ABSTRACT

With a prevalence of 0.3-0.5/1000 births, congenital diaphragmatic hernia (CDH) remains a serious, poorly understood abnormality with a high mortality rate that cannot always be effectively managed. Its reported frequency in Spain is 0.69‰ with a yearly decreasing trend of 0.10‰ during the period 1980-2006. Up to 5% of cases are incidentally identified in adults undergoing studies for other reasons.

We report the case of a 74-year-old woman with vomiting for three months due to parasternal diaphragmatic hernia of Morgagni-Larrey (retrochondrosternal, retrocostoxyphoid, retrosternal, subcostal, substernal, or substernal hernia), which allowed us to report an update on this condition in the adult, and on thoracoabdominal diaphragm morphogenesis. It is in the embryology of the diaphragm where an explanation may be found for some morphological changes and clinical manifestations, even though a number of uncertainties remain. We also analyze the extent of controversy persisting on some aspects of surgical treatment (access routes, mesh use, hernia sac reduction). Overall, minimally invasive techniques predominate. We consider laparoscopy the approach of choice for adult patients with parasternal hernia eligible for surgery.

Key words: Diaphragmatic hernia. Diaphragm. Laparoscopy. Embryology. Anatomy.

INTRODUCTION

A weak area in a portion of the diaphragm or diaphragmatic hernia (DH) may allow abdominal contents to enter the thorax. A diaphragmatic hernia may be located in the esophageal hiatus (hiatal hernia), in its proximity (paraesophageal hernia), at a posterolateral level –Bochdalek’s hernia (BH)—, or at a parasternal level –parasternal (PDH), retrochondrosternal, retrocostoxyphoid, retrosternal, subcostal, substernal, or substernal hernia—.

Etiologically, hernias may be acquired or congenital. Up to 7% of patients suffering from closed thoracoabdominal trauma have a post-traumatic diaphragmatic tear, most often on the left side (1). Increased intra-abdominal pressure and thoracic depression may be significant factors for the development of later hernias (in the adult or the elderly) (2). Thus, in obese patients or subjects with kyphoscoliotic deviation, repeat high abdominal pressure events, as in vomiting or coughing (acquired substrate), may affect reduced-resistance areas in the diaphragm (congenital substrate) (3).

A congenital origin may be demonstrated when symptoms develop in the newborn, but cases have been reported outside the neonatal period. The prevalence of congenital diaphragmatic hernia (CDH) is 0.3-0.5/1,000 births, and hernias are more often located on the left side and predominate in women (2:1) (4). A frequency of 0.69‰ has been reported in Spain, with an annual decreasing trend of 0.10‰ for the period 1980-2006 (5). BH has a prevalence of 1/2,200 births, and PDH has a prevalence of 1/1 million births (6).

In practice CDH is a serious, poorly understood abnormality with a high mortality rate—resulting from underlying pulmonary hypoplasia—whose management is not always effective (7). Most of these hernias are found and repaired in children, but 5% are incidentally diagnosed in adults studied for other conditions (8).

A potential hereditary connection was reported in two instances—a patient who had a mother with PDH and a daughter with congenital pulmonary hernia (9), and a case of CDH in twins (10). Around 10% of all individuals with CDH have chromosomal abnormalities (11). Table I lists some of these syndromes.
Since the studies by Bremer (12) and Wells (13), a widely accepted theory is that CDH stems from diaphragmatic dysgenesis; however, the embryology of the diaphragm remains obscure to this very day.

DIAPHRAGM MORPHOGENESIS

It takes place between the 4th and 12th weeks of gestation. It is a complex process that starts in the cervical region and proceeds in a caudal direction. Table II and figures 1 and 2 schematically illustrate diaphragm development. The diaphragm is made up from four embryonic structures (14-18): 1) Septum transversum (ST) of His or transverse mass of Uskow: a mesodermal bridge representing the primordium of the diaphragm’s phrenic center. It grows dorsally from the body’s ventrolateral wall. 2) Pleuroperitoneal membranes (PPMs) or pleuroperitoneal laminae of Brachet or pillars of Uskow: folds developing on each side of the coelom’s dorsal wall. They grow ventrally and progress closely parallel along regional veins. The costovertebral trigone does not represent the closing of PPMs (19). 3) Dorsal esophageal mesentery (DEM): represents the muscle bundles of the diaphragmatic crura. PPMs converge on DEM and fuse with ST’s dorsal portion (primordial diaphragm). The right hemidiaphragm consolidates earlier than the left one; this, together with the liver’s position on the right side, would explain why BH is more common here; regarding PDH, its higher frequency on the right side would result from the pericardium protecting the left flank, which would hinder its development in this area. 4) Muscle grows medially from the body’s wall: from week 9 to week 12 (fetal period), as lungs increase in size, pulmonary caudal apices open up additional spaces on the body’s wall. The latter’s associated mesenchyma, separated from the wall proper, constitutes a thin ring of tissue along the dorsolateral borders. The diaphragm’s muscular component is made up by myotomes invading the mesenchyma following a dorsal-to-ventral direction, and the anterior diaphragm is formed last. Myoblasts derive from the third, fourth, and fifth cervical somites, with innervation also stemming from these segments. This is the common origin of both anterior diaphragmatic muscle fibers and the suprapleural membrane (Sibson’s fascia) (9). The circumferential portions of the diaphragm are sensitively supplied from caudal intercostal nerves.

Concurrently with the caudal migration of the diaphragm, the sternum fuses together in a craniocaudal direction, and abdominal contents increase rapidly.

<table>
<thead>
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<th>Characteristic</th>
<th>Carnegie stage</th>
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<td>Anlage of ST</td>
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<td>Cervical ST</td>
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<td>Thoracic ST</td>
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<tr>
<td>Anlage of PP membranes</td>
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<td>PP channel closure</td>
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<td>ST fusion- PP membranes</td>
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Several theories have been put forward since pre-hernial lipoma, which is always present in early stages, was considered to penetrate the sternocostal trigon dragging the peritoneum along (20).

No environmental pathogenetic factors have been reported for humans. CDH (always BH) has been experimentally induced with thalidomide (21), vitamin A deficiency (22), polybromobiphenyls (23) or nitrophen (24).

Attributions to the primary role of the lung, phrenic nerve, myotube formation, and pleuroperitoneal channel closure are currently considered false (25). Appropriate primordial diaphragm development has been shown not to depend on lung tissue signals, and diaphragm malformation has been seen to be a primary defect in CDH, resulting from a malformed, non-muscular mesenchymal diaphragm component prior to myogenesis (26).

In practice (18), when a dysontogenetic cause occurs during the embryonic phase (false hernia) the membranous diaphragm completion process comes to a halt, which conditions a persistent gap. Abdominal viscera will not be covered by peritoneum. Hernial sacs will lack peritoneal evaginations, as in BH. In contrast, when such a cause occurs during the fetal period (true hernia), with a closed pleuroperitoneal hiatus but incomplete muscle migration, the defect takes place in the muscular diaphragm. In such cases an increase in abdominal pressure may push abdominal viscera into the thoracic cavity. The hernial sac will have a peritoneal evagination, as in PDH. Exceptionally, as a result of lacunar diaphragmatic aplasia and persistent pericardio-peritoneal shunt, hernias may have no sac, which constitutes the extremely rare diaphragmatic peritoneal-pericardial hernia (27).

CASE REPORT

CDH is clinically asymptomatic in the adult in 30 to 50% of cases, and mainly affects overweight women (28). However, it may also present in thin individuals, as in the following case report.

A 74-year-old woman was admitted for intermittent food vomiting within 30 minutes and 24 hours after ingestion, and weight loss (10 kg) for the last three months. She had no history of abdominal trauma.

Physical examination only reveals a mildly reduced respiratory noise in the right hemithorax. There is dysphonia without odynophagia, and dysmotility-type dyspeptic manifestations.

Chest x-rays (Fig. 3) revealed the right pulmonary area to be occupied by a smoothly opaque, well-delimited, paracardial, antero-inferior mediastinal mass with air-fluid levels on the PA view, suggestive of intestinal loops.

MRI (Fig. 4) showed a parasternal mass and a wide defect in the anterior diaphragm, through which a hernial sac containing most of the gastric body and antrum, omentum, and right colon, consistent with PDH, ascends into the antero-lateral thorax.

The patient was operated upon using laparoscopic surgery under general anesthesia. A hernial hiatus 5 x 10 cm in size was found - hernial contents reduction and hernial sac resection were performed. The defect was
DISCUSSION

PDH is a hernia through the sternocostal triangle (trigonum sternocostale), the space across which superior epigastric vessels and lymph vessels from the liver’s diaphragmatic aspect travel. A multifactorial etiology is currently posited that includes hereditary factors associated with other malformation syndromes (Table II). However, when PDH is diagnosed in children and adults, it is only rarely accompanied by other congenital malformations (29).

PDH represents 3-5% of all DHs (30). It was first described in 1761 by Giovanni Battista Morgagni (1682-1771) on the right sternocostal triangle an Italian stonecutter at necropsy. In 1829 Dominique J. Larrey (1766-1842), Napoleon’s surgeon, described the retrosternal space as an access route for the management of pericardial tamponade. Some authors have designated the right sternocostal triangle “hiatus of Morgagni”, and the left sternocostal triangle “hiatus of Larrey”. Most authors consider the term “anterior diaphragmatic hernia of Morgagni-Larrey” to be the most adequate name (31). However, between the two muscular bundles of the diaphragm there is a tiny intersternal slit designated hiatus of Marfan, after the French pediatrician Antonin Marfan (1858-1942) (32). Should there be agenesis in either sternal bundle – right or left – the hiatus of Marfan may favor the hernial orifice and render its topography more challenging to recognize, hence we consider the term “parasternal diaphragmatic hernia of Morgagni-Larrey” more appropriate.

Clinically, most patients (72%) present with hernia-related symptoms, and pulmonary manifestations are most common (36%) (33). PDH is usually diagnosed with x-rays (chest PA and lateral views). Waelli was first to diagnose the condition in 1911 (34).

It must be differentiated from other masses in the anterior mediastinum (35). Pericardial diverticulum and cyst contain fluid, and are clearly related to the heart. Pericardial hematoma usually shows hyperdensity, is uncommon, and is usually associated with a history of chest trauma. In PDH, air contained in the mass – resulting from the passage of intestinal loops into the thorax – facilitates identification, which is difficult when only omentum is present. Differential diagnosis with lipoma is considered for fatty contents, which are perfectly distinguishable with CT.

CT is the primary diagnostic method, but may be inconclusive should the hernial sac be empty at imaging. Cases diagnosed with MRI and US have also been reported. MRI is a useful method to assess PDH – no radiation exposure, movement arctifact reduction, and potential multiplanar reconstruction (36). Ultrasonography may facilitate detection for PDH near the heart (37). PDH preferentially develops on the right side in 70% (38) to 91% (33) of cases.
MANAGEMENT

Given the low prevalence of this condition no conclusive treatment-related studies or guidelines are available. We consider surgery the therapy of choice even for asymptomatic patients in view of potential complications. Therapy selection will depend on individual patient characteristics, presence or absence of manifestations, and hernia location and size.

Some controversy remains regarding a few aspects of surgical treatment for PDH, including access routes (thoracic vs. abdominal), hernial sac reduction, and use of mesh.

During the 1950s (39) PDH was the one condition where an abdominal approach was favored; thoracic access was only required in the presence of hernial sac to sac contents adhesions. An abdominal approach is currently recommended for children (40), and is a requirement for complications such as hernial strangulation, incarceration (41) or perforation (42) regardless of patient age.

Thoracotomy has remained the most commonly used and/or reported access to this day because of benefits in terms of intraoperative diagnosis (33), greater exposure, easier repair (43), and mediastinal mass characterization (44).

An abdominal route will facilitate hernial reduction, and an identification of associated lesions at both the diaphragmatic level and anywhere else in the abdominal cavity at the expense of more postoperative complications and longer hospital stays. On the other hand, a thoracic route will facilitate hernial sac, pleural, and mediastinal dissection.

Authors perform hernia reduction in 100% of thoracotomies and 82% of laparotomies reported (33), but its benefits have not been demonstrated (45). In children, perteironal sac dissection has resulted in some fatal pneumopericardium events. Other authors recommend excision only for small sacs with no intrathoracic adhesions, as the potential for injury to thoracic structures would be small. However, some believe that sac resection is a safe procedure and adhere to traditional surgical principles. This opinion may explain why thoracotomy has been the most commonly used surgical approach (42). In cases where the sac was not withdrawn some spontaneous reductions were seen, while others persisted with a fluid content. To prevent cyst formation some authors leave a drainage tube in the hernial sac. We consider that sac resection should not be attempted universally but selected on an individual basis.

On the other hand, most case reports do not consider using a mesh, which is usually of non-resorbable material when used (46).

However, since the first laparoscopic approach was reported in 1992 (47), surgeons have been using this technique increasingly. Benefits include ease of use, excellent access to the parasternal area (48), outstanding surgical field allowing appropriate manipulation with minimal trauma and complications (around 5%), and mean hospital stay of 3 days. Most of those who use this technique use mesh for defect repair (64%) and do not resect hernial sacs (69%) (42).

We consider laparoscopy the procedure of choice for adult patients with PDH who are eligible for surgery, and believe that surgeons should become familiar with this technique.

REFERENCES