



CLINICAL GUIDELINE

Congenital Heart Disease: Overview

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| Scope (Staff): | Nursing and Medical Staff |
| Scope (Area): | NICU KEMH, NICU PCH, NETS WA |

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

CHD can be benign and not require any treatment to life threatening requiring immediate life saving action. This protocol will concentrate 'duct' (PDA) dependent CHD lesions most often presenting in the neonatal period. These conditions are often asymptomatic in the early stages but may be detected by routine post-natal SaO₂ protocols and day 1 baby checks (refer to appropriate guidelines). Immediately report any baby who either fails the SaO₂ protocol or has a soft murmur with poor femoral pulses, tachypnoea, tachycardia or an active precordium.

Duct dependent lesions are divided into **Right heart lesions** e.g. Transposition of the Great Arteries, Tetralogy of Fallot, Pulmonary Atresia/Stenosis and **Left heart lesions** e.g. Hypoplastic Left Heart Syndrome, Coarctation of the Aorta, Critical Aortic Stenosis/Interrupted Aortic Arch. In general, right heart lesions present with cyanosis and left heart lesions with heart failure.

Cyanosis

Differentiating cardiac from respiratory causes for cyanosis can be problematic, especially as some infants have mixed causes. The following assessments are suggested:

- A thorough clinical assessment.
- Check pre and post ductal SaO₂ levels.
- Hyperoxia Test (HT). (Discuss with neonatologist if infant is preterm). HT involves placing the infant in 100% oxygen which will result in a significant increase SaO₂ and PaO₂ in lung disease but NOT in Right Heart lesions due to the Right to Left shunts through the PDA. Monitor pre and post ductal SaO₂ throughout and check an ABG.
 - PaO₂ > 100 mmHg or SaO₂ increase by 15%: pulmonary disease likely.
 - PaO₂ < 70 mmHg, rise by < 30 mmHg or SaO₂ unchanged: cardiac cause or PPHN likely (Mild hyperventilation to decrease PaCO₂ and correction of metabolic acidosis can help differentiate PPHN from Right Heart Lesions).
- CXR to rule out lung disease and check heart size and shape.
- ECG.
- Cardiac echo is diagnostic.

Congestive Cardiac Failure (CCF)

CCF reflects the inability of the myocardium to meet the metabolic requirements of the body. Early CCF presents with tachycardia, tachypnoea, hepatomegaly and usually poor pulses. Advanced CCF presents with a collapsed infant with cyanosis, hypotension and severe metabolic acidosis. The following assessments are suggested:

- Clinical assessment for murmurs, gallop rhythm and especially femoral pulse.
- Check pre and post ductal SaO₂ levels.
- Blood gas for metabolic acidosis.
- Check 4 limb BP - an upper to lower limb systolic difference of > 10 mmHg is significant and suggestive of Coarctation of the Aorta.
- CXR to rule out lung disease and check heart size and shape.
- Check for secondary organ dysfunction e.g. renal, coagulation etc.

Murmurs and Femoral Pulses

Murmurs may indicate CHD but can be soft, non-specific or absent in duct dependent lesions at presentation. Weak or absent femoral pulse with a strong right brachial pulse is virtually diagnostic of left heart lesions. However, femoral pulses will be present whilst the PDA is wide open in left heart lesions.

Management





- Monitor vital signs and pre and post ductal SaO₂.
- Support respiration: supplemental O₂, CPAP or ventilation especially for left heart lesions.
- Venous and arterial access depending on severity.
- Maintain adequate calorific intake. How this achieved depends on the stage of management. TPN only or manipulation of feeds e.g. fortified, frequent small feeds or continuous milk feed.
- Parental antibiotics may be indicated.
- **Prostaglandin E1 (PG)** infusion to maintain ductal patency is important for both right (maintain pulmonary flow) and left (maintain systemic flow) heart lesions. Common **side effects of PG E1** are apnoea, especially in 'small', acidotic babies; hypotension-can be profound with bigger doses or inadvertent boluses; and fever. Prophylactic assisted ventilation, volume expansion or inotrope infusion maybe required. The threshold to intubate an infant on a PG infusion will be lower during NETS retrieval and individual requirements will be discussed with the NETS and cardiac consultant at the time of retrieval.

Related CAHS internal policies, procedures and guidelines

Neonatal Postnatal Ward Guidelines – [Pulse Oximetry Screening to Detect Critical Congenital Heart Disease](#)

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