

Title:	Persistent Pulmonary Hypertension of the Newborn (PPHN)		
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Definition

Persistent Pulmonary Hypertension (PPHN) of the newborn is defined as delay in the normal postnatal decline in pulmonary vascular resistance resulting in arterial hypoxemia, with associated shunting of de-oxygenated blood across to the systemic circulation resulting in persistence of the foetal circulation.

Three main types

1. PPHN with initially normal lungs
e.g. perinatal asphyxia, sepsis, congenital heart disease, maternal diabetes, Down's Syndrome
2. PPHN associated with hypoplasia of lung tissue
e.g. congenital diaphragmatic hernia, Potter's syndrome, oligohydramnios
3. PPHN associated with pulmonary parenchymal disease or primary pulmonary hypertension
e.g. RDS, meconium aspiration syndrome, chronic fetal hypoxia with increased smooth muscle, pneumonia

Guide to Diagnostic Features

- Usually presents in term or post-term infants
- Onset at birth or few hours after birth
- Signs
 - Cyanosis
 - Tachypnoea
 - Low arterial pO₂ levels which may be lower in post ductal samples (similar patterns may be found in interrupted aortic arch, coarctation of the aorta) - presentation often mimics cyanotic congenital heart defect
- pCO₂ normal or low in absence of parenchymal lung disease
- CXR normal (idiopathic PPHN) or abnormal related to lung pathology
- Evidence of shunt may be present with difference in pre- and post-ductal SaO₂

Management aims

- Lower pulmonary vascular resistance & maintain or increase systemic blood pressure
- Reverse right-to-left shunting
- Minimise barotrauma
- Improve arteriolar oxygen saturation and oxygen delivery to the tissues

Investigation

1. Pre & postductal pulse oximetry screening
2. Arterial blood gas sampling and oxygenation index calculation
3. Chest radiography
4. ECHO – if possible

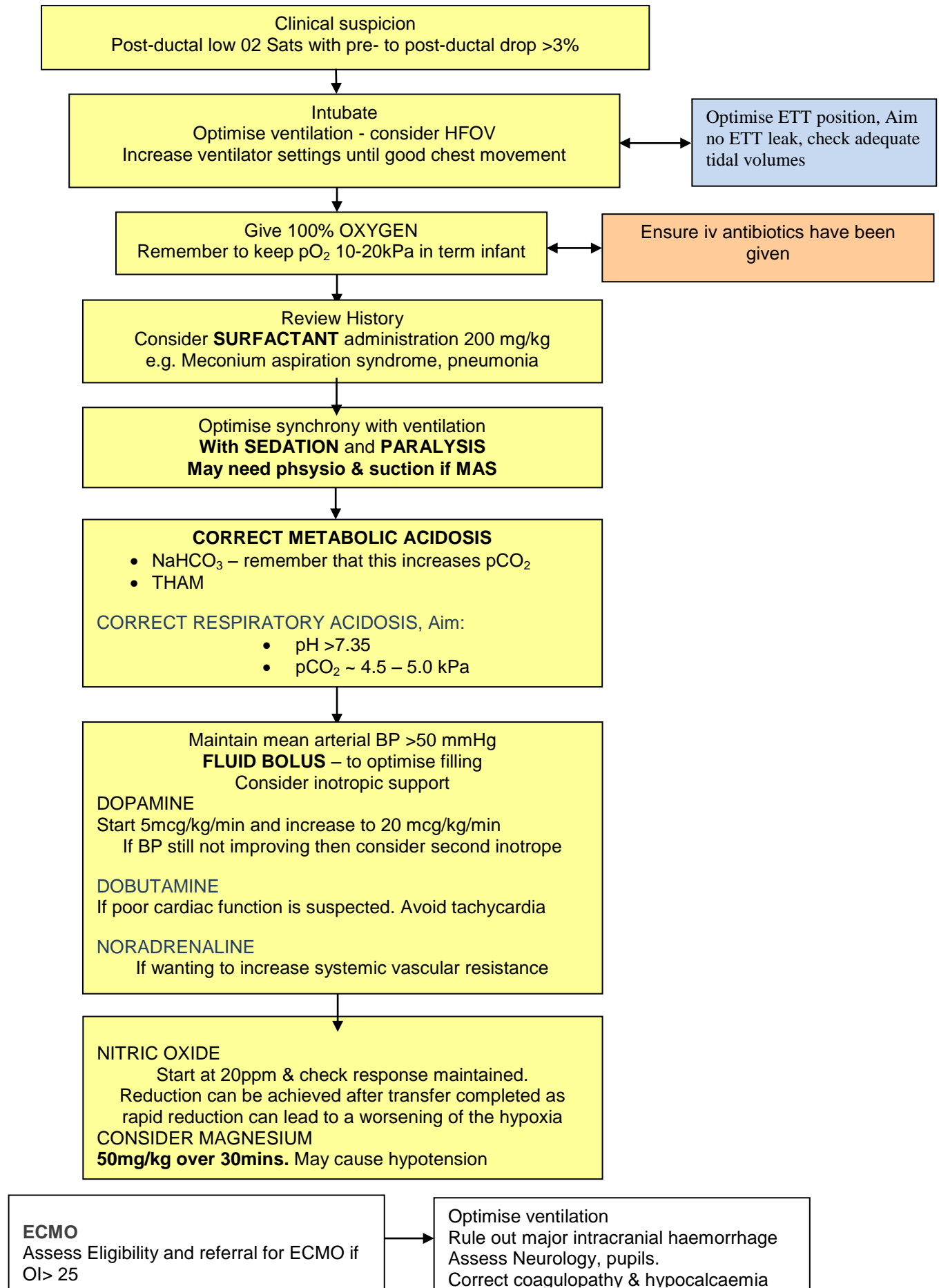
Gold standard is Echocardiogram to differentiate between PPHN and cyanotic congenital heart defect
It also helps

- Assess adequate filling

- Define the pulmonary artery pressures by:
 - Determining R→L shunt across ductus arteriosus +/- foramen ovale
 - Calculating right ventricular (RV) systolic pressures by peak velocity of tricuspid regurgitation (TR)
 - Define myocardial contractility
5. Cranial ultrasound scan
- To exclude significant intra-cranial haemorrhage
 - This is essential prior to considering or referring for Extra Corporeal Membrane Oxygenation (ECMO)
6. Correct coagulopathy

The differential diagnosis of PPHN includes cyanotic congenital heart disease, primary pulmonary disorders, and sepsis. However congenital pneumonia and sepsis can itself lead to PPHN

Treatment



Indications for ECMO

- Oxygenation index > 40.
- Birthweight >2kgs
- Gestation > 34 weeks
- Severe but reversible cardiac or pulmonary disease unresponsive to optimal ventilation and pharmacological therapy

Contraindications for ECMO include:

- Major intracranial haemorrhage (> Grade 2 PVH)
- Prolonged asphyxia predicted to cause brain damage
- Lethal congenital abnormality

Calculation for mean airway Pressure

$$M_{PAW} = MAP (cmH_2O) = \frac{(PIP \times T_i) + (PEEP \times T_e)}{T_i + T_e}$$

Where PIP = Peak inspiratory pressure cmH₂O
 PEEP = Positive end expiratory pressure cmH₂O
 T_i = Inspiratory time s
 T_e = Expiratory time s

Calculation of the Oxygenation Index (OI)

$$OI = \frac{F_i O_2 \times MAP(cmH_2O) \times 100\%}{P_a O_2(mmHg)}$$

UK gas machines usually give PaO₂ in kPa so to convert PaO₂ (mmHg) = PaO₂ (kPa) x 7.5

In the context of this transport service, referrals should normally be made to Great Ormond Street Hospital. Referrals are via CATS on 0207 430 5850. Following discussion with the ECMO team, it can be decided whether the NTS team or the CATS team would be most appropriate to expedite the transfer. The main considerations should be optimisation of treatment and timely movement of the baby before secondary hypoxic deterioration and damage occurs.

References:

1. University of Washington website: <http://neonatal.peds.washington.edu/NICU-WEB/pphn.stm#Dx>
2. Prince Alfred Hospital Sydney Australia:
<http://www.cs.nsw.gov.au/rpa/neonatal/html/newprot/pphn.htm>
3. University of California website:
http://www.ucsfhealth.org/childrens/health_professionals/manuals/27_PersPulmHyper.pdf
4. Women and Children's Hospital Melbourne Australia Neonatal Handbook website:
www.wch.org.au/nets/handbook/index.cfm?doc_id=448
5. Lancet 1996; 348:75-82