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

Clinical suspicion
Post-ductal low 02 Sats with pre- to post-ductal drop >3%

Intubate
Optimise ventilation - consider HFOV
Increase ventilator settings until good chest movement

Give 100% OXYGEN
Remember to keep pO₂ 10-20kPa in term infant

Review History
Consider SURFACTANT administration 200 mg/kg
e.g. Meconium aspiration syndrome, pneumonia

Optimise synchrony with ventilation
With SEDATION and PARALYSIS
May need physio & suction if MAS

CORRECT METABOLIC ACIDOSIS
- NaHCO₃ – remember that this increases pCO₂
- THAM

CORRECT RESPIRATORY ACIDOSIS, Aim:
- pH >7.35
- pCO₂ ~ 4.5 – 5.0 kPa

Maintain mean arterial BP >50 mmHg
FLUID BOLUS – to optimise filling
Consider inotropic support

DOPAMINE
Start 5mcg/kg/min and increase to 20 mcg/kg/min
If BP still not improving then consider second inotrope

DOBUTAMINE
If poor cardiac function is suspected. Avoid tachycardia

NORADRENALINE
If wanting to increase systemic vascular resistance

NITRIC OXIDE
Start at 20ppm & check response maintained.
Reduction can be achieved after transfer completed as rapid reduction can lead to a worsening of the hypoxia
CONSIDER MAGNESIUM
50mg/kg over 30mins. May cause hypotension

ECMO
Assess Eligibility and referral for ECMO if OI> 25

Optimise ETT position, Aim no ETT leak, check adequate tidal volumes

Ensure iv antibiotics have been given

Barts Health
NHS Trust

Printed/uploaded: 01/03/2015
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 \ (cmH_2O)
- PEEP = Positive end expiratory pressure \ (cmH_2O)
- T_i = Inspiratory time \ (s)
- T_e = Expiratory time \ (s)

Calculation of the Oxygenation Index (OI)

\[ OI = \frac{F_iO_2 \times MAP(cmH_2O) \times 100\%}{P_aO_2(mmHg)} \]

UK gas machines usually give PaO_2 in kPa so to convert PaO_2 (mmHg) = PaO_2 (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:
5. Lancet 1996; 348:75-82