



Refer EARLY to KIDSNTS for advice - 0300 200 1100

Persistent Pulmonary Hypertension of the Newborn (PPHN) = failure of the pulmonary circulation to follow normal postnatal adaptation and achieve reduced pulmonary vascular resistance. This leads to Right to Left shunting and subsequent hypoxia.

Primary causes: a) **Idiopathic** - Placental insufficiency/chronic fetal hypoxia with vascular remodelling; b) **Peripartum** (Intrapartum hypoxia)
Secondary causes: a) **Parenchymal Lung Diseases** - Meconium aspiration syndrome/Pneumonia; b) **Abnormal Transition** - Sepsis/Asphyxia/Perinatal Stress/Hypothermia; c) **Developmental Lung Disease** - CDH/CCAM/Lobar emphysema; d) **Maternal drug ingestion** - i.e SSRI's

Signs & Symptoms: Hypoxia; Pre/post ductal difference >5-10%; Minimal respiratory distress comparative to degree of hypoxia; Pulmonary Oligaemia on CXR; Loud 2nd Heart Sound; Hypotension.

If commencing Ventilation + considering Nitric Oxide—please Contact KIDSNTS early for discussion.

Key aims and plan for uplift:

Aims - Improve PaO₂; Reduce PVR; Reverse Right to Left shunting; Increase SVR

Plan - 1) Intubate and Ventilate, 2) Central Access ASAP, 3) Sedation and Muscle Relax, 4) Connect Infusions early.

**Airway/
Breathing**

- Intubation and ventilation with muscle relaxation - optimise as per blood gases.
- CXR to confirm ETT position and assess lung pathology/expansion.
- **Monitor pre and post ductal saturations.**
- If Oxygenation Index (OI) > 10-15 consider starting inhaled Nitric Oxide.
- Consider **High Frequency Oscillatory Ventilation** + Higher Mean airway pressures.
- Meconium Aspiration - additional surfactant therapy.
- Judicious use of O₂ to maintain PaO₂ 6-12kPa.



Circulation

- Early central venous and arterial access.
- **Echo** - To exclude congenital heart disease (including TAPVD); Assess pulmonary hypertension - direction of shunt at PDA/Atrial level, Tricuspid regurgitation and right sided pressures, RV function and size. Assess Left heart function and chamber sizes.
- Assess volume status and give fluid bolus as required.
- **Early inotropic support** - mean arterial pressure target will depend on estimated pulmonary arterial pressure. Generally consider adrenaline or noradrenaline where increases in SVR are required (+ vasopressin). Dobutamine improves contractility but may cause excessive tachycardia and vasodilatation. Avoid high dose Dopamine - worsens PVR. **Try to avoid excessive tachycardia.**
- **Give Hydrocortisone** early to up-regulate Alpha receptors.
- Consider Prostin to maintain PDA/Offset Right Sided Pressures - **specialist advice only.**
- Try to maintain normothermia. (If cooled consider re-warming due to effect on PVR)
- Consider correcting calcium and magnesium levels.
- Ensure antibiotics have been given.



Disability

- Ensure adequate sedation - Morphine 10-20 micrograms/kg/hr (+ Rocuronium).
- CrUSS important if considering ECMO referral.
- Maintain glucose > 3.0mmol/L.

If OI remains above 20 despite starting the above therapies – conference call with KIDSNTS and PICU to discuss management and ECLS criteria

If any delay to Nitric Oxide therapy – i.e. in local SCBU/LNU can discuss alternative management strategies with KIDSNTS Consultants.