Management of Duct-dependant congenital heart defects

For more detailed information, please read the complete guideline

Clinical presentation:

- Persistent cyanosis despite supplemental oxygen
- Significant O2 requirement with apparently normal CXR/work of breathing
- Acute cardiorespiratory collapse with cardiogenic shock and/or hypoxia
- Apnoea
- Feeding difficulties due to increased breathlessness
- Signs of heart failure: eg tachycardia, tachypnoea, hepatomegaly
- Murmur (not always present)
- Absent/ weak femoral pulses

FEMORAL PULSES

Present

Suspect duct-dependant Pulmonary circulation:

- Tetralogy of Fallot
- Pulmonary Atresia/stenosis
- Tricuspid Atresia

Absent

Suspect duct-dependant systemic circulation:

- CoAo
- HLHS
- Critical Ao stenosis
- TAPVD

d-TGA

Initial management & Stabilisation

- Monitor pre & post ductal saturations
- Add supplemental O2 to achieve **SatO2 in the 75-85% range**
- Blood gases: monitor pH, lactate, pCO2, Hb, Ca++, glucose
- 12-lead ECG and Echo as soon as feasible
- 4-limb blood pressure (>20 mmHg difference between upper and lower limbs is abnormal)
- If Duct dependant lesion start **Prostin infusion** @ 5-10 ng/kg/min
  - Ensure dedicated line (ideally via Long line, UVC or central line)
  - Consider doubling dose every 20 mins if no improvement after discussion with Cardiology and PaNDR Consultant
- If **persistent hypoxemia** despite prostin infusion consider:
  PPHN→ consider trial of NOi (see full guideline PPHN separately)
  TGA with intact ASD→ ***needs emergency balloon Atrioseptostomy***
- **If persistent systemic hypoperfusion:**
  Likely due to critically obstructed systemic circulation
  Increase prostin infusion rate
  Review need for fluid bolus +/- inotropic support

Complications during stabilisation & transfer

- Apnea (due to high rate prostin infusion)
  Acute desaturation in ventilated child assess for DOPES
  Ensure prostin infusion running appropriately & line patent
  If duct-dependant Pulm circulation treat as Cyanotic spell (morphine, fluid bolus and increase systemic vascular resistance)
- Hypotension: give 3-5 ml/kg fluid and reassess
- Arrhythmias: keep K+>3.8, Ca2+ >1.2 and Mg2+ >1

Differential diagnosis of cyanosis in newborn:

PPHN
Primary pulmonary disease
Sepsis
Inborn errors of metabolism
Methemoglobinemia

Indications for intubation:

- Apnoea needing bag-valve-mask ventilation
- Shock causing profound metabolic acidosis
- Severe respiratory failure
- High dose Prostaglandin (typically >30nanograms/kg/min)

Side effects of Prostin infusion

Hypotension
Hypoglycaemia (check blood sugar 2 hourly initially)
Apnoea (usually at doses > 10 nanograms/kg/min)
Fever

Prostin infusion maker:

Use dinoprostine (PGE2)
Weight (in kg) x 15 micrograms = (micrograms) to total volume 50ml of 5% glucose
Dose range 5-50 nanograms per kg per minute (1-10 ml /hour)
Doses over 50 ng/kg/min to be discussed with Paediatric Cardiologist

References:

1)Penny DJ. Management of the neonate with symptomatic heart disease. Arch Dis Child Fetal Neonatal Ed 2001
2)Auckland District Healthboard Newborn Guidelines
3)NCCU Clinical Guidelines, Neonatal Cardiac Conditions, King Edward Memorial/Princess Margaret Hospitals, Perth, Western Australia
4)Marino BS, Bird GL, Wernovsky G. Diagnosis and management of the newborn with suspected congenital heart disease. Clin Perinatol. 2001

Joint PIC/NIC guideline on the Management of Duct Dependant Congenital Cardiac Lesions

1 Scope

For use within the Paediatric and Neonatal Decision Support and Retrieval Service (PaNDR) for the East of England.

2 Purpose

This is a departmental guideline written to guide the initial management of Duct Dependant Congential Cardiac Lesions within the PaNDR Transport Service.

This is a jointly written guideline and applies to both the Neonatal and Paediatric Transport Teams. In-hospital management may follow a similar process but this guidance is written primarily from a transport perspective.

3 Definitions

PaNDR – The Paediatric and Neonatal Decision Support and Retrieval Service
ECG – electrocardiograph
Echo – echocardiograph
CXR – Chest X-ray
TGA – Transposition of the Great Arteries
PPHN - Persistent Pulmonary Hypertension
ECMO – Extracorporeal Membrane Oxygenation
PVR – Pulmonary vascular resistance
iNO – inhaled nitric oxide

4 Introduction

PaNDR is commissioned to undertake transfers of infants with suspected duct-dependant cardiac lesions to a cardiac centre for further management. Stable transfers could be performed either by the Neonatal or the Paedatric team. However, the Paediatric team should be involved for unstable transfers.

All referrals for neonates or young children with suspected or confirmed duct dependent congenital heart disease should be discussed with the cardiology team and cardiac intensive care consultant at the receiving hospital should the clinical status of the patient requires respiratory and/or cardiovascular support.

In the North Thames region , including the East of England, the reference units would be Great Ormond Street Hospital or the Royal Brompton

Hospital and in the South Thames region it would be the Evelina Children's Hospital. Alternatively, if the patient's condition and diagnosis have already been made and discussed elsewhere, Leicester Royal Infirmary and the Birmingham Children's Hospital are the second closest cardiac centers for the East of England.

If ECMO is required (*see ECMO guideline separately*), referral to the ECMO Consultant on call should be made promptly.

The presence of a duct dependent cardiac lesion may have been suggested by antenatal
ultrasound scanning, or by clinical presentation in the first few days of life.

5 Differentials

Duct dependent systemic circulation:

- Coarctation of the aorta
- Critical aortic stenosis
- Hypoplastic left heart syndrome

Duct dependent pulmonary circulation:

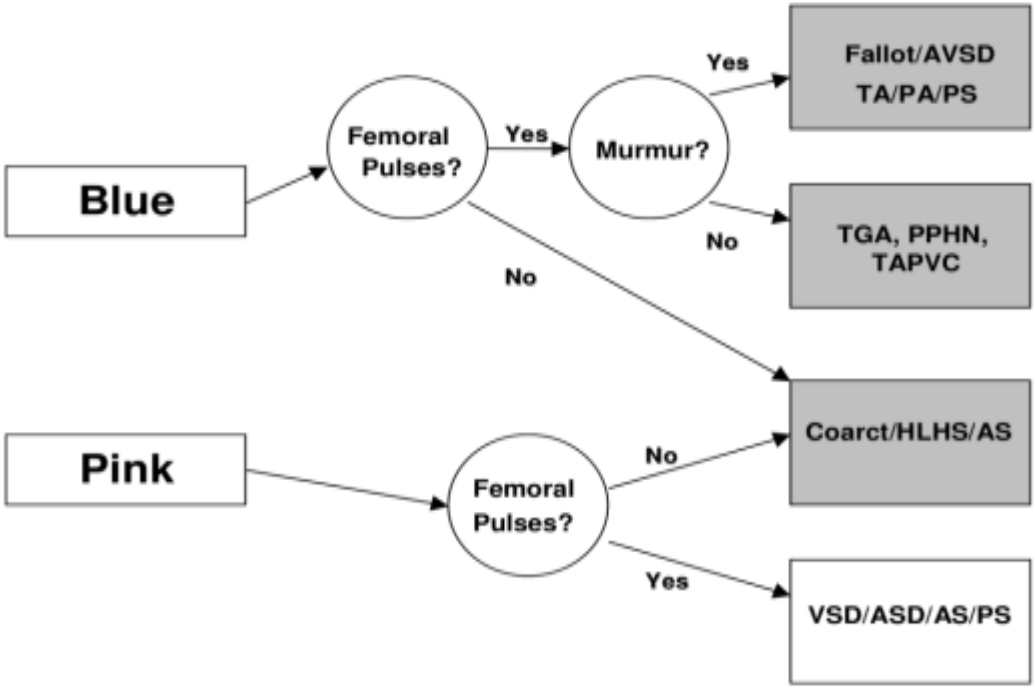
- Pulmonary atresia
- Critical pulmonary stenosis
- Tricuspid atresia
- Tetralogy of Fallot

Duct dependent pulmonary and systemic circulation:

- Transposition of great arteries (TGA)

Differential diagnosis of cyanosis in newborn:

- Persistent Pulmonary Hypertension of the Newborn (PPHN)
- Primary pulmonary disease
- Sepsis
- Metabolic disorders
- Methaemoglobinaemia



Grey box = potential duct dependent lesion: START PROSTIN
Consider iNO if pulmonary hypertension likely

6 History and Examination

Duct-dependant cardiac lesions are a varied group of conditions and present in a variety of ways. Some clues may include:

- Persistant cyanosis despite supplemental oxygen
- Significant oxygen requirement with apparently normal CXR/work of breathing
- Acute cardiorespiratory collapse with cardiogenic shock and/or hypoxia
- Apnoea
- Feeding difficulties due to increased breathlessness
- Signs of heart failure: eg tachycardia, tachypnoea, hepatomegaly
- Murmur (not always present)
- Absent/ weak femoral pulses

Important details to be taken at the time of referral include:

- Antenatal scans and family history
- History of labour, delivery and resuscitation
- Time course of signs/ symptoms
- Cyanosis - present from birth or time of onset
- Perfusion, pulses and four limb blood pressures
- Presence of murmur

- Presence of hepatomegaly
- Blood gases and lactate levels
- Chest X-ray: cardiac ratio, contour and vasculature
- ECG if suspicion of arrhythmia (rarely diagnostic of individual lesions)
- Echo findings if skills and experience locally available

7 Stablisation - ABC

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

Airway and Breathing

Indications for intubation:

- Apnoea needing bag-valve-mask ventilation
- Shock causing profound metabolic acidosis
- Severe respiratory failure
- High dose Prostaglandin (typically >30nanograms/kg/min)

- Monitor pre and post ductal saturations (right hand and either foot)
- Add supplemental oxygen to achieve **saturations in the 75-85% range**
  Discuss saturation target with a paediatric cardiologist and PaNDR team– the appropriateness of supplemental oxygen may vary depending on the underlying diagnosis.
- A **hyperoxia test (see appendix 2)** can be used to support a likely diagnosis of congenital cyanotic heart disease
- Blood Gas: monitor pH, pCO_2, lactate, blood sugar, haemoglobin, ionised calcium.

Circulation

- Ensure at least two good and reliable IV access, especially if prostin is being infused (site umbilical lines if possible or 2 x IV cannulae)
- Treat hypotension initially with 10mls/kg of crystalloid to a maximum of 30ml/kg. Use blood products when indicated
- If persistent hypotension despite initial fluid resuscitation, rationale to start adrenaline should be following discussion with PaNDR Consultant.
- Adrenaline can be started peripherally, then further inotropes guided by response and discussion with PaNDR consultant/paediatric cardiologist
- Echocardiogram if possible at referring centre
- 4 limb blood pressure (>20 mmHg difference between upper and lower limbs is abnormal)
- 12-lead-ECG

- If arrhythmia is part of the clinical presentation, ensure electrolytes are within normal range (Ca^{2+} > 1.1, K^{+} > 3.8, Mg^{2+} > 1).

7.1 Use of Prostaglandin E2 (Dinoprostone)

- Dinoprostone is used to reopen or maintain a patent duct and hence support duct dependent lesions
- Therefore it should be used immediately after birth in patients with known duct-dependant cardiac lesions
- Commence at 5-10 nanograms/kg/minute to maintain duct patency
- If baby is deeply desaturated or with circulatory collapse (severe left heart obstruction) start at 20 nanograms/kg/min
- Consider doubling dose every 20 mins if no improvement after discussion with Cardiology and PaNDR Consultant
- Can be run centrally or peripherally but needs dedicated line (ideally not to be
  run with other infusions)
- Higher doses (up to 100 nanograms/kg/minute) may be needed to re-open a duct that has closed
- Side effects include:
  - Hypotension
  - Hypoglycaemia (check blood sugar 2 hourly initially)
  - Apnoea (usually at doses > 10nanograms/kg/minute – often necessitating intubation prior to transfer)
  - Fever

DO NOT USE PROSTACYCLIN (PGI2) / EPOPROSTENOL / FLOLAN
these are used as a pulmonary vasodilator, NOT to maintain ductal patency

Other:

- If intubated, ensure baby is well sedated e.g. with a morphine infusion at 10-30mcg/kg/hr
- Monitor blood glucose, correct hypoglycaemia
- Take blood cultures and start antibiotics according to local guidelines
- Appropriate thermoregulation, avoiding pyrexia
- Correct hypocalcaemia and hypomagnesaemia
- If persistent acidosis (pH <7.2) despite treating all reversible causes, consider sodium bicarbonate infusion (over 30min) specially if signs of PPHN and/or using high dose inotropes

8 Management following intubation

- Ventilate in air when indicated. Add supplemental oxygen if needed to achieve saturations in the 75-85% range
- Ensure infant is sedated and paralysed to reduce the risk of endotracheal tube dislodgement on transfer and to reduce energy consumption.
- iNO may be needed and should be started if pulmonary hypertension is likely or has been documented
- Keep patient NBM
- Start IV fluids according to local guidelines

9 Troubleshooting

It is essential to rediscuss with the PaNDR team any ongoing problem during stabilisation or transfer

9.1 Persistent systemic hypoperfusion (oliguria, signs of shock, metabolic acidosis and myocardial dysfunction)

- Most common cause due to critically obstructed systemic circulation
- Increase Dinoprostone infusion at rate sufficient to maintain ductal patency after discussion with PaNDR Consultant and Cardiology
- Reduce pulmonary circulation
  - by ventilating with a moderate PEEP (minimum 6 cm H_2O)
  - Maintain systemic saturations 75% – 85%
  - Avoiding respiratory alkalosis with pCO2 5 – 6 kPa
- If signs persist, review need for volume expansion and correction of anaemia (aim Hb above 10 g/dL in cyanotic heart disease)
- Discuss with PaNDR Consultant need for low dose inotropes

9.2 Persistent hypoxemia despite high infusion rate of Dinoprostone

- First of all, check that prostin infusion is running appropriately with no signs of extravasation at the IV access site. Ensure the prescription is correct
- Consider presence of **Persistent Pulmonary Hypertension** (PPHN) associated with cyanotic heart disease (ie premature closure of PDA in utero)
  - Echocardiogram to demonstrate presence of raised PVR
  - Trial of inhaled nitric oxide (iNO) at 20ppm
  - Consider sodium bicarbonate infusion to optimise pH > 7.4 (to be discussed with PaNDR consultant)
  - Increase inotropes or consider vasopressor to increase systemic vascular pressure

- In case of **TGA with restricted ASD or an intact atrial septum,** an urgent discussion with Cardiology is required as emergency balloon Atrioseptostomy can be lifesaving

9.3 Cyanotic heart disease masquerading as lung pathology

Total anomalous pulmonary venous drainage (TAPVD) when obstructed can present with an abnormal CXR suggesting a primary lung disease with pulmonary hypertension.
Although a classical X Ray appearance is described, more frequently, it is indistinguishable from common lung diseases such as pneumonia and meconium aspiration.

10 Potential Complications during stabilisation and transfer

- **Apnoea**
  - Rarely occurs with prostaglandin doses <10nanograms/kg/min
  - Usually responds to stimulation/airway positioning but may ultimately require additional respiratory support

- **Acute desaturation**
  - Assess chest movement/air entry for respiratory causes e.g. blocked ETT, secretions, pneumothorax. Use **DOPES algorithm.**
  - Check that the prostin infusion is running at the correct dose and rate, with no leaks, and that the amount already infused is consistent with the expected amount.
  - Is there an associated drop in blood pressure? This may be the primary problem and the saturations may improve once this is rectified – consider a fluid bolus or increase in inotrope dose
  - Use caution when increasing the FiO_2 in response to desaturation as this may actually be deleterious in some cardiac lesions. It can cause rapid changes in pulmonary vascular resistance, leading to pulmonary overcirculation and a subsequent drop in systemic blood pressure

- **Hypotension**
  - Check all infusions are running and at the correct dose
  - Consider a fluid bolus and/or increase in inotrope dose
  - Consider repeating a gas to exclude significant acidosis as a contributing factor – improving the pH by optimising ventilation (whilst avoiding hypocarbia $PaCO_2$ <4) and/or giving sodium bicarbonate may improve efficacy of the inotropes
  - Ensure electrolyte abnormalities are corrected

- **Cardiac arrhythmias** (*see guideline " Management of cardiac arrhythmias"*)
    - Infants with significant/ prolonged arrhythmias are likely to be moved by the Paediatric team but if encountered unexpectedly during transfer, seek urgent advice from the PaNDR consultant who will liaise with the paediatric cardiologist
    - Ensure defibrillator and pads are attached to the patient's chest during transfer should an acute episode happens

11 Monitoring compliance with and the effectiveness of this document

The PaNDR team will monitor compliance with this document by undertaking regular audits which will be reported back to the consultants and lead nurse. The effectiveness of the document will be monitored by review of any reported incidents via the lead nurse for risk. These incidents will be shared with the team and consideration given to adjusting the guideline if concerns are identified.

12 References

1) Penny DJ. Management of the neonate with symptomatic heart disease. Arch Dis Child Fetal Neonatal Ed 2001;84:F14-145
2) Children's Acute Transport Service guideline: http://site.cats.nhs.uk/wp-content/uploads/2016/01/cats_chd_2015.pdf
3) Auckland District Healthboard Newborn Guidelines: http://www.adhb.govt.nz/newborn/Guidelines/Cardiac/AntenatallyDiagnosedCHD.htm
4) NCCU Clinical Guidelines, Neonatal Cardiac Conditions, King Edward Memorial/Princess Margaret Hospitals, Perth, Western Australia: http://www.kemh.health.wa.gov.au/services/nccu/guidelines/index.htm#sec14

5) Marino BS, Bird GL, Wernovsky G. Diagnosis and management of the newborn with suspected congenital heart disease. Clin Perinatol. 2001 Mar;28(1):91-136. doi: 10.1016/s0095-5108(05)70071-3. PMID: 11265513.

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Appendix 1: Dinoprostine infusion maker

- Make up 15 micrograms per kg body weight of dinoprostone (prostaglandin E2) to a total volume of 50 ml of 5% dextrose
    - Weight (in kg) x 15 micrograms = (micrograms) to total volume 50ml of 5% glucose
- When running at 1 ml/hour = 5 nanograms per kg per minute
- Dose range 5-50 nanograms per kg per minute (1-10 ml /hour)
- Doses over 50 ng/kg/min to be discussed with Paediatric Cardiologist
- Can run via a peripheral intravenous line (dedicated line)

Appendix 2: Hyperoxia test

- Place the baby in 100% oxygen for 10 minutes
- Persistently low oxygen saturations support the diagnosis of congenital cyanotic heart disease (but does not exclude primary pulmonary pathology)
- An increase in 15% of systemic saturations would make cardiac disease unlikely
- Caution: **TAPVD** and **HLHS** may respond when associated with high pulmonary vascular resistance; lung disease with large intrapulmonary shunt may not.

Hyperoxia test results in neonates with cyanosis

	PaO_2 (percent saturation) when FiO_2 = 0.21		PaO_2 (percent saturation) when FiO_2 = 1	$PaCO_2$
Normal	>70 (>95)		>300 (100)	35
Pulmonary disease	50 (85)		>150 (100)	50
Neurologic disease	50 (85)		>150 (100)	50
Methemoglobinemia	>70 (<85)		>200 (<85)	35
Cardiac disease				
Parallel circulation*	<40 (<75)		<50 (<85)	35
Mixing with reduced PBF¶	<40 (<75)		<50 (<85)	35
Mixing without restricted PBFΔ	40 to 60 (75 to 93)		<150 (<100)	35
	Preductal	**Postductal**		
Differential cyanosis◇	70 (95)	<40 (<75)	Variable	35 to 50
Reverse differential cyanosis§	<40 (<75)	>50 (>90)		

Hyperoxia test: The typical results of PaO_2 and percent of oxygen saturation following administration of room air (FiO_2 = 0.21) or 100% oxygen (FiO_2 = 1) to neonates with different causes of cyanosis.

PaO_2: partial pressure of oxygen; FiO_2: fraction of inspired oxygen; $PaCO_2$: partial pressure of arterial carbon dioxide; PBF: pulmonary blood flow.
* D-transposition of the great arteries with or without ventricular septal defect.
¶ Tricuspid atresia with pulmonary stenosis or pulmonary atresia, pulmonary atresia or critical pulmonary stenosis with intact ventricular septum, tetralogy of Fallot.
Δ Truncus arteriosus, total anomalous pulmonary venous connection without obstruction, hypoplastic left heart syndrome, single ventricle without pulmonary stenosis or pulmonary atresia.
◇ Persistent pulmonary hypertension of the newborn, interrupted aortic arch, severe coarctation.
§ D-transposition of the great arteries associated with either coarctation or suprasystemic pulmonary vascular resistance.

Marino BS, Bird GL, Wernovsky G. Diagnosis and management of the newborn with suspected congenital heart disease. Clin Perinatol 2001; 28:91. Copyright © 2001.