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 or in shock.

Presenting as Cyanosis

Differential Diagnosis

- Transposition of the great vessels.
- Pulmonary atresia / critical pulmonary stenosis / Tetralogy of Fallot / other forms RV outflow tract obstruction and Total or Partial Anomalous pulmonary venous return.

Management principles

It may be difficult to distinguish cyanotic CHD from PPHN in early neonatal life. Hyperoxia test may help. See Guideline on Persistent Pulmonary Hypertension of the Newborn (PPHN).

- Asymptomatic, non-acidotic infants: minimal intervention.
- Unstable and/or acidotic: consider intubation, ventilation, PGE1 infusion ± Sodium bicarbonate infusion (rarely).
- To consult with the on call cardiologist before commencing PGE1 (Alprostadil) infusion.
- PGE1 improves systemic circulation in obstructive left heart conditions, and the pulmonary circulation in cyanotic CHD.
- Common side effects of PGE1 include:
    - Vasodilation (and therefore hypotension). May need fluid bolus ± inotrope.
    - Apnoea (at higher doses,). May need ventilation.
    - Typical starting dose is 25-50 ng/kg/min. At lower doses (≤ 15 ng/kg/min) apnoea is less likely, please refer to Neonatal Monograph PGE1 infusion
    - For longer transports, sick infant, or higher dose of PGE1, consider elective intubation. Discuss with the on-call neonatologist/Cardiologist.

Consider sedation in infants who are ventilated. Preferably Fentanyl as Midazolam or Morphine as can cause hypotension.

Please carry inhaled Nitric Oxide for all transport where an infant is suspected to have cardiac disease.

Cardiac Failure

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

Differential Diagnosis

- 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)

Management principles

- Administer Oxygen to maintain normal SpO_2. (To discuss with the cardiologist/neonatologist to get a target saturation level).
- CPAP or ventilation (positive pressure reduces afterload).
- Diuretics (Frusemide 1 mg/kg).
- Inotrope support (Dobutamine or Dopamine). See Guideline on Shock.

- Consider Sodium bicarbonate infusion in cases of severe acidosis.
- PGE1 (Alprostadil) infusion may be indicated, to be discussed with on-call neonatologist and/or cardiologist:
    - PGE1 might be deferred until arrival at PCH in a stable patient and/or short trip back to PCH (< 30 minutes) after discussion with the neonatologist/cardiologist.

Related CAHS internal policies, procedures and guidelines
NETS WA Guidelines • Cardiac Dysrhythmias • Persistent Pulmonary Hypertension of the Newborn (PPHN) • Shock. Neonatal Medication Monograph • PGE1 infusion

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

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