Parahiatal hernia: A rare type of hernia and the answer to an anatomical challenge

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SUMMARY

Parahiatal hernias are rare. They are difficult to diagnose preoperatively, as the clinical symptoms may be similar to hiatal and paraoesophageal hernias. Here, we report two cases of parahiatal hernia that were preoperatively diagnosed and successfully repaired laparoscopically; using the particular anatomic characteristics of this hernia, we also review the controversial oesophageal hiatal anatomy, as the surgical community often refers to the left bundle of the right crus as the left crus. There is no consensus on the indication or surgical technique to repair them.

The first case is a 59-year-old woman with non-specific abdominal symptoms, in whom the preoperative gastroscopy and computed tomography (CT) raised the suspicion for parahiatal hernia. The second case is a 68-year-old woman who presented to the emergency department with abdominal distention and nausea, but no vomiting. Preoperative CT raised the suspicion of an incarcerated parahiatal hernia. Both patients underwent laparoscopic repair of the parahiatal hernia and a Toupet fundoplication. They had an uneventful postoperative course. After more than 4 years of follow-up, they are both asymptomatic. Parahiatal hernias are a rare form of diaphragmatic hernia that occur through a diaphragmatic defect lateral to an anatomically normal oesophageal hiatus, with herniation of contents between the left portion of the right crus and the left crus. Up to five different anatomical variations have been described. The knowledge of these anatomical variations has an impact on the type of surgical repair that will need to be performed if a parahiatal hernia is found.

Key words: Parahiatal hernia – Left crus – Oesophageal hiatus – Gastroesophageal reflux

INTRODUCTION

Parahiatal hernia (PH) is a rare form of diaphragmatic hernia that occurs from muscular defects separate from the oesophageal hiatus and the foramina of Morgagni (Demmy et al., 1994; Scheidler et al., 2002; Palanivelu et al., 2008; Ohtsuka et al., 2012; Lew and Wong, 2013; Takemura et al., 2013; Akiyama et al., 2017; Staerkle et al.,

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Submitted: April 26, 2023. **Accepted:** June 16, 2023

https://doi.org/10.52083/YJGE5299

2018; Preda et al., 2019; Li et al., 2020). The anatomical studies of the oesophageal hiatus reported by Collis et al. in 1954 demonstrated that there is a large right crus and a smaller left crus which takes no part in the formation of the oesophageal hiatus (Collis et al., 1954). PH result from the herniation of intraabdominal contents between the left portion of the right crus and the left crus lateral to, but distinct from, an intact oesophageal hiatus (Koh et al., 2016; Li et al., 2020). PH are often misdiagnosed as paraoesophageal hernias (POH), as they may have similar radiological findings (Koh et al., 2016).

Although the symptoms of PH are similar to POH, the herniation through a parahiatal defect is generally associated with a high risk of developing perforation and strangulation of the involved organs, which can be life-threatening (Li et al., 2020). There is no consensus regarding the diagnosis and treatment of PH (Li et al., 2020) .

It is common in surgical practice to call the "left crus" the bundle of muscles situated medially to the oesophagus, when in actuality both medial and lateral muscles are part of the right crus. The importance of this anatomical variation lies on the higher risk of complications associated to PH, advocating for an early repair, and on the different surgical approach required for PH with no reflux symptoms in comparison to those that do have acid reflux symptoms and POH.

The aim of this paper is to present two cases of PH treated in our unit and also to review the anatomy of the oesophageal hiatus, clarifying which is the true left crus, and highlighting the importance of its anatomical knowledge when performing a surgical repair of a PH.

CLINICAL CASES

The first case is a 59-year-old woman, with no remarkable past medical history and no previous abdominal surgeries, who presented with dyspepsia, nausea, vomiting and abdominal pain. An oesophago-gastro-duodenoscopy (OGD) was performed, showing a patulous oesophagogastric junction (OGJ) and a herniation of the fundus in a parahiatal fashion with the OGJ at the correct position (36cm from incisor) (Fig. 1A). The CT scan

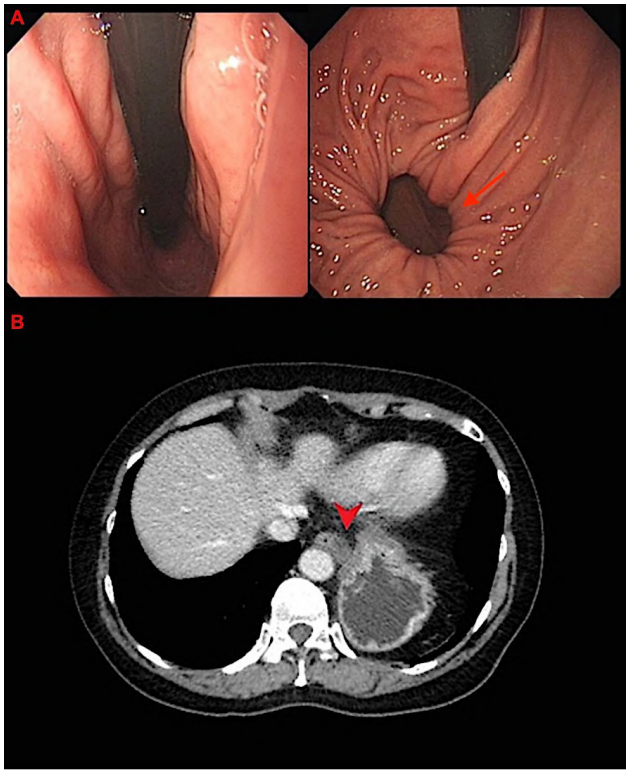

Fig. 1.- Preoperative study case 1. **A:** gastroscopy (arrow: herniation in parahiatal fashion). **B:** CT scan (arrowhead: parahiatal defect).

revealed a left PH (Fig. 1B) with a clear bundle of muscle between the oesophagus and the herniated stomach (Fig. 1B, arrow). The patient subsequently underwent elective surgery.

The second case is a 68-year-old woman, with no other comorbidities, who presented to the emergency department with a short history of abdominal pain, distention and nausea, but no vomiting. She underwent a CT abdomen pelvis that suggested an incarcerated PH (Fig. 2). Decision was made to take her for emergency surgery.

In the first case, a PH between the left portion of the right crus and the left crus was identified intraoperatively (Fig. 3. 1: right portion right crus; 2: left portion right crus; 3: left crus; O: oesophagus; PH: parahiatal hernia). A laparoscopic tension-free parahiatal-defect repair, reinforced with a biological mesh and a Toupet fundoplication, was performed.

In the second case, an incarcerated PH was identified. A laparoscopic Toupet fundoplication was also performed, together with a primary repair of the parahiatal defect and closure of the hiatus. A 28Fr chest drain inserted transabdominal-

ly through one of the laparoscopic ports was left in the left chest. It was removed on post-operative day 3.

The post-operative course was uneventful in both patients, being both able to tolerate a soft diet upon discharge. More than four years after surgery they both were asymptomatic. Both patients were followed up with post-operative barium swallow, with no evidence of gastroesophageal reflux or hernia recurrence.

DISCUSSION

PH is a rare form of diaphragmatic hernia. They are distinctly different from POH, such that they occur through a diaphragmatic defect lateral to an anatomically normal oesophageal hiatus (Rodefeld and Soper, 1998; Scheidler et al., 2002; Lew and Wong, 2013; Takemura et al., 2013; Preda et al., 2019) . The hiatus is structurally normal and both crura are intact (Rodefeld and Soper, 1998). Their exact incidence is unknown, being estimated in 0.2-0.35% from different case series (Scheidler et al., 2002; Palanivelu et al., 2008; Koh et al., 2016; Akiyama et al., 2017; Staerkle et al., 2018; Preda et al., 2019; Li et al., 2020). In our series, the incidence is of 0.23% (2 out of 850 fundoplications for hiatus hernia and gastroesophageal reflux). They are characterized by the presence of a separate extrahiatal diaphragmatic defect between the left portion of the right crus and the left crus with an intact oesophageal hiatus (Koh et al., 2016; Lew and Wong, 2013; Akiyama et al., 2017) (Fig. 4).

They can be classified based on their aetiology (primary or secondary), complications (complicated and uncomplicated), and association with the OGJ (normal OGJ or displaced OGJ) (Palanivelu et al., 2008; Lew and Wong, 2013). Congenital or primary PH develop as a result of a failure of the embryonic pleuroperitoneal canal to obliterate during embryogenesis, resulting in a persistent pneumoenteric recess that is located immediately to the left of the oesophageal hiatus (Demmy et al., 1994; Rodefeld and Soper, 1998; Palanivelu et al., 2008; Ohtsuka et al., 2012; Lew and Wong, 2013; Takemura et al., 2013; Li et al., 2020). Although these hernias may arise from both sides of the pneumoenteric recess, they are usually found

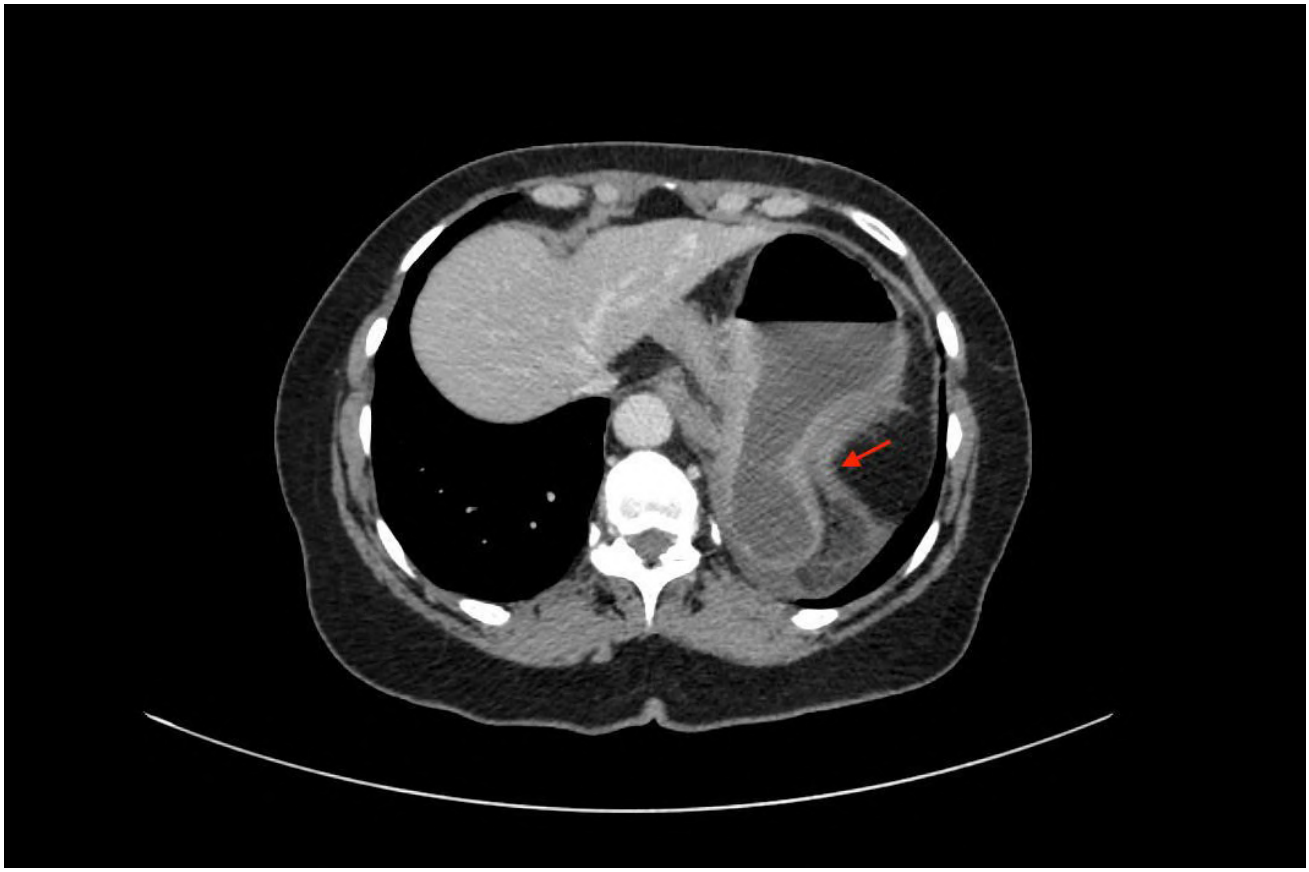

Fig. 2.- Preoperative CT scan case 2 showing incarcerated parahiatal hernia (arrow).

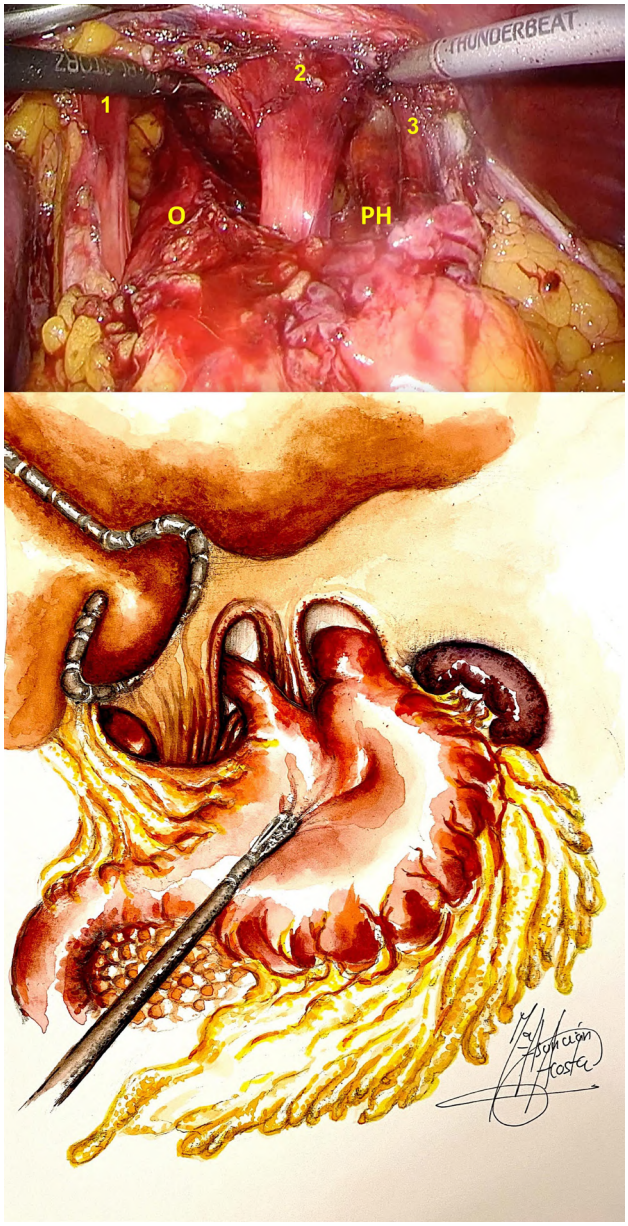

Fig. 3.- Intraoperative findings in case 1 showing the oesophageal hiatus (O) and the parahiatal defect (PH), in comparison to drawing of intraoperative appearance of parahiatal hernias. 1: right portion of the right crus. 2: left portion of the right crus. 3: left crus.

on the left side, possibly due to the presence of the liver on the right side (Takemura et al., 2013; Koh et al., 2016). Acquired or secondary PH can be a result of traumatic injury to the diaphragm or iatrogenic injury from previous surgery in the left upper quadrant of the abdomen (Lew and Wong, 2013). Secondary PH are known to occur after oesophagectomy or cardiomyotomy, probably due to excessive manipulation of the crura or incision on the diaphragm, or while dissecting the GOJ (Palanivelu et al., 2008). The content of the sac is most often the gastric fundus, prone to volvulation (Palanivelu et al., 2008).

Unlike the mechanism of POH, which occur as the stretching of the phreno-oesophageal membrane, there are no morphological changes of the phreno-oesophageal membrane in PH (Li et al., 2020).

The fibres of the right crus arise from the main tendon and in varying degree from the median arcuate ligament (Collis et al., 1954). Some of these fibres may arise from this latter ligament to the left of the midline, but in all cases they can readily be separated from the left crural fibres (Collis et al., 1954). There is no decussation of muscle fibres in front or behind the oesophageal orifice, but varying degrees of muscle overlap are a constant feature (Collis et al., 1954). The fibres on the left part of the right crus, which often arise from the median arcuate ligament, pass upwards above the fibres already mentioned to reach the left side of the hiatus. This produces an effect of a double-breasted coat, more or less well marked from case to case. In the standard type, the median arcuate ligament is always present, although in many cases it is poorly developed (36%) (Collis et al., 1954).

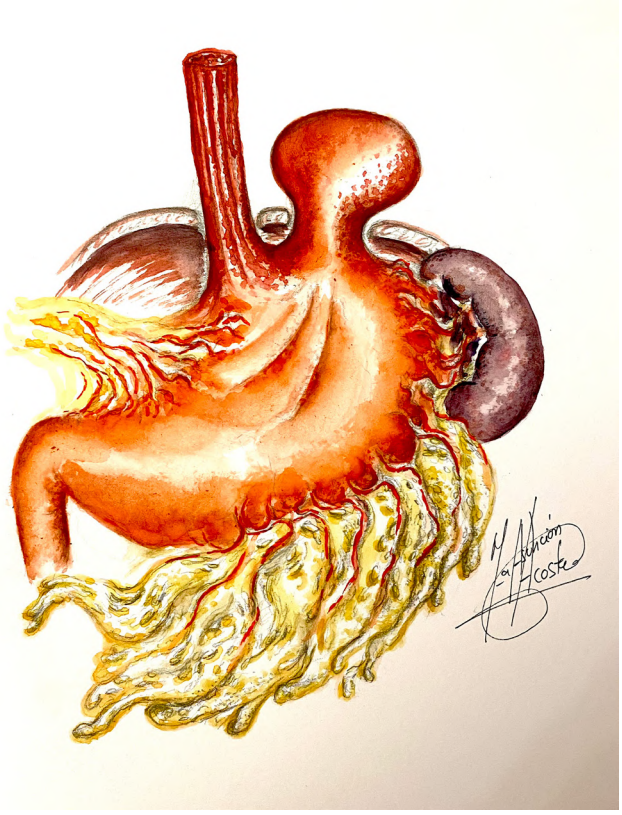

Fig. 4.- Drawing of anatomical appearance of parahiatal hernias.

John Leigh Collis, an English thoracic surgeon, et al. described in 1954 the different anatomical variations of the crura of the diaphragm based on cadaveric anatomical studies (Collis et al., 1954). Up to five different variations of the anatomy of the hiatus were described (Fig. 5). In most cases the left part of the right crus is medial to the oesophagus, and during the hiatal hernia repair the two portions of the right crus are the muscles included in the repair. This is often mistaken in surgical practice, quoting the suturing of the right pillar to the left pillar.

In the type one or standard type of muscular arrangements at the oesophageal hiatus, the muscle quality is good, with a good overlap of the fibres from the right across to the left. The fibres pass inferiorly to the similar band passing from the left to the right. A good median arcuate is present. The fibres of the left crus take no part in the boundaries of the oesophageal orifice (Collis et al., 1954).

Type two is a weak variety of the standard type. The muscle is quite good, but the overlap of fibres between the two parts of the right crus is poorly developed. No median arcuate ligament is present (Collis et al., 1954).

In the type-three variant, a shift to the left is described. A strong median arcuate ligament is present, and many fibres destined for the right of the oesophageal orifice arise from this ligament to the left of the mid-line. The left crus is independent from these fibres and plays no part in forming the oesophageal orifice (Collis et al., 1954).

The type-four variant is characterized by the absence of the median arcuate ligament. The band of fibres on the right side of the oesophagus arises wholly from the left crus and crosses underneath, in scissor fashion; a corresponding band from the right crus going to the left of the oesophageal orifice (Collis et al., 1954).

In the last variant (type five), a complete shift to the left is observed. It is the most uncommon variant (2%). The left crus supplies all muscle fibres taking part in the formation of the oesophageal orifice. The overlapping fibres pass in the same direction as with a standard type diaphragm. This would be reversed in a congenital transposition (Collis et al., 1954).

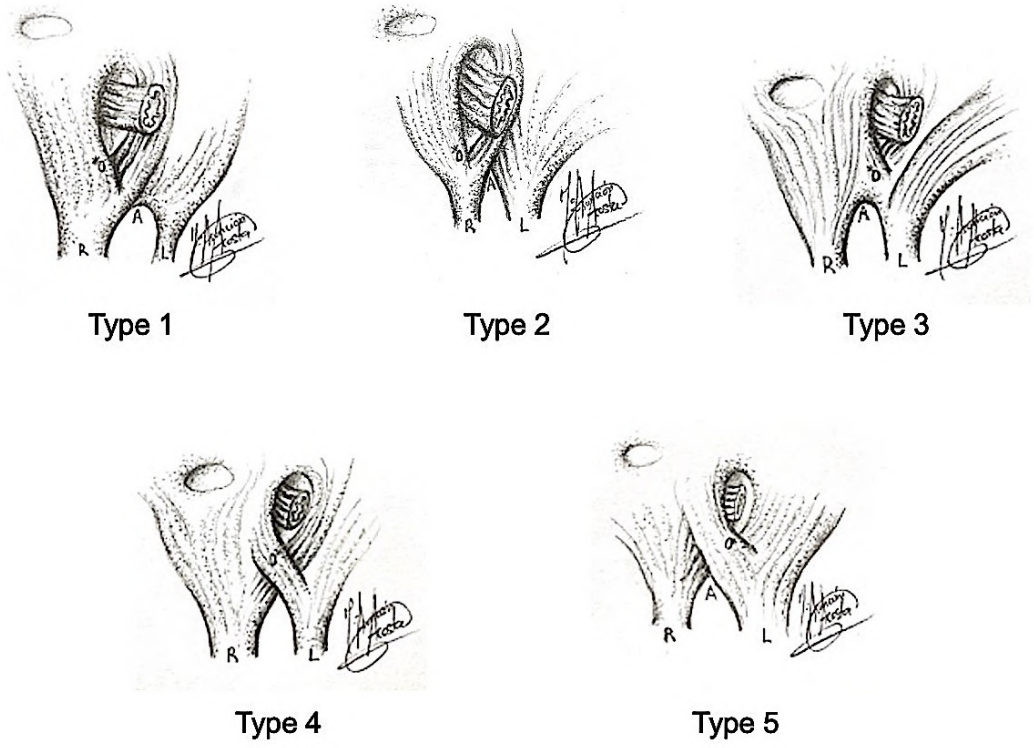

Fig. 5.- Anatomical variations of the oesophageal hiatus depending on the disposition and insertions of the diaphragmatic crura. R: right crus. L: left crus. O*: oesophageal hiatus. A: Aortic hiatus.

Unlike in our cases, it is difficult to diagnose a PH preoperatively (Ohtsuka et al., 2012; Akiyama et al., 2017). Most PH are diagnosed intraoperatively, during the repair of preoperatively presumed hiatal or POH (Lew and Wong, 2013). Clinically, it is difficult to differentiate between a parahiatal and a hiatal or paraoesphageal hernia, as they can present with epigastric pain, nausea, vomiting, heartburn and post-prandial bloating (Scheidler et al., 2002; Lew and Wong, 2013; Akiyama et al., 2017; Staerkle et al., 2018). Radiologically, if the crural musculature between the hiatus and the hernia orifice can be identified on an abdominal computed tomography (CT), as in our cases, it might aid in the diagnosis of PH (Akiyama et al., 2017). Preoperative studies in suspected PH should be the same as for hiatal or POH, including appropriate history looking for gastroesophageal reflux symptoms, preoperative OGD, and pH studies and manometry if indicated.

The detailed knowledge of the hiatal anatomy is crucial given the complications associated to PH and the type of hiatal repair required. Because of a high risk of perforation, incarceration and strangulation of involved organs, when preoperatively identified, surgery is always indicated to correct the parahiatal defect (Scheidler et al., 2002; Lew and Wong, 2013; Staerkle et al., 2018). Intraoperatively, in cases where a PH is suspected, focused dissection should be performed at the left crus; the right crus should be left alone, as unnecessary dissection might disrupt an otherwise normal hiatus (Koh et al., 2016). Fundoplication is also typically not required unless the patient has symptomatic reflux or there is a hiatal defect (Koh et al., 2016; Staerkle et al., 2018; Preda et al., 2019). In our cases, we associated a fundoplication to the parahiatal defect repair, as a hiatal defect was seen intraoperatively in both cases. In the first case, the fundoplication was also indicated for the gastroesophageal reflux symptoms that the patient presented preoperatively. Moreover, extensive hiatal dissection with the subsequent destruction of the natural antireflux mechanism, required to achieve the parahiatal defect repair, is also an indication for associating an antireflux procedure. Deep anatomical knowledge is fundamental for appropriate choice of surgical repair. The differentiation between the oesophageal hiatus, formed by the two portions of the right crus, and the parahiatal space found between the left and the right crus, can be referred by surgeons as a challenge, as the hiatal anatomy is not clearly described in most surgical papers that refer to this type of condition: usually the left portion of the right crus is referred as the "left crus", which is anatomically incorrect. This misunderstanding of the hiatal anatomy may lead to suboptimal surgical repairs. An incorrect repair of the hiatus may lead to long-term post-operative complications, such as recurrence of the hernia, gastroesophageal reflux and all its complications, as well as stricturing of the hiatus with dysphagia that may require endoscopic dilatation, re-do of the hiatal repair, or even oesophagectomy in cases with severe strictures resistant to less invasive treatments.

The repair of these hernias can be performed through an open or a laparoscopic approach. Laparoscopic repair provides many benefits, including better visualization of the operative field, faster recovery and shorter hospital stay, and can be performed safely by laparoscopic surgeons familiar with the repair of paraoesophageal and hiatal hernias (Lew and Wong, 2013). The surgical principle of tension-free repair should be also applied to PH. This can be done either by primary repair, with a prosthetic mesh, or both (Lew and Wong, 2013). In circumstances where large defect size and fibrosis prevent tension-free primary repair, the use of a composite mesh can provide effective repair of the hernia with good outcome (Lew and Wong, 2013). As seen in our patients, the few cases published to date had no post-operative complications; recurrence in the long term after surgery was not reported.

CONCLUSIONS

PH are an uncommon type of diaphragmatic hernia. These hernias arise between the left portion of the right crus and the left crus, existing up to five different types of configurations of the hiatal anatomy. And the two bundles of muscle that are repaired during antireflux surgery in most cases belong to the right crus. Preoperative diagnosis is challenging, given the similarity with

hiatal hernias in imaging and symptoms. Surgical repair of these diaphragmatic defects is advocated in all cases, given the high risk of complications; and deep knowledge of the anatomy of the hiatus is fundamental to perform an appropriate repair with or without an anti-reflux procedure, depending on the intraoperative findings and the patient's preoperative symptoms. An anti-reflux procedure should be performed if there are any reflux symptoms, if there is a hiatal defect, or after an extensive mobilization of the oesophagogastric junction. This can be done by an open or a laparoscopic approach, with or without mesh reinforcement of the repair.

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