Congenital Diaphragmatic Hernia with Massive Abdominal Contents

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Abstract

Introduction: Congenital diaphragmatic hernia (CDH) is a congenital defect that occurs in about 1: 3000 to 5000 live births leading to communication between the abdominal and thoracic cavities during critical stages of development. The displacement of abdominal contents into the thoracic space during this time can cause pulmonary hypoplasia due to the mass effect of abdominal contents. The lung not only suffers from hypoplasia, but also lacks a normal vascular and bronchial branching pattern. This abnormal development leads to increased pulmonary vascular resistance and subsequent significant pulmonary hypertension. These two pathological conditions are the main source of morbidity and mortality in patients with CDH.

CDH can occur either on the right side of the diaphragm, on the left side or even in the least of cases may be bilateral. Within the most frequent hernias are left-sided and there are two main types: Morgani (anteromedial) and Bochdalek (posterolateral). Bochdalek hernias occur much more often and are usually associated with physiological disorders in the newborn.

While open surgery (thoracotomy or laparotomy) has traditionally been performed for patients with CDH, minimally invasive surgery (laparoscopy or thoracoscopy) has emerged as a safe and feasible alternative. Each technique has advantages and disadvantages. Minimally invasive surgery has a shorter duration of postoperative ventilation, less pain and short-term use of narcotics, faster recovery, shorter hospitalization, and less morbidity, such as thoracic wall deformities and small bowel obstruction unlike open surgery.

Purpose: Presentation of a clinical case at the Pediatric Hospital of Peralvillo, of the Health Secretariat in Mexico City, on a case of congenital diaphragmatic hernia with massive abdominal contents to the surgical community.

Results: We present a clinical case of a male patient with 35 weeks of pregnancy who experiences respiratory distress, having at physical examination, absence of respiratory noise in left hemithorax, heart noises predominantly in right hemithorax, as well as concave abdomen without auscultating peristaltic noises, is taken a chest X-ray, in which is evidenced a diaphragmatic hernia, as well as displacement of the cardiac silhouette to the right, and abdomen with ground glass image, so surgical management is decided, having as a finding a hernial defect in the posterolateral left face of the diaphragm with massive abdominal content, diaphragmatic plasty with tension is performed. Post surgery, its postoperative period is carried out in the neonatal ICU where gasometry is taken in which severe respiratory acidosis is reported, requiring invasive mechanical ventilation with increase in ventilatory parameters, use of aminergics and life support with drug products, with death 15 hours after surgery.

Discussion: Congenital diaphragmatic hernia (CDH) is a condition in which partial or complete agenesis of the diaphragm occurs, resulting in continuity between the chest and abdominal cavities, usually more common in males, occurs in about 1: 3,000 to 5000 live births. Left side CDH is more common than on the right side. The posterolateral left side of the diaphragm is the most common localization in 75 to 90% of cases (Bochdalek’s hernia) and 10% occurs in the anteromedial portion (Morgani’s hernia); the defect on the right side occurs in 10 to 15%, it can even be bilateral in 1 to 2% of cases. The prevalence of CDH does not appear to be associated with maternal age.

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The pathogenesis of CDH is not fully understood, however, the most accepted theory is failure in the closure of the pleuroperitoneal membrane or environmental factors altering the differentiation of mesenchymal cells during the formation of the diaphragm and other somatic structures.

Prenatal diagnosis of CDH is based on ultrasound. Usually, diagnosis is based on the presence of a mediastinal displacement and a fluid-filled stomach next to or just behind the heart. In some cases, the fetal liver may herniate into the chest and appear as an intrathoracic homogeneous mass at heart level. The defect in the right hemidiaphragm is more difficult to diagnose since on ultrasound the liver is similar in appearance to the fetal lung.

Patients with CDH management before surgery may take several different forms, but all maintain several basic principles: permissive hypercapnia (PaCO₂, 45 - 60 mm Hg), maintaining oxygen saturation between 85% - 95%, minimizing the volutrauma index and barotrauma with conventional ventilatory strategies aimed at maintaining a positive inspiratory pressure less than 25 cm H₂O with a positive pressure at the end of expiration between 2 and 5 cm H₂O. The addition of inhaled nitric oxide can further decompress the pulmonary vascular bed. If these goals cannot be achieved with maximum medical therapy, extracorporeal membrane oxygenation (ECMO) is required to deliver oxygen and eliminate CO₂. However, in terms of hernia repair surgically, it has several modalities, with an open or minimally invasive technique, with or without mesh and approached from the abdomen or chest.

Conclusion: Congenital diaphragmatic hernia is a rare entity, whose clinical presentation varies from mild symptoms to incompatible with life, depending on the degree of pulmonary hypoplasia and concomitant diseases. One of the main problems related to anesthetic management of these patients is ventilation. According to the severity of symptoms, it is used from pressure-controlled respiratory assistance to high-frequency oscillatory respiratory assistance and extracorporeal membrane oxygenation. Congenital diaphragmatic hernia remains a challenging condition with a little-known etiology and pathogenesis.

Keywords: Hernia; Congenital; Massive; Viscera; Bochdalek; Morgani; ECMO; FETO

Introduction

Congenital diaphragmatic hernia (CDH) is a congenital defect that occurs in about 1: 3000 to 5000 live births leading to communication between the abdominal and thoracic cavities during critical stages of development. The displacement of abdominal contents into the thoracic space during this time can cause pulmonary hypoplasia due to the mass effect of abdominal contents. The lung not only suffers from hypoplasia, but also lacks a normal vascular and bronchial branching pattern. This abnormal development leads to increased pulmonary vascular resistance and subsequent significant pulmonary hypertension. These two pathological conditions are the main source of morbidity and mortality in patients with CDH.

CDH can occur either on the right side of the diaphragm, on the left side or even in the least of cases it can be bilateral. Among the most frequent hernias are the left-sided hernias and there are two main types: Morgagni (anteromedial) and Bochdalek (posterolateral). Bochdalek hernias occur much more often and are usually associated with physiological disorders in the newborn.

While open surgery (thoracotomy or laparotomy) has traditionally been performed for patients with CDH, minimally invasive surgery (laparoscopy or thoracoscopy) has emerged as a safe and feasible alternative. Each technique has advantages and disadvantages. Minimally invasive surgery has a shorter duration of postoperative ventilation, less pain and short-term use of narcotics, faster recovery, shorter hospitalization, and less morbidity, such as thoracic wall deformities and small bowel obstruction unlike open surgery.

Complications resulting from thoracoscopy repair of CDH are divided into early and late, among early complications the most frequent are pneumothorax and hydrothorax which may require a chest tube as treatment for drainage.
Thoracoscopy aims to centralize the mediastinum, improve venous return to the heart and maintain systemic perfusion. Occasionally, an abdominal compartment syndrome may result from the relocation of the viscera in the abdominal cavity as result of abdominal domain loss, so the ideal treatment would be a decompressive laparotomy.

**Purpose of the Study**

Presentation of a clinical case at the Pediatric Hospital of Peralvillo, of the Health Secretariat in Mexico City, on a case of congenital diaphragmatic hernia with massive abdominal contents to the surgical community.

**Case Report**

This is a male patient with 24 hours since birth, premature, 35 weeks of pregnancy, with ABGAR rating of 8 at birth and 7 at 10 minutes. Physical examination is observed normocephalous skull, without endostosis or exostosis, adequate pupillary response, permeable nostrils, absent respiratory noises in left hemithorax, however, peristaltic noises are observed in said hemithorax, rhythmic cardiac noises with good tone and intensity of predominance in the right hemithorax, concave abdomen, peristaltic noises are not observed in abdomen, genitals according to age and sex, permeable anus, integral and symmetrical limbs without alterations, with respiratory difficulty data, therefore, umbilical gases are taken, which reports: pH 7.15, pCO₂ of 42 mm Hg, PO₂ of 6.6 mm Hg, HCO₃ of 18.3 mEq/liter, O₂ saturation of 61% and excess base of -8 mEq/l. He is then sent to the neonatal ICU where advanced airway management is given. New gases control is taken post intubation, finding: pH 7.36, pCO₂ of 40.2 mm Hg, PO₂ of 30.1 mm Hg, HCO₃ of 19.7 mEq/l, O₂ saturation of 69.2% and excess base of -6.8 mEq/l. Likewise, hematic biometry is taken which reports: leukocytes of 8,000; 36.6% neutrophils, 52.5% lymphocytes, 38.7% hematocrit, 13.5 g/dl hemoglobin and 218,000 platelets. Simple thoracoabdominal X-ray is taken in which intestinal loops in left hemithorax are seen, as well as deviated cardiac silhouette to the right, and abdomen with ground glass image (Figure 1), showing a diaphragmatic hernia so its surgical intervention is decided, following clinical and metabolic stabilization of the patient.

![Figure 1: Chest X-ray, where abdominal contents are seen in the thoracic cavity.](image)

Once the patient is stable, it is decided admission to the OR, the patient is placed in the supine position and under general anesthesia an incision is made on the supraumbilical midline with scalpel, dissected by planes until reaching the abdominal cavity, observing it "empty" as well as hernial defect in the posterolateral face of the left hemidiaphragm of approximately 8 x 6 cm with contents of the small intestine, colon, appendix, stomach, spleen and left lobe portion of the liver (Figure 2). Likewise, left pulmonary hypoplasia of about 90% is evidenced. Performing diaphragmatic plasty without mesh (Figure 3).
Post-surgery, its postoperative period is carried out in the neonatal ICU where new gas test is taken in which severe respiratory aci-
dosis is reported, with increased ventilatory parameters, use of aminergics and life support with drug products, with death occurring 15
hours post-surgery.

Discussion

Congenital diaphragmatic hernia (CDH) is a condition in which partial or complete agenesis of the diaphragm occurs, resulting in
continuity between the chest and abdominal cavities, usually more common in males, occurs in about 1: 3,000 to 5000 live births [1].
Left-side CDH is more common than on the right side, with a ratio of 6:1 [2]. The posterolateral left side of the diaphragm is the most
common location in 75 to 90% of cases (Bochdaleck's hernia) and 10% occurs in the anteromedial portion (Morgani's hernia); the defect
in the right hemidiaphragm occurs in 10 to 15%, may even be bilateral in 1 to 2% of cases. The prevalence of CDH does not appear to be
associated with maternal age [3].
Although CDH pathogenesis is not fully understood, the most widely accepted theory is failure in the closure of the pleuroperitoneal membrane or environmental factors altering mesenchymal cells differentiation during diaphragm and other somatic structures formation [4].

Multiple authors consider it is secondary to defective retinoid signaling in the uterus usually occurring before 10 gestational weeks, secondary to vitamin A deficit. As fetal development continues, the diaphragmatic defect allows herniation of abdominal viscera into the chest. Abdominal viscera in the chest cavity generate an increase in intrathoracic pressure, inhibiting normal lung development and disrupting normal respiratory movements of the fetus. The most important physiological and developmental disorders are hypoplasia and pulmonary hypertension [5]. CDH has long been recognized as a disease characterized by a broad spectrum of severity. Major or minor degrees of hypoplasia and pulmonary hypertension, the contribution of additional abnormalities and particular anatomy of the diaphragm defect, along with general cardiopulmonary function, combine to result in a highly variable clinical phenotype. Gestational changes, the transition of fetal circulation and iatrogenic factors after birth can greatly affect the clinical condition of the patient from one moment to another [6].

Bochdalek was the first to predict that children with CDH would benefit from surgical correction. In 1902, Heidenham performed the first successful surgery on a baby. It was in 1940 that surgery became an accepted treatment for CDH when Ladd and Gross reported 9 out of 16 survivors after surgery. After this, in 1946, Gross reported on the first successful repair in a newborn under 24 hours of birth and early repair became the basis of treatment [7].

It is assumed that congenital diaphragmatic hernia usually occurs as an isolated finding, but in 10 - 30% of cases there may be an association with chromosomal defects, the most frequent being trisomies 13 and 18. Structural defects are found in 25 - 57% of all cases of CDH and include congenital heart defects, kidney, brain and gastrointestinal abnormalities [8].

Prenatal diagnosis of CDH is based on ultrasound. In the vast majority of cases it is detected during routine examination, so the mean gestational age at the time of diagnosis is about 22 to 24 weeks [9]. The diagnosis is usually based on the presence of a mediastinal displacement and a fluid-filled stomach next to or just behind the heart. In some cases, the fetal liver may herniate into the chest and appear as an intrathoracic homogeneous mass at heart level. The defect in the right hemidiaphragm is more difficult to diagnose since on ultrasound the liver is similar in appearance to the fetal lung. Doppler ultrasound can be useful to determine liver position by viewing the venous duct and the course of intrahepatic vessels [10].

Magnetic resonance imaging (MRI) allows easy evaluation of hepatic hernia. Unlike ultrasound, MRI is not limited by maternal obesity or oligohydramnios and provides better soft tissues contrast.

Affected newborns have respiratory distress at birth or in the first hours of life, may have cyanosis that in mild cases may occur in the perioral region, palms or feet soles, or in severe cases it may be generalized cyanosis. There is a decrease in air intake on the affected side with the presence of audible intestinal noises and a flat or concave abdomen may be evidence of intestinal loops displaced towards the chest.

In radiographic data can be observed, intestinal loops in the chest; with shortage of loops in the abdomen, mediastinal displacement. With CDH on the right side, a radiopaque space occupying a solid shade representing the liver replaces lung tissue in the lower part of the chest [2].

The current clinical strategy to promote lung growth in severe cases is percutaneous fetoscopic endoluminal tracheal occlusion (FETO) [11]. FETO is performed under endoscopic guidance and local anesthesia. A thin flexible cannula is inserted avoiding the placenta and pointing towards or above the tip of the fetus’s nose. A 1.3 mm fetoscope is moved inside a slightly curved 3.3 mm cannula towards the...
trachea until the carina is displayed. A detachable balloon is placed between the vocal cords and the carina with a specifically designed catheter. The balloon can be visualized on ultrasound as an anechoic structure. Mean duration of FETO is 10 minutes; longer operating times are associated with a higher rate of premature membrane rupture (PMR) [12].

FETO increased survival from 24% to 49% in patients with Bochdalek’s hernia and appeared to improve early respiratory morbidity [13,14], while in right-sided CDH FETO increased survival from 17% to 42% [15]. In survivors, fetal tracheomegaly has been documented, but it does not appear to have a clinical impact other than crup, which decreases over time [16,17]. Adverse secondary tracheal side effects of FETO are rare and usually occur in case of a very early occlusion and at the time of emergency removal of the balloon [18].

CDH patient management prior to surgery may take several different forms in terms of repair, but all maintain several basic principles: permissive hypercapnia (target range of PaCO₂ 45 - 60 mm Hg), maintenance of preductal oxygen saturation between 85% and 95%, minimize volutrauma and barotrauma with conventional ventilation strategies aimed at maintaining a positive inspiratory pressure of less than 25 cm H₂O with a positive pressure at the end of expiration between 2 and 5 cm H₂O. The addition of inhaled nitric oxide can further decompress the pulmonary vascular bed [19]. If these goals cannot be achieved with maximum medical therapy, extracorporeal membrane oxygenation (ECMO) is required to deliver oxygen and eliminate CO₂. Common ECMO indications include an oxygenation index > 40, increased lactate despite ventilation, inotropic support, hypoxemia and associated severe hypercapnia [20]. 4 weeks duration has been discussed [21].

Historically, a time limit has been suggested for the duration of ECMO execution in patients with CDH between 2 and 4 weeks. At present, it has not been proven that prolonged need for ECMO is associated with increased morbidity and mortality [22]. However, establishing the ideal duration of ECMO in patients with CDH is complicated. In patients with severe CDH, ECMO time may be more than 4 weeks, with satisfactory pulmonary results possible [23].

Surgical repair during ECMO is associated with an increased bleeding incidence [24]. One study, although it did not deter ECMO repair, found that survival rate is higher if surgery is delayed until ECMO is stopped [25]. Therefore, the repair has traditionally been performed after ECMO decannulation. However, complications of post-ECMO bleeding have been significantly reduced with anticoagulant and tranexamic acid treatment during the perioperative period [26]. Among the possible intestinal complications of attempting to delay surgery is ischemia or volvulus [27]. Theoretically, ECMO repair can improve respiratory function by restoring a more normal anatomy. Some reports suggest that early ECMO repair may decrease complications and tend towards better survival [28].

The optimal time of surgery for patients on ECMO is difficult to establish. There seems to be a developing consensus that repair at an early stage (within 2 weeks) may help to get out of ECMO or make decisions about withdrawal later and potentially improve the outcome.

While open surgery (thoracotomy or laparotomy) has traditionally been performed for patients with CDH, minimally invasive surgery (laparoscopy or thoracoscopy) has emerged as a safe and feasible alternative. Each technique has advantages and disadvantages. Minimally invasive surgery has a shorter duration of postoperative ventilation, less pain and short-term use of narcotics, faster recovery, shorter hospitalization, and less morbidity, such as thoracic wall deformities and small bowel obstruction unlike open surgery.

However, CO₂ insufflation is used both during laparoscopy and thoracoscopy, for the repair of CDH and careful handling of the additional CO₂ load during the transoperative period is crucial [29]. CO₂ absorbed during chest insufflation and the creation of a capnothorax can cause significant acidosis [30,31]. Alteration of respiratory capacity imposed by pulmonary collapse has important implications for CO₂ oxygenation and excretion and can lead to a marked increase in CO₂ at the end of expiration. CO₂ increase during thoracoscopy is generally higher than that observed during laparoscopy, and higher in younger children undergoing pulmonary ventilation [32].

Most studies have confirmed a significant increase in CO₂ and/or acidosis excretion during thoroscopic repair of CDH during transoperative period [33]. Another situation occurring during thoracoscopy is a higher rate of recurrence with 8% compared to open surgery.

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(2.7%). This incidence increases even more in those cases where mesh is used during minimally invasive surgery with an incidence of up to 8.8% [34].

Complications resulting from thoracoscopy repair of CDH are divided into early and late, among early complications the most frequent are pneumothorax and hydrothorax which may require a chest tube as treatment for drainage.

This procedure centralizes the mediastinum, improves venous return to the heart and maintains systemic perfusion. Occasionally, an abdominal compartment syndrome may result from the relocation of the viscera in the abdominal cavity as a result of an abdominal domain, so the ideal treatment would be a decompressive laparotomy [19].

The most noticeable late complication is recurrent hernia. Signs of recurrence may include dyspnea, tachypnea, and poor diet; however, some recurrences are hidden and only occur on the chest X-ray. Recurrences can be addressed with laparoscopy, thoracoscopy, laparotomy, thoracotomy or a hybrid approach [35]. A key to recurrence management is to provide tension free repair, often this means the use of a prosthetic mesh to close the gap in muscle coverage.

A postoperative chest X-ray is done to check the diaphragm position. Mediastinal changes may occur in the postoperative period, as pressure and volume changes occur due to reduced abdominal contents. Mediastinal displacement caused by CDH does not usually return to the center position immediately after surgery, but does so slowly. The space left by the return of the intestine is initially filled with air, and then with liquid.

Early postoperative care of patients with CDH is supportive and aims to disconnect the ventilator and control pulmonary hypertension. Diet begins with special attention to problems related to gastroesophageal reflux disease. Patients are monitored every 3 months on an outpatient basis during the first year with simultaneous chest X-rays to assess recurrence. Most recurrences occur during the first 12 months after repair; probably related to the dramatic growth experienced by babies in the first year of life. During the second year of the postoperative period, visits are spaced every 6 months, again with chest X-rays. Thereafter, patients are seen annually with simultaneous chest X-rays [19].

Conclusion

Congenital diaphragmatic hernia is a rare entity, whose clinical presentation varies from mild symptoms to incompatible with life, depending on the degree of pulmonary hypoplasia and concomitant diseases. There are various treatment schemes depending on the time of diagnosis, depending on the severity of the symptoms, it is used from pressure-controlled respiratory assistance to high frequency oscillatory respiratory assistance and extracorporeal membrane oxygenation, as well as the definitive management that is surgical either by a minimal technique invasion or open technique.

The thoracoscopic approach of CDH is currently the most used and best prognostic technique, but it must be performed by people with sufficient experience in minimally invasive surgery; however, in our environment there are not enough supplies so the open technique is still performed equally with results satisfactory in most cases. Congenital diaphragmatic hernia remains a challenging condition with a little-known etiology and pathogenesis, hence, the importance of presenting this case to the medical community.

Bibliography


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