



CLINICAL GUIDELINE	
Congenital Heart Disease (CHD)	
Scope (Staff):	Nursing and Medical Staff
Scope (Area):	NICU KEMH, NICU PCH, NETS WA

This document should be read in conjunction with this [DISCLAIMER](#)

Key Point: Symptomatic patients with CHD either present with cyanosis, or in cardiac failure.

Cyanosis Presenting Day 1

Differential Diagnosis

- Transposition of the great vessels.
- Pulmonary atresia / critical pulmonary stenosis / Tetralogy of Fallot / other forms RV outflow tract obstruction.

Management

- May be difficult to distinguish cyanotic CHD from PPHN. Hyperoxia test may help. See Guideline on [Persistent Pulmonary Hypertension of the Newborn \(PPHN\)](#).
- **If unsure whether the infant has CHD or PPHN, it is safer to administer oxygen, and commence PGE1(Alprostadil) infusion**
- Asymptomatic, non-acidotic infants: minimal intervention.
- Unstable and/or acidotic: consider intubation, ventilation, PGE1 infusion ± Sodium bicarbonate 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.
 - For longer transports, sick infant, or higher dose of PGE1, consider elective intubation. Discuss with the on-call neonatologist.

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).

Management


- Administer Oxygen to maintain normal SpO₂.
- 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 PMH in a stable patient and/or short trip back to PMH (< 30 minutes) after discussion with the neonatologist/cardiologist.

Related CAHS internal policies, procedures and guidelines

NETS WA Guideline

- [Persistent Pulmonary Hypertension of the Newborn \(PPHN\)](#)
- [Shock](#)

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