PERSISTENT PULMONARY HYPERTENSION OF THE NEWBORN (PPHN)

PPHN is not a single disease entity but a complex multifactorial disorder that causes arterial or venous pulmonary hypertension in the newborn.

PRIMARY PPHN
Mild or no lung disease, but has hypoxaemia that is disproportionate to the X-RAY and clinical signs.

SECONDARY PPHN
There may be associated pulmonary or other systemic problems precipitating the PPHN:
- Severe intrapartum asphyxia.
- Group B Strep. Sepsis (or other sepsis).
- Pneumonia of any cause.
- Meconium aspiration syndrome.
- Polycythemia.
- Pulmonary hypoplasia.
- Maternal drug use/ fetal duct closure.
- Alveolar capillary dysplasia.
- Iatrogenic PPHN secondary to overventilation.
- Congenital heart disease: conditions which obstruct the venous outflow from the lungs or cause myocardial failure.

ALL HAVE:
- Severe hypoxaemia, usually a PaO2 < 37.5-45 mmHg in FiO2 of 1.0 and ventilation if necessary.
- Evidence of a R-to-L ductal shunt.
- ECHO confirmation of a structurally normal heart.

EPIDEMIOLOGY
In the mid 80’s, the incidence was 1-2/1000 live births with a mortality around 50%, and high morbidity. With the advent of additional supports, this has decreased.

PATHOPHYSIOLOGY
Pulmonary hypertension arises when the pulmonary vascular resistance does not decrease at birth. Blood is shunted R-to-L at the level of the ductus arteriosus and the foramen ovale. In addition, high pulmonary vascular resistance results in right ventricular overload which, when severe, leads to displacement of the interventricular septum and impaired left ventricular function. In some rare instances there may be an anatomical abnormality of the pulmonary vessels (alveolar capillary dysplasia and congenital diaphragmatic hernia).
CLINICAL PRESENTATION

In primary PPHN, the primary presenting feature is cyanosis. The presentation is often more subtle and may mimic cyanotic congenital heart disease. The infant remains cyanosed even when high oxygen concentrations are administered by IPPV. Respiratory distress is often mild, but with the respiratory rate increased to 60-100/min, higher rates seen in term infants. Retraction is mild and grunting rare. Ventilation or PaCO₂ may be normal to low in primary pulmonary hypertension. Air entry is usually normal and added sounds are rarely present. Infants with primary PPHN virtually always present within 12 hours of birth and very rarely after 24 hours age.

In PPHN secondary to pre-existing lung disease, the clinical features will be that of the underlying disease eg. RDS, GBS sepsis or MAS, together with cyanosis. Secondary effects from hypoxia such as hypotension and acidemia may be present. The age at diagnosis is related to the underlying problem and its severity. In GBS, severe asphyxia and congenital diaphragmatic hernia, PPHN will appear shortly after birth usually within 6 hours.

Hyperoxia test usually demonstrates improved oxygenation, except in severe cases. The heart rate is normal or slightly increased. All pulses, including the femoral pulses, are normal. The second heart sound is often single and loud because of the rise in pulmonary artery pressure. There can be a parasternal heave and a soft systolic murmur signifying functional tricuspid incompetence. Heart failure is not usually present but the infant may be hypotensive.

Examination of the abdomen, genito-urinary system and CNS in the absence of predisposing factors is usually normal.

MANAGEMENT

This is the same as that for MAS and needs to be modified for the particular underlying cause in the presence of secondary PPHN. In those requiring long term ventilation, PIE and Pneumothorax are common, as is CLD.