

Title:	Duct dependent Cardiac lesions		
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Foreword

This document provides guidance for the recognition and management of duct dependant cardiac lesions and neonatal arrhythmias.

Infants with known cardiac lesions / arrhythmias who weigh ≥ 1.5 kg, should normally be transferred by the Children’s Acute Transport Service (CATS). However, it is possible that NTS may become involved in the management of these infants if CATS are unable to undertake the transfer, or if a cardiac lesion comes to light during the referral or assessment process.

Suspected duct dependant cardiac lesions not responding to Prostin are UK Neonatal Transport Group Time critical transfers (meaning the transport team should dispatch dispatches from base or previous mission within one hour from the start of the referring call). For NTS this dispatch time is within 30 minutes.

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.

Quick reference

The main differential diagnoses of duct dependent congenital heart disease are:

- Persistent Pulmonary Hypertension of the newborn (PPHN)
- Sepsis
- Metabolic disease
- Pulmonary disease

Immediate management

ABC approach

Airway and Breathing

- Consider immediate intubation and ventilation if:
 - Recurrent apnoea
 - Shock
 - Severe respiratory failure
 - Prostin dose exceeding 10 nanograms/kg/min
- Monitor pre and post ductal saturations (right hand and either foot)

Circulation

- At least 2 IV cannulae / Consider Umbilical venous catheter
- 4 limb BP, ECG, ECHO if possible
- If hypotension:
 - 10ml/kg 0.9% saline and consider further boluses based on clinical response
 - If remains hypotensive see Management of hypotension guideline for further management)
- If suspect duct dependent cardiac lesion commence Prostin (dinoprostone-prostaglandin E2) to open and/or maintain ductal patency at 5-10 nanograms/kg/min (can be increased up to 20 nanograms/kg/min).

Higher doses (max 100 nanograms/kg/minute) may be necessary to open a closed duct but this should be re-discussed with NTS and Cardiology Consultants

NB: do not use Prostacyclin (PGI2) / Epoprostenol / Flolan

These are used as pulmonary vasodilators and NOT to maintain ductal patency

Doses for Dinoprostone (Prostaglandin E2)

- 1mg/ml solution dinoprostone
- Draw up 15 micrograms X weight (Kg)

- Make up to 50mls of 0.9% Sodium Chloride
- At 1ml per hour – 5 nanograms/kg/min
- Run at 1 – 2ml/hr = 5-10 nanograms/kg/min (higher rates should be discussed with Paediatric Cardiologist)

Monitor for:

Hypotension – prepare saline boluses and dopamine in anticipation

Apnoea – consider intubation if long journeys or where dose exceeds 10 nanograms/kg/min

Fever

Jitteriness, myoclonic jerks, irritability

On-going management

- Post-intubation: Paralyse and sedate to reduce ETT dislodgement risk
- Cautious O₂ use in cyanotic duct dependent cardiac lesions: Aim for saturations 75-85%
- Inhaled Nitric oxide: Should be started if pulmonary hypertension is likely
- Monitor: Hypotension, hypoglycaemia and apnoea are all side effects of Prostin

Background

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

Common presenting features

- Cyanosis – often unresponsive to supplemental oxygen
- Feeding difficulty due to breathlessness
- Congestive cardiac failure: Hepatomegaly, tachypnoea, tachycardia
- Cardiogenic shock: absent / weak femoral pulses, severe hypoxia

Many of the signs mimic other neonatal emergencies and consideration should be given to possible differential diagnoses: PPHN, Sepsis, metabolic disease, Respiratory disease

Important questions at referral

- Antenatal scans and family history
- Labour, delivery, and resuscitation details
- Time course of presentation including timing of cyanosis
- Perfusion: Pulses and 4-limb blood pressure
- Examination findings: Cardiac murmur, hepatomegaly, heart rate, respiratory rate
- Blood gases and lactate levels
- ECG / Echocardiogram findings
- Chest x-ray: Cardiac ratio, contour, vasculature
- Other relevant findings e.g. evidence of sepsis

Classification

Duct dependant lesions may be classified in to 3 main categories:

Duct dependant <i>systemic</i> circulation	<ul style="list-style-type: none">▪ Coarctation of the aorta▪ Critical aortic stenosis▪ Hypoplastic left heart syndrome
Duct dependant <i>pulmonary</i> circulation	<ul style="list-style-type: none">▪ Pulmonary atresia▪ Critical pulmonary stenosis▪ Tricuspid atresia▪ Tetralogy of Fallot
Duct dependant <i>systemic and pulmonary</i> circulations	<ul style="list-style-type: none">▪ Transposition of the great arteries