Management of Acute Persistent Pulmonary Hypertension (aPPHN) for Neonatal Transfers

Refer EARLY to KIDSNTS for advice - 0300 200 1100

aPPHN is a severe hypoxemia due to failure of the normal postnatal drop in pulmonary vascular resistance. Can be primary (idiopathic) or secondary.

Primary/Idiopathic aPPHN

Degree of hypoxia is disproportionate to degree of hypercarbia.

Maternal (IDMD, NSAID's, Pre-eclampsia), ***Peripartum*** (Intrapartum hypoxia), ***Rare*** (Alveolar dysplasia), ***Idiopathic*** (Vascular remodelling in utero).

Secondary aPPHN

Lung parenchymal disease—Pneumonia, Meconium Aspiration
Abnormal transition at birth—Hypoxia/Asphyxia (vasoconstriction)
Developmental lung disease—Congenital diaphragmatic hernia (CDH)

Presentation

Cyanosis , Hypoxia with/without hypercarbia, $Pa0_2$ < 5kPa when on $Fi0_2$ 1.0, $Sp0_2$ difference >10% in preductal and postductal saturations.
Respiratory distress in secondary aPPHN, additional respiratory signs of underlying pathology (i.e. Meconium, CDH)
CXR: Black lung in idiopathic aPPHN, specific presentation of underlying pathology in secondary aPPHN.
CVS: Tricuspid regurgitant murmur, right ventricular heave, loud 2nd sound with/without systemic hypotension.
ECHO (if available): Tricuspid regurgitation, Dilatation of right side of heart, intra-septum bowing

Aims of management **1)** Adequate pulmonary blood flow (PBF) + systemic blood flow (SBF) **2)** Decrease pulmonary vascular resistance (PVR) and decrease pulmonary artery pressure (PAP) **3)** Increase systemic vascular resistance (SVR) **4)** Optimise heart function.

Immediate actions at referral 1) Check Temp + Glucose 2) Intubate + ventilate 3) Central Access 4) Sedate + Muscle Relax 5) Connect Inotropes early

Airway/ Breathing

Conventional ventilation (TTV 5mls/kg) - 'gentle approach' in Black lung. Consider High Frequency (HFO)+ higher MAP in rescue scenarios. Assess Oxygen Index (OI) If > 10 consider Nitric Oxide if available.

Order CXR early while awaiting KIDSNTS.

Maintain $Pa0_2$ 7-10 kPa (avoid hyperoxia)

Monitor pre/post ductal Sp02
(Pre ductal 90-95% /Post ductal >70%)

Maintain $PaC0_2$ 6-8 kPa (avoid hypocapnia)

Consider additional surfactant therapy.

Attach transcutaneous monitoring if available

RSI intubation drugs - followed by **Morphine infusion 10-40mcg/kg/hr and Rocuronium 0.6mcg-1mg/kg/hr**.

Circulation

Central access ASAP

- **Hypotension** > One bolus of Saline 10mls/kg over 20-30 minutes.
- If **low pulse pressure** (-/+ LV dysfunction on ECHO), start Adrenaline (0.05-0.2mcgs/kg/min), if no adequate response add Dobutamine (5-10mcgs/kg/min).
- If **normal pulse pressure** (-/+ LV dysfunction on ECHO), start **Adrenaline** (0.05-0.2mcg/kg/min) and **Noradrenaline** (0.05-0.4mcgs/kg/min). If inadequate response, add Dobutamine (5-10mcgs/kg/min).
- If **hypotension with high pulse pressure** (normal LV function), use **Vasopressin** and/or **Noradrenaline.**

If no response to initial Inotropes - Add Hydrocortisone (2.5mg/kg)

- Consider adding **Prostin** to off load RV + support systemic circulation.
- If normal blood pressure (RV dysfunctions) - consider **Milrinone** (discuss with KIDSNTS).
- Monitor Lactates and avoid Tachycardia

If **ECHO** available—try and exclude CHD. Assess pulmonary hypertension + direction of shunt at PDA/Atrial level, Tricuspid regurgitation and right sided pressures. Assess LV function. Assess fluid status and give fluid bolus as required.

Disability

Ensure normal temperature + normal glucose range. Check **Calcium** and **Magnesium** levels—correct out of range.
CrUSS important if considering ECLS referral. Check Clotting parameters ASAP.

If OI remains >20 despite the above therapies—consideration for ECLS can be discussed with KIDSNTS.

ECLS Criteria **1)** Born >34 weeks or >2kg with aPPHN **2)** Reversible lung disease **3)** No lethal congenital malformation
ECLS Exclusion **1)** Major intracranial haemorrhage **2)** Lethal congenital or chromosomal anomalies **3)** Severe encephalopathy **4)** Major CHD

Author - BG - Oct '24. Review Oct '27