CONGENITAL HEART DISEASE (CHD) OVERVIEW

CHD can be life threatening and require immediate action or need no intervention until later in life, or not at all. For a detailed description of neonatal circulation, epidemiology and pathophysiology of individual lesions (see appropriate textbook). This protocol will concentrate on signs and symptoms and recommended management practices of the duct dependent CHD lesions most often presenting in the neonatal period, namely transposition of the great arteries, Fallots, hypoplastic left heart syndrome, coarctation of the aorta, critical aortic stenosis/interrupted aortic arch, pulmonary atresia/stenosis.

CYANOSIS

Is one of the most common signs of CHD. Differentiating cardiac from respiratory causes can be problematic in infants with mixed causes. Carry out the following:

1. A thorough clinical assessment and resuscitation of the infant.
2. Confirm cyanosis with an ABG to check Pao2 (right arm if possible) in room air.
3. Hyperoxia Test (HT). (Discuss with neonatologist if infant preterm). Due to the right-to-left shunting, placing the infant in 100% oxygen will not increase arterial saturation. Monitor pre and post ductal SaO2 throughout and then repeat ABG should only be used after a thorough clinical assessment and resuscitation of the infant.
   - PaO2 > 100mmHg or SaO2 increase by 15%: pulmonary disease likely
   - PaO2 < 70mmHg, rise by < 30mmHg or SaO2 unchanged: cardiac cause or PPHN likely.
4. Ascertain if there is metabolic acidosis and poor perfusion. If present treat with fluid boluses and inotropes / bicarbonate as needed.
5. Take four limb BP - an upper to lower limb systolic difference of > 10mmHg is significant and suggestive of Coarctation of the Aorta. Hypotension in a cyanotic infant is a serious finding.
6. Request a Cardiac Team consultation. Cardiac echo is diagnostic. Obtain a CXR, and ECG if possible.
7. Two IV lines ideally and/or UVC.
8. Parental antibiotics (preferably after blood cultures taken).
9. Prostaglandin E1 (PG) infusion to maintain the patency of the ductus arteriosus if a duct dependent cardiac disease is suspected. The aim is to stabilise the infant by increasing the
pulmonary blood flow and hence the Pao2 whilst preparing the infant for a balloon septostomy or surgery. Close observation is needed following commencement of PG infusion, and assisted ventilation and volume expansion or inotrope infusion are sometimes required. The threshold to intubate an infant on a PG infusion will be lower if the infant needs transporting. Factors to consider include:
- presence of apnoea
- the distance to the receiving hospital (if applicable)
- gestation of the infant
- clinical state of the infant (metabolic acidosis, shock, severe distress and tachypnoea)
- high PG dose required to achieve ductal patency

In general, a stable infant with a PG responsive ductal lesion at low levels of PG, can be transported without intubation if the distance is short. Conversely, infants requiring high dose PG should generally be electively intubated (these infants may require respiratory support for other reasons).

MURMURS
Murmurs may indicate CHD but can be completely absent.

PULSES
Examinations performed within 6 hours of birth are more likely to miss lesions as the duct may still be widely patent. Lower limb pulses in infants with lesions of the left side of the heart can be normal if the duct is still wide open with a good right to left shunt through it. Examinations performed after 24hrs may have a better chance of detecting an abnormality.

CONGESTIVE CARDIAC FAILURE (CCF)
CCF is diagnosed on the presence of certain clinical signs and symptoms and reflects the inability of the myocardium to meet the metabolic requirements of the body. In the early stages some compensation is seen, later CCF presents as cardiorespiratory collapse.

CAUSES
- Volume overload
- Pressure overload
- Cardiomyopathy
- Dysrhythmias

FEATURES
1. Tachycardia – in an attempt to increase cardiac output.
2. Cardiac enlargement – in response to volume or pressure overload.
3. Tachypnoea – as pulmonary oedema progresses there is an increase in intercostal retractions, grunting, flaring, dyspnoea, rales and cyanosis.
4. Gallop rhythm – heard as an abnormal filling sound due to dilation of the ventricles.
5. Mottling / decreased perfusion (capillary refill) and decreased pulses – decreasing cardiac output results in a compensatory redistribution of blood flow to vital organs and tissue.
6. Decreased urine output and oedema – this is a later sign and normally manifested as weight gain and periorbital oedema.

**MANAGEMENT**

Treat CCF by treating the cause and maintaining close observation of the infant.

1. Diuretics (such as frusemide) help to decrease total body water, which is increased as a result of CCF, but be aware of electrolyte balance and long term use is associated with renal calcification.

2. Monitor vital signs. Watch for tachycardia, arrhythmias, decreasing Sao2 and dysnoea, apnoea or tachypnoea.

3. Maintain adequate calorific requirements. May need changes to feeds if unable to feed adequately. For example, more frequent smaller feeds or tube feeds if having trouble sucking. Weight gain is often slower in infants with cardiac defects so fortified feeds may be needed.

4. Cardiac drugs that may be used include, Digoxin, Captopril, Propranolol (see drug manual).