BILATERAL MORGAGNI HERNIA IN INFANT, A RARE TYPE IN CONGENITAL DIAPHRAGMATIC HERNIA: A CASE SERIES

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Introduction. Congenital diaphragmatic hernia (CDH) is a rare congenital anomaly characterized by a defect in diaphragm development with subsequent herniation of abdominal contents into the chest during fetal life. The two most CDH are posterolateral (Bochdalek) and anteriorly (Morgagni) hernias.1-6 First described by Vincent Bochdalek in 1848, these hernias usually become evident during the neonatal period with signs and symptoms of respiratory failure. In 1769, Giovanni Battista Morgagni first described the substernal herniation of abdominal contents into the thoracic cavity based on observations made during autopsies examinations.3,4

Incidence of CDH varies from 1 in 2200 to 1 in 5000 births. Morgagni hernia is a rare pathologic finding, representing approximately 3% to 5% of diaphragmatic hernias. Its bilateral presentation is an absolute rarity, and its description is based only on isolated case reports. 80%-90% of Bochdalek hernia occur on the left side. Between 5% to 25% of all posterolateral hernias present after the newborn period. Numerous approaches have been described and, particularly the significance of
laboratory has been emphasized as an operative technique.\textsuperscript{1,2,5-8}

The mortality rate of infants born with CDH remains high, despite optimal perinatal care, has been attributed to pulmonary hypoplasia and associated persistent pulmonary hypertension. Newer management strategies such as permissive hypercapnia, high frequency oscillatory ventilation (HFOV), inhaled nitric oxide (NO), extracorporeal membrane oxygenation (ECMO) and delayed surgical repair have emerged in the care of high-risk CDH patients, which offer some hope of improving overall survival.\textsuperscript{1,2,5-6}

We present two case series with congenital diaphragmatic hernia. First case was a symptomatic 4-month-old baby girl with intrathoracic bowel and liver associated with bilateral anterior diaphragmatic defects. Second case was a 22-days neonates girl who presented with respiratory distress, with intrathoracic bowel and spleen, lung hypoplasia in the affected site who suspected as anterior hernia, but found posterolateral diaphragmatic defects intra operatively. Those patient were successfully treated with abdominal approach. We describe this case because of the absolute rarity of bilateral localization in Morgagni hernia and unsuspected posterolateral diaphragmatic defects found intra operatively.

**Case Report**

**Case 1**

A 4-month-old baby girl was referred to our centre with the diagnosis of the bilateral anterior diaphragmatic hernia. Presented with non specific symptoms of failure to thrives, decreased of body weight, feeding difficulties, often vomiting when breast milk, and respiratory tract infection. Vital sign was stable. No associated anomalies was found. Physical examination found scaphoid +abdomen, epigastric, intercostal and suprasternal retraction, with normal breath sounds on the affected side. Chest radiography revealed a multiple fibro infiltrate in the right lung and possible anterior diaphragmatic hernia consist of bowel intrathoracic. In barium enema shows bowel placed in left intrathoracic. Laboratory findings shows leucocytosis 18.460/\textmu l, respiratoric acidosis in blood gas analysis with pH 7.345, PO\textsubscript{2}52.9 mmHg, PCO\textsubscript{2} 48.4 mmHg and oxygen saturation 85.6\%. Patient was admitted to neonatal intensive ward, spontaneous breathing with 3 liters nasal oxygen and bowel decompression with nasogastic-tube for stabilize her condition. During hospitalization, she got fever up to 38.5°C caused by bronchopneumonia and suspicious of sepsis, was given double broad spectrum antibiotics. IT ratio was 0.16 revealed borderline to sepsis. Blood culture result was Acinetobacter spp. Her serial blood gas analysis remain good without any worsening. Also performed brochoscopy with normal result.

After the condition was stable with normal leucocyte 8.580/\textmu l and no fever, patient then scheduled for surgery. Surgery was performed through abdominal approach. Left Sub costochondral insicion was made. During exploration, ileum and transverse colon are heading to cranially. These organs were withdrawn into the abdomen and returned to normal color. Found defect in bilateral anterior diaphragm with teres hepatic ligament between it, size 6x3 cm, consist of ileum, transverse colon and left lobe liver, located medial to the esophageal hiatus. Lateral and posterior parts of the diaphragm were intact. Teres hepatic ligament was excise, due to defect closure, and was closed using Gore-tex patch 6-0 with interrupted suture. Patient then transferred to ICU, the condition improved dramatically. Blood gas analysis shows good results with pH 7.385, PO\textsubscript{2} 80.3 mmHg, PCO\textsubscript{2} 37 mmHg, Oxygen saturation 95.8 %. He was extubated on the 1\textsuperscript{st} postoperative day, and was discharged home on the 10\textsuperscript{th} postoperative day, with the chest radiograph showing good lung expansion.
Figure 1. Case 1(A). Pre Op AP Chest X-Ray. Reveals shifting of the mediastinum to the right, look bowel enema in the left chest and a paucity of gas in the abdomen. (B) Intra Op : Finding ileum, transverse colon and left lobe liver, in the anterior chest were withdrawn into the abdomen. (C) Defect in bilateral anterior part of diaphragm with treitz ligament (arrow) between it, size 6x3 cm, located medial to the esophageal hiatus.

Figure 2. Case 1(A). Intra Op : Shows the defect already closed by gore-texpacth 6-0. (B) Post Op Pleuroscopy; Finding both lung were in good expansion, no mediastinal shift and air-filled loops bowel in the chest. (C) Post op clinical picture, shows good wound healing and no scaphoid abdomen.

Case 2
A 22-days neonates girl was came to our center with the diagnosis of the left posterolateral diaphragmatic hernia. Presented with a sign of respiratory distress, shortness of breath, feeding difficulties, and vomiting when breast milk since one week ago. Vital sign was stable, except increasing of respiratory rate of 80 times per minutes. No associated anomalies was found. Physical examination found scapoid abdomen, epigastric, inter costal and suprasternal retraction, with decreased breath sound on the affected side. Lateral view on chest radiography demonstrating a sign of anterior diaphragmatic hernia with air-filled loops of the bowel in the chest, dextocardia. Laboratory findings shows leucocytosis 11.060/µl, severe respiratoric acidosis in blood gas analysis with pH 7.285, PO₂41.9 mmHg, PCO₂54.0 mmHg and oxygen saturation 69.7 %. Patient was admitted to neonatal intensive ward, spontaneous breathing with 5 liters nasal oxygen and bowel decompression with naso-gastric-tube for stabilize her condition. During hospitalization, she got stable condition, was given double broad spectrum antibiotics to prevent sepsis. Her serial blood gas analysis remain good without any worsening. Also performed brochoscopy with edematous in left main stem bronchus.
Patient then scheduled for surgery. Surgery also performed through abdominal approach. Left Sub costochondral incision was made. During exploration, ileum, jejunum, transversal colon are heading to posteriorly. These organs were withdrawn into the abdomen and returned to normal color. Found defect in left posterolateral diaphragm with size 3x3 cm. Hernial sac content of ileum, jejunum, transversal colon, and spleen. Anterior part of the diaphragm were intact. Also there is lung hypoplasia in the affected site, measurement of intra abdominal pressure was 8 cmH$_2$O. Defect was closed using Gore-texpacth 6-0 with interrupted suture.

![Figure 3. Case 2(A). Pre Op AP Chest X-Ray. Reveals shifting of the mediastinum to the right, air-filled loops bowel in the left chest and a paucity of gas in the abdomen and dextrocardia (B) Lateral Chest X-Ray; shows numerous cystic and tubular lucencies filling the anterior left chest consisted with herniated bowel.(C) Intra Op Finding: ileum, transverse colon in the left posterolateral chest were withdrawn into the abdomen.](image)

![Figure 4. Case 2(A). Intra Op Finding :Spleen also herniated in the left posterolateral chest. (B) Defect in the left posterolateral of diaphragm size 3x3 cm.(C) Shows the defect in already closed by gore-texpacth6-0.](image)

Patient then transferred to ICU, the condition improved dramatically. Blood gas analysis shows good results and no respiratoric acidosis with pH 7.364, PO$_2$ 101.4 mmHg, PCO$_2$ 42.9 mmHg, Oxygen saturation 97.4 %. He was extubated on the 1$^{st}$ postoperative day, and was discharged home on the 8$^{th}$ postoperative day, with the chest radiograph showing good lung expansion. Postoperatively, both of patient enjoyed an uneventful course and was discharged home without any further events.

**Discussion**

*Congenital diaphragmatic hernia (CDH)* is an abnormal opening in the diaphragm, characterized by a defect in diaphragm
development with subsequent herniation of abdominal contents into the chest during fetal life. Still remains a challenging problem for both surgeons and neonatologists. Improved knowledge in pathophysiology mechanisms explaining the respiratory failure has led to changes in the management of newborn infants with CDH. The mortality rate of infants born with CDH first 12 hours of life remains high, although survival can be as close as 90% at advanced perinatal centers. The primary cause of mortality is refractory pulmonary hypertension because of pulmonary hypoplasia with reduced alveolar surface area, reduced pulmonary perfusion, persistent shunting from right to left, hypoxia and surfactant deficiency.

Newer CDH management strategies are “gentle ventilation” (preservation of spontaneous respiration, permissive hypercapnia and avoidance of high ventilation pressure) HFOV, ECMO and delayed surgical repair to stabilize it for at least 72 hours, which offer some hope of improving overall survival.

There are several types of congenital diaphragmatic hernia, which includes Bochdalek, Morgagni and Central (septum transversum) diaphragmatic hernia. Embryologically the diaphragm has four components. (1) Septum transversum (central tendon) developed from the anterior thorax, (2) Pleuroperitoneal membranes developed from the posterior thorax, (3) Dorsal mesentry of the esophagus, (4) Muscular components developed from the body wall

Diaphragmatic hernias due to embryological deficiencies of the diaphragm are usually of three main types. (1) Those through the pleuro peritoneal hiatus, (2) Those due to the lack of formation of the posterior portion of the diaphragm (3) Those through the foramen of Morgagni (Larrey’s spaces).

Described by Bochdalek in 1848, CDH usually become evident during the neonatal period with signs and symptoms of respiratory failure in first hours or day of life. Bochdalek hernia account for 85% -90% of all diaphragmatic defects; 80%-90% of these defects occur on the left side. Between 5% and 25% of all posterolateral hernias present after the newborn period. Bochdalek hernia results from incomplete closure of the normal pleuro peritoneal canal during fetal development. Our second case was neonatal, who presented with respiratory distress and suspected as anterior hernia, but intraoperative found that defect was left posterolateral. This misleading was caused by preoperative chest radiograph, the bowel was placed anteriorly in the chest.

In 1769, Morgagni first described the substernal herniation of abdominal contents into the thoracic cavity, is a rare case, less than 5% of all types of CDH. Bilateral Morgagni it may rarely be found and its description is based only on isolated case reports. Two different theories exist about its origin: congenital vs acquired. A symptomatic in most cases, but it can also manifest with abdominal or thoracic symptoms and nonspecific presentations, contributes to the delay in diagnosis. Our first case also present with bilateral Morgagni hernia and non symptomatic symptoms of failure to thrive and respiratory tract infection. Caused when normal development of the diaphragm and of the digestive tract do not occur; there is an improper fusion of structures during fetal development. Diagnosis is based on findings from conventional radiography and computed tomography.

The majority of hernias (about 85%) are left sided and they may contain any or all of the following: stomach, small bowel, colon, liver, spleen, kidney and supra adrenal gland. The size of the defect varies from small (2 or 3 cm) to very large, involving most of the hemidiaphragm. In our case, the Morgagni defect was bilateral and consist of bowel, colon and left lobe liver, but in Bochdalek it was the same as the literature said.

Table 1. Shows Bochdalek and Morgagni hernia symptoms in infant
Extra diaphragmatic anomalies such as congenital heart defects, neural tube defects, genitor urinary, skeletal, and cranio facial defect are present in 25% to 57% of all cases of CDH. Position of the liver in the chest, polyhydramnions, fetal abdominal circumference < 5th 0/00, size of contra lateral lung measured as a ratio of lung area to head circumference, lung volume, Mc Goon Index < 1.31, Pulmonary Artery Index (PAI) < 90 and associated anomaly have been made to identify prognostic factors based on prenatal ultrasound findings in infant with CDH.

Ultrasound allows prenatal diagnosis of CDH in the first trimester. For left CDH mediastinal shift and rightwards displacement of the heart can be seen, a fluid-filled stomach or bowels are later on present within the thoracic cavity. An important feature to look for is presence of (a portion of) the liver in the thorax. With right CDH, the right lobe of the liver usually herniates into the chest, combined with mediastinal shift to the left. Postnatal diagnosis made by symptoms which are cyanosis, tachypnoea and grunting respirations within minutes or hours after birth. Physical examination reveals a scaphoid abdomen, an increased anteroposterior diameter of the thorax and mediastinal shift. Breath sounds are absent on the affected side. Chest radiography and abdomen by demonstration of air-filled loops of the bowel in the chest and a paucity of gas in the abdomen.

Both our patient were infant and neonates. Found scaphoid abdomen, epigastric, intercostal and suprasternal retraction, with decreased breath sounds on the affected side. Chest radiography shows hernia with air-filled loops of the bowel in the chest. No history of polyhydramnios in mother pregnancies. Age, symptoms and radiographic findings were the same as in the literature mentioned above, except for their mother history of pregnancies, we have got no data about it.

### Management Strategies

The antenatal and postnatal management of patients with CDH is still evolving. Given the wide variations in the management, it is not surprising that there are no predictors that are reliable. After the newborn infant with CDH is transferred to a center with ECMO capabilities, an effort is made to stabilize it for at least 72 hours before operative repair is considered. To achieve this goal, a new management strategy for antenatal diagnosed CDH including:

<table>
<thead>
<tr>
<th>Bochdalek</th>
<th>Morgagni</th>
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<tr>
<td>- Respiratory distress → Life threatening</td>
<td></td>
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<td>- Feeding intolerance.</td>
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<td>- Some patients have no symptoms.</td>
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<td>- Older Infant→GI symptoms</td>
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<td>- 1/3 patients asymptomatic.</td>
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<td>- Most frequent : cramping pain, constipation from partial colonic obstruction.</td>
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<td>- Rare case : Gastric volvulus or GI obstruction/incarceration/strangulation</td>
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<td>- Cardiorespiratory symptoms &lt; Gastro-Intestinal symptoms.</td>
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<td>- Trauma, exercise and pregnancy can cause the occurrence.</td>
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(1) Delivery of a “depressed baby” (no breathing and swallowing) by c-section;  
(2) Exclusive use of HFO in the NICU prior to surgery;  
(3) Delayed surgery following short-term stabilization

### Prenatal Management

Advances in prenatal assessment, lung-sparing ventilation, and extracorporeal interventions have improved survival and decreased morbidity associated with CDH. Once a
Prenatal diagnosis of CDH was confirmed by ultrasound and the parents expressed their wish to continue their pregnancy. They will received genetic counseling including fetal karyotyping, and repeated ultrasound examination to determine the following:\textsuperscript{30,32}

| (1) Lung-thorax transverse area ratio (LT ratio), which is calculated by a simple ratio of right and left lung area to thorax area in a cardiac four-chamber view, to assess the severity of the pulmonary hypoplasia; | (2) Presence of congenital abnormalities, particularly those affecting the cardiovascular system, the central nervous system, and airway structures. |

Serial measurements of LT ratio are also important because it changes with the progress of pregnancy. LT ratio $< 0.25$ indicates that the fetus has a severely hypoplastic lung and requires close attention to determine the timing of delivery.\textsuperscript{30}

Despite regular progress in NICU, CDH diagnosed antenatal is still associated with up to 80\% mortality. It is impossible to predict which fetus with CDH will survive or not. Antenatal parameters included: gestational age at diagnosis, herniated organs, associated malformations and presence of polyhydramnios and LV hypoplasia.\textsuperscript{33}

Management during delivery

Antenatal diagnosis of CDH has not been thought to mandate the necessity for cesarean section for the delivery of CDH babies. Although they are routinely intubated on birth in the delivery room and are immediately transferred to the NICU, some neonates have already developed pneumothorax and/or intestinal distention on admission to the NICU. These complications are postulated to be attributed to the struggle or gasp during the initial resuscitation, bucking after intubation, and inadvertent high peak inspiratory pressure during transport. Since the neonates with CDH have pulmonary hypoplasia and a fixed number of alveoli, a single pneumothorax requiring a chest tube or mediastinal shift due to intestinal distension compressing the contra-lateral lung will result in a lung that is less capable of performing its function than it was prior to injury.\textsuperscript{30}

Postnatal management

Regardless of the clinical presentation, HFOV was initiated on admission to the NICU, and was exclusively used before surgery. The patient received extensive medical therapies to achieve preoperative stabilization, which included; infusion of sedatives and muscle relaxants; inotropic support and volume infusion to maintain stable circulation; administration of pulmonary vasodilators such as prostaglandin E$_1$, or inhaled nitric oxide; administration of surfactant to improve oxygenation. Preoperative stabilization was defined by the following criteria:\textsuperscript{30}

| (1) Normal hemodynamic variables with minimal circulatory support, | (2) No signs of persistent pulmonary hypertension (PPHN) evidenced by no difference in pre-/post-ductal oxygen saturation; |
| (3) Adequate oxygenation and ventilation by minimal setting of HFOV ($F_{O_2} < 0.3$, mean airway pressure $< 8$ cmH$_2$O, peak to peak pressure level $< 50$ cmH$_2$O). | |

Surgical correction was attempted only when the preoperative stabilization was achieved. According to the literature said, what occurred in both our cases also shows that they have respiratoric acidosis in blood gas analysis at the first time they arrived in our center, not doing emergency surgery, but stabilize them at neonatal intensive ward. During hospitalization, first case got fever up to 38.5$^\circ$C due to bronchopneumonia and suspicious of sepsis, was given double broad spectrum antibiotics.

Timing of Surgery and Surgical Approach

Delayed surgical repair is now widely placed for at least 72 hours before operative repair is considered.\textsuperscript{19} Most institutions perform surgery in NICU when the patient was stable. The correction of CDH can be performed abdominal through paramedian or subcostal insicion or thoracic approach through thoracotomy. Also can be performed with laparoscopy or thoracoscopy.\textsuperscript{11,21,33} The significance of laparatomy has been emphasized as an operative technique. This allows easy reduction and inspection of contents, allows access and repair of bilateral hernias, and corrects an associated malrotation
if present. When diaphragmatic tissue is adequate, primary repair with non absorbable suture can be done. But if too large, prosthetic material can be used for a tension-free repair. Implantation of foreign material can expose patients to the risks of infection, displacement, or erosion into adjacent structures.16,34

Both our patient also performed abdominal approach through subcostal incision. In first case it was bilateral Morgagni hernia, abdominal incision was the best approach, especially for bilateral hernias, it allows easier reduction of the hernia. And in second case because it was unsuspected posterolateral defect, the defect can be seen through abdominal incision. We didn’t performed ECMO for both our patient, because no indication was attempted, patients were stable with spontaneous breathing, despite that, in our center don’t have ECMO facilities if those infant need that treatment.

Abdominal compartment syndrome as a complication after CDH repair has not been reported. These conditions, are associated with insufficient room in the abdominal cavity (ie, loss of domain) to accommodate all of the organs without elevation of intraabdominal pressure. Defined as intraabdominal pressure > 25 to 30 mmHg. If pressure is high, the abdomen may be closed with a constructed silo of prosthetic material, patch, or VAC system. With fluid shifts and abdominal wall stretching for several days, the abdomen can be closed in the operating room. Abdominal compartment syndrome didn’t found in both our patient after abdominal closure.

Because of degree of pulmonary hypoplasia determines survival in infant with CDH, another strategy has developed to reverse the pulmonary hypoplasia. Open fetal surgery which called tracheal occlusion (TO) leads to increased levels of lung tissue stretch, triggers lung growth, and reverses pulmonary hypoplasia before birth. Timing and duration of the occlusion period are crucial for the quality and response. Oral sildenafil also can be give to infant with CDH in appear to reduce pulmonary vascular resistance as measured by functional echocardiography.15,19,35,36

The most common surgical outcomes in infants with CDH include diaphragmatic hernia recurrence, chest hernia incarceration, gastro esophageal reflux disease, midgut small bowel obstruction, volvulus, chest deformity, wall and spinal deformities.37,38

Conclusion
Bilateral Morgagni hernia is a rare type congenital anomaly, Nonspecific presentations, contributes to the delay in diagnosis, and preoperative chest radiography can be possible misleading for diagnosis in Morgagni hernia. Numerous approaches have been described and, particularly the significance of laparatomy has been emphasized as an operative technique. This allows easy reduction and inspection of contents, allows access and repair of bilateral hernias, and corrects an associated malrotation if present

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