

Intensive Care Nursery House Staff Manual

Pulmonary Hypoplasia, including Congenital Diaphragmatic Hernia

INTRODUCTION: As survival for other pulmonary conditions has improved in recent years, pulmonary hypoplasia (P-Hyp, inadequately sized lungs) has become an increasingly important cause of neonatal morbidity and mortality. P-Hyp has been described as the most common anomaly in infants who die in the neonatal period. P-Hyp has several causes, and it can often be suspected on the basis of historical factors or ultrasound findings during the current pregnancy. Appropriate resuscitation of infants with P-Hyp requires special considerations and techniques.

ETIOLOGY AND PATHOGENESIS: The lungs are unique in that their growth is dependent primarily on extrinsic factors. The factors which lead to P-Hyp and the clinical conditions associated with these factors are:

- Inadequate intra-thoracic space:** Congenital diaphragmatic hernia (CDH); intrathoracic tumors; pleural effusions
- Prolonged oligohydramnios:** Renal agenesis; prolonged rupture of fetal membranes
- Decreased or absent fetal breathing movements:** CNS lesions; phrenic agenesis

With P-Hyp, there is inadequate pulmonary parenchymal tissue and pulmonary blood flow for normal gas exchange.

CLINICAL FEATURES of P-Hyp include:

- Immediate respiratory distress with tachypnea, cyanosis, retractions, hypercarbia, acidosis
- With specific conditions, there are typical clinical signs, for example:
 - CDH: scaphoid abdomen
 - Oligohydramnios: “Potter’s facies,” arthrogryposis
 - CNS lesions: other signs of abnormal CNS function
- P-Hyp can also occur as part of certain syndromes (*e.g.*, Pena-Shokier Syndrome)

DIAGNOSIS can be made with certainty only at autopsy by measurement of total lung DNA content. Because of this, the incidence of P-Hyp is probably underestimated. The diagnosis should be suspected in the presence of:

- Historical factors suggestive of P-Hyp (*e.g.*, prolonged oligohydramnios)
- Unexpected respiratory distress
- Specific clinical findings (*e.g.*, scaphoid abdomen, “Potter’s facies”)

MANAGEMENT: Adequate care begins with a high level of suspicion that P-Hyp is the cause of the infant’s respiratory problems. If there is concern that P-Hyp is present, follow the guidelines below in addition to basic resuscitation techniques (P. 1):

- Intubate infant’s trachea and start assisted ventilation.** Unless P-Hyp is very mild, infant will require assisted ventilation.
- Use low ventilatory pressures (PIP <25 cmH₂O) and high rates (60-90/min) with very short Ti.** Infants with P-Hyp have ↑ incidence of **pneumothorax**.

- Insert **umbilical arterial catheter**, especially for measurement of **pH and PaCO₂**. Because these infants often have ↓ peripheral perfusion, capillary samples for pH and blood gas tensions are not adequate.
- Obtain **chest radiograph** immediately.
- Place pre- (right hand) and post-ductal (lower extremity) saturation monitors to help evaluate for pulmonary hypertension complicating P-Hyp.
- Consider early use of **inhaled nitric oxide** therapy (see P. 89).
- Even in cases of apparently severe P-Hyp, **continue vigorous resuscitative efforts for at least several hours**. In some cases, there will be rapid resolution of the respiratory distress, as the infant does not actually have P-Hyp, despite historical factors and severe respiratory distress. The reason for the severe respiratory distress mimicking P-Hyp is not evident.

MANAGEMENT OF INFANT WITH KNOWN OR SUSPECTED CDH: In addition to the above:

- Intubate the infant's trachea immediately. Do not use bag and mask ventilation.** This will cause distension of stomach and small bowel (which are in the chest) and worsen respiratory status.
- If CDH is known prenatally, be prepared to insert umbilical venous catheter to administer morphine and pancuronium (0.1 mg/kg of both) to facilitate assisted ventilation and prevent the infant swallowing air. Because this mixture is not damaging to the vasculature, it can be given as soon as the UVC has been inserted, even if the tip is in the portal circulation.
- Place pre- and post-ductal SpO₂ monitors.** Use pre-ductal to judge oxygenation.
- Insert Replogle tube to suction to decompress stomach and small bowel.**
- In addition to low pressures for PIP, use **low PEEP levels (2-3 cmH₂O)**. Infants with CDH do not tolerate higher PEEP levels for reasons that are not apparent
- Do not attempt to correct hypoxemia and hypercarbia rapidly.** Infants with CDH should receive ventilation with low pressures and rapid rates. The aim of assisted ventilation should be to provide adequate oxygenation (SpO₂ >75%) and ventilation (PaCO₂ ≤60 mmHg). Oxygenation and ventilation almost always improve over the first few hours after birth.
- Notify the Pediatric Surgeons immediately if there is a CDH. Surgical repair of the CDH should be delayed until after stabilization of the respiratory status, which may take several days. The Surgeons should be notified early in case of the need for ECMO and so that they can follow the patient closely with the ICN team.
- Look for other anomalies.** Approximately 40% of infants with CDH have congenital heart disease or other significant anomalies.

OUTCOME of infants with P-Hyp is related mainly to lung size and presence of other anomalies. Because of the difficulty of diagnosing P-Hyp clinically, accurate mortality statistics are not available. Prenatal ultrasound can help predict outcome of infants with CDH (Lung:Head Ratio at 22-27 weeks). Severe P-Hyp due to renal causes of oligohydramnios generally has a poor prognosis for survival. Survivors of P-Hyp often have chronic lung disease. Infants with CDH also have associated problems with feeding, growth and development.