NHS Children's Acute Transport Service

Clinical Guidelines

PPHN (see also ECMO guideline)

Document Control Information

Author	R.Paget	**Author Position**	Consultant
Document Owner	E. Polke	**Document Owner Position**	Service Coordinator
Document Version	Version 3	**Replaces Version**	January2016
First Introduced		**Review Schedule**	2 Yearly
Active Date	January 2018	**Next Review**	January 2020
CATS Document Number			
Applicable to	**All CATS employees**		

Introduction

Persistent Pulmonary Hypertension of the Newborn (PPHN) is defined as a failure of the normal postnatal fall in pulmonary vascular resistance, which leads to persisting right to left shunts across the fetal channels and resultant hypoxia. PPHN may be primary or secondary.

1. Assessment

- **Primary PPHN** – typically idiopathic but may be in response to in utero foetal stress/hypoxia/pulmonary hypertension (eg premature Ductus Arteriosus closure 2° to maternal NSAID exposure)
- Idiopathic PPHN ("black lung" PPHN)-**2nd most common cause**

- **Secondary PPHN** - acute pulmonary vasoconstriction 2° to hypoxia/hypoglycaemia/hypothermia
- MAS (meconium aspiration syndrome)
- Sepsis +/- pneumonia
- Neonatal Respiratory Distress Syndrome
- Congenital diaphragmatic hernia (degree of pulmonary hypoplasia, pulmonary hypertension + pulmonary vascular remodelling)
- Pulmonary Hypoplasia
- Congenital Lung Dysplasias e.g. Congenital Cystic Adenomatous Malformation (CCAM)

Echocardiography (if available) to:

- Exclude congenital heart disease as a cause of hypoxia.
- Define the pulmonary artery hypertension by:
  - Defining R→ L shunt across Ductus Arteriosus and/or Foramen Ovale
  - Estimation of RV systolic pressures via peak velocity of regurgitant flow across the Tricuspid Valve
- Define myocardial contractility and chamber dilatation

Cranial ultrasound scan to exclude significant IVH if ECMO is being considered.

2. Immediate management

- Oxygen is a potent pulmonary vasodilator**.** Aim to maintain PaO_2 above 8 kPa.(8-10 kPa if possible).
- Confirm endotracheal tube position and size (listen for an ETT leak). Consider regular chest physiotherapy and suctioning to reduce areas of atelectasis and for airway clearance.
- Sedate and use muscle relaxation.
- Consider early surfactant therapy if significant lung disease or MAS.
- HFOV if available to optimise lung volume whilst minimizing risk of lung injury. **Avoid overexpansion of lungs** (aim <9 ribs on CXR)
- If HFOV unavailable, conventional ventilation with high PEEP (8-10cmH2O) may improve oxygenation whilst reducing requirement for high PIP which is associated with lung injury.
- Aim to reduce $PaCO_2$ to normal levels to aid alkalinisation and pulmonary vasodilation. Avoid hypocapnia.
- Alkalinise with sodium bicarbonate/THAM to maintain pH >7.35 if gas exchange permits.
- Inhaled nitric oxide at 20 ppm (if available) if:
    - Oxygenation index >15
    - Oxygenation index = (mean airway pressure x Fi02x 100)/Pa02 (in mmHg)
    - Difference in pre to post-ductal SaO2 >5% in the absence of CHD **(+/- evidence of significant pulmonary hypertension on echo)**
    - NB Monitor methaemoglobin levels closely, which can aggravate hypoxia **(adjust nitric oxide range 5-20 ppm (to keep methaemaglobin <5%).**

3. Drain pneumothoraces.

- Optimise circulating volume.
- Use inotropes/vasopressors to maintain ventricular function and pulmonary blood flow. Aim for a Mean Arterial Pressure > estimated Pulmonary Pressure (may need MAP >60) using dopamine, adrenaline and/or noradrenaline.

- Consider a magnesium bolus of 50mg/kg over 30 min if MAP maintained (watch for hypotension). This may be repeated as tolerated if effective, or administered as an infusion (suggested maximum serum Mg level of 3mmol/l).
- Correct hypocalcaemia.
- Correct hypoglycaemia.
- Maintain normothermia.
- Investigate and treat for infection with appropriate antibiotic cover.
- The CATS Consultant may suggest an Adenosine or Milrinone infusion and/or Bosentan if oxygenation is not improving.

4. Transport considerations

- Ensure ETT well secured/good position/no leak.
- Run continuous infusions of sedation and muscle relaxant.
- Ensure adequate peripheral venous, intraosseous or central venous and arterial access.
- Discuss with CATS Consultant and ECMO Consultant re: consideration for ECMO**.**