NETS WA
Newborn Emergency Transport Service

Congenital Heart Disease (CHD)

Scope (Staff):	Nursing and Medical Staff
Scope (Area):	NETS WA

Child Safe Organisation Statement of Commitment

CAHS commits to being a child safe organisation by applying the National Principles for Child Safe Organisations. This is a commitment to a strong culture supported by robust policies and procedures to reduce the likelihood of harm to children and young people.

This document should be read in conjunction with this disclaimer

Aim

Summarise the considerations for the safe retrieval of neonates with congenital heart disease.

Risk

Delays in recognition and/or management of CHD can place neonates at increased risk of deterioration and adverse events. A standardised approach to assessment and management aims to minimise these risks.

Key point

Symptomatic patients with CHD either present with cyanosis, or in cardiac failure/ shock.

General Management Principles

- On the NETS call conference, ask for the neonatal management plan and the cardiac scan reports from the antenatal and postnatal period. In sick cases, consider patching the Cardiology and the PCH NICU/PICU consultant early into the call-conference system.
- Ask about cardiac examination with HR, murmur (type, grade, location), palpable femorals and other systemic examination (resp distress, respiratory support, hepatomegaly, capillary refill, perfusion, neurological examination)
- Request 4 limb blood pressures, pre-post ductal saturations and arterial blood gas

report with paO2 and lactates. Difference in pre-post ductal sats difference (>10%), BP difference (>10 mmhg) with lower limb BP lesser than upper limb BP, absent femorals, and/or high lactates (> 4mmol/L) are helpful markers pointing towards a cardiac pathology in sick infants.

- Request the referring team to establish venous access for administering iv fluids and medications (inotropes, prostaglandins, antibiotics)
- Request for chest xray and ECG. Some cardiac conditions (Ebsteins anomaly, cardiomyopathy) can be associated with arrhythmias.
- Keep NBM for transport, especially those infants on respiratory support.
- For duct dependent cardiac conditions consider urgent (P1) transfer back to PCH for definitive diagnosis and management.
- Look for signs of dysmorphism as many genetic conditions can present with cardiac manifestations.
- Look for other non-cardiac causes such as Vein of Galen Malformation, AV fistula that can present with PPHN or cardiac failure.

1. Presenting as Cyanosis

Differential Diagnosis	
• Transposition of the great vessels. • Pulmonary atresia / critical pulmonary stenosis • Tricuspid Atresia	• Epstein's anomaly • Tetralogy of Fallot • Double Outlet Right Ventricle • Total or Partial Anomalous pulmonary venous return.

Management

It may be difficult to distinguish cyanotic CHD from PPHN in early neonatal life in undiagnosed cases. A simple bedside test ie Hyperoxia test might be helpful to distinguish between respiratory and cardiac causes of cyanosis.

If the oxygen saturation improves to normal after increasing FIO2 to 100% transiently or the paO2 values rise above 100, it usually would point towards respiratory pathology for cyanosis ie PPHN. However, echocardiography is the only gold standard means to diagnose congenital cardiac conditions. See Persistent Pulmonary Hypertension of the Newborn (PPHN).

- Asymptomatic, non-acidotic infants can be considered for minimal intervention after discussion with NETS WA and Cardiology consultants, especially those from KEMH (as per neonatal management plan and post-natal echo) to PCH.
- Unstable and/or acidotic: consider intubation, ventilation in an acidotic or infant in shock with suspected duct dependent congenital heart disease.
- Strongly consider starting PGE1 (Alprostadil) infusion. PGE1 improves pulmonary circulation in cyanotic CHD. Discuss with on-call NETS WA and Cardiology consultants for the starting dose of PGE1 infusion. Standard doses

of starting PGE1 are from 10-20 ng/kg/min. Higher doses (50 ng/kg/min) can be considered if late presentation for opening the ductus arteriosus. Common side effects of PGE1 include:

  - Vasodilation (and therefore hypotension). May need fluid bolus ± inotrope.
  - Apnoea (at higher doses,). Consider assisted respiratory support (eg. intubation and ventilation) if frequent apnoea's (≥2/hour) or 1 episode needing bag and mask ventilation. At lower doses (≤ 15 ng/kg/min) apnoea is less likely, refer to Neonatal Monograph.
  - For longer transports, sick infant, or higher dose of PGE1, consider elective intubation and ventilation. Discuss with the on-call NETS WA consultant.
  - Consider additional peripheral or central venous access especially when needing medications such as PGE1 +/- inotropes associated with long duration transfers. In a known case of Transposition of great vessels, avoid the umbilical venous access if possible as Cardiologist would consider using umbilical venous access for balloon atrial septostomy in PCH.

- Consider sedation in infants who are ventilated. Prefer Fentanyl. Midazolam and Morphine can cause hypotension and hence are less preferred options.
- Target oxygen saturations should be discussed with the on-call Cardiologist for known cyanotic congenital heart disease.
- For unknown/suspected congenital heart disease, to target normal neonatal saturations till arrival at PCH for formal diagnosis.
- Carry inhaled Nitric Oxide for all transport where an infant is suspected to have cardiac disease.
- For infants with suspected cardiac cause and moderate to severe respiratory distress in metro areas, consider using the FABIAN cot that has an additional option for High Frequency Oscillatory ventilation.

2. Presenting as Cardiac Failure

Often present > Day 3 life (when PDA closes).

Differential Diagnosis	
• Left outflow tract obstructive lesions: Coarctation of aorta / interrupted aortic arch. • Hypoplastic left heart. • VSD and other large L-R shunts (usually present much later).	• Arrhythmias (SVT, heart block) (See Cardiac Dysrhythmias management) • Non-cardiac conditions (eg AV fistula, Vein of Galen Malformation)

Management

- Target oxygen saturations should be discussed with the on-call Cardiologist for known congenital heart disease presenting with cardiac failure. For unknown/suspected congenital heart disease, to target normal neonatal saturations till arrival at PCH. (Caution for high oxygen concentrations that can cause pulmonary vasodilatation and reduce the flow from pulmonary to systemic circulation via the ductus arteriosus, counterproductive for duct dependent left sided cardiac conditions such as Hypoplastic left heart syndrome).
- Consider appropriate assisted ventilation modalities (non-invasive or invasive) CPAP or ventilation (positive pressure reduces afterload).
- Strongly consider PGE1 (Alprostadil) infusion for suspected duct dependent left sided congenital cardiac conditions e.g. Coarctation of aorta, hypoplastic left heart syndrome. Discuss with on-call NETS WA and Cardiology consultants for the starting dose of PGE1 infusion. Standard doses of starting PGE1 are from 10-20 ng/kg/min. Higher doses (50 ng/kg/min) can be considered if late presentation for opening the ductus arteriosus.
- Consider Inotrope support (Dobutamine +/- Nor-epinephrine) in cases of neonatal shock secondary to congenital heart disease. The choice of inotropes and dosage should be discussed with the on-call NETS WA and Cardiology consultants. Inotropes that increase the systemic peripheral vascular resistance and afterload (e.g. Noradrenaline, Vasopressin) to elevate blood pressures might be sometimes counterproductive for obstructed left ventricular outflow tract obstruction (coarctation of aorta, hypoplastic left heart syndrome). See Neonatal Medication Monographs and Sepsis and Shock Guideline.
- Diuretics (Frusemide) might be helpful in those infants with congenital heart disease (VSD, PDA) presenting with congestive cardiac failure.
- Look for signs of severe anaemia or blood loss that can worsen the cardiac failure. Consider packed red cell transfusion for those with low Hb after discussion with NETS WA consultant.

Related CAHS internal policies, procedures and guidelines
NETS WA Guidelines • Cardiac Arrhythmias • Persistent Pulmonary Hypertension of the Newborn (PPHN) • Sepsis and Shock CAHS Neonatal Guidelines Congenital Heart Disease Anaemia Neonatal Medication Monograph

This document can be made available in alternative formats on request.

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