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REVIEW

Presentation and management of Morgagni hernias in adults: a review of 298 cases

John D. Horton · Luke J. Hofmann · Stephen P. Hetz

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Abstract
Background Morgagni hernias are a very rare form of diaphragmatic hernias. No robust studies have been performed to show the true natural history of this disease process. This study aimed to summarize clinically relevant data with respect to Morgagni hernias in adults. These data should help surgeons workup, diagnose, and treat Morgagni hernias in adult patients.
Methods A literature search was performed using PubMed, Google scholar, and the following key words: Morgagni, Larrey, retrosternal, retrocostoxiphal, retrochondrosternal, parasternal, substernal, anterior diaphragmatic, and subcostosternal. All case reports and series after 1951 that pertained to adults were included in the review. The following data points were queried: age, sex, presentation, studies used during workup, laterality, surgical approach, hernia sac management, specific laparoscopic techniques, and follow-up evaluation.
Results These criteria were met by 135 articles representing 298 patients. Based on the data provided, several conclusions regarding this disease process can be drawn. Most patients (72%) present with symptoms related to their hernia. Pulmonary complaints are the most common symptoms (36%). Men present earlier in life than women. Thoracotomy is the most widely used surgical approach (49%). However, laparoscopic repair has gained popularity since its first report in 1992. Laparoscopic surgeons usually repair the defect with mesh (64%) and do not remove the hernia sac (69%). Laparoscopic repair can be performed with a low complication rate (5%) and a short hospital stay (3 days). Outcomes of other surgical approaches also are reported.
Conclusions Using modern surgical techniques including laparoscopy, repair of Morgagni hernia can be performed safely with a short hospital stay and with little morbidity or mortality.

Keywords Diaphragmatic hernia · Hernia · Laparoscopy · Morgagni · Review

A Morgagni hernia (MH) is a congenital defect found in the anterior aspect of the diaphragm between the costal and sternal portions of this muscle. Morgagni hernias make up 3% of all diaphragmatic hernias [1]. Because of its congenital nature, MH often is considered to be a pediatric condition. However, there have been many case reports and small series of MH involving adults. There have been no large retrospective or prospective studies on this topic. Morgagni hernia is a rare clinical entity among adults without a well-described prevalence or natural history. The clinical presentation of this hernia may be confusing, and definitive management strategies have not been well established. As surgical technology has evolved, new techniques for repairing these defects have been developed. In this review, we aim to provide the clinician with a primer on the presentation, workup, and surgical management of MH. The review pays special attention to the role of laparoscopic surgery in the treatment of MH. The analysis represents the most comprehensive review on the topic of MH in adults.

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J. D. Horton (✉) · L. J. Hofmann · S. P. Hetz
Department of Surgery, William Beaumont Army Medical Center, 5005 North Piedras Street, El Paso, Texas 79920, USA
e-mail: John.horton2@amedd.army.mil

Materials and methods

We conducted an English language PubMed search for every case report, series, and literature review relating to MH in adults. The key words used were Morgagni, Larrey, retrosternal, retrocostoxiphal, retrochondrosternal, parasternal, substernal, anterior diaphragmatic, and subcostosternal hernias. For the purpose of this review, patients older than 16 years were considered adults. The search included all articles from 1951 to November 2006. We excluded articles pertaining only to MH in children. Morgagni hernias discovered at autopsy were not included. Finally, we reviewed the reference lists of every included and relevant article to identify additional articles.

A single author extracted data from each article in a standardized manner. If available, the author recorded age, sex, laterality, presenting symptoms, acuity of symptoms, preoperative diagnostic studies, presence of preoperative diagnosis, predisposing factors, surgical repair technique, hernia sac contents, complications, postoperative stay, 30-day mortality, recurrence, follow-up data, and presence of preoperative symptoms postoperatively. Due to the large numbers of small reports, we performed only descriptive statistics.

Results

The inclusion criteria were met by 135 articles reporting clinical data for 298 patients. Of these 135 articles, 106 were single-patient case reports. Despite an English language only search strategy, we included authors from around the world, including the United States, Europe, Turkey, Japan, India, Pakistan, Nairobi, New Zealand, Australia, Israel, and Greece. The disciplines of general surgery, minimally invasive surgery, thoracic surgery, internal medicine, and radiology all contributed articles for analysis.

Women made up 62% of the patients in this review. The average age of the patients was 53 years (Table 1). The women had a rather normal distribution, with 15% presenting between 56 and 60 years of age. Men, on the other hand, had a more bimodal distribution, with peaks at ages 31–35 years and 66–70 years. The average age at presentation was 58 years for the women and 50 years for the men.

The anatomic distribution of MH was found to be 91% for the right, 5% for the left, and 4% for bilateral cases (Table 1). Predisposing conditions, present in 41% of the cases, included pregnancy, trauma, obesity, chronic constipation, chronic cough [2–5], and any condition the author thought contributed to the patient's clinical presentation.

Table 1 Demographics and laterality

	Result
Average age (year) ($n = 282$)	53
Female (%) ($n = 298$)	62
Predisposing factor (%) ($n = 213$)	41
Right sided (%) ($n = 258$)	91
Left sided (%) ($n = 258$)	5
Bilateral (%) ($n = 258$)	4

The analysis showed that only 28% of the patients were truly asymptomatic at the time of presentation. For this analysis, symptoms were broken down into the following categories: bowel obstruction, pulmonary symptoms, pain/pressure, dysphasia, bleeding, gastroesophageal reflux disease (GERD), and other. The percentage of patients presenting with each symptom category is listed in Table 2. The pain/pressure category included patients with chest or abdominal discomfort not related to a bowel obstruction. Pulmonary symptoms were assigned to patients with dyspnea, cough, and shortness of breath that could reasonably be explained by their MH. According to the findings, 28% of patients presented within 1 month after the onset of symptoms.

Several diagnostic methods have been used to evaluate patients with MH. The most common study reported is chest X-ray (93%), followed by computed tomography (CT) scan (47%), contrast enema (24%), upper gastrointestinal (GI) study (23%), upper GI endoscopy (8%), and magnetic resonance imaging (MRI) (5%).

Repair of MH has been accomplished primarily through four procedures: laparotomy, thoracotomy, thoracoscopy, and laparoscopy (Table 3). Only two cases using the thoracoscopic approach have been reported in the English literature [6, 7]. Analyses of the surgical technique used in the repair of MH were included in this review. Table 3 also displays the percentage of cases in which the hernia sac was excised for each surgical approach.

Table 2 Presenting symptoms

	%
Asymptomatic ($n = 295$)	28
Pulmonary symptoms	36
Pain/pressure (chest or abdomen, not obstruction)	37
Obstruction	20
Dysphagia	3
Bleeding	1
GERD	1
Other (HTN, fatigue, indigestion)	1
Symptoms for less than 1 month ($n = 112$)	28

GERD, gastroesophageal reflux disease; HTN, hypertension

Table 3 Comparison of surgical approaches

	Laparotomy % (*n*)	Thoracotomy % (*n*)	Laparoscopy % (*n*)	Thoracoscopy % (*n*)
Choice of surgical approach	30 (81/269)	49 (132/269)	17 (46/269)	0.7 (2/269)
Diagnosis before surgery	87 (67/77)	66 (46/70)	91 (42/46)	0 (0/2)
Cases requiring emergency surgery	26 (17/65)	6 (3/49)	3 (1/35)	0 (0/2)
Resection of hernia sac	82 (32/39)	100 (63/63)	31 (13/42)	100 (1/1)
Primary repair	88 (68/77)[a]	92 (110/119)	29 (13/45)[b]	100 (2/2)
Interposition mesh graft	6 (5/77)[a]	8 (9/119)	64 (29/45)[b]	0 (0/2)
Complications	17 (9/53)	6 (6/94)	5 (2/44)	0 (0/2)
30-Day mortality	4 (3/76)	0 (0/123)	0 (0/46)	0 (0/2)
Mean hospital stay (days)	11 ($n = 22$)	8 ($n = 34$)	3 ($n = 39$)	14 ($n = 2$)
Recurrence	0 (0/78)	1 (1/129)	0 (0/46)	0 (0/2)
Mean follow-up (months)	28 ($n = 23$)	85 ($n = 42$)	15 ($n = 35$)	12 ($n = 2$)

[a] Four cases used a primary repair reinforced with mesh. These cases were not classified as either a primary repair or an interposition mesh graft

[b] Three cases used a primary repair reinforced with mesh. These cases were not classified as either a primary repair or an interposition mesh graft

The review identified 46 laparoscopic cases in the literature. During laparoscopy, the hernia sac usually was removed (69%). Repair generally was performed using an interposition mesh graft (64%), with 29% of cases receiving primary repair and 7% receiving primary repair reinforced with mesh. The reported outcomes for the laparoscopic approach have been excellent (Table 3). Due to the small cohort, conclusions regarding the superiority of primary repair versus interposition mesh graft versus primary repair with mesh reinforcement cannot be substantiated. However, no appreciable differences in outcomes are evident with the available data. The complication rate is comparable with that for other surgical approaches. Hospital stay appears less than after thoracotomy or laparotomy. Additionally, there have been no reported recurrences or deaths after laparoscopic repair.

Authors reported a wide variety of complications. Examples include pneumonia, arrhythmias, pneumothorax, wound infection, and pleural effusion. Only one recurrence and three deaths were reported.

Discussion

The hernia that now bears his name was first described by Morgagni after reading an autopsy of an Italian stonecutter and published in his book *Seats and Causes of Diseases* (1761) [8, 9]. This defect also is referred to as the space of Larrey, Napoleon's surgeon, who described the retrosternal space as an avenue through which pericardial tamponade could be treated [9]. Some authors refer to the potential retrosternal space on the right as "Morgagni's gap" and the space on the left as "Larrey's gap" [10].

In 1911, Waelli made the first radiologic diagnosis of an MH [11]. The formation of the diaphragm, which takes place between the 4th and 12th weeks of gestation, is a complex process. The diaphragm begins formation in the cervical region and proceeds in a caudal direction [12]. The muscular component of the diaphragm is formed by myotomes that invade the mesenchyma in a dorsal to ventral orientation [13]. Thus the anterior aspect of the diaphragm is the last to form. As the diaphragm migrates caudally, two other processes are occurring. The sternum is fusing in a cephalad to caudal direction, and there is a rapid increase in the abdominal contents. Errors in the coordination of this process lead to congenital defects or weakness in the diaphragm.

Morgagni hernias are far more common on the right despite protection from the liver. One hypothesis for this disparity is that the more extensive pericardial attachments on the left provide additional support for that side of the diaphragm [14]. The defect itself usually has a greater transverse dimension than the anteroposterior dimension [2].

Almost any nonretroperitoneal abdominal organ can be found within an MH. However, omentum exclusively (31%) or colon and omentum (29%) constitute the most common contents of the hernia sac (Fig 1). Stomach (15%), small bowel (11%), and liver (4%) also may be found within the hernia sac. Some authors believe that patients with a herniated stomach are more likely to have serious symptoms than those with herniated colon and/or omentum [14]. Occasionally, patients may have a coexisting paraesophageal hernia [15–19]. There are two reported cases of a right Morgagni defect containing both pleural and peritoneal contents [20]. One of these patients reported a long history of asthmatic-type attacks and poor exercise tolerance, which subsided after surgery [21]. Dawson and Jansing [22] reported a case of a patient who underwent exploration and was found to have colon cancer within the hernia sac.

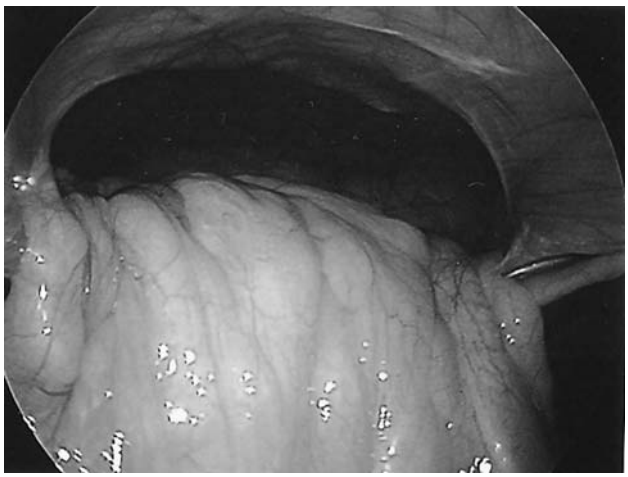

Fig. 1 Laparoscopic view of a right-sided Morgagni defect with omentum and colon herniating into the chest

A hereditary link was hypothesized by Catalona et al. [3] in a report that described a mother with an MH and her daughter with a congenital lung hernia. These authors observed that both the anterior muscle fibers of the diaphragm and Sibson's fascia have a common embryologic origin in the third, fourth, and fifth segments of the cervical mesenchyme. Tazuke et al. [23] provided additional data when they documented a case of congenital diaphragmatic hernia in identical twins. The condition also has been associated with the following syndromes and congenital defects: Down's syndrome, Turner's syndrome, Noonan syndrome, Prader Willi syndrome, tetralogy of Fallot, ventricular septal defects, scoliosis, Morquio syndrome, connective tissue disorders, dextrocardia, chest wall deformities, genitourinary abnormalities, and omphalocele [24–33].

Although the prevalence of MH in the adult population has not been studied, Mullins et al. [34] reviewed 13,138 CT scans and found the prevalence of Bockdalek hernias to be 0.17%. It is important to understand that the statistics in the following discussion describe only the patients reported in the literature. Older series report that 59% of all patients who present with this condition are older than 50 years [35]. Our study supports this finding because the average age of all the patients was 53 years. Based on our analysis, it appears that men may present earlier than women. This may reflect the fact that men are more likely to sustain mild to moderate trauma at a younger age. Whereas our review found that 62% of the patients were female, it is reported that the prevalence among infants is equal between the sexes [36].

This review found that a significant proportion of patients had predisposing conditions possibly contributing to the development of an MH. Pregnancy, trauma, obesity, chronic constipation, and chronic cough are common predisposing conditions cited in the literature [2–5]. The common thread in these conditions is increased intraabdominal pressure. Authors have reported several unusual cases resulting from a presumed increase in intraabdominal pressure. Two cases of herniation after peritoneal dialysis have been reported [37, 38]. Furthermore, herniation has been attributed to pancreatitis that caused increased intraabdominal pressure [4].

A common misconception is that these patients often are asymptomatic at the time of presentation [2, 13, 16, 39–41]. This analysis found that 72% of patients present with symptoms related to their MH. Pain and pulmonary complaints were the most commonly reported symptoms (Table 2). Intuitively, it is reasonable to assume that there are many asymptomatic people with MH. Unfortunately, there are no data to support this assumption. Physiologic consequences of MH are not always apparent, as illustrated by one patient who had resolution of refractory hypertension after repair of massive bilateral MH [42]. Slaetis [43], who published one of the larger series, was unable to make an association between the size of the hernia and symptoms. In a review of 24 cases, Sirmali et al. [44] found that small hernias ($<5 \times 5$ cm) were asymptomatic. However, small hernias in several cases did produce symptoms.

The differential diagnosis of MH is quite extensive. The diagnosis can be especially challenging when the only radiographic abnormality is an anterior cardiophrenic angle abnormality with no evidence of bowel gas patterns in the chest. Based on this chest X-ray finding, the differential diagnosis could include lipoma, liposarcoma, lymphoma, teratoma, neurofibroma, pericardial cyst, Bochdalek hernia, tuberculoma, lung cancer, pleural mesothelioma, atelectasis, pneumonia, thymoma, localized anterior segmental eventration of the diaphragm, empyema, hydatid cyst, and sequestration of the lung [45]. Radiographic abnormalities in this location may be the consequence of medical treatment. For example, high doses of steroids can cause enlargement of the pericardial fat pad [9].

The workup for a suspected MH begins with a thorough history and physical examination. Providers should query patients for obstructive or pulmonary symptoms. Patients may report an experience of trauma in the remote past. At the physical examination, auscultation of the chest may reveal bowel sounds. Radiographic studies begin with a chest X-ray, which may reveal a bowel gas pattern within the chest (Fig. 2) [46]. On a lateral chest X-ray, the hernia may resemble the profile of a cane handle located just beneath the sternum. This finding has come to be known as the "sign of the cane" [47]. Abnormal findings usually are followed up with a CT scan of the chest and abdomen. The CT scan may show contrast-filled bowel in the chest (Fig. 3) or a solid-appearing mass that can be confused with a lipoma.

Fig. 2 Preoperative chest X-ray showing bowel gas within the right chest

Fig. 3 Coronal image of a computed tomography three-dimensional reconstruction showing contrast-filled bowel within the right chest

A diagnosis of MH is strongly suspected if fine curvilinear or linear densities are seen within the mass [48–52]. Hounsfield units ranging from −80 to −120 suggest that the lesion is composed of fat [53]. These curvilinear densities most likely represent vessels within the omentum. Additionally, a CT three-dimensional reconstruction may be helpful [54]. In cases that have no abdominal viscera located within the chest, MRI may be able to discern between a chest process and one that originates from within the abdomen. Some features that make MRI a useful method for evaluating diaphragmatic hernias are the absence of ionizing radiation, the reduction in motion artifacts by cardiac and respiratory gating, and the potential for multiplanar imaging [55]. Echocardiography provides another diagnostic option that has been used to identify MH when it closely abuts the heart [56, 57]. Pfannschmidt et al. [58] reported a series of four patients in whom they measured pre- and postoperative spirometric values. Their study showed that some spirometric values improved 3–21 months after surgery. Regardless of the method, prompt diagnosis is imperative because a late diagnosis or misdiagnosis can be fatal [59].

Surgery provides definitive management for patients with MH. However, because the prevalence of MH has not been reported, it is impossible to compare operative and nonoperative management. Hence, the true benefit of surgery is unknown. In the opinion of many authors, surgery is indicated for both symptomatic and asymptomatic presentations [13, 44, 60–63]. It is easier to justify surgery for symptomatic patients, so the real quandary lies with the management of asymptomatic patients. Whereas one report describes successful nonoperative management of a MH [64], several examples exist of patients who have failed this management strategy [63, 65–71]. One of these patients was followed for 18 years before requiring surgery [71].

Another factor that supports the routine repair of MH is the addition of laparoscopy to the surgeon's armamentarium. Cholecystectomy, Nissen fundoplication, paraesophageal hernia repair, gastrectomy, abdominal aortic aneurysm repair, and coronary artery bypass graft (CABG) have been performed simultaneously with MH repair [17, 61, 72–76]. Successful repair during pregnancy also has been reported. Kurzel et al. [77] recommend elective repair if MH is discovered in the first or second trimesters. If MH is discovered in the third trimester, a mature fetus should be ensured before simultaneous cesarean section and hernia repair are performed.

Several surgical approaches have been used to repair MH (Table 3). The abdominal approach allows for easier reduction of the hernia contents, evaluation of the contralateral diaphragm for additional defects, and concomitant evaluation and repair of intraabdominal pathology. One report describes a patient who underwent thoracotomy then subsequently experienced an intestinal obstruction 2 years later through the contralateral defect that was not visualized [75]. Also, in the event that the abdomen cannot accommodate the hernia contents, use of the abdominal approach allows the surgeon to repair the defect and leave a ventral hernia, which can be addressed at a later time [46]. A higher complication rate and a longer hospital stay after laparotomy may be explained by the fact that most emergency operations are performed through this approach (Table 3). The primary advantage of the thoracic approach

Fig. 4 Chest X-ray taken 10 days postoperatively with evidence of fluid accumulation in the unexcised hernia sac above the right hemidiaphragm

is that it provides easier dissection of the hernia sac off the mediastinal and pleural structures [43]. Despite the more widespread use of laparoscopy, thoracotomy continues to be the most widely used surgical approach for MH reported in the literature.

Management of the hernia sac is a key decision point for many surgeons, and their opinion on this issue may affect their surgical approach. The natural history of an unexcised hernia sac has not been studied extensively [10, 78]. Ramachandran and Arora [72] reported complete resolution of an unexcised sac by CT scan 1 month postoperatively.

However, during the same time interval, the patient studied by Contini et al. [79] still had a small residual cyst as seen by CT scan. Figure 4 is an example of a postoperative chest X-ray showing a fluid collection within the unexcised hernia sac. Many authors feel the hernia sac should not be resected [12, 19, 80, 81]. In children, dissection of the peritoneal sac has resulted in fatal cases of pneumopericardium [82]. Other surgeons prefer a more selective approach and recommend removing the sac only when it is small, without intrathoracic adhesions, and when the chance of injuring thoracic structures is small [72, 83, 84]. Still, some feel resection of the sac is safe and adhere more strictly to classic surgical principles [2, 60, 85]. This opinion may explain why thoracotomy continues to be the most widespread surgical approach.

Minimally invasive surgery has changed the way many surgeons approach MH. Only two cases of thoracoscopy have been reported in the English literature, so the following discussion is based primarily on laparoscopy. Although incarceration and even strangulation of hernia contents have been reported [48, 86, 87], hernia contents usually are not incarcerated and reduce without much difficulty [14]. To avoid tension, Thoman et al. [10] recommend using prosthetic mesh to cover defects larger than 20–30 cm^2. Others recommend using mesh repairs in all cases except those involving neonates [88]. Several different types of materials have been used: polypropylene, expanded polytetrafluoroethylene (ePTFE) (dual mesh; Gore Medical, Flagstaff, AZ, USA), Vypro (Vypro II, Ethicon Endosurgery, Cincinnati, OH, USA), Composix (C. R. Bard, Inc, NJ, USA), bovine pericardium (Tutopatch; Tutogen Medical, Neunkirchen, Bavaria, DE, USA), and Parietex (Parietex Composite; Sofradim, Trevoux, France), to name a few [29, 68, 89–92]. Another technique used by several authors to avoid cyst formation is to leave a drain in the hernia sac [24, 60, 72, 79, 84, 93].

The technique for laparoscopic repair of MH has been well described. The patient is placed in reverse Trendelenburg position and three to five ports are placed with an orientation similar to that for a laparoscopic Nissen fundoplication or other upper abdominal surgery. It is important not to place trocars too close to the costal margin [94]. Management of the hernia sac has already been discussed. Based on the opinions of several authors, achieving at least 1.5 to 2.5 cm overlap of the mesh is preferred [89, 91, 92]. Of those authors who have used polypropylene mesh to repair the defect, 29% (5/17) do not cover the mesh [25, 72, 95] because they think the round ligament, liver, and omentum provide ample protection for the bowel [12, 70]. This concept may not apply to left-sided hernias. However, tissue coverage of the mesh can be achieved with omentum [93], peritoneal flap [81,83], falciform ligamament/ligamentum teres [85], or nothing [25, 72, 95]. Mesh fixation can be performed with surgical tacks or various laparoscopic suturing techniques. Additionally, repair has been accomplished laparoscopically without pneumoperitoneum by using an abdominal wall-lifting device [96]. As with any laparoscopic case, the surgeon should be prepared to convert to an open procedure because cases of failed laparoscopy have been reported [48].

Conclusion

Based on our analysis, MH is a rare clinical entity that presents earlier in life for men than women. Patients usually present with symptoms, often of the pulmonary or pain variety. The differential diagnosis can be quite extensive, and advanced imaging technology may prove helpful. Surgical repair should be attempted for healthy individuals regardless of symptoms. Resection of the hernia sac is not universally performed. Laparoscopy is a safe surgical approach with a low morbidity and a short hospital stay. Surgeons familiar with laparoscopic techniques should consider this approach when treating patients with a Morgagni hernia.

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