St. Peter's Hospital, Neonatal Unit
Diagnosis & Investigation of Congenital Heart Disease in the Neonate

Congenital heart disease (CHD) is one of the commonest congenital malformations, affecting 7 to 8 per 1000 live births[1,2]. Although many of the structurally significant heart lesions will present with symptoms early on, a significant proportion may demonstrate no symptoms until they later collapse[3]. At the same time, however, symptoms such as murmurs are very common in the newborn period and often do not require any therapeutic intervention.

Common Presentations & Their Management

The common ways in which CHD presents are with cyanosis; respiratory distress; a murmur; cardiovascular collapse; feeding difficulties; or a combination of the above. Assessment of potential CHD must include a thorough pre- and postnatal history, as well as review of any antenatal scans performed.

The routine newborn check must include:
- assessment of feeding and any respiratory distress
- auscultation of the praecordium
- palpation of femoral pulses

Cyanosis (+/- respiratory distress)
Causes include:

- CHD
- Respiratory disease
- Sepsis
- Persistent pulmonary hypertension of the newborn
- Methaemoglobinaemia (rare)

Clinically, cyanosis is best seen in the tongue and the perioral mucosa, but the clinical pick-up rate of cyanosis is low. Therefore, if there is any doubt at all, lower-limb oxygen saturations (**normal >95%**) must be obtained. **All babies with cyanosis in the postnatal ward MUST be admitted to the neonatal unit** for further investigation, directed at the above causes. A hyperoxia (nitrogen washout) test (see Appendix 1), together with chest X ray & ECG, is still a robust means of distinguishing between cardiac and non-cardiac causes of cyanosis, where echocardiography is not immediately available.

The classical presentation of a cyanotic lesion in the neonatal period is due to the closure of the ductus in one of the duct-dependent circulations, the treatment for which would be prostaglandin E2– see Neonatal Formulary. This must be discussed with the consultant.

Once the cause is confirmed as a duct-dependent cardiac lesion, oxygen therapy must be minimised (to keep saturations at 70% -75%) as hyperoxia can stimulate closure of the PDA and subsequent clinical deterioration. Paediatric cardiology opinion must be sought urgently to advise on further management.

Murmurs:
Murmurs are relatively common in the newborn period, affecting 0.5 – 1% of all newborns[5]. Of these, a significant proportion (46% - 62%) are likely to have a clinically insignificant murmur, commonly a PDA or peripheral pulmonary artery stenosis[5,6]. In babies with a murmur a full cardiovascular examination, and lower-limb oxygen saturations is a minimum requirement[7].

Features of a clinically insignificant murmur are:

1) Soft (no heaves/thrills) & Systolic (diastolic murmurs are NEVER innocent)
2) Localised
3) Normal pulses & limb oxygen saturations (>95%)[4]

A) If the murmur is clinically insignificant, saturations >95% & baby is asymptomatic:
- re-examine in 24 hours' time, ideally by the same person.
- If murmur still present, examination by a senior colleague to confirm that the murmur is clinically insignificant.
- If so, arrange an appointment in 4 to 8 weeks' time, provide a parent information sheet on heart murmurs & advise to return to hospital if there is:
  - Central cyanosis
  - Respiratory distress and/or excessive sweating
  - Poor feeding (>2 consecutive feeds of <50% usual volume) or poor weight gain

B) If there is any doubt about the clinical significance of the murmur:
Any potentially-significant murmurs must be reviewed by a senior colleague. In addition, a chest X ray and ECG must be performed as well as 4-limb blood pressures. Discuss early echocardiography with the consultant.

C) If limb saturations <95% and/or baby is symptomatic:
See "Cyanosis (+/- Respiratory distress)"

Cardiovascular collapse

Circulatory collapse, with diminished peripheral pulses and poor systemic perfusion, is a classic presentation of a systemic outflow tract obstruction, such as coarctation of the aorta, critical aortic stenosis, interrupted aortic arch and hypoplastic left heart syndrome[2,3]. The decompensation occurs as a result of closure of the ductus arteriosus. Cyanosis is not usually present. Care must be taken to distinguish this picture from that of sepsis, which may present in a very similar fashion, and if in doubt treatment for sepsis should be commenced alongside investigation and management of suspected CHD.

The **emergency management** of any baby presenting in the above manner is:

- Assess & resuscitate according to A(Airway), B(Breathing) & C(Circulation)
- Full examination, noting presence of murmurs & quality of femoral pulses, 4-limb blood pressures, respiratory distress, hepatomegaly and central & peripheral perfusion
- Support ventilation if needed
- Secure vascular access x2
- Admit to the neonatal unit (or other appropriate unit) and inform consultant
- Bloods for FBC/U&E/CRP/blood culture/blood gas including blood sugar
- Commence first-line antibiotics
- Correct metabolic acidosis as indicated
- In addition, inotropic support may be required (particularly in hypoplastic left heart syndrome where, once the ductus has re-opened, the ratio of blood flow in the pulmonary and systemic circulations must be balanced)
- CXR; urgent echocardiography if available
- Consider invasive blood pressure monitoring
- If duct-dependent CHD is suspected, the mainstay of treatment is to re-open the ductus arteriosus with prostaglandin E2– see neonatal formulary for details of administration.
- Early discussion with a cardiology unit is mandatory, both for management advice and potential transfer

Appendix 1:

Hyperoxia (Nitrogen Washout) Test

This test differentiates between cardiac and respiratory causes of cyanosis.

1. Place the baby in 100% oxygen for 10 minutes, in order to completely fill the alveoli with oxygen.

2. If the paO2 rises above 100mmHg (14kPa) (or oxygen saturations to 100%) then the cause of the cyanosis is more likely to be respiratory or central (e.g. sepsis).

3. If there is no/little change in the paO2 (<4kPa), or if it rises but stays below 100mmHg/14kPa, the cause is more likely to be due to an intracardiac right-to-left shunt.

Exceptions are TAPVD (which may respond to high-flow oxygen due to the large pulmonary blood flow), or conversely in very severe respiratory disease, or with large intrapulmonary shunting (which will NOT respond to high-flow oxygenation).

References

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