CONGENITAL HEART DISEASE: DUCT-DEPENDENT LESIONS

[Including hypoplastic left heart syndrome (HLHS) and left-sided outflow tract obstructions]

INTRODUCTION

Ductal-dependent congenital heart lesions are dependent upon a patent ductus arteriosus (PDA) to supply pulmonary or systemic blood flow, or to allow adequate mixing between parallel circulations

Duct-dependent congenital heart disease can be broadly divided into 3 categories

1	Mixing lesions e.g. transposition of great arteries (TGA)	Usually presents as cyanosis ('blue baby')
2	Obstruction to pulmonary circulation e.g. pulmonary or tricuspid atresia, Fallot's tetralogy, critical pulmonary stenosis	Usually presents as cyanosis ('blue baby')
3	Obstruction to systemic circulation e.g. HLHS, critical aortic stenosis, coarctation of aorta, interrupted aortic arch	Usually presents as poor perfusion (shock)

Differential diagnosis of central cyanosis ('blue baby') or persistently low SpO_2 (<95%)

- Cyanosis is the abnormal blue discoloration of skin and mucous membranes

Without echocardiography, clinical distinction between significant persistent pulmonary hypertension (PPHN) and a duct-dependent pulmonary circulation can be extremely challenging.
If cause in doubt and echocardiogram cannot be obtained, discuss starting prostaglandin urgently with on-call consultant, as can also be beneficial in PPHN

Cardiac causes of central cyanosis

- Duct-dependent lesions (see above)
- Other cardiac conditions e.g. anomalous pulmonary venous drainage, Fallot's tetralogy, truncus arteriosus etc.

Respiratory causes of central cyanosis

- Persistent pulmonary hypertension
- Other respiratory conditions, e.g. congenital pneumonia, pneumothorax, meconium aspiration, congenital diaphragmatic hernia, respiratory tract obstruction

Other rare causes of central cyanosis

- Methaemoglobinemia

Differential diagnosis of babies presenting with poor perfusion (shock)
Cardiac causes of shock

- Duct-dependent lesion (see above)
- Other cardiac causes e.g. arrhythmias (supraventricular/ventricular tachycardia), cardiomyopathy etc.

Other causes of shock

- Sepsis, bleeding, dehydration, metabolic

RECOGNITION AND ASSESSMENT OF DUCT-DEPENDENT LESIONS

In-utero (antenatal) diagnosis

- If diagnosed in-utero, see management plan in mother's healthcare record
- Deliver at local NNU or NICU equipped for the degree of congenital heart disease.

Stabilise before non-urgent transfer to regional paediatric cardiac centre for full cardiology assessment

- For all antenatally diagnosed cases of TGA, antenatal care to be transferred to fetal medicine in a regional NICU attached to the tertiary cardiac centre (e.g. Birmingham Women's Hospital – refer to local pathway) with a plan to be delivered there (as may need atrial septostomy)
  - if urgent septostomy indicated e.g. postnatally diagnosed TGA, contact KIDS NTS urgently (see **Transport and retrieval** guideline)
- Neonatal team meet parents pre-delivery
- In some cases of HLHS or complex congenital heart disease, comfort care plan may be in place antenatally – clarify with cardiac team and parents before delivery
- When delivery expected, notify on-call neonatal consultant, NNU and paediatric cardiology team at local referral centre

Postnatal

- Some babies, particularly if left heart lesion developed later in gestation, will present when duct closes
  - can happen at any time during neonatal period and early infancy
  - baby often asymptomatic before duct closes

A baby presenting with cyanosis or shock is a neonatal emergency requiring consultant input. These babies can deteriorate very quickly

Signs of duct-dependent cardiac disease

- Central cyanosis and/or SpO_2 <95%
- Poor perfusion and shock
- Weak or absent femoral pulses
- Usually limited signs of respiratory distress
- Murmur (in some) (see **Cardiac murmurs** guideline)
- Hepatomegaly or other signs of cardiac failure

Investigations

- Chest X-ray
  - oligaemia/plethora/congenital anomaly
  - 'classic' appearance (e.g. 'boot-shaped' heart) is unusual
- Blood gas including lactate
- Echocardiogram if available
- Blood pressure in right upper limb and a lower limb (>20 mmHg difference between upper and lower limb is abnormal)
- Preductal (right upper limb) and postductal (lower limb) saturations (SpO_2 <95% in both limbs or >2% difference is significant) (see **Pulse-oximetry screening** guideline)
- Modified hyperoxia test (carries risk of duct closure: discuss with consultant first) to differentiate between respiratory (parenchymal) and cardiac cause of cyanosis including baseline saturation (and blood gas if arterial line *in situ*)
  - place baby in 100% ambient oxygen for 10 min
  - if there is respiratory pathology, SpO_2 usually rises to ≥95%

IMMEDIATE MANAGEMENT

A suspected cardiac baby presenting collapsed, shocked and/or cyanosed is a challenging neonatal emergency, discuss commencement of prostaglandin infusion urgently with consultant.
Discuss urgently with cardiac centre and KIDS NTS (see* Transport and retrieval *guideline)

Immediate post-delivery and resuscitation

- If antenatally diagnosed duct-dependent lesion, tier 2 neonatal staff to be present at delivery
- Do not delay resuscitation of baby if required (see **Resuscitation** guideline)

- Check SpO_2 using pulse oximetry
- Once stable, transfer baby to NNU immediately in transport incubator (if on saturation monitor, SpO_2 75–85% should be acceptable for babies with antenatal diagnosis of duct-dependent cyanotic heart lesion)
- if cyanotic heart lesions suspected but not confirmed postnatally, manage initially by trying to achieve maximum SpO_2 possible

Stable babies with normal breathing and SpO_2 ≥75% may not require intubation

Management in NNU

- Aim to maintain patency of (or open a closed) ductus arteriosus, and optimise systemic perfusion
- Commence prostaglandin infusion (as per antenatal plan if known) through peripheral IV line, or long line (see **Prostaglandin infusion** guideline)
- two venous access lines recommended to ensure reliable infusion
- Unless access difficult, avoid umbilical venous line [cardiac centre may need umbilical venous catheterisation (UVC) for septostomy]; if multiple infusions (e.g. inotropes) required, discuss UVC with on-call consultant/cardiac team
- Use **dinoprostone** (prostaglandin E2, prostin E2) (see **Prostaglandin infusion** guideline)
- start IV infusion at 5–15 nanogram/kg/min as indicated; dose may be increased up to 50 nanogram/kg/min if no response within 1 hr
- oral dinoprostone used temporarily on very rare occasions when IV access is extremely difficult (see **Neonatal Formulary**)
- if dinoprostone not available, use prostaglandin E1 (alprostadil) (see **Neonatal Formulary** for dose)
- make fresh solution every 24 hr
- **Be vigilant**: if apnoea occurs secondary to a prostaglandin infusion, intubate baby but do not reduce infusion dose (see **Intubation** guideline)
- Discuss management with cardiac team at regional paediatric cardiac centre
- Echocardiogram if available
- If any evidence of hypoperfusion (e.g. base deficit >5, lactate >3, hypotension, cool peripheries), give sodium chloride 0.9% 10 mL/kg IV fluid bolus

Monitor

- SpO_2
- Heart rate and ECG
- Blood gases (including lactate) and avoid acidosis
- Blood pressure (preferably using a peripheral arterial cannula – avoid umbilical lines – if UAC required, discuss with on-call consultant)
- Avoid hypothermia

Ventilation (see also **Ventilation** guidelines)

Indications

- If intubation not needed as emergency, discuss with KIDS NTS/cardiac centre (see **Transport and retrieval** guideline)
- Severe hypoxaemia, acidosis and cardiorespiratory failure
- Apnoea after starting prostaglandin infusion
- dose >20 nanogram/kg/min (review need for such a high dosage in stable baby)
- Features of high pulmonary flow in case of HLHS
- Elective ventilation, if preferred by paediatric cardiologist or retrieval team lead

Technique

- Use sedation/muscle relaxants as needed
- Avoid hyperventilation – can increase pulmonary blood flow
- Use supplemental oxygen judiciously if SpO_2 <75%
- Initial settings: PEEP 4–5 cm H_2O, low mean airway pressure, tidal volume 4–6 mL/kg and FiO_2 0.21, adjusted accordingly
- Aim for:
- $PaCO_2$ 5–7 kPa

- PaO_2 4–6 kPa
- pH 7.30–7.40
- SpO_2 75–85% (although many will run higher in room air)

Inotropes
- If signs of peripheral under-perfusion, discuss using fluid boluses and inotropes (e.g. dobutamine, milrinone etc.) with cardiac centre
- Arrange local echocardiography (if available) to assess contractility

Restrictive atrial septum
- Signs:
- severe cyanosis
- cool peripheries
- pallor
- respiratory distress
- X-ray signs of pulmonary oedema with relatively normal heart size. In contrast, if atrial septum is non-restrictive, pulmonary congestion with cardiomegaly and prominent right heart border is likely
- May require balloon atrial septostomy as an urgent procedure at cardiac centre. If too unstable for transfer or no beds at cardiac centre, discuss with KIDS NTS and cardiac team about the possibility of this emergency septostomy procedure being done by transferring directly to theatres at cardiac centre (see **Transport and retrieval** guideline)
- due to change in septostomy equipment, outreach septostomy not possible in a neonatal unit

High pulmonary blood flow (especially in left-sided lesions such as HLHS)
Presentation
- If there is too much pulmonary blood flow due to pulmonary 'steal' phenomenon, baby may have:
- high or near normal saturations
- metabolic acidosis with a rising lactate
- low blood pressure (especially low diastolic)
- cool peripheries
- tachycardia

Management
- Aim is to improve perfusion and acidosis by balancing systemic versus pulmonary circulation
- Discuss urgently with cardiac centre
- Intubate and ventilate (technique as above)
- Fluid boluses and inotropes as needed

PARENT COMMUNICATION
- It is important that parents are kept informed and updated regularly during management
- Parent leaflets for specific heart conditions are available from British Heart Foundation website www.bhf.org.uk/