

Congenital Diaphragmatic Hernia

Congenital diaphragmatic hernias occur as a result of intra-abdominal contents protruding into the thoracic cavity through a defect in the diaphragm. This condition occurs during intrauterine development. Symptoms typically become apparent after birth. Patients present with loops of bowel in the chest, and may also present with pulmonary hypoplasia or respiratory distress at birth. These types of hernias can be classified by where they occur in the diaphragm, namely Bochdalek and Morgagni. The diagnostic workup includes chest x-ray and/or ultrasound. Treatment consists of corticosteroids, surfactant, mechanical ventilation, and surgery. An important consideration to remember is that the herniated bowel may become incarcerated or even perforate.



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Clinical Features

Bowel in the Chest

Bowel-bowl in Treasure-chest

In congenital diaphragmatic hernias, loops of bowel or other intra-abdominal contents protrude into the thoracic cavity.
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Pulmonary Hypoplasia

Lungs and Hippo-plates

The protruded intra-abdominal contents occupy space in the thoracic cavity and can prevent the lungs from expanding and properly developing. This can lead to the development of pulmonary hypoplasia.

Respiratory Distress

Lungs Shooting Flare-gun

As a result of the intra-abdominal contents compressing on the lungs as well as the associated pulmonary hypoplasia, neonates can develop severe respiratory distress immediately after birth. Since the fetal pulmonary respiratory function is not vital for survival during the intrauterine period, patients are typically asymptomatic up until birth.

Classification

Bochdalek Hernia

Book-to-left with Hermit-crab

Bochdalek hernia is a subtype of congenital diaphragmatic hernia which accounts for more than 90% of all cases. This type of hernia occurs on the posterior left-side of the chest. This type of hernia is not typically seen on the right side of the chest because the liver prevents herniation of intra-abdominal contents. Bochdalek hernias occur because of the failure of closure of the pleuroperitoneal membrane in utero. This eventually leads to a defect in the posterior attachment of the diaphragm.

Morgagni Hernia

Captain-Morgan with Hermit-crab

Morgagni hernia is one subtype of congenital diaphragmatic hernia. It is a rare type of hernia and accounts for approximately 3% of all cases. In contrast to Bochdalek hernias, Morgagni hernias typically occur on the anterior right-side of the chest wall. Morgagni hernias occur as a result of intra-abdominal contents protruding through the foramen of Morgagni. This anatomic area serves as a passing point of the internal thoracic vessels into the abdomen and constitutes a weak point through which abdominal contents can herniate.

Diagnostic Workup

Chest X-ray

Chest X-ray

Newborn chest X-rays can be used to diagnose congenital diaphragmatic hernias. Chest X-rays will typically show bowel loops on one side of the chest.



Ultrasound

Ultrasound

Prenatal ultrasound can be used to diagnose congenital diaphragmatic hernias.

Treatment

Corticosteroids ± Surfactant

Quarter-on-steroids and Surf-Surfactant-Surfboard

Steroids may be used to treat CDH is diagnosed during the prenatal period. Steroids help expedite pulmonary development and reduce the risk of preterm labor and delivery. If the patient has significant respiratory distress i.e. NRDS, then surfactant can be administered to keep alveoli open to allow for adequate gas exchange.

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Mechanical Ventilation

Machine Vent Ventilating

Patients may require intubation immediately after birth, mechanical ventilation, and stabilization. In some cases, patients may require extracorporeal membrane oxygenation (ECMO).

Surgery

Surgeon

Patients typically require surgery to correct the anatomic defects. In some cases, surgery is delayed for 3-4 days after birth to allow for lung maturation. Surgery involves reducing the bowel contents back into the abdomen and closing the diaphragmatic defect.

Considerations

Bowel Incarceration or Perforation

Bowel-bowl Incarcerated with Perforations

In congenital diaphragmatic hernias, the protruded loops of bowel may become incarcerated or perforate. This is a serious complication of CDH and requires immediate treatment.

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