possible lesions, they usually present with cyanosis, acute heart failure or shock and can easily be mistaken for having neonatal sepsis or lung disease.
- Knowledge of the changes in the circulation after birth is essential to understand the pathophysiology of acute neonatal cardiac presentations (including PPHN), particularly the impact of a closing PDA on duct-dependent pulmonary and systemic circulations.
- The hyperoxia test remains a useful tool to differentiate cardiac from respiratory cyanosis.
- 'Typical' chest X-ray patterns are only occasionally seen and should not be relied on for diagnosis.
- An alert but shocked neonate has an aortic arch obstruction until proved otherwise.
- Prostaglandin therapy to re-establish and maintain ductal patency is not only indicated for cyanotic CHD (i.e. duct-dependent pulmonary blood flow), but also for critical left-sided obstructive lesions causing duct-dependent systemic blood flow.
- Prostaglandin (IV or oral) must always be readily available and should be regarded as an emergency life-saving drug which should be commenced immediately if there is any possibility of a critical CHD.
- The majority of neonates with critical CHD present without a known diagnosis, even if an antenatal scan was done (as detection rates for CHD are at best about 67%); hence a high index of suspicion is required in any acutely ill neonate.
- Many of these neonates will require ICU admission for stabilisation and, once this is achieved, prompt and safe transfer to a tertiary centre is imperative for definitive diagnosis and management.

SINGLE SUTURE

Fizzy drink ban has little effect on students

Nothing, apparently, can come between US students and sugary drinks. In states where schools banned sugary soft drinks to reduce calorie counts, children simply brought in their own, consuming equal amounts to those in states that didn't implement the ban.

Dan Taber at the University of Illinois in Chicago analysed 6 900 questionnaires sent to students in 40 states in 2006 and 2007. Irrespective of availability in schools, around 85% of respondents said they consumed sweetened drinks at least once a week, and a quarter to a third imbibed daily.

The policy still has worth, Taber says. '[The bans] have improved the school food environment.' He hopes they will now be supplemented with measures such as a tax on sweet drinks and distancing fast-food outlets from schools.

'The study clearly shows that isolated efforts are insufficient to improve nutrition to prevent obesity,' says Arne Astrup of the University of Copenhagen in Denmark, who has investigated links between sugary drinks and weight gain. 'What's needed is a much more comprehensive strategy that attacks the problem from several angles,' he says.

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