

CYANOTIC CONGENITAL HEART DISEASE

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 seen as white peripheries

INVESTIGATIONS

- Chest X-ray
- oligoemia/plethora/congenital anomaly
- 'classic' appearance (e.g. 'boot shaped' heart) is unusual
- Echocardiogram
- 4-limb BP (>20 mmHg difference between an upper and lower limb is abnormal)
- Pre- and postductal saturations (>3% difference is abnormal)
- 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 RESUSCITATION

***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

Stable babies found to be desaturated during monitoring for a murmur do not require intubation

- If apnoea occurs secondary to a prostaglandin infusion, intubate baby but do not alter infusion

Breathing

- If ventilation required for prostaglandin-induced apnoeas, ventilate in air with PEEP 4-6 cm plus compliant lung ventilation PIP, inspiratory times and rate. See **Ventilation** guideline
- Adjust ventilation to maintain
 - PaCO₂ 5-7 kPa
 - SaO₂ 80-90%
 - pH ≤7.4

Circulation

- Vascular access with 2 IV cannulae or UVC. See **Umbilical venous catheterisation** guideline

Presence of cyanosis and a murmur suggests baby likely to respond to prostaglandin infusion

- Prostaglandin infusion to maintain ductal patency
- open duct with **dinoprostone** (prostaglandin E₂, prostin E₂), see **Neonatal Formulary**. Start at 5 nanogram/kg/min, may be increased to 40 nanogram/kg/min, but only on advice of cardiologist

Monitor

- Monitor blood pressure invasively using a peripheral arterial cannula, not UAC
- Titrate infusion to keep to SpO₂ >75% and **mean BP no greater than** gestation in weeks for first 48 hr (a 34 week baby should have a mean BP of 34 mmHg)
- need to balance pulmonary and systemic circulations:
too high an SpO₂ compromises LV output and worsens hypotension
too high a BP can reduce pulmonary blood flow and SpO₂
- Assess cardiac output, likely to be low when:
 - tachycardia persists
 - BP remains low
 - acidosis persists
 - **lactate elevated or rising**
 - peripheral perfusion poor (white peripheries)
- When cardiac output low:
 - **ensure prostaglandin infusion adequate**
 - ensure adequate intravascular volume
 - correct anaemia
 - inotropes (**dobutamine**) may be required for hypotension

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
- inotropic support needed
- apnoea occurring
- recommended by cardiac team

DISCHARGE FROM CARDIAC CENTRE

Patient may go home or return to a paediatric ward or neonatal unit, possibly [undergoing](#) a prostaglandin infusion whilst awaiting surgery or for continuing care after a palliative procedure (e.g. septostomy)

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