

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

 $Cyanosis\,, Hypoxia\,with/without\,hypercarbia, Pa02 < 5kPa\,when\,on\,Fi02\,1.0, Sp02\,difference > 10\%\,in\,preductal\,and\,postductal\,saturations.$

 $Respiratory\ distress\ in\ secondary\ a PPHN,\ additional\ respiratory\ signs\ of\ underlying\ pathology\ (i.e.\ Meconium,\ CDH)$

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

<u>Aims of management</u> 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 Pa02 7-10 kPa (avoid hyperoxia)

Monitor pre/post ductal Sp02

(Pre ductal 90-95% /Post ductal >70%) Maintain PaCO₂ 6-8 kPa (avoid hypocapnia)

Consider additional surfactant therapy. Attach transcutaneous monitoring if available

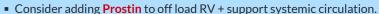
RSI intubation drugs - followed by Morphine infusion 10-40mcg/kg/hr and Rocuronium 0.6mcg-1mg/kg/hr.



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)



- 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
ECLS Exclusion

1) Born > 34 weeks or > 2kg with aPPHN 2) Reversible lung disease 3) No lethal congenital malformation

1) Major intracranial haemorrhage 2) Lethal congenital or chromosomal anomalies 3) Severe encephalopathy 4) Major CHD