

Congenital Diaphragmatic Hernia

Anesthetic Pearls: Anesthetic Management of Congenital Diaphragmatic Hernia (CHD)

Occurrence:

- 1 in 5000 births
- High mortality as a result of lung hypoplasia and associated birth defects

Site Classification:

1. 80% posterolateral (Bochdalek) defects
2. Left-sided defects are 5 times more common than right-sided lesions.
3. Bochdalek hernias are the largest with the greatest degree of pulmonary hypoplasia.
4. Approximately 2% of CDH are small anterior defects of the foramen of Morgagni (the remainder are through the esophageal hiatus).

Embryology:

A common pleuroperitoneal cavity present during the first fetal month becomes 2 separate cavities during the second fetal month with the formation of the pleuroperitoneal membrane. The developing gut may return from the yolk sac to the peritoneum before the pleuroperitoneal closure is complete, the stomach, spleen, liver, and the small and large bowel may migrate into the chest cavity. The normal development of the lung is affected by the space-occupying mass of migrated abdominal contents. The ipsilateral lung is affected the most, but both lungs are abnormal. Bronchial and bronchiolar development arrest occurs, the number of airway generations can be decreased by 50%. The pulmonary artery is smaller in proportion to the size of the lung. Arterial branching is decreased. There is a normal ratio of alveoli to capillaries, but the total vascular cross-sectional area is decreased leading to pulmonary hypertension.

Pathophysiology:

The primary cause of death in CDH is progressive hypoxemia and acidosis caused mostly by elevated pulmonary vascular resistance (PVR) and persistent pulmonary hypertension of the newborn (PPHN). Pneumothorax complication of mechanical ventilation is most clinically significant occurring on the contralateral side.

Diagnosis:

Prenatal ultrasound. Postnatal, the newborn presents with immediate respiratory compromise, scaphoid abdomen, and barrel-shaped chest. Early onset of symptoms reflects degree of lung hypoplasia and defect size. Respiratory failure in first 6 hours of life considered higher risk (those presenting in first hour of life have highest mortality).

Medical treatment:

Maintain normoxia, and slight hypocarbia, and a normal to high pH. ECMO may be necessary. Pulmonary vasodilators (Milrinone, Epinephrine, and inhaled Nitric Oxide) are helpful.

Intra-Operative Care:

Maintenance of normoxia ($\text{PaO}_2 > 80 \text{ mmHg}$) and normo to hypocarbia ($\text{PaCO}_2 < 30 \text{ mmHg}$) using minimal airway pressure ($< 30 \text{ cm H}_2\text{O}$) and rapid respiratory rates (60 – 120 breaths per minute). Pulmonary mechanics change during the operative manipulation and pulmonary compliance may decrease necessitating hand ventilation with close attention to the progress of the surgery. Sudden deterioration in lung compliance, oxygenation, or blood pressure suggests a developing pneumothorax. Be prepared for immediate chest tube insertion. Monitor ABG frequently and correct acidosis with sodium bicarbonate infusion. Opioids (Fentanyl and Sufentanil) and muscle relaxants are used. Inhaled anesthetics are used as tolerated by the cardiovascular stability of the child. Avoid nitrous oxide which may cause distention of intestinal loops.

