Management of Persistent Pulmonary Hypertension of the Newborn

Reference:	1764v2
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Purpose

To recommend evidence-based or best practice for the management of persistent pulmonary hypertension in newborn term and preterm infants.

Intended Audience

The guideline is intended for use by the Embrace clinical staff.

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1. Introduction

Persistent pulmonary hypertension is the failure of transition from intra- to extra-uterine life, resulting in high pulmonary pressures leading to extra-pulmonary right to left shunting (across a patent ductus arteriosus/patent foramen ovale) causing hypoxaemia.

2. Guideline Content

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Background: To recommend evidence based or best practice for management of persistent pulmonary hypertension (PPHN) in the newborn term and preterm infant. PPHN is failure of the circulatory transition from intra- to extra-uterine life, resulting in high pulmonary pressures leading to extra-pulmonary right to left shunting (across PDA/PFO) causing hypoxaemia.

Consider PPHN diagnosis:

- Often term or near term infant
- Onset at birth or few hours after
- Profound hypoxia – low arterial PaO_2 on blood gas may be associated with low, normal or high $PaCO_2$
- **Always consider congenital cardiac disease presenting with a right to left shunt**

Investigations:

- **Echo** – gold standard; exclude congenital cardiac disease (TAPVD difficult); diagnose PPHN with raised pulmonary pressures and tricuspid regurgitation
- **Oxygen saturations** – Pre (right hand) and post-ductal (feet) differential of 10-15%; measure simultaneously
- **CXR** to exclude other pathology and guide respiratory management – lung expansion, RDS requiring surfactant etc. may be normal
- **Bloods** – FBC, coag, U&E, LFT, lactate, Ca^{2+}, Mg^{2+}
- **Cranial ultrasound** – if ECMO referral

General measures

- Minimal handling, reduce noise (minimuffs) and light (goggles)
- Normothermia
- Central access – UAC and double lumen UVC
- Antibiotics
- Sedation and muscle relaxation if intubated

Specific measures to consider if iNO not available or effective – on advice of tertiary/ECMO centre

- Milrinone
- Adenosine
- Alprostadil
- Epoprostenol
- Tolazoline
- Magnesium sulphate

Outcomes

- Mortality 10-20% - highest with hypoplastic pulmonary vasculature
- Often neurodevelopmental, cognitive and hearing problems (approx. 25%)

Oxygen Index = $\frac{\text{MAP (cmH20) x FiO2 x 100}}{\text{Post-ductal } PaO_2 \text{ (mmHg)}}$
(1KPa = 7.5mmHg)

OI > 15 consider iNO OI > 25 refer to ECMO

Take: the nitric tray and spare nitric cylinder
Additional saturation monitor

Use a multi-system, simultaneous approach to A,B, C

A - Intubate

B – Optimise oxygenation

- Give oxygen to achieve arterial PaO_2 >10 or saturations >95% (pre-ductal)
- Increase mean airway pressure as required
- Give surfactant if indicated (RDS, MAS)
- Optimise haemoglobin (transfuse if <140)
- Give sedation and muscle relaxation

Maintain normal PCO_2 4.5-5.5 KPa

Maintain normal pH 7.35 – 7.45

- Use sodium bicarbonate or THAM as needed

C – Optimise systemic blood pressure

- Term baby aim mean BP >50mmHg (may need to be higher, saturation gap should start to close)
- Give volume 10-20ml/kg 0.9% sodium chloride
- Give 'useful' volume eg. Packed cells, FFP if appropriate
- Optimise Ca^{2+}
- Start **dopamine** 5 microgram/kg/min, assess after 10 mins, increase to max 10 microgram/kg/min
- Start second agent early (dopamine @10 microgram/kg/min)
- **Dobutamine** 5-20 microgram/kg/min
- **Adrenaline** 0.05 microgram/kg/min to 1 microgram/kg/min

If poor response discuss with tertiary and/or ECMO centre

- **Hydrocortisone 2.5mg/kg/ iv QDS**
- **Noradrenaline** start at 0.1microgram/kg/min

OR

- **Vasopressin (Argipressin)** start at 0.009-0.03 units/kg/hr (seek ECMO centre advice for starting dose and use of higher doses)

Start iNO at 20ppm

- Consider use in preterm infants >26 weeks on an individual basis where there is evidence of PPHN

Reassess every 20 minutes
Consider referral to ECMO if OI>25

ECMO – refer via Embrace (through CenTre)
Exclusions:

- Known non-reversible underlying condition
- <34 weeks gestation
- <2kg
- Significant intracranial injury (need cranial ultrasound)

Nair J, Lakshminrusimha S. Update on PPHN: Mechanism and Treatment Semin Perinat. 2014 March; 38 (2) 78-91
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