LEFT HEART OBSTRUCTION - NETS

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

Please observe the following key points:

- These cases are urgent until Prostaglandin E_1 (PGE1) is running – transport is then as per usual practices

CHANGE SUMMARY

Updated with referencing

READ ACKNOWLEDGEMENT

- All NETS clinical staff are to read and acknowledge they understand the contents of this guideline.

Approved by:	SCHN Policy, Procedure and Guideline Committee	NETS Executive
Date Effective:	1st September 2024	**Review Period:** 3 years
Team Leader:	Staff Specialist	**Area/Dept:** NETS

Introduction

Antenatal diagnosis of congenital heart lesions in NSW approaches 80% of cases however left sided lesions can be missed at the 18-week fetal anomaly scan as they are not obvious until postnatal closure of the ductus. This leads to infants with suspected left heart obstruction (LHO) presenting to NETS in the first two weeks of postnatal life at least once per month on average[1]. Symptoms range from effortless tachypnoea with one or more from poor feeds to complete shock from heart failure. This is due to significant or complete obstruction to left sided (systemic circulation) flow with closure of the ductus arteriosus (DA).

Signs of heart failure may include:

- Tachypnoea
- Enlarged liver - highly suggestive[1]
- Cardiomegaly with pulmonary plethora on CXR[2]
- Weak or absent femoral pulses or symmetrically reduced pulses (depending on the level of obstruction)
- Colour discrepancy between upper and lower body
- Pre/Post ductal saturation difference (right arm higher than a lower limb)
- Poor urine output
- Cardiac murmur (although this may not be present or prominent in infants)

Laboratory tests

Laboratory tests may help confirm the diagnosis prior to a diagnostic echocardiogram:

- Elevated lactate
- Metabolic acidosis on arterial blood gas
- Right ventricular hypertrophy and right axis deviation on ECG in the early period
- Arrhythmias may be present on ECG

Anomalies

Congenital left sided obstructive anomalies of the heart include:

- Coarctation of the aorta
- Interrupted aortic arch
- Critical aortic stenosis
- Hypoplastic left heart syndrome

Primary call

- Resuscitate the infant
- As severe metabolic acidaemia is common, don't be put off intubating and ventilating an infant despite what may be some effort at compensation with lowered $PaCO_2$
- Discussion on the first call with the NETS consultant, neonatologist or cardiologist will guide the commencement of Prostaglandin E_1 (PGE1).
- The moribund and severely acidotic baby should be intubated and ventilated and as early as possible. The most experienced team member should perform the intubation such an extremely sick patient
- Poor cardiac output is supported with ventilation
- Infants with sepsis will **not usually** have an enlarged liver or an active precordium
- All Infants presenting in shock need to be treated with antibiotics for infection; even if a duct-dependant heart lesion seems likely
- An early presentation may occur while in the nursery while a late presentation may be from the community to an ED. These environments are different and support of acute care may need to be different. Consider requesting neonatal support locally to support ED care; especially when neonatal ICU skills in umbilical catheterization and clinician cardiac ultrasound may be available.

Team assessment and Stabilisation

- Review primary call discussion, interventions and process
- If no discussion has occurred regarding the use of PGE_1 and the team feels it is warranted, then discuss at the earliest opportunity:
    - In the infant who is diagnosed early prior to acidosis commence PGE_1 1at0 nanograms/kg/min;
    - Where there is significant acidosis (pH<7.2), PGE_1 should be commenced at 50 nanograms/kg/min with simultaneous low dose adrenaline to support the circulation from further collapse on initiation of PGE_1
    - Also see PGE_1 guideline

Airway and Breathing

- If metabolic acidosis is present, the provision of mechanical ventilation will reduce cardiac workload and treat pulmonary oedema.
- The acidotic infant with decompensated left heart obstruction will require mechanical ventilation initiated prior to transport. Occasionally, infants are diagnosed prior to the

development of significant acidosis and these infants may or may not require mechanical ventilation for transport. PGE_1 infusion is not an absolute indication for ventilation– see PGE_1 guideline.

- Apnoea is more likely at PGE_1 infusion rates > 15 nanograms/kg/min and these infants will likely require mechanical ventilation[5].
- Oxygenation should be maintained at a PaO_2 of between 60-80 mmHg and saturations 88-92%.
    - Oxygen saturation above and below the level of the ductus arteriosus (DA) may vary if the obstruction is principally at arch level. This may produce lower saturations in the lower limbs (post-ductal) than in the right arm (pre-ductal) - but not always. Assessment of oxygen saturation is most reliably made using the right arm.
    - Be mindful of targeting a post-ductal PaO_2 if arterial blood is sampled from an umbilical arterial line – pre-ductal (right arm) oxygen saturations may be more representative of cerebral oxygenation in this setting if the DA is open, due to right to left ductal shunting.
    - **Be wary of oxygen administration as it will drop the pulmonary vascular resistance, reducing right to left flow across the ductus arteriosus and further compromising the systemic circulation in the setting of HLHS**.
    - If oxygen saturations remain high (>92%) without supplemental oxygen and systemic perfusion remains poor, discuss ventilation options for modulating pulmonary blood flow with the NETS consultant and receiving neonatologist.

Circulation

- **The goal of circulation in these infants is to reopen and maintain the patency of the DA to encourage and balance the flow across the ductus; mainly right to left** (deoxygenated blood crosses the DA from the pulmonary artery to the aorta, providing a source of blood flow to the lower body in the setting of left-sided obstruction)
- In the shocked infant, double lumen umbilical venous catheter (UVC) access should be established. This is often possible up to two weeks of postnatal life[3]. If not possible then two peripheral cannulae should be placed in the upper limbs. If unable to gain access with UVC or peripheral cannulae within minutes of presentation, then intraosseous access is an option. It is important to remember that the lower limbs are not well perfused whilst the DA remains closed.
- These Infants are not usually volume depleted as the clinical onset is usually over a few hours. Reduced oral intake over a longer period might potentially make the baby dehydrated.
- After commencing mechanical ventilation, if dehydration is present, a single 10 mL/kg bolus of 0.9% saline might be appropriate prior to considering and commencing an inotrope such as dopamine or low-dose adrenaline (0.05 - 0.1 micrograms/kg/minute).

In the severely acidotic infant (pH <7.2), an inotrope should be commenced together with the PGE_1 to help support the circulation.

- If LHO is suspected, fluid boluses of >10 mL/kg should not be administered without discussion with the NETS consultant.
- Give half corrections of 4.2% $NaHCO_3$ whilst BE remains less than -10. This helps to improve cardiac function.
  - ½ correction mL of $NaHCO_3$ = 0.3 x wt in kg x BE
- It should be remembered that the best treatment for improving lactic acidosis in this setting is to establish adequate systemic perfusion, best achieved by administration of PGE_1 for ductal patency and appropriate cardiorespiratory support (as above)
- Intra-arterial monitoring should be commenced when possible, either via the umbilicus or one of the upper limbs (preferably the right radial artery)

Important issues to consider

- **Time is critical in these cases until the PGE1 is commenced, so the team +/aircraft will normally be tasked Priority 1 for any patient needing urgent treatment (such as PGE_1 or intubation/ventilation) which can't be started locally. Road ambulance response for the sickest patients would normally be R1**
- Once a PGE_1 infusion is commenced, stabilise the infant in the normal manner
- This is a life-threatening condition and the parents will require clear, honest and unambiguous information about the severity of the problem; which might range from surgically treatable to untreatable; with full recovery or incomplete recovery; especially if there has been severe acidaemia or hypoperfusion – eg. Lactate > 10. The precise diagnosis may not be clear initially and is likely to be confirmed only at the receiving hospital. Discuss the need for transport to a paediatric cardiology centre and the likely need for surgery.
- Ensure that both team members have calculated and checked the PGE_1 dose and infusing solution
- Minimal handling is a priority in Infants with these conditions, so be efficient with procedures

Conference

- Have a recent arterial blood gas available – not capillary (perfusion is usually poor and arterial gases are more accurate)
- Discuss the ventilation strategy and PGE_1 dose
- Discuss the use of inotropes - indication(s), choice and dose
- Discuss use of supplemental oxygen at altitude if travelling at altitude or high elevation

Pre-transport checklist

- TOPS (Temperature, Oxygen, Perfusion, Sugar)
- Adequate vascular access
- Orogastric tube
- Indwelling urinary catheter may be required
- PGE1
- Maternal blood sample (with completed documentation)

En-route

- Plan for a transport that involves as little disruption to the baby as possible.
- If the infant is ventilated, provide adequate morphine for sedation/analgesia.
- In a stable infant on PGE_1 there is no need to increase FiO_2 at altitude or high elevation as hypoxic vasoconstriction in the pulmonary vasculature encourages right to left ductal flow and therefore improves systemic flow.
- In an unstable infant or one who is still acidotic maintain the oxygen saturations that you had prior to aircraft take-off with judicious use of oxygen at altitude.

References

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3. Legge N, Browning Carmo K. Umbilical Venous access in neonatal emergencies – indication, timing and outcome. *[Unpublished data – article under review]*
4. J.Cloherty, E.Eichenwald, A.Stark. Cloherty and Stark's Manual of Neonatal Care, 6th edition.
5. Browning Carmo K, Barr P, West M, Hopper NW, White JP, Badawi N Transporting infants with suspected duct dependent congenital heart disease on low dose prostaglandin E1 without routine mechanical ventilation; ADC 2007; 92
6. Keane JF,Fyler DC,Lock JE.Nadas. Pediatric cardiology, 2nd edition 2006.