NHS Children's Acute Transport Service

# Clinical Guidelines

# Duct Dependant Congenital Heart Disease

***This guideline has been agreed by both NTS & CATS***

## Document Control Information

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## Introduction

*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 it is possible that ECMO will be required, this must be discussed with the ECMO consultant on call.

## 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 presenting features include

- Difficulty feeding secondary to increased breathlessness
- Cyanosis – often unresponsive to supplemental oxygen
- Acute cardiorespiratory collapse with shock.
- Examination may reveal features of heart failure with tachycardia, tachypnoea and hepatomegaly.
- There is not always an audible murmur.
- In severe cases a neonate can present in severe **cardiogenic shock** with absent or weak femoral pulses or with severe **hypoxia**.

## Important things to ask about at referral:

- Antenatal scans and family history
- History of labour, delivery and resuscitation
- Time course
- Cyanosis- present from birth or time of onset
- Perfusion, pulses and four limb blood pressures
- Cardiac murmur
- 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<br>2. Critical aortic stenosis<br>3. Hypoplastic left heart syndrome |
| **Duct dependent pulmonary circulation** | 1. Pulmonary atresia<br>2. Critical pulmonary stenosis<br>3. Tricuspid atresia<br>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)<br>2. Primary pulmonary disease<br>3. Sepsis<br>4. Metabolic disorders<br>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 & 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 isotonic 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
- 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**.

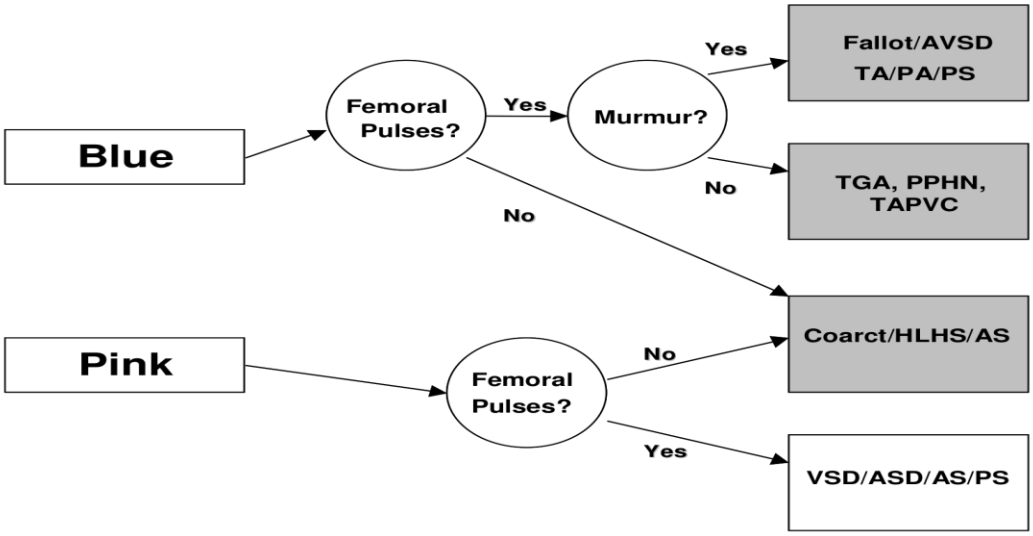
## Indications for intubation

- Apnoea
- Shock
- Respiratory failure

## Management following intubation

- Muscle relax and sedate well. Use a morphine infusion. Muscle relaxants should be used to reduce the risk of endotracheal tube dislodgement on transfer.
- 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 more dilute than has been traditionally used in neonatal practice but is the concentration used in the receiving cardiac intensive care units. This is to ensure that the drug is delivered to the neonate and reduce the lag time to effect due to dead space in the intravenous line.***

For example an infusion running at 0.1 ml per hour would take 4 hours to traverse a 24G Neoflon and T-piece connector (total dead space 0.4 ml).

Using this concentration from the outset also reduces the chance of an inadvertent bolus administration of concentrated prostaglandin, causing apnoea, on changing the infusion in the receiving unit.

## Inotrope Infusion Worksheet

### Dopamine

Make up 15mg per kg body weight of dopamine to a total volume of 50 ml 5% glucose.
The resulting solution run at 1 ml per hour delivers 5 micrograms per kg per minute.

Dose range is 0-20 micrograms per kg per minute (0-4 ml per hour).
Weight (kg) _____ x 15 mg = _____ (mg) to total volume 50 ml 5% glucose
Can run via a peripheral intravenous line for infants <5kg

### Adrenaline

Make up 0.3mg per kg body weight of adrenaline to a total volume of 50 ml 5% glucose.
The resulting solution run at 1 ml per hour delivers 0.1 micrograms per kg per minute.

Dose range is 0-0.5 micrograms per kg per minute (0-5 ml per hour).

Weight (kg) _____ x 0.3 mg =_____ (mg) to total volume 50 ml 5% glucose

MUST be given via a **central venous line or intraosseus needle.**