NHS Children's Acute Transport Service

Clinical Guidelines

Duct Dependant Congenital Heart Disease

Document Control Information

Author	CATS/NTS	**Author Position**	CC Transport Services
Document Owner	E. Polke	**Document Owner Position**	Service Coordinator
Document Version	Version 4	**Replaces Version**	January 2016
First Introduced		**Review Schedule**	2 Yearly
Active Date	January 2018	**Next Review**	January 2020
CATS Document Number			
Applicable to	**All CATS employees**		

The purpose of this guideline is to standardise the initial management of neonates with duct dependent congenital heart disease across the region.

All referrals for neonates with suspected or confirmed duct dependent congenital heart disease should be discussed with the cardiology registrar and cardiac intensive care consultant at the receiving hospital. In the North Thames Region this 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. If ECMO is required, a referral to the ECMO consultant on call should be made.

Presentation and assessment

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.

Common acute presenting features include

- Cyanosis – often unresponsive to supplemental oxygen (see hyperoxia test below)
- Acute cardiorespiratory collapse with shock.
- In severe cases a neonate can present with severe **cardiogenic shock** and with severe **hypoxia**.
- Associated features:
    - Examination may reveal features of heart failure with tachycardia, tachypnoea and hepatomegaly.
    - Absent or weak femoral pulses
    - Cardiomegaly on chest X-ray
    - Difficulty feeding secondary to increased breathlessness
    - There is not always an audible murmur.

Important things to ask about at referral:

- Antenatal scans and family history
- History of labour, delivery and resuscitation
- Cyanosis- time course and response to supplemental oxygen
- Perfusion, pulses and four limb blood pressures
- Presence of a cardiac murmur or hepatomegaly
- Blood gases and lactate levels
- Chest X-ray: cardiac ratio, contour and vasculature
- ECG
- Echocardiograph

Differential diagnosis

Duct dependent systemic circulation	1. Coarctation of the aorta 2. Critical aortic stenosis 3. Hypoplastic left heart syndrome
Duct dependent pulmonary circulation	1. Pulmonary atresia 2. Critical pulmonary stenosis 3. Tricuspid atresia 4. Tetralogy of Fallot
Duct dependent systemic and pulmonary circulations	1. Transposition of the great arteries
Other potential diagnoses	1. Persistent Pulmonary Hypertension of the Newborn (PPHN) 2. Primary pulmonary disease 3. Sepsis 4. Metabolic disorders 5. Methaemoglobinaemia

Immediate management

Resuscitation of a rapidly deteriorating neonate must take immediate priority. Emergency intubation and ventilation may be indicated for recurrent apnoea, shock or severe respiratory failure, otherwise a more detailed assessment should take place:

Airway and Breathing

- Intubate if indicated.
- Monitor pre and post ductal saturations (right hand/fingers and either foot).
- A hyperoxia test can be used to support a likely diagnosis of congenital cyanotic heart disease.
    - To do this, place the baby in 100% oxygen for 10 minutes. Persistently low oxygen saturations support the diagnosis of congenital cyanotic heart disease (does not exclude primary pulmonary pathology).
- Administer supplemental oxygen as required to maintain oxygen saturations in the range 75-85%.

Circulation

- Site 2 intravenous cannulae (consider umbilical venous access)
- Treat hypotension with 10 ml/kg crystalloid fluid bolus up to a maximum of 30 ml/kg.
- Treat resistant hypotension with dopamine, which may be started peripherally. Add adrenaline as a second line agent if needed. (Appendix 1)
- Commence dinoprostone (prostaglandin E2) to open and/or maintain ductal patency at 5-10 nanograms/kg/minute. (Appendix 2)
- Side effects of dinoprostone infusion are
    - hypotension
    - hypoglycaemia
    - apnoea (More common with doses >10 nanograms/kg/minute and should prompt serious consideration of intubation for any transfer)
    - fever
    - higher doses may be necessary to open a closed duct but this should be discussed with the Paediatric Cardiologist/Cardiac Intensivist/CATS Consultant
- 4 limb blood pressures
- Echocardiogram (if able)
- ECG

DO NOT USE PROSTACYCLIN (PGI2) / EPOPROSTENOL / FLOLAN

-these are used as a pulmonary vasodilator, NOT to maintain ductal patency.

Other

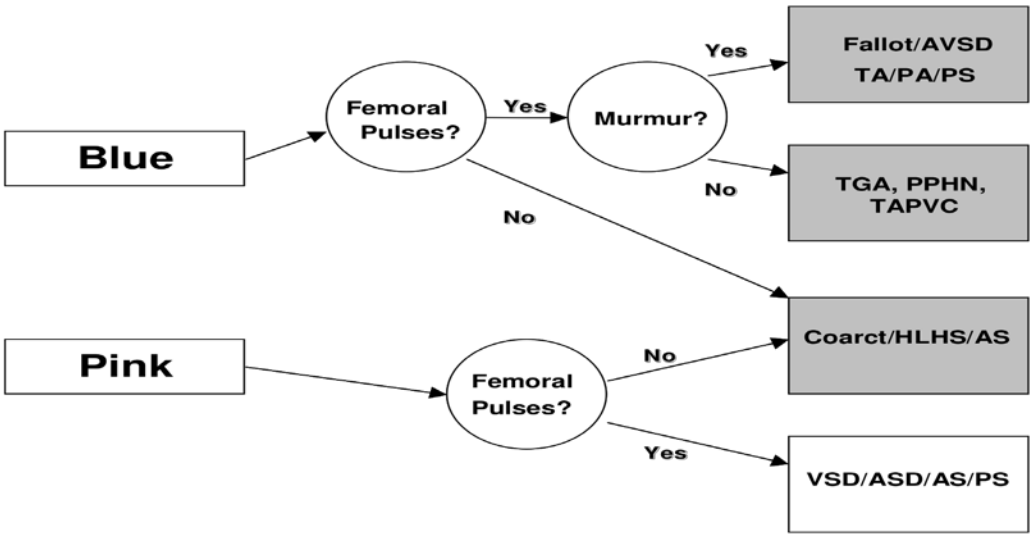
- **Check blood glucose regularly**.

Indications for intubation

- Apnoea
- Shock
- Respiratory failure

Management following intubation

- Sedate well with a morphine infusion. Muscle relaxants should be used to reduce the risk of endotracheal tube dislodgement on transfer and to reduce energy consumption.
- Ventilate in air. Add supplemental oxygen if needed to achieve saturations in the 75-85% range. This is to reduce pulmonary over circulation.
- Inhaled nitric oxide may be needed and should be started if pulmonary hypertension is likely.



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

Dinoprostone worksheet

DO NOT USE PROSTACYCLIN (PGI2)/EPOPROSTENOL/FLOLAN

Make up 15 micrograms per kg body weight of **dinoprostone (prostaglandin E2)** to a total volume of 50 ml 5% glucose.

The resulting solution run at 1 ml per hour delivers 5 nanograms per kg per minute.

Dose range is 5- 50 nanograms per kg per minute (1-10 ml per hour).

Weight (kg) _____ x 15 micrograms =_____(micrograms) to total volume 50ml of 5% glucose

Can run via a peripheral intravenous line

This is a more dilute solution than specified in some local neonatal protocols but is the concentration advocated by the receiving cardiac intensive care units. It reduces the delay caused by intravenous line dead space. Thus the drug is thus delivered to the bloodstream more quickly,

- A more concentrated infusion running at 0.1 ml per hour for example would take a potentially impactful 4 hours to traverse a 24G Neoflon and T-piece connector (total dead space 0.4 ml).
- Local variance in prostaglandin preparation also risks bolus administration when the above recommended dilute infusion is eventually connected to the same line by the receiving or transport teams. Apnoea may result.