Congenital Diaphragmatic Hernia

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Congenital diaphragmatic hernia (CDH) – This condition is often severe due to an accompanying pulmonary hypoplasia. Through advanced neonatal intensive care and select application of extracorporeal membrane oxygenation, most children survive to correction.

1. Epidemiology:
   (a) One in 2,000–5,000 births.
   (b) May be higher in stillborn children (1/3 of CDH).
   (c) Higher in females if stillborn children are included.
   (d) Two percent in first-degree relative.
   (e) Eighty percent left-sided.
   (f) Linked to phenmetrazine, thalidomide, quinine, nitrofen, vitamin A deficiency.
2. Associated anomalies are present in 28–50 % of infants with CDH and in 100 % of stillborn infants with CDH. Common defects are neural tube, cardiac, and midline defects, such as pentalogy of Cantrell. CDH is more common in trisomy 13, 18, and 21.
3. Pathophysiology:
   (a) Midgut returns to abdomen week 9–10.
   (b) If diaphragm is open, abdominal viscera herniate to thoracic cavity.
   (c) Occurs by week 12.
   (d) Malrotation occurs.
   (e) Hernia sac present in 10–15 %.
   (f) Left more common than right.
   (g) Bochdalek (posterior lateral) hernia is more common than Morgagni (anterior) hernia.

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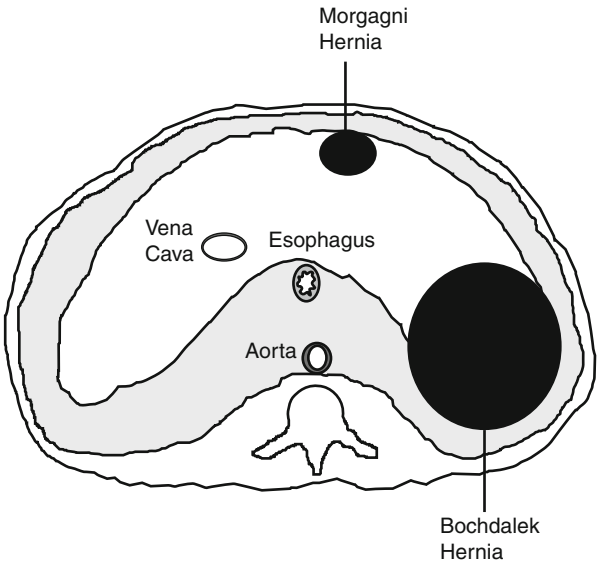

Fig. 1 Types of congenital diaphragm hernias. Morgagni hernias are defects in the anterior diaphragm while the more common Bochdalek hernias are in the posterolateral diaphragm (*Source*: Christopher Coppola)

(h) Can contain liver, spleen, gastrointestinal tract, and kidney.
(i) Associated pulmonary hypoplasia.

(i) Decreased lung mass.
(ii) Decreased bronchial branching.
(iii) Decreased alveoli.
(iv) Decreased total vascular diameter.
(v) Hypertrophied vascular smooth muscle.
(vi) Smooth muscle in alveolar vessels.
(vii) Bilateral lungs affected.

(j) Factors resulting in persistent fetal circulation in CDH.

(i) Pulmonary hypoplasia.
(ii) High pulmonary vascular resistance.
(iii) Hypoxia.
(iv) Acidosis.
(v) Stress.

(k) Diagnosis:

(i) Prenatal ultrasound (CDH, polyhydramnios).
(ii) Respiratory distress after delivery.
(iii) Scaphoid abdomen/funnel chest.
(iv) Abdominal x-ray with nasogastric tube, showing tube in chest.
(v) Ten – 20 percent with delayed presentation.

(l) Predicting severity of CDH:

(i) Prenatally diagnosed CDH as a group has worse prognosis than CDH diagnosed after birth.
(ii) Presence of cardiac defects worsens mortality.
(iii) Smaller pulmonary artery size (which can be quantified by the McGoon Index) carries worse prognosis.
(iv) Defects large enough to allow herniation of the stomach into the chest have worse prognosis and have reported to have survival as low as 30 %.
(v) Right-sided hernia represents more severe disease occurring earlier in gestation.
(vi) After delivery, the Oxygenation Index (OI) = (MAP × FiO2/PaO2) is used to quantify severity of pulmonary hypoplasia.

1. OI <0.06 carries a survival of 98 %.
2. OI >0.175 predicts survival of 0 %.
3. OI can be used to predict need for extracorporeal membrane oxygenation (ECMO).

(m) Prenatal care:

(i) When a patient is prenatally diagnosed with CDH, parents should receive genetic counseling, and karyotyping should be performed on the fetus to detect any associated anomalies or syndromes.
(ii) Prenatal care should be given to support gestation as close as possible to term to minimize the effect of premature delivery on an already compromised pulmonary function.
(iii) When possible, delivery should occur in a center with access to high level NICU care and a plan for access to ECMO therapy if it becomes necessary.

(n) Treatment:

(i) Initial resuscitation:

1. Patient should be assessed by neonatologist and surgeon after deliver. IV fluid should be started and a nasogastric tube is placed to decompress the stomach, which may be in the chest, and to reduce the volume of gas and fluid in the gastrointestinal tract.
2. Pulmonary function should be monitored for signs of pulmonary hypoplasia and failure. The patient may worsen over the course of the initial 48 h “honeymoon” period.
3. Obtain chest x-ray to detect abdominal organs in the chest.
4. When intubation and mechanical ventilation is needed, use a strategy of “gentle ventilation” to avoid barotrauma to the alveoli.

(a) High frequency/low pressure ventilation, with use of an oscillating ventilator when needed.

(b) Permissive hypercapnea.
(c) Medications to reduce pulmonary hypertension.

(i) Inhaled nitric oxide.
(ii) Sildenafil.
(iii) Tolazoline.

(d) When high frequency ventilation and inhaled nitric oxide fail to provide adequate oxygenation, evaluate patient for suitability for ECMO therapy.

(ii) Surgical repair of CDH:

1. Timing of operation: Repair should be delayed until after the initial 48 h "honeymoon period" to allow for stabilization and assessment for extent of pulmonary hypoplasia, rather than add the stress of repair to a child with worsening pulmonary function. When ECMO is needed, it is advantageous to delay repair until after completion of the ECMO run to avoid operation while the child is heparinized and more prone to hemorrhage. Rarely, it is necessary to repair CDH while on ECMO because child is failing to progress.
2. Technique:

(a) Subcostal incision or thoracotomy: On the left, the approach via abdomen allows for convenient reduction of viscera into abdominal cavity. For a right sided CDH, thoracotomy avoids the difficulty of working around the liver that is encountered with an abdominal approach (though a right subcostal incision is a viable option.)

(i) Reduce hernia contents back into abdomen.
(ii) Excise hernia sac when present as it can interfere with visualization of lungs or viscera while closing defect.
(iii) Identify muscular rim of diaphragm circumferentially. Some areas along posterolateral ribs and mediastinal structures may have no diaphragm and require careful placement of sutures.
(iv) Determine if primary closure is possible or if a patch of exogenous material (artificial or biologic mesh) will be needed (approximately 1/3 of cases).
(v) Consider addressing malrotation/non-fixation of bowel, if present, by performing a Ladd's procedure. An inversion appendectomy, rather than an amputation of appendix, will preserve the operative field as clean.
(vi) Close abdomen if possible. If there is insufficient domain in abdomen after reducing viscera from chest to abdomen, a silo or mesh closure of abdomen may be needed on a temporary basis.

(o) Outcome:

(i) Through the twentieth century, CDH has historically had 50 % survival, however advancements in neonatal intensive care unit techniques and select use of ECMO have produced a survival range of 39–95 % (mean, 69 %), depending on setting of care.
(ii) There is a risk of both early and late recurrence. Early recurrence can be due to technical error and increased abdominal pressure. Placement of mesh reduces tension across the repair, but approximately 1/3 of patients will need a late revision of repair due to recurrence of hernia or due to tension on the thoracic wall and deformation of the ribcage.
(iii) Chronic lung disease may result from the accompanying pulmonary hypoplasia.
(iv) Gastroesophageal reflux is common after CDH repair and some patients will require an anti-reflux procedure.
(v) Development/cognitive deficits occur and can be due to associated anomalies or as sequela from ECMO: