NHS Children's Acute Transport Service

Clinical Guidelines

PPHN (see also ECMO guideline)

Document Control Information

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Introduction

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) - **most common cause**
  - Sepsis +/- pneumonia
  - Neonatal Respiratory Distress Syndrome
  - Congenital diaphragmatic hernia (degree of pulmonary hypoplasia, pulmonary hypertension + pulmonary vascular remodelling)
  - Pulmonary Hypoplasia
  - Congenital Lung Dysplasias eg CCAM

Echocardiography (if available) to:

- Exclude congenital heart disease.
- Define the pulmonary artery pressures by:
  - Determining R→ L shunt across Ductus Arteriosus and/or Foramen Ovale
  - Calculating RV systolic pressures by looking at peak velocity of regurgitant flow across TV
- Define myocardial contractility

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

2. Immediate management

2.1 Oxygen **(potent pulmonary vasodilator).** Aim to maintain PaO_2 above 8 kPa.(8-10 kPa if possible).

2.2 Optimise endotracheal tube position and size (aim for no ETT leak).

2.3 Consider chest physio/suction to reduce areas of atelectasis.

2.4 Sedate and paralyse.

2.5 Consider early surfactant therapy if significant lung disease or MAS.

2.6 HFOV if available to optimise lung volume whilst minimizing risk of lung injury. **Avoid overexpansion of lungs** (aim <9 ribs on CXR)

2.7 If HFOV unavailable, conventional ventilation with high PEEP (8-10) may improve oxygenation whilst reducing requirement for high PIP and ΔP which are associated with lung injury.

2.8 Aim to reduce $PaCO_2$ to normal levels to aid alkalinisation and pulmonary vasodilation. Avoid hypocapnia.

2.9 Alkalinise with sodium bicarbonate/THAM to maintain pH >7.35 if gas exchange permits.

2.10 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%).**

2.11 Optimise circulating volume.

2.12 Institute inotropes/vasopressors to maintain ventricular function and pulmonary blood flow. Aim for a MAP>Pulmonary Pressure (may need MAP >60) using dopamine as a first line agent and consider other agents (eg noradrenaline) if dopamine exceeds 10 mcg/kg/min.

2.13 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).

2.14 The CATS Consultant may suggest Adenosine infusion at 50mcg/kg/min, Milrinone and/or Bosentan if oxygenation not improving.

2.15 Correct hypocalcaemia.

2.16 Correct hypoglycaemia.

2.17 Maintain normothermia.

2.18 Investigate and treat for infection with appropriate antibiotic cover.

2.19 Drain pneumothoraces.

3. Transport considerations

3.1 Ensure ETT well secured/good position/no leak.

3.2 Run continuous infusions of sedation and muscle relaxant.

3.3 Ensure adequate peripheral venous, intraosseus or central venous and arterial access.

3.4 Discuss with CATS Consultant and ECMO Consultant re: consideration for ECMO. **See ECMO SOP.**

References

Bendapudi P, et al. Diagnosis and management of persistent pulmonary hypertension of the newborn. Paediatr. Respir. Rev. (2015)

Yaseen H, Darwich M, Hamdy H. Is sildenafil an effective therapy in the management of persistent pulmonary hypertension?. J Clin Neonatol [serial online] 2012 [cited 2015 May 15];1:171-5

Bassler D, Kreutzer K, McNamara P, Kirpalani H. Milrinone for persistent pulmonary hypertension of the newborn. Cochrane Database of Systematic Reviews 2010, Issue 11. Art. No.: CD007802. DOI: 10.1002/14651858.CD007802.pub2.

Shah PS, Ohlsson A. Sildenafil for pulmonary hypertension in neonates. Cochrane Database of Systematic Reviews 2011, Issue 8. Art. No.: CD005494. DOI: 10.1002/14651858.CD005494.pub3.

Intravenous sildenafil in the treatment of neonates with persistent pulmonary hypertension.
Steinhorn RH, Kinsella JP, Pierce C, Butrous G, Dilleen M, Oakes M, Wessel DL
J Pediatr. 2009 Dec; 155(6):841-847.e1.

Intravenous sildenafil for postoperative pulmonary hypertension in children with congenital heart disease.
Fraisse A, Butrous G, Taylor MB, Oakes M, Dilleen M, Wessel DL
Intensive Care Med. 2011 Mar; 37(3):502-9.

Pharmacokinetics, safety, and efficacy of bosentan in pediatric patients with pulmonary arterial hypertension.
Barst RJ, Ivy D, Dingemanse J, Widlitz A, Schmitt K, Doran A, Bingaman D, Nguyen N, Gaitonde M, van Giersbergen PL
Clin Pharmacol Ther. 2003 Apr; 73(4):372-82.

Long-term efficacy of bosentan in treatment of pulmonary arterial hypertension in children.
Hislop AA, Moledina S, Foster H, Schulze-Neick I, Haworth SG
Eur Respir J. 2011 Jul; 38(1):70-7.

Persistent Pulmonary Hypertension of the Newborn: Recent Advances in the Management International Journal of Clinical Pediatrics, ISSN 1927-1255