CLINICAL GUIDELINE	
Persistent Pulmonary Hypertension in the Newborn (PPHN)	
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 Points

- Most often seen in association with:
  - Parenchymal lung disease (pneumonia, Meconium aspiration, HMD, pulmonary hypoplasia, diaphragmatic hernia).
  - Or in a sick neonate (severe asphyxia, fulminant sepsis, shock).
- Need to differentiate from cyanotic congenital heart disease (CHD).
- All have severe hypoxaemia, usually PaO_2 < 45mmHg in 100% O_2.

Management

- If PPHN suspected:
  - Discuss with the on-call neonatologist.
  - Manage the underlying cause.
- Intubate and ventilate.
- Judicious use of oxygen to maintain preductal SpO_2 ≥ 94%.
- Stabilise circulation: treat hypotension with **fluid boluses** (to increase pre-load on the heart) and/or inotropes (to increase systemic blood pressure). Aim for mean BP 55-60mmHg. Consider **dopamine** or **adrenaline.**
- Consider central venous access and arterial access (if short journey these may be reserved for insertion at tertiary centre).
- Reduce stress and movement: Avoid excessive handling and sedate with Morphine/Midazolam. Some may require muscle relaxation (e.g. Vecuronium).
- Aim for pH ~7.4 (help to vasodilate pulmonary vasculature): may require sodium bicarbonate infusion if $PaCO_2$ 30-35.
- Pulmonary vasodilators: First line therapy is **Inhaled Nitric Oxide (iNO)**.
- Other pulmonary vasoldilators include **Alprostadil** (**PGE_1)** and **Milrinone**. BEWARE: Both PGE_1 & Milrinone can cause systemic hypotension!

Use of Inhaled Nitric Oxide (iNO) During Retrieval

Consideration for using iNO during retrieval **requires greater clinical judgment** as to the possible underlying causes for hypoxia, since investigations such as echocardiogram and even pre- and post-ductal oxygen saturation measurements may not be available. Consequently, a 'trial of therapy' may be indicated on the grounds of the clinical history and examination findings alone. In general, decisions pertaining to the use of iNO during transport should be made in conjunction with the on-call neonatologist.

1. The following **clinical conditions** may be potentially responsive to iNO;
    - Meconium aspiration syndrome.
    - Severe hyaline membrane disease.
    - Congenital pneumonia / sepsis.
    - Congenital respiratory malformation (e.g. diaphragmatic hernia).
2. **Clinical evidence** of pulmonary hypertension:
    - SpO_2 < 85% despite FiO_2 > 80%.
    - Pre- and post-ductal SpO_2 difference > 10%.
    - Hypoxia despite high ventilation settings (e.g. PIP ≥ 30 cmH_2O).

Contraindications

- Coagulopathy / Active bleeding.
- Stage III Hypoxic-ischaemic encephalopathy.
- Imminent death.
- Lethal congenital malformation.

Optimise Other Strategies

- Ventilation parameters.
- Consider Surfactant (depending on underlying respiratory disease).

Prior to Commencement

- Arterial Blood Gas to calculate Oxygenation Index* (as a baseline to monitor response).
- Document pre and post-ductal SpO_2 (if available) and FiO_2.

$$\text{*Oxygenation index (OI)} = \frac{\text{Mean Airway Pressure x \% oxygen}}{PaO_2}$$

- **Term Infants (≥ 37w):** Commence iNO at **10-20ppm.** Doses in excess of 40ppm are not recommended, and rarely effective if there is no response to the original dose.
- **Preterm Infants (< 37w):** Commence iNO at **5-10ppm.** Doses above 20ppm are not recommended.
- **Repeat blood gas** 30 minutes after commencement.
- Document FiO_2, pre and post-ductal SpO_2 (if available).

Maintenance and Weaning on iNO

As a general rule, iNO commenced during a transport should be continued at the effective dose (and not weaned) until return to the accepting hospital.

Potential Complications of iNO Use

Risks to Health Caregivers

- As NO is a corrosive gas, there is a **small** potential risk to staff if an entire cylinder of NO was discharged in a confined space (such as in an aircraft or ambulance).
- Special charcoal filters are used to scavenge Nitric Oxide and its by-products (NO_2) although many centres do not scavenge at all.
- There is excellent evidence to show that
- complete discharge of a 'D' cylinder of NO in a well ventilated enclosed space, **does not result in dangerous levels of either NO or NO_2** due to the usual turnover of air in transport vehicles and aircraft.

Nevertheless, should a full cylinder discharge, the following safety precautions should be employed:

1. **Eyes: Corrosive to the eye, mostly for those wearing contact lenses**

   This is mostly an issue for health workers receiving a direct stream of NO to the eyes. Wear protective goggles if in close proximity to the cylinder. Immediately flush with large quantities of tepid water or saline for 15-20 minutes.

   Remove contact lenses if exposed.

2. **Inhalation: Immediate effects**

   Irritation of the nose and throat.

3. **Suggested late effects (up to 72 hours)**

   Cough, chest pain, nausea, dyspnoea and haemoptysis (Note: none of these side effects have been noted in any trials of NO).

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