NTS Clinical/Operational Guideline

Title:	Duct dependant Cardiac Lesions		
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Scope	For use within London Neonatal Transfer Service		
Applies to	*All London NTS staff, All London NTS middle grade doctors, All London NTS Nursing staff, All Neonates transferred by NTS, London Ambulance Vehicles*, St John's Vehicles		

1. Introduction

Infants with known cardiac lesions who weigh ≥ 1.5kg, should normally be transferred by the Children's Acute Transport Service (CATS) in North London, or >2kg should be transferred with South Thames Retrieval Service (STRS) in South London. 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.

2. 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
- 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 (but does not exclude primary pulmonary pathology)
- 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

Referrals

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.

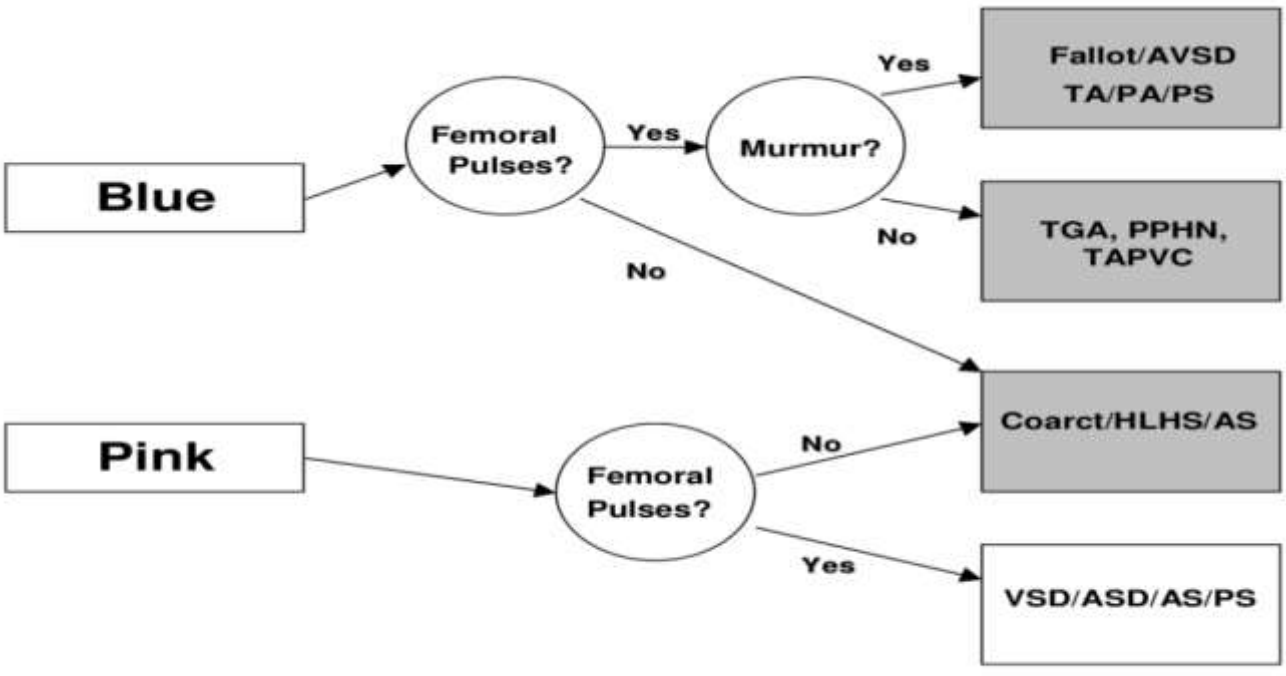
For babies in the North Thames Region who require transfers, this would be
Great Ormond Street Hospital or the Royal Brompton Hospital and

For babies in the South Thames Region who require transfers, this would be
The Evelina Children's Hospital.

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

Flowchart for assessment of baby with suspected Congenital Heart Disease



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

Type of cardiac circulatory malformation and diagnoses

Duct dependant systemic circulation	Duct dependant pulmonary circulation	Duct dependant systemic and pulmonary circulation
• Coarctation of aorta • Critical aortic stenosis • Hypoplastic left heart syndrome	• Pulmonary atresia • Critical pulmonary stenosis • Tricuspid atresia • Tetralogy of Fallot	• Transposition of great vessels with restrictive circulation
Aim for: palpable pulses, resolving acidosis.	*Aim for:* sats 75 – 85%, lactate <2mmol/L.	
Differential diagnosis • Pulmonary hypertension • Sepsis • Metabolic disorders • Primary lung pathology • Obstructed TAPVD – CXR plethoric		*Aim for:* improved oxygenation, BP and acidosis.

3. Process

Take the same approach as all NTS *transfers – see Stabilisation and Preparation for transfer guideline.* These are additional specific considerations to the transfer:

Initial assessment	**Antenatal and family history** Scans +/- antenatal diagnosis - there should be a plan for care following birth if antenatally diagnosed **Labour and delivery** Risk factors for sepsis, resuscitation, condition at birth Time frame to presentation Care plans & discussions with cardiology to date
A	Mostly will be SVIA or receiving non-invasive respiratory support **Indication for intubation:** • Recurrent apnoeas, shock, respiratory failure, Consider intubation if dinoprostone dose >10nanograms/kg/min or higher or potential long transfer.
B	Ventilate in air O2 to achieve sats 75-85% to avoid pulmonary over circulation. Aim PaO2 5kPa, PaCO2 5kPa Monitor pre and post ductal sats

C ***NB there may be no murmur**	At least 2 IV cannula - one for a designated dinoprostone cannula. Umbilical venous catheter +/- UAC if clinical condition requires. 4 limb BP, ECG, ECHO if possible Treat hypotension: IV 10mL/kg 0.9% saline and consider further boluses based on clinical response (max 30mL/kg) Treat resistant hypotension with Dopamine. Use adrenaline second line. **Dinoprostone (Prostaglandin E2) – see under Drugs heading below**
DEF	Consider PPHN and treat accordingly Monitor blood glucose, correct hypoglycaemia Blood cultures + antibiotics Appropriate thermoregulation, avoid pyrexia Correct acidosis Correct hypocalcaemia and hypomagnesaemia
Transfer of the baby may be required despite still being hypoxic and/or acidotic. Keep a close dialogue with NTS Consultant and Cardiology Consultant throughout.	

4. Drugs

Dinoprostone (Prostaglandin E2)
IV infusion, can be given peripherally or centrally

- Antenatal diagnosis, well baby = start 5nanograms/kg/min
- If presents with absent femoral pulses but otherwise well and not acidotic = start 10-20nanograms/kg/min
- If presents acidotic and unwell (late presentation) start 20-50nanograms/kg/min *on discussion with Cardiology. Higher doses may be required
- *(max 100nanograms/kg/min) may be necessary to open a closed duct.*

Assess every 20mins, double dose if no improvement.

Dinoprostone (Prostaglandin E2)

- 1mg/mL solution dinoprostone
- Draw up 15 micrograms X weight (Kg)
- Make up to 50mL with 0.9% Sodium Chloride
- 1mL/hr = 5 nanograms/kg/min
- Run at 1 – 2mL/hr = 5-10 nanograms/kg/min (higher rates should be discussed with Paediatric Cardiologist)

Potential side effects:
Hypotension – *prepare saline boluses and dopamine in anticipation*
Apnoea – *consider intubation if long journeys or where dose exceeds10nanograms/kg/min*
Fever

Do not use Prostacyclin (PGI2) / Epoprostenol / Flolan
These are used as pulmonary vasodilators and NOT to maintain ductal patency.

5. Recordkeeping

Complete the NTS transfer form and BadgerNet as contemporaneous as possible.

6. References

Akkinapally S, Hundalani SG, Kulkarni M, et al. Prostaglandin E1 for maintaining ductal patency in neonates with ductal-dependent cardiac lesions. Cochrane Database Syst Rev 2018; 2:CD011417.

Browning Carmo KA, Barr P, West M, et al. Transporting newborn infants with suspected duct dependent congenital heart disease on low-dose prostaglandin E1 without routine mechanical ventilation. Arch Dis Child Fetal Neonatal Ed 2007; 92:F117.

Lewis AB, Freed MD, Heymann MA, et al. Side effects of therapy with prostaglandin E1 in infants with critical congenital heart disease. Circulation 1981; 64:893.