GUIDELINE

Congenital Heart Disease: Presentation and Initial Management of Duct-Dependent Lesions

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

CHD can be benign and not require any treatment to life-threatening requiring immediate life-saving action. This protocol will concentrate on duct -dependent cardiac lesions which most often present in the neonatal period. The majority of these lesions are now picked up on antenatal screening; however, some are still diagnosed postnatally.

Background

These conditions may be asymptomatic in the early stages and sometimes get detected by routine postnatal SaO_2 protocols or by Day 1 baby checks. Refer to Pulse Oximetry Screening to Detect Critical Congenital Heart Disease (CHD). Some lesions present a few hours to days later when the ductus arteriosus starts to close.

Immediately report any baby who either fails the SaO_2 protocol or has a soft murmur with poor femoral pulses, tachypnoea, tachycardia, an active precordium or an incidental finding of a high lactate on a gas taken for another reason.

Duct dependent lesions are divided into:

- **Cyanotic lesions** e.g. Transposition of the Great Arteries (TGA), Pulmonary Atresia/Stenosis, Tricuspid atresia, Severe Tetralogy of Fallot,
- **Left heart obstructive lesions** e.g. Hypoplastic Left Heart Syndrome (HLHS), Coarctation of the Aorta, Interrupted Aortic Arch Critical Aortic Stenosis.

In general, right heart lesions/ TGA present with cyanosis and left heart lesions with poor peripheral perfusion with difficult to feel pulses and lactic/ metabolic acidosis +/- multiorgan failure. These become more pronounced as the ductus arteriosus closes.

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.
- Continuous pre and post ductal SaO_2 levels.
- Blood gas.
- CXR to rule out lung disease and check heart size and shape.
- ECG.
- Referral to cardiology for echocardiogram which is diagnostic.
- Hyperoxia Test (HT) is not usually done in the tertiary setting these days, but may be helpful in the community setting where diagnosis is unsure and transport to the tertiary centre may take hours
    - HT involves placing the infant in 100% oxygen which will result in a significant increase SaO_2 and PaO_2 in lung disease but NOT in Right Heart lesions due to the Right to Left shunts through the PDA. Monitor pre and post ductal SaO_2 throughout and check an ABG.
        - PaO_2 > 100 mmHg or SaO_2 increase by 15%: pulmonary disease likely.
        - PaO_2 < 70 mmHg, rise by < 30 mmHg or SaO_2 unchanged: cardiac cause or PPHN likely (Mild hyperventilation to decrease $PaCO_2$ and correction of metabolic acidosis can help differentiate PPHN from Right Heart Lesions).

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 pulses.
- Continuous pre and post ductal SaO_2 levels.
- Blood gas.
- 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 bloods for evidence 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.

Initial Management of Suspected Duct-Dependent Cardiac Lesions

- Early involvement of neonatal consultant with any suspected duct-dependent lesion and early referral to cardiology for assessment and echocardiogram.
- Monitor vital signs and pre and post ductal SaO_2.
- Support respiration: supplemental O_2 (judicious use in any lesion prone to pulmonary overcirculation eg, HLHS refer to Cardiac: Neonatal Circulation Changes / Unbalanced Circulation, CPAP or ventilation especially for left heart lesions.
- 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, refer to NETS WA Congenital Heart Disease Guideline .
- 2 points of venous access (PGE runs alone, and do not want any interruption to delivery)
- UVC/ UAC ideally for any infant with a duct-dependent lesion except for TGA (may need Balloon Atrial Septostomy (BAS) which is usually done through UV).
- Initially keep nil by mouth and on intravenous fluids until diagnosis and plan made.
- Parental antibiotics may be indicated.

Related CAHS internal policies, procedures and guidelines
Balloon Atrial Septostomy (BAS)
Cardiac: Neonatal Circulation Changes / Unbalanced Circulation
Coarctation of the Aorta
Pulse Oximetry Screening to Detect Critical Congenital Heart Disease (CHD)
Prostaglandin E1
NETS WA Congenital Heart Disease Guideline

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

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