Review Article

Natural history and non-operative treatment of paraesophageal hernias

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Abstract: Paraesophageal hernias (PEHs) are a commonly encountered, anatomical process that result from enlargement of the diaphragmatic hiatus. PEHs are characterized by projection of abdominal organs into the mediastinum. They may be asymptomatic and noted incidentally on imaging studies or present with a variety of symptoms, such as reflux, post-prandial fullness or discomfort, dysphagia, vomiting, regurgitation, cough or shortness of breath. The main components of a diagnostic work-up and characterization of PEHs are a contrast swallow study and esophagogastroduodenoscopy (EGD). Modern treatment paradigms involve surgical repair for symptomatic hernias or large asymptomatic hernias in those without significant operative risk. Asymptomatic patients require no specific treatment but are at risk for progression over time. Lifestyle modifications and medical treatment of reflux are important for management of mild to moderate symptoms. While the risk of acute symptom development is low, volvulus/obstruction is associated with significant worsening morbidity and mortality. Although surgical correction is the optimal treatment, when indicated, conservative or endoscopic management in high-risk surgical patients may be options. This, however, has a likely high rate of recurrent symptoms. Non-operative approaches include symptom control and watchful waiting in those without severe symptoms or high-risk surgical candidates. In severe cases, obstruction and organ ischemia from volvulus can occur, which require urgent intervention and surgery. Patients who present with these symptoms have a high rate of recurrence with non-operative management.

Keywords: Paraesophageal hernia (PEH); non-operative management; natural history; gastric volvulus

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Introduction

Paraesophageal hernias (PEHs), also known as hiatal hernias, are a common and important anatomical disease and represent a wide spectrum of variations and symptoms (1). They are characterized by projection of abdominal organs, most commonly the intra-abdominal esophagus and gastroesophageal (GE) junction, into the mediastinum through abnormal splaying of the diaphragmatic hiatus. Understanding the natural history and non-operative management is important for general and thoracic surgeons in this traditionally surgically managed disease. This review explores the classification, pathophysiology, diagnosis, natural history and non-operative treatment of PEH.

Classification

The nomenclature and classification of PEHs has changed over the last half-century. Terms such a “giant” or “sliding” have been replaced with a more anatomically defined system. Early literature with varying descriptions of PEHs has made it difficult to interpret and apply to patients. The modern

Table 1 Classification of paraesophageal hernia type

Type	Details
Type I	Gastroesophageal junction slides above the diaphragm
	Stomach remains in its usual position
	Fundus remains below the gastroesophageal junction
Type II	Gastroesophageal junction remains in its normal anatomic position
	Fundus herniates through the hiatus
Type III	Gastroesophageal junction and fundus herniate through hiatus
Type IV	Herniation of other abdominal organs through the hiatus +/– stomach

classification system (see *Table 1*) categorizes the herniation of the stomach and/or other abdominal organs in relation to the GE junction from type I to IV (2). Type I is defined as elevation of the GE junction above the diaphragm but does not involve herniation of the stomach or other organs. This is often described as a "sliding" hiatal hernia. Because type I hernias do not involve paraesophageal herniation, these are not true PEHs. Type II PEHs involve herniation of the fundus of the stomach next to the esophagus into the chest but the GE junction remains in its native position. This is a true paraesophageal hernia. Type III hernias involve herniation of both the GE junction and the fundus of the stomach through the esophageal hiatus and is often referred to as a "combined" hernia. Finally, type IV are those that include other abdominal organs such as the liver, colon, small bowel, or pancreas in conjunction with the stomach. Although not part of this classification, the extent or percentage of herniation of the stomach or involvement of other organs is helpful both for understanding associated symptoms, risks and potential difficulty of surgical repair (3).

Etiology and presentation

Although the pathophysiology of PEHs is not completely known, several studies have sought to understand the changes that occur at the hiatus. Biopsies of the gastrohepatic ligament, gastrophrenic ligament and phrenoesophageal ligament of those with PEH compared to control groups have found differences in the connective tissue matrix architecture, with higher ratios of type I collagen to type III collagen present in those with PEH (4). In addition, elastin fibers have been noted to be significantly reduced in the phrenoesophageal and gastrohepatic ligaments of patients with hiatal hernias compared to those without hiatal hernias with gastroesophageal reflux disease (GERD) symptoms (5). This may lead to relaxation of the ligaments and allow herniation of the stomach or abdominal contents through the hiatus. High intra-abdominal pressure with a relatively low intrathoracic pressure from obesity, pregnancy, or chronic cough can also lead to hernia formation over time. Additionally, PEHs can develop from bony changes such as degenerative disc disease and kyphosis with distortion of the hiatus (6).

There is no clearly identified genetic or somatic mutations that underlie the formation of PEHs. Most hernias are sporadic and are related to risk factors such as increased age, central obesity, and smoking (7). Hereditary clustering is unusual but can be seen in some genetic disease such as Down syndrome (Trisomy 21) or tissue collagen diseases such as Ehlers-Danlos and Marfan syndromes, which have increased incidences compared to healthy populations (8).

The vast majority of hiatal hernias are type I, which account for >95% of cases. As noted, these are not true PEHs, but are a common cause of GE reflux symptoms. Of the true PEHs, type III is the most common and account for >90% of these hernias, followed by type IV at 8% and type II at 2% (9).

With the increasing use of imaging and screening protocols, PEHs are commonly found incidentally on chest X-rays, computerized tomography (CT) scans and echocardiograms performed for other reasons (6). Most of these patients are asymptomatic, but a focused history may reveal symptoms related to the PEH extending back years that were overlooked or ignored. Others may present with symptoms including reflux, post-prandial fullness or discomfort, dysphagia, vomiting, regurgitation, cough or shortness of breath. Focused questioning to rule out concern for gastrointestinal (GI) bleeding, incarceration, volvulus, obstruction, strangulation and perforation should be performed. CT scans for other pathology that incidentally show small, asymptomatic PEHs typically do no need further imaging or testing.

Those with type I hernias may be younger and tend to have predominantly reflux symptoms (if symptomatic), whereas those with type II–IV tend to be older and have heartburn, chest pain/pressure, cough, early satiety, dysphagia, nausea, vomiting, weight loss, or anemia and may or may not have reflux (10,11). Chronic pneumonias from aspiration can be a common symptom, especially in the elderly. A high clinical suspicion is required for recurrent respiratory tract infections that cannot be explained by other pathologies.

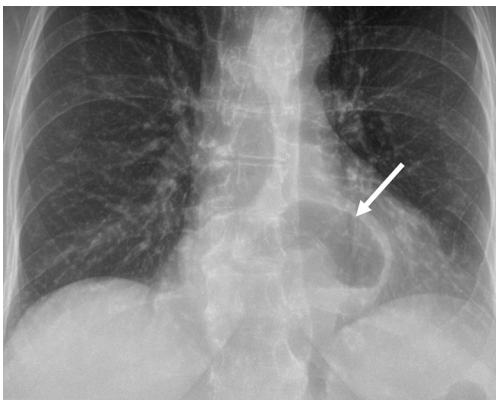

Figure 1 X-ray demonstrating retrocardiac lucency (arrow) from a type III paraesophageal hernia.

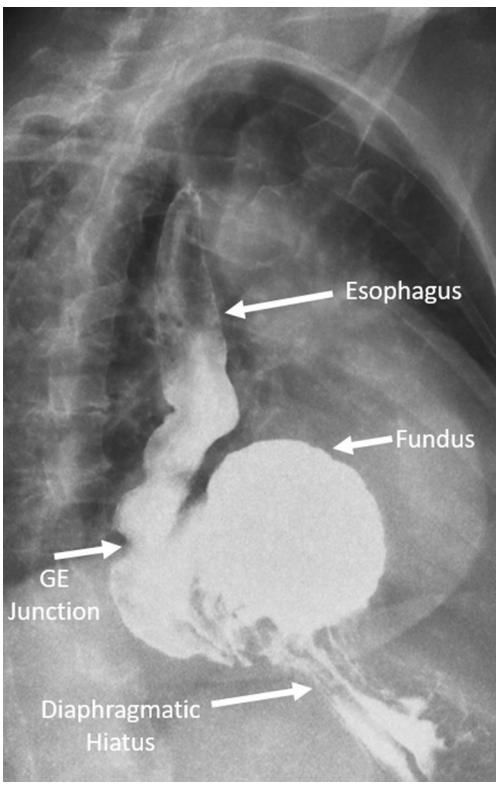

Figure 2 Barium swallow study demonstrating findings consistent with a type III paraesophageal hernia. GE junction, gastroesophageal junction.

Diagnosis

The main components of a diagnostic work-up and characterization of PEHs are a contrast swallow study and EGD. Chest X-rays may reveal a lucency in the retrocardiac space indicative of herniated stomach (see *Figure 1*) or pneumonia indicative of aspiration and these abnormalities are often the first clue to diagnosis. A chest X-ray alone, however, is not typically adequate in the work-up, as the differential diagnosis includes a mediastinal cyst, abscess, dilated/obstructed esophagus or other type of diaphragmatic hernia. CT scans of the chest and abdomen are helpful to understand the extent of herniation, accurately measure crural separation, and may be important in the acute setting. However, they are not always necessary in the elective setting, with contrast swallow studies and EGD being commonly used (12). Contrast swallow studies help classify the type of hernia by delineating the location of the GE junction and showing the extent of herniated stomach (see *Figure 2*). They can also help identify functional abnormalities of the esophagus, such as poor peristalsis or tertiary contractions, and may reveal reflux (either spontaneous or provoked).

EGD is an important diagnostic tool to help characterize the extent and type of hernia. This also provides the added advantage of visualizing the mucosa for ulcers, Barrett's esophagus or other GE pathology, like strictures or malignancy. Esophageal manometry and/or pH testing may be useful in the elective setting, particularly in determining the need for an anti-reflux procedure at the time of hernia repair, but are often abnormal and may not necessarily change the approach to the operation (13). They are not needed in the urgent/emergent setting for management.

Natural history and management

Truly asymptomatic type I hernias do not require either medical or surgical management (14). Controversy exists over the optimal management of asymptomatic or mildly symptomatic types II–IV hernias, given the historical concern for progression to acute symptoms. As such, individual patient factors including presence of symptoms, size of the hernia, risk of progression, and patient co-morbidities all play a factor in deciding when to offer an operation. Surgical repair of PEH alleviates presenting symptoms, but also prevents the rare development of gastric incarceration and subsequent obstruction, strangulation and volvulus (15). Mechanical ulceration of the gastric mucosa at the site of the stomach protruding through the hiatus (Cameron's ulcer) can lead to anemia and acute gastric bleeding. Cameron's ulcers were seen in <1% of a large study of all patients undergoing EGD, but noted in 1.2% of patients with a small (<3 cm hiatal hernia) to 13% of patients with a large hiatal hernia (>5 cm) (16). Persistent

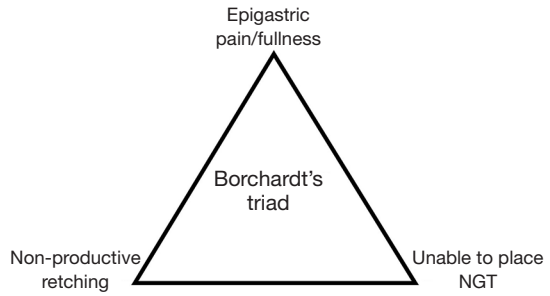

Figure 3 Borchardt's triad. NGT, nasogastric tube.

non-steroidal anti-inflammatory medication use may increase the incidence of Cameron's ulcers in PEH.

Obstruction can result when the stomach rotates around an axis along its mid-portion, resulting in a volvulus (6). Organoaxial rotation involves rotation of the stomach along the longitudinal axis on a line from the GE junction to the pylorus, leading to a closed-loop obstruction. A mesenteroaxial volvulus involves rotation along the transverse axis that bisects the greater and lesser curves. Mixed rotations along both axes can occur. Organoaxial is more common and typically presents with symptoms of retching with inability to vomit, epigastric pain and distention. Inability to pass a nasogastric tube in addition to these symptoms represents Borchardt's triad (see *Figure 3*) and signifies a high-grade gastric obstruction (17). Gastric ischemia and resultant necrosis can occur with volvulus formation, if not corrected, and lead to perforation and subsequent mediastinitis, sepsis and death.

There is a paucity of published contemporary data related to the natural history of symptomatic PEH without surgical correction, making it difficult to fully understand the untreated natural history of this disease process. Most surgeons generally agree that if left untreated paraesophageal hernias have the potential to enlarge and/or become symptomatic over time. The generally accepted rate of progression from an asymptomatic to a symptomatic hernia is approximately 14% per year (6). The risk of developing acute, severe symptoms requiring urgent surgery has been estimated at 1–2% per year (18,19). A more recently published retrospective review of 186 patients with "giant" PEH who were treated conservatively with an average follow up of 58 months noted that 64% had no change in their clinical course or hernia-related symptoms (20). In this study 21% developed progression of symptoms that could be managed conservatively, and 7% had elective surgery due to symptom progression. A hernia-related complication was noted in 8.1% of patients, with 2% having semi-elective surgery, 1% requiring emergency surgery, and 5% undergoing conservative treatment or endoscopic desufflation. While observation is a reasonable approach to the asymptomatic or mildly symptomatic patient (18), those with acute symptoms often require emergent repair with resultant higher rates of morbidity and mortality compared to elective surgery, particularly in elderly patients (21). However, historical rates of high mortality with emergent operation (>17%) are likely significantly over-estimated and likely closer to 5% (18). Of note, patients undergoing foregut surgery for PEH repair tend to have an associated higher morbidity with the risk of pulmonary, cardiac, and thromboembolic complications and 30-day mortality being significantly increased compared to those only having an anti-reflux procedure (22). Moreover, octogenarians undergoing PEH repair have a mortality of 2.4% with elective repair and 15.7% with "non-elective repair" (21).

Non-operative management options are limited for PEHs. They will not resolve and may have progressive symptoms. Lifestyle modifications such as weight loss, physical activity, avoidance of constipation or other sources of increased intra-abdominal pressure could be helpful in reducing symptom formation or progression. For those with mainly reflux symptoms, medical management of reflux with proton pump inhibitors or H2 blockers may be sufficient. In patients who present with more severe symptoms such as dysphagia, postprandial pain or mild/intermittent obstructive symptoms, surgical management is often indicated (23). However, for poor surgical candidates dietary modifications including smaller and more frequent meals may provide some relief. Patients with progressive dysphagia, significant weight loss or failure to thrive from their PEH who remain poor surgical candidates for definitive repair can be treated with feeding jejunostomy or parenteral nutrition to provide necessary caloric intake. Symptomatic anemia related to ulceration or GI bleeding should be treated with transfusions, anti-acid therapy and iron supplementation. Pneumonias from aspiration should be treated when diagnosed.

While relatively rare, patients with acute symptoms of volvulus can present with a combination of gastric obstruction, GI bleeding and/or gastric ischemia with or without perforation (24). These patients require immediate evaluation by a surgeon and should have large bore peripheral intravenous catheters placed with intravenous fluid resuscitation. A nasogastric tube should

be attempted to provide emergent decompression, but can often be difficult to do without endoscopic or imaging assistance (25). Emergent endoscopic decompression and directed placement of a nasogastric tube may be required. Following the above measures and appropriate resuscitation, patients who are candidates for surgical management should undergo repair (26). For those with prohibitive surgical risk, endoscopic decompression can provide relief of the acute symptoms, though the risk of recurrence is high (27). Percutaneous endoscopic gastrostomy or endoscopic/laparoscopic gastropexy can be helpful for those hernias that are anatomically favorable. This can provide gastric fixation and reduce the risk of rotation and also provide enteral access. A gastrojejunal tube may be needed to provide venting of the stomach and allow enteral feeds if significant reflux and aspiration is a concern.

A Finnish study demonstrated an in-hospital mortality of 16.4% for patients hospitalized with severe symptoms of PEH treated conservatively (28). Patients who present with acute volvulus with failed attempts of nasogastric decompression or who do not undergo endoscopic or surgical intervention rarely have spontaneous resolution. Progression to ischemia and gastric necrosis will likely occur with ongoing obstruction. For patients that decline or are unable to undergo endoscopic or surgical intervention, discussion of palliation with end of life goals should be done as mortality without treatment of an acute volvulus, especially with ischemia, is high.

Conclusions

Paraesophageal hernias have a broad spectrum of symptom presentation. Classifying these hernias is an important step in understanding the risks and treatment options. Endoscopy and upper GI contrast studies remain the best modalities for diagnosis. Surgical repair, as indicated by type and symptoms, is an important cornerstone of treatment. As a result, there has been little high-quality literature on the natural course and non-operative management of large, symptomatic hernias. Asymptomatic patients typically require no specific treatment but are at risk for progression over time. Lifestyle modifications and medical treatment of reflux are important for management of mild to moderate symptoms. While the risk of acute symptom development is low, volvulus/obstruction is associated with significant worsening morbidity and mortality. Although surgical correction is the optimal treatment, conservative or endoscopic management in high-risk surgical patients may be options. This, however, has a likely high rate of recurrent symptoms.

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Footnote

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Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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