**CYANOTIC CONGENITAL HEART DISEASE**
**INCLUDING HYPOPLASTIC LEFT HEART SYNDROME (HLHS)**
**AND OTHER DUCT-DEPENDENT LESIONS DIAGNOSED ANTEnatalY**

**DEFINITION**

'Blue' refers to central cyanosis (e.g. colour of tongue and gums). It is very difficult to see with the naked eye before the ductus arteriosus reaches a critical closing point

*Any baby presenting as blue has a critically small or closed duct and is a neonatal emergency requiring consultant input. These babies can deteriorate very quickly*

**Differential diagnosis**

Without echocardiography, clinical distinction between persistent pulmonary hypertension (PPHN) and a duct-dependent pulmonary circulation can be extremely challenging

**Lungs**
- Persistent pulmonary hypertension of newborn
- Congenital diaphragmatic hernia
- Congenital lung lesions (e.g. pulmonary lymphangiectasia)

**Heart**
- Obstruction of blood flow from heart to lungs
- Obstruction of blood flow from lungs to heart
- Mixing oxygenated and deoxygenated blood
- Total disconnection of pulmonary and systemic circulations

**SYMPTOMS AND SIGNS IN CARDIAC DISEASE**

- Central cyanosis
- Usually limited signs of respiratory distress
- Murmur, in the minority
- Hepatomegaly
- Poor perfusion

**INVESTIGATIONS**

- Chest X-ray
- 'classic' appearance (e.g. 'boot shaped' heart) is unusual
- Blood gas including lactate
- Echocardiogram
- 4-limb BP (>20 mmHg difference between an upper and lower limb is abnormal)
- Pre- and postductal saturations (saturation of <95% in both limbs or >3% difference is significant)
- Nitrogen washout test (carries risk of duct closure: discuss with consultant first) to differentiate between respiratory (parenchymal) and cardiac cause of cyanosis including PPHN
  - baseline saturation (and blood gas if arterial line in-situ)
  - place baby in 100% ambient oxygen for 10 min
  - if there is respiratory pathology, saturations should rise to >95%

**IMMEDIATE MANAGEMENT**

Call consultant

*A cardiac baby presenting collapsed and/or cyanosed is a challenging neonatal emergency*
Airway
- Intubate and ventilate all babies presenting profoundly cyanosed or collapsed
- If apnoea occurs secondary to a prostaglandin infusion, intubate baby but do not alter infusion

Maintain ductal patency
- Open duct with dinoprostone (prostaglandin E₂, prostin E₂)
- Start at 5 nanogram/kg/min, may be increased to 40 nanogram/kg/min on advice of cardiologist
- if dinoprostone not available, use prostaglandin E₁ (Alprostadil); see local formulary
- Make fresh solution every 24 hr
- Be vigilant: 10% will develop apnoeas on this low dose regimen

Ventilation
Indications
- Significant apnoeas
- Congestive cardiac failure
- Respiratory failure

Technique
- Avoid hyperventilation, which can increase pulmonary blood flow
- initial settings: PEEP 4–5 cm H₂O and FiO₂ 0.21, adjusted accordingly
- Aim for:
  - PaCO₂ 5–7 KPa
  - PaO₂ 4–6 KPa
  - pH 7.35–7.40
  - SpO₂ 75–90%, although many will run higher in room air

Inotropes
- If signs of peripheral underperfusion persist, arrange local echocardiography (if available) to assess contractility
  - if confirmed, start dobutamine 5–10 microgram/kg/min by continuous IV infusion
  - if no improvement to peripheral circulation, consider nitroprusside 0.5–8 microgram/kg/min by continuous IV infusion after discussion with cardiac team

Monitor
- Monitor blood pressure invasively using a peripheral arterial cannula, not UAC
- Titrate infusion to keep to SpO₂ >75–90%
- need to balance pulmonary and systemic circulations:
  - too high an SpO₂ compromises LV output and worsens hypotension
- Assess cardiac output, likely to be low when:
  - tachycardia persists
  - BP remains low
  - acidosis persists
  - lactate elevated or rising
  - poor peripheral perfusion
  - When cardiac output low:
    - ensure prostaglandin infusion adequate
    - ensure adequate intravascular volume
    - correct anaemia
  - inotropes (dobutamine) may be required for tissue oxygen delivery and hypotension (as usually started for elevated lactate and reduced perfusion long before hypotension arises)

SUBSEQUENT MANAGEMENT
TRANSFER

It is imperative that baby is kept warm and normoglycaemic
Discuss further management and transfer with regional cardiac centre
Babies who respond to a prostaglandin infusion do not need to be transferred out-of-hours
Appropriately skilled medical and nursing staff are necessary for transfer

Intubation

**Do not intubate routinely for transfer**
*An intubated baby requires a cardiac centre ITU bed*

- Intubate if:
  - continuing metabolic acidosis and poor perfusion
  - long-distance transfer necessary
  - significant inotropic support in sick baby needed
  - apnoea
  - recommended by cardiac team

**DISCHARGE FROM CARDIAC CENTRE**
Patient may go home or return to a paediatric ward or neonatal unit

**Management plan**
- Regardless of outcome, obtain a management plan from cardiac centre, defining:
  - medication, including dosage
  - acceptable vital signs (e.g. saturations)
  - follow-up arrangements

**HYPOPLASTIC LEFT HEART SYNDROME (HLHS)**
*(AND OTHER DUCT-DEPENDENT LESIONS DIAGNOSED ANTENATALLY)*

**BACKGROUND**
- Malformation affecting left ventricle, aorta and valves
- Degree of hypoplasia varies considerably and, in many cases, lesion is dynamic with progression during pregnancy
- Recently palliative/corrective surgery (Norwood procedure and its modifications) has become increasingly successful in prolonging survival with a view to later heart transplantation

**ANTENATAL MANAGEMENT**
- Record management plan in mother’s notes and in antenatal counselling letter (with additional copy of both on neonatal unit)
- After delivery and stabilisation at local maternity unit, arrange non-urgent transfer to regional cardiac surgery centre for full cardiology assessment
- If closed or small (restrictive) atrial septum puts fetus at risk of severe pulmonary congestion and hypoxaemia shortly after birth, urgent septostomy immediately after delivery at maternity unit may be recommended, despite a generally poor outcome

**Postnatal diagnosis**
- Some babies will present when duct closes, anytime during neonatal period and early infancy, particularly where a left heart lesion has developed later in gestation

*It is important to consider diagnosis of HLHS in any baby presenting with 'shock'*
MANAGEMENT

Aim to maintain patency of (or open a closed) ductus arteriosus, and optimise systemic perfusion

- In HLHS, single right ventricle supplies blood to systemic and pulmonary circulations. Excess flow in one circulation will lead to a significant decrease in blood supplied to other circulation
- Greatest risk is excessive blood flow through pulmonary circulation with systemic underperfusion, presenting with:
  - tachypnoea;
  - poor peripheral perfusion; and
  - metabolic acidosis
- Poor peripheral perfusion can lead to cerebral injury and necrotising enterocolitis (NEC)

Resuscitation
- Neonatal team must be present at delivery to ensure baby establishes effective respirations and becomes pink
- Although no increased requirement for resuscitation, it is extremely important to avoid hypoxia and hyperoxia
- If stable, allow short cuddle with parents before transfer to neonatal unit, but keep warm to avoid hypothermia
- Admit to unit for routine monitoring and investigations including blood gases and avoid acidosis

FURTHER MANAGEMENT
See Cyanotic congenital heart disease section of this guideline