

NHS CATS
Children's Acute Transport Service



Clinical Guidelines

Management of PPHN (also see ECMO guideline)

Document Control Information

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Document Version	Version 6	Replaces Version	Version 5
First Introduced	2006	Review Schedule	2 Yearly
Active Date	2026	Next Review	2028
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, with persistent right to left shunting across the fetal channels, and profound hypoxia.

Assessment

- **Primary PPHN** is typically idiopathic but may occur in response to *in utero* foetal stress and vascular remodelling (e.g. premature ductus arteriosus closure secondary to maternal NSAID exposure).
- **Secondary PPHN** - acute pulmonary vasoconstriction secondary to:
 - Hypoxia/hypoglycaemia/ hypothermia
 - Meconium aspiration syndrome (MAS)
 - Sepsis
 - Neonatal RDS
 - Congenital diaphragmatic hernia (CDH). Degree of pulmonary hypoplasia, pulmonary hypertension + pulmonary vascular remodelling.
 - Pulmonary hypoplasia
 - Congenital lung dysplasia's e.g. congenital pulmonary airway malformation (CPAM)

Echocardiography, if available, is useful to:

- Exclude congenital heart disease (CHD) as a cause of hypoxia
- Define the pulmonary artery hypertension by describing the shunt across the ductus arteriosus and/or foramen ovale, and estimating RV systolic pressures via peak velocity of the regurgitant flow across the tricuspid valve
- Define myocardial contractility and chamber dilatation

A cranial ultrasound scan to exclude significant intraventricular hemorrhage (IVH) if ECMO is being considered, and to exclude a Vein of Galen Malformation (VGAM).

Management strategy

- Consider whether ECMO might be required and escalate accordingly. See CATS ECMO guideline.
- Confirm that the endotracheal tube is correctly sited, and of appropriate size (assess for an ETT leak).
- If there is a significant leak around the ETT, it is likely it will need exchanging for a larger and/or cuffed tube.

- Consider regular chest physiotherapy and suctioning to reduce areas of atelectasis and for airway clearance.
- Oxygen is a pulmonary vasodilator. Aim to maintain PaO_2 above 8 kPa (8-10 kPa if possible).
- Sedate and muscle relax with continuous infusions of morphine and vecuronium.
- HFOV, if available, can optimise lung volume whilst minimizing risk of lung injury. Avoid overexpansion of the lungs (aim <9 ribs on CXR).
- If HFOV is unavailable, conventional ventilation with high PEEP (8-10cm H_2O) may improve oxygenation whilst reducing requirement for high PIP which is associated with lung injury.
- Inhaled nitric oxide at 20 ppm (if available), if:
 - Oxygenation index (OI) >15
(Oxygenation index = (mean airway pressure x FiO_2 x 100)/ PaO_2 (in mmHg)
<https://www.mdcalc.com/calc/80/oxygenation-index>)
 - Clinical or echocardiographic evidence of right ventricular failure
 - Difference in pre- and post-ductal SpO_2 >5%, in the absence of CHD.
(Monitor methaemoglobin levels on blood gases closely; methaemoglobinemia can aggravate hypoxia; adjust nitric oxide range 5-20 ppm to keep methaemoglobin <5%).
- Aim to reduce PaCO_2 to normal levels to prevent pulmonary vasoconstriction.
- Use sodium bicarbonate to maintain pH >7.35 if gas exchange permits.
- Drain pneumothoraces. PPHN with pneumothorax is likely to require transport for ECMO assessment.
- Optimise circulating volume using 10ml/kg fluid boluses, reassessing response to each bolus.
- Treat hypotension with fluid resuscitation, inotropes (adrenaline) and vasopressors (noradrenaline). Consider targeting a mean arterial pressure > estimated pulmonary artery pressure in order to reverse the shunt from right-to-left to left-to-right. Using very high doses of inotropes and vasopressors to achieve supra-physiological blood pressure may be harmful.
- Consider a magnesium bolus of 50mg/kg over 30 min if mean arterial pressure is maintained (and watch carefully for hypotension). This may be repeated as tolerated if

effective or administered as an infusion (suggested maximum serum Mg level of 3 mmol/l).

- Correct hypocalcaemia, hypomagnesaemia, and hypoglycaemia.
- Maintain normothermia.
- Investigate and treat for infection with appropriate antibiotic cover.

Other therapies to consider in cases of refractory hypoxia

- *Discussion with CATS consultant is required.*
- Other therapeutic options include:
 - Hydrocortisone, if refractory septic shock is suspected.
 - Milrinone infusion, used as a pulmonary vasodilator and inotrope. Systemic vasodilation is likely to result in increased vasopressor requirement.
 - Prostaglandin (PGE) infusion for ductal patency to preserve right ventricular systolic function and limit afterload, and as a pulmonary vasodilator.
 - Alternative ventilation modes, such as APRV.