Role of imaging in congenital diaphragmatic hernias

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Learning objectives

To explore the spectrum of different types of diaphragmatic hernias in pediatrics population.

Main radiological findings of CDH.

Post procedural images and outcome if applicable.
Background

Congenital diaphragmatic hernias are seen in 1 of every 2000-4000 live births. 84% are left-sided, 13% are right-sided and 2% bilateral (usually fatal).

There are 3 basic types of congenital diaphragmatic hernia: posterolateral Bochdalek hernia, anterior Morgagni hernia and hiatus hernia. Hiatal hernias account for only 9% of diaphragmatic hernias in infants younger than 1 year.

Congenital Diaphragmatic hernia (CDH) is characterized by a defect in the diaphragm leading to the protrusion of abdominal contents into the thoracic cavity affecting the normal development of the lungs. The condition may present as an isolated lesion or as part of a syndrome. It is one of the most common non-cardiac fetal intrathoracic anomalies. It accounts for 8% of all major congenital anomalies.

Left-sided hernias allow herniation of both the small and large bowel and intraabdominal solid organs into the thoracic cavity. In right-sided hernias, only the liver and a portion of the large bowel tend to herniate.

Infants with CDH most commonly present with respiratory distress and cyanosis in the first minutes or hours of life, although a later presentation is possible. The respiratory distress can be severe and may be associated with circulatory insufficiency, requiring aggressive resuscitative measures. The degree of respiratory distress is dependent on the severity of lung hypoplasia and the development of persistent pulmonary hypertension of the newborn (PPHN).

The most common content of hiatus hernia is the stomach.

It is divided into four types:

Type 1: sliding hiatal hernia (95%)

Type 2: para-esophageal hiatal hernia with the gastro-esophageal junction in a normal position.

Type 3: mixed or compound type, para-esophageal hiatal hernia with displaced gastro-esophageal junction.

Type 4: mixed or compound type hiatal hernia with additional herniation of viscera.

Differential diagnosis
Other thoracic lesions - congenital pulmonary airway malformation, bronchopulmonary foregut malformation, bronchogenic cysts, bronchial atresia, enteric cysts, and teratomas.

Diaphragmatic eventration - Diaphragmatic eventration refers to elevation of a portion of the diaphragm that is intact but thinned because of incomplete muscularization.
Findings and procedure details

Prenatal diagnosis of Bochdalek and Morgagni hernia:

Ultrasound:

Using ultrasonography, congenital diaphragmatic hernia (CDH) may be prenatally diagnosed as early as the second trimester. Ultrasound detects more than 50% of CDH cases at a mean gestational age of 24 weeks.

The definitive sonographic diagnosis of fetal CDH relies on the visualization of abdominal organs in the fetal chest.

Left-sided CDH is characterized by the presence of a heterogeneous lesion (small bowel) in the left chest that often results in right mediastinal shift. Fluid and peristalsis in the heterogeneous mass help to distinguish CDH from other intrathoracic masses. The fluid-filled stomach may be absent from the abdomen. The liver may be herniated, as well, appearing as a homogeneous mass in the chest at the level of the heart and continuous with the intraabdominal liver. Color Doppler ultrasound can be used to document the location of the liver by demonstrating the course of the intrahepatic vessels. The gallbladder and hepatic or umbilical veins may be abnormally located within the abdomen, which can be scaphoid. The abdominal circumference may be smaller than expected for gestational age.

Right-sided CDH is characterized by the presence of a homogeneous mass (liver) in the right chest that often results in left mediastinal shift. Pleural fluid is often present and bowel may herniate with the liver. The left shift of the heart is a key finding since, sonographically, the liver is similar in appearance to fetal lung and bowel does not reliably herniate. As discussed above, color Doppler ultrasound can be used to document the location of the liver. Sometimes the gallbladder can be seen in the chest, which when present is another key finding diagnostic of right-sided CDH.

Right CDH is more frequently missed or misdiagnosed than left CDH because the herniated viscera consist predominantly of the right lobe of the liver, which may have similar echogenicity to the lung or be confused with a solid mass in the chest.

With either left- or right-sided CDH, esophageal compression secondary to mediastinal shift can result in polyhydramnios, which is common. Obstruction of venous return due to mediastinal shift rarely occurs and can result in hydrops fetalis.
Three-dimensional ultrasound imaging, fetal echocardiography and fetal magnetic resonance imaging (MRI) are other prenatal diagnostic modalities used.

**MRI:**

MRI confirm presence of CDH and differentiate CDH from other chest masses. It is superior to ultrasonography in demonstrating position of fetal liver above or below diaphragm. It can clearly depict diaphragmatic discontinuity, fetal compressed lung, and connecting bowel segments between abdomen and chest.

Fetal MRI protocol should include T2WI in all three planes and T1WI breath hold gradient recalled echo images in the coronal plane to assess for liver and meconium positions. Meconium-filled bowel loops are low signal on T2-weighted images and high signal on T1-weighted images, making location of the bowel easy. The liver is high signal on T1-weighted images and intermediate signal on T2-weighted images, also making it easy to separate from bowel and adjacent lung. MRI is most helpful in the evaluation of right and bilateral CDH.

In these cases, with the stomach often located below the diaphragm in the abdomen, it may be difficult to differentiate a CPAM from a CDH by ultrasound. MRI easily distinguishes abdominal contents within the chest from cystic lesions and provides specific information on hernia content, size of diaphragmatic defect, and amount of ipsilateral and contralateral lung.

**Postnatal diagnosis of Bochdalek and Morgagni hernia:**

**Chest X Ray:**

**Bochdalek hernia:** X-Ray initially, hemi thorax appear opaque because loops are fluid-filled, paucity of bowel loops beneath the diaphragm. Once air swallowing begins, multiple cystic lucencies and mediastinum shifted to opposite side. In the case of intraabdominal solid organ herniation such as the liver and spleen, the hemi thorax can remain homogeneously opacified.

Deviation of lines: endotracheal tube, nasogastric tube, umbilical arterial and venous catheters. The nasogastric (NG) tube deviates to the side opposite to the hernia in the chest. If the stomach is herniated within the hemi thorax, the tip of the NG tube can project in the chest. The position of umbilical venous catheters also is affected according to the location of the liver, which is shifted either in the abdomen or chest. In contrast, the position of umbilical arterial catheters is rarely affected because of their retroperitoneal location.
**Morgagni hernias:** Most cases are discovered incidentally on chest radiographs that are obtained for evaluation of other conditions in older children and adults.

When solid organs such as the liver or spleen are involved, the appearance may not be specific and can resemble focal diaphragmatic eventration, lymphadenopathy, or a foregut duplication cyst.

Ultrasound, CT, or MRI can be helpful in the diagnosis when solid organs are herniated.

**Fluoroscopy:** in elective cases, contrast gastrointestinal studies performed to confirm diagnosis and delineate the contents.

**MRI:** has been found to be useful in detecting fetal anomalies and can be a valuable adjunct to evaluate the position of the liver and estimating lung volume. Associated cardiac and neural tube defects may affect the outcome of infants with CDH.

**CT:** for better delineated of position of herniation and help to rule out differential diagnosis like mediastinal masses, bronchogenic cyst, and other congenital lung malformations.

**Postoperative assessment of Bochdalek and Morgagni hernia:**

However, chest radiographs often are obtained routinely for confirmation of an intact diaphragm and early detection of a possible CDH recurrence. On follow-up chest radiographs, abnormalities including persistent lung hypoplasia, decreased pulmonary vascularity, and mediastinal shift may be observed.

An ipsilateral pneumothorax is a common finding and should not be rapidly evacuated. A rapid evacuation of a pneumothorax in this situation may cause mediastinal rotation and subsequent venae obstruction because of the increased mobility of the neonatal mediastinum. The pleural air subsequently reabsorbs by itself and sometimes is replaced by fluid.

**Hiatal hernia:**

Radiographic features:

**Plain radiograph:**
Retro-cardiac opacity with air-fluid level or lucency.

Fluoroscopy

- Numerous coarse thick gastric folds within the supra-hiatal pouch.
- Tortuous esophagus with an eccentric gastro-esophageal junction.

CT:

- Focal fat collection in the middle mediastinum

Omentum herniates through the phrenico-esophageal ligament.

May see an increase in the fat surrounding the distal esophagus.

- Para-esophageal hernia through a widened esophageal hiatus

Visualize contents, size, orientation of herniated stomach within the lower thorax.

Herniated contents lie adjacent to the esophagus.

- Widening of esophageal hiatus

Dehiscence of diaphragmatic crura (>15 mm): increased distance between crura and esophageal wall.
**Fig. 1:** Normal barium swallow

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**Fig. 2:** Combined Congenital Hiatal and small Morgagni Hernias

Radiograph of chest and upper abdomen; frontal (A) and cross table (B) projections show gastric air bubble seen within lower posterior mediastinum (arrows) with nasogastric tube coiled within (arrowhead).

Barium meal (C) and follow through of the same patient show stomach (arrows) is seen within lower posterior mediastinum.
Fig. 3: Combined Congenital Hiatal hernia and small Morgagni Hernias

(A and B) Chest radiograph show cystic lucency (arrow) in relation to the left posterolateral aspect of the diaphragm. (C, D and E) upper GIT study and follow through how herniating gastric fundus through diaphragmatic hiatus into the left hemithorax with associated herniating part of transverse colon anteriorly.

Combined Congenital Hiatal hernia and small Morgagni Hernias

Fig. 4: Combined Congenital Hiatal hernia and small Morgagni Hernias

(F and G) CT chest axial cut shows hiatus hernia and Morgani hernia show widening of the GEJ suggest hiatus hernia of the stomach (red arrow) and multicystic lesion (white arrow) between the left liver lobe and anterior chest wall represent herniating part of transverse colon.

Combined Congenital Hiatal hernia and small Morgagni Hernias
Fig. 5: Congenital Right Sided Diaphragmatic Hernia

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Fig. 6: Hiatus Hernia

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Fig. 7: Hiatus Hernia Type 1

(A) Upper GI study barium swallow showing a sliding hiatus hernia (asterisk). The gastroesophageal junction (arrowheads) is above the diaphragm (arrow) and the gastric folds are above the diaphragm.

(B and C) barium meal for the same patient, contrast through PEG tube shows gastric folds above the diaphragm with no GER.

Hiatus Hernia Type 1

Fig. 8: Hiatus Hernia Type III

(A) Chest radiograph shows right para-cardiac opacity (arrow).

(B, C, and D) Shows stomach (arrows) and GEJ (arrowheads) are seen above the diaphragm (dashed line).

Para-esophageal hernias Type III
Fig. 9: Hiatus Hernia Type III

(A) Chest radiograph shows cystic lucency projecting over the cardiac shadow.
(B and C) Barium meal show that the whole stomach is above the left hemidiaphragm.
(D) CT chest with IV contrast show that the stomach herniating posterior to the heart.

Para-esophageal hernias Type III

Fig. 10: Hiatus Hernia Type IV

(A) Radiograph of chest shows cystic lesion (arrows) overlapping the heart show towards the right side.
(B, C) Upper GIT study barium sallow and follow-through series shows the stomach (arrow) and GEJ (arrowheads) are seen above the diaphragm.
(D) Delayed images shows transvers colon is also herniated to the chest.

Para-esophageal hernias Type IV
Fig. 11: Congenital diaphragmatic hernia of the liver

Fig. 12: Right diaphragmatic hernia containing liver and right kidney
Fig. 13: Congenital diaphragmatic hernia of the bowel

35 years old female patient, pregnant 35 weeks gestation, prenatal US shows abnormal chest to R/O CDH. (A,B,C, and D) Coronal HASTE of fetal MRI shows cardiac dextro-postion (red star). The right hemi-diaphragm is intact (dash line). The right lung appears well inflated with uniform moderately high signal intensity (arrow). The left hemi-diaphragm is deficient and the left side of the chest is occupied by herniated colonic and small bowel loops, stomach and probably spleen (arrowhead). The left lung is compressed.

Morgagni and pericardial hernia

(A and B) Chest radiographs in frontal and lateral projections show cyst lucency in left anterior diaphragmatic region (arrow). (C and D) Barium meal and follow through show normal position of stomach with herniation of part of transverse colon (unfortunately no lateral images in follow-through). Operative findings showed pre-cardial location of the hernia.

Fig. 14: Morgagni and pericardial hernia
Fig. 15: Morgagni hernia

6 months old C/O cough
(A) Frontal and (B) lateral radiograph of the chest show cystic lucency (arrow) in the right paracardiac and retrocardiac regions. In the lateral chest x-ray showed high position of the bowel loop in the right precordial retrosternal space. No follow-up.

Morgagni hernia

Fig. 16: Morgagni hernia

6 months old boy C/O cough and fever
(A) Frontal and (B) lateral radiograph of the chest show cystic lucency (arrow) in the right paracardiac and retro-cardiac regions.
(B) Lateral chest x-ray shows high position of the bowel loop in the right precordial retrosternal space.

Morgagni hernia
Fig. 17: Morgagni hernia

(A) Frontal and (B) lateral radiograph of the chest show cystic lucency (arrow) in the right para-cardiac and retrocardiac regions. In the lateral chest x-ray showed high position of the bowel loop in the right precordial retrosternal space.
Conclusion

Diagnosis of CDH and more importantly the position and contents of the herniation is achieved by different radiological modalities.
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