Laparoscopic surgery for achalasia and other primary esophageal motility disorders (PEMD)—indications, preoperative investigations and patient's selection

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Abstract: Primary esophageal motility disorders (PEMD) including achalasia cardia are relatively rare disorders and diagnosed with relative accuracy using high-resolution manometry applying the new Chicago Classification v3.0. The role of additional investigations such as endoscopy and barium swallow play a vital role in the diagnosis of achalasia, however their inclusion in diagnosing other PEMD is of no value as the esophagus is anatomically normal. Surgical treatment in the form of Heller myotomy and partial fundoplication is considered the gold standard for achalasia but there is uncertainty regarding the surgical treatment of other PEMD even when the patient experiences obstructive symptoms of dysphagia due to unpredictable outcome. It is therefore imperative that the patient selection and preoperative counseling should take the lead role in these patients before embarking on the surgical treatment. In recent days Heller myotomy has been challenged by a newer endoscopic technique of peroral endoscopic myotomy for achalasia treatment. However, the long-term results are still not available and caution is required. This review scrutinizes both the new and old literature regarding the diagnostic features, preoperative investigations, indications and patient’s selection for the laparoscopic treatment of achalasia and other PEMD. It also aims to provide argument for and against various aspects of Heller myotomy in combination with antireflux procedures. It is hoped that further refinement of Chicago Classification may subgroup some of these PEMD and therefore provide clarity regarding the ones requiring definite surgical treatment for the improvement of their symptoms in long term.

Keywords: Esophageal motility disorders; achalasia cardia; Heller cardiomyotomy

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Introduction

Primary esophageal motility disorders (PEMD) are relatively rare motor disorders and may occur in the absence of gastroesophageal reflux disease (GERD). They present with specific manometric characteristics and classified as: (I) achalasia cardia (AC); (II) hypertensive lower esophageal sphincter (HLES); (III) Nutcracker esophagus (NE) or Jackhammer esophagus (JE) or hypercontractile or...
hypercontracting esophagus and (IV) diffuse esophageal spasm (DES) (1-3). These rare conditions of unknown etiology are being increasingly recognized with the use of high resolution manometry (HRM). However, their classification has been a source of much debate and the new Chicago Classification v3.0 (CC3) continue to provide much needed clarification on this subject (4). This review scrutinizes both the new and old literature regarding the diagnostic features, preoperative investigations, indications and patient’s selection for the laparoscopic treatment of achalasia and other PEMD.

**Achalasia and other PEMD—a brief overview**

**Achalasia**

Achalasia is by far the commonest PEMD with a prevalence of 10 per 100,000 individuals (5,6). It is characterized by absence of peristalsis and a defective relaxation of lower esophageal sphincter (LES) on HRM (7). Radiologically “bird-beak” or “rat tail” appearance, esophageal dilation and poor emptying on a contrast swallow study is evident. Endoscopically esophagus appears dilated with retained saliva, liquid and undigested food in the esophagus in the absence of mucosal stricturing or tumor suggesting impaired bolus transport through the LES (2,8). Histologically, the affected esophagus demonstrates loss of ganglion cells in its musculature effecting the esophageal motility resulting in dysphagia for both solids and liquids and loss of weight as a consequence. Although the exact etiology remains unclear, it is thought to be due to interplay between autoimmune and inflammatory processes in genetically susceptible patients (9). The symptoms vary from dysphagia to solids and liquids associated with regurgitation of bland undigested food or saliva and/or substernal chest pain during meals, weight loss, and even heartburn which often lead to erroneously misdiagnosis of achalasia as GERD.

**Hypertensive lower esophageal sphincter (HLES)/ esophagogastric junction outlet obstruction (EGJOO)**

HLES or EGJOO is defined as a resting pressure of the lower esophageal sphincter (LES) exceeding 3 standard deviations above the upper limit for normality (45 mmHg for conventional manometry (Richter, 2001 #2093) and 41 mmHg for HRM with normal peristalsis (10). Symptoms for HLES or EGJOO include chest pain and dysphagia (11). Multichannel intraluminal impedance demonstrated outflow obstruction at the LES but normal esophageal body bolus clearance (12). With HRM, EGJOO is characterized by preserved peristalsis in conjunction with an elevated integrated relaxation pressure (IRP) (13) of >15 mmHg (4,13). EGJOO may result from specific anatomic abnormalities (e.g., paraesophageal hernia, Nissen fundoplication, esophageal stricture, prior laparoscopic band placement and diverticulum) or functional variants or may be a prestige of classic achalasia (14). A recent retrospective analysis by Triadafilopoulos and Clarke (15) analyzing 478 HRM studies, found 116 patients with EGJOO and a vast majority of them had a coexisting motility disorder such as ineffective esophageal motility (IEM), DES or JE suggesting more work is needed to subgroup this disease especially with a view of distinguishing those cohort of patients requiring further treatment.

**Nutcracker or jackhammer or hypercontractile esophagus**

It is defined as esophageal contractions with high amplitude and normal peristalsis. The criteria include mean distal body contraction pressures exceeding 2 standard deviations above normal values (180 mmHg for conventional manometry) (1) and 216 mmHg for HRM along with normal IRP of <15 mmHg with hypertensive peristalsis (10,13). Based on CC3 (4), 20% distal contractile integral (DCI) should exhibit >8,000 mmHg-s-cm and normal latency in context of normal esophageal bolus transit. These patients present with chest pain more often than they do with dysphagia. Recent observations indicated a progressive nature of the disease in 25% of patients with further development to type II and III achalasia (16,17).

**Diffuse esophageal spasm DES**

According to CC3 the diagnosis of DES is considered in the presence of premature contractions defined as having a distal latency (DL) (essentially the time between upper esophageal sphincter relaxation and when the deglutitive contraction reaches the distal esophagus) of <4.5 seconds for 20% or more of wet swallows compared to rapid contractions defined as contractive front velocity (CFV) pf >9 cm/s (4). Therefore, DES is defined by the occurrence of ≥20% of premature contractions in a context of normal esophagogastric junction. Chest pain and dysphagia are the main symptoms.
Preoperative investigations and patients' selection

Manometry, barium swallow and endoscopy form the essential list of investigations in a suspected patient with achalasia and other PEMD prior to embarking on either a surgical or any other treatment. Unlike achalasia or DES, identification of other PEMD requires manometry because the esophagus is anatomically and radiographically normal. HRM has become the standard practice over the last 10 years compared to conventional manometry for diagnosing and classifying various types of achalasia. Additionally, HRM distinguishes achalasia from other forms of PEMD that were poorly recognized using conventional manometry. In a recent randomized control trial consisting of 245 patients with unexplained dysphagia, Roman et al. (18) showed that HRM was associated with a significantly higher detection rate of achalasia compared to conventional manometry i.e., 26% compared to 12%. In CC3, a diagnosis of achalasia is based on an elevated median IRP (13) in combination with failed peristalsis or spasm (4). The use of HRM has led to the subclassification of achalasia into three clinically relevant subclasses based on the pattern of contractility in the esophageal body; in type I, no pressure waves are recorded in the distal esophagus; type II is characterized by panesophageal pressurizations; whereas in type III, at least 20% of swallows reveal rapidly propagating or spastic simultaneous contractions (7). In addition, the RCT by Roman et al. (18) also demonstrated that with HRM the detection rate of non-specific motility disorders was significantly lower compared to conventional manometry.

Barium swallow, which has been used extensively, historically, provides less clarity in terms of differentiating between different esophageal motility disorders. It demonstrates a characteristic dilated proximal esophagus with a bird beak appearance distally with achalasia but that typical finding may not be appreciated every time. Furthermore, it is even less sensitive in demonstrating any abnormality with DES or JH. In JH, the barium swallow is most often normal. Almansa et al. (19) found that only 4% of the patients with DES proven on HRM had typical corkscrew appearance while majority had non-specific findings.

Endoscopy not only demonstrates features suggestive of achalasia as mentioned earlier, but also more importantly eliminates causes of pseudoachalasia such as malignancy, strictures or webs, eosinophilic esophagitis and hiatus hernia and therefore plays an important role as a diagnostic tool.

Surgical treatment—Heller myotomy with fundoplication

The main goal of treatment of achalasia or other PEMD is symptom relief and in case of dysphagia to establish an ability for the individual to tolerate solids and liquids. There exist medical, endoscopic and surgical treatment options. Medical therapy is reserved for elderly or those unfit for endoscopic or surgical treatment. In the recent years endoscopic treatment such as peroral endoscopic myotomy (POEM) has been said to obtain similar results to surgical treatment based on a short-term follow-up data (8,20,21). Nevertheless, the surgical treatment i.e., Heller cardiomyotomy (HM), remains the gold standard (22,23).

HM with fundoplication has been performed as the operation of choice for the treatment of achalasia (24,25). Various approaches have been reported; laparoscopy (26,27), laparotomy (28), left thoracotomy (29,30) and thoracoscopic (26,27). Currently laparoscopic approach via transabdominal route is a technique of choice and preferred by most surgeons (31) compared to a thoracoscopic approach. This is because it provides excellent exposure of GEJ so the myotomy could be extended for 2 cm on the gastric wall. Furthermore, it allows the formation of a partial anterior fundoplication i.e., Dor which is important to limit postoperative reflux. The open approach i.e., laparotomy, would be limited to patients who may not be able to tolerate pneumoperitoneum, in the event of complication during laparoscopic approach or rarely due to surgeons’ choice.

Although rare, the most effective surgical treatment, for HLES, JH and DES is still HM and partial fundoplication. Effects of myotomy and partial fundoplication reported by both Tamhankar et al. (n=16) (32) and Nastos et al. (n=16) (Nastos et al. 2002) showed that the surgical treatment for HLES resulted in complete resolution of symptoms (dysphagia or chest pain) in the long term in a substantial number of patients. Furthermore, the authors also noted complete satisfaction with the outcome of this surgery by their patients (30,32). However, the diagnostic accuracy and the surgical outcome of the so called esophageal spastic disorders in the above two studies based on conventional manometry has to be questioned due to its inability to properly diagnose and classify some of these patients who may not even be suffering from PEMD.

Patti et al. (31) reported the effects of myotomy in NE in 12 patients. They noted an improvement in dysphagia in 80% of their patients and in chest pain in 50% on a long term follow-up. The authors recommend a HM only
when dysphagia is the leading symptom and when LES or IRP pressure is above normal. Champion et al. (33) in their follow up study on patients undergoing a HM and fundoplication for NE reported a recurrence of dysphagia or chest pain in 75% of their patients indicating that those with no initial dysphagia may not benefit form surgical management.

Due to the functional obstructive nature of DES, a laparoscopic HM and partial fundoplication have been advocated in the past with good results between 70 and 95% in most reports (28,29,31,33-35). The largest reported series of a HM for DES comprising of 65 patients was reported by Henderson and Ryder over 3 decades ago (36). The patients were treated with long myotomy and different types of fundoplication through a thoracotomy. Reasonable postoperative GER control was obtained with partial fundoplication without dysphagia, whereas postoperative dysphagia was more of an issue after total fundoplication. Patti et al. (31) reported their results for 19 patients with DES where dysphagia was relieved in 80% of the patients after thoracoscopy myotomy and in 86% of patients after laparoscopic approach. Chest pain was relieved in 65% and 80% of the patients after thoracoscopic and laparoscopic approach respectively. Regurgitation and heartburn scores were also significantly improved after both approaches. Similarly, Eypasch et al. in their 15 patients with a mean follow up of 24 months (8 months to 13 years) (34) and Leconte et al. in 20 patients with a median follow up of 50 months (6–84 months) (28) found significant improvement for symptom scores regarding chest pain, dysphagia, regurgitation and heartburn after a long esophageal myotomy and antireflux procedure. The authors therefore concluded the beneficial effect of this surgical approach in selected patients with DES who were incapacitated with dysphagia and chest patient and unresponsive to conservative management.

**Extent of myotomy**

A myotomy is usually performed anteriorly avoiding the vagus nerve exposing the mucosa and submucosa (37). The length of myotomy on the esophageal side has varied in literature. It ranges from 4–8 cm (38-40). On the other hand, the myotomy on the gastric side is shorter i.e., between 1–4 cm and some have even argued for not crossing the LES (41,42). With the myotomy length quoted above, various studies have shown a significant relief of dysphagia in patients ranging from 82–98% (38,40-43). The consensus is that the extent of myotomy on the esophageal side should be guided by manometry and must be at least 1 cm beyond the proximal extent of abnormal contraction spanning the whole segment of a dysmotive esophagus (29). On the gastric side most authors agree that a myotomy of 1.5–2 cm across and below the LES is adequate (44). Attempts to exclude LES, has only resulted in significant number of patients developing recurrent dysphagia or achieving no relief from dysphagia in the first place (27,44).

**To wrap or not**

HM resolves dysphagia symptoms by dividing the distal esophageal muscular layers, thereby limiting lower esophageal sphincter contraction (43). Although the LES disruption along with myotomy will relieve dysphagia in 77–89% (45), it will also induce gastroesophageal reflux (GER) (25) ranging from 31–100%. A meta-analysis published in early part of 21st century recommended against routine use of a partial fundoplication surprisingly (46). In their analysis consisting of 15 individual studies without any comparative arms or randomization, the pathological reflux was identified in only 10% of patients without fundoplication compared to 7.9% in patients with a partial fundoplication based on a very small number of patients undergone ambulatory pH studies. The authors erroneously concluded that reflux is not necessarily eliminated with the addition of a partial fundoplication and they recommended against adding any antireflux procedure to laparoscopic HM. Since then many researchers have supported the role of fundoplication in controlling GER following HM (25,45,47).

Falkenback et al. (47) compared 20 patients with 10 undergoing a HM alone and 10 with a HM and Nissen fundoplication. They reported 100% pathological reflux on 24 hr pH study in those who had a HM alone compared to 25% in those who had a HM and Nissen fundoplication over a median follow up of 8 years. 60% of HM patients were noted to have esophagitis on endoscopy, including 20% with Barrett’s esophagus compared to none in a HM and Nissen group. The median percentage of time with pH of <4 was 13.1% in a HM group compared to 0.15% in a HM and Nissen fundoplication group. They reported on a single treatment failure in a HM and fundoplication group who needed esophagectomy at 3 years due to recurrent dysphagia.

Richards et al. (25), in their RCT compared 43 patients with 21 undergoing a HM alone and 22 undergoing a HM with Dor fundoplication. They reported significantly higher
risk of GER of 47.6% in patients without a fundoplication compared to 9.1% in those who had a fundoplication. The follow up, however was only 6 months, which is the main criticism of this study. At 6 months the median distal esophageal acid exposure time was significantly lower in the a HM plus Dor group (0.4%; range, 0 to 6.7%) compared with the a HM group only (4.9%; range, 0.1% to 43.6%). Dysphagia rates were not reported at 6 months. Recently the same group published their long term outcome with a follow up rate of only 66% of patients (48). At a mean follow up of 11.8 years outcomes after a HM alone and a HM plus Dor group for achalasia was reported to be comparable, but there was significant drop out of 34% of patients in their follow up. Median Dysphagia Scores and gastroesophageal reflux disease health related quality of life (GERD-HRQL) scores were slightly worse for a HM than a HM plus Dor patients but were not statistically different.

Campos et al. (45) who considered 39 studies of laparoscopic HM in their review reported high rates of symptom improvement following myotomy whether it was associated with fundoplication or not (90.3% vs. 89.9%) but observed a significant difference in GER rates (8.8% vs. 31.5%, P=0.001). In their regression analysis for the subset of articles providing 24 pH study results, they found that patients who had fundoplication in addition to a HM had lower rates of abnormal distal esophageal reflux compared to ones who did not have fundoplication (14.5% vs. 41.5%).

**Kind of wrap**

A partial fundoplication is usually performed in association with the myotomy (27,28,30,33,49), although some cases of total fundoplication (50) have been reported (26,30). Henderson et al. (35) compared the outcomes of 34 patients submitted to a HM and different types of fundoplication (Belsey, gastroplasty + partial wrap, gastroplasty + total wrap, and Nissen). Not surprisingly, better postoperative GER control was obtained with partial fundoplications, whereas postoperative dysphagia was more of an issue after total fundoplication at a follow period that varied between 5–10 years. Based on the clinical literature, it is now well established that total fundoplication such as Nissen is contraindicated as it can cause total dysphagia and therefore a partial wrap either posterior 270° Toupet or anterior 180° Dor is generally recommended (51). There are proponents and opponents of both types of partial fundoplication. Some authors have suggested that a potential disadvantage of the posterior approach is an angulation of the gastroesophageal junction, which may cause bolus obstruction and disruption of the periesophageal ligament and its attachments leading to reflux (52). However, others are of the opinion that Toupet may keep the edges of the myotomy separated therefore preventing stricturing of the myotomy scar and also provide better antireflux control (53,54). Advocates of the Dor argue that this procedure preserves the periesophageal ligament and attachments, thereby decreasing the risk of reflux, it is less complex to perform and covers the exposed mucosa, which is an advantage in case of either inadvertent micro or macro perforation (55,56).

There have been few comparative studies comparing Toupet and Dor fundoplication after HM since 2006 (38-41,54,57,58). The first comparative study by Richardson et al. (38) reported a significant difference in postoperative dysphagia between the groups; 20% Dor vs. 66% Toupet, however, it was a questionnaire based study and included even minor dysphagia. The only study to report any significant difference between the two groups when considering only significant postoperative dysphagia is the one by Wright et al. (17% for Dor vs. 5% in Toupet) (57). The most recent meta-analysis by Siddaiah-Subramanya et al. (59) analyzing seven comparative trials of Dor vs. Toupet partial fundoplications totaling 486 patients found that approximately 1 in 10, 8.5% in Dor and 9.1% in the Toupet group experienced treatment failure during the follow up period. Almost all significant dysphagia resolved with either endoscopic dilatation (70% of the study population) or surgical extension of myotomy (30%) further suggesting that it is the extent of the myotomy most like contributing to postoperative residual or recurrent dysphagia rather than the degree of wrap.

Treatment of achalasia related dysphagia has to be addressed in conjunction with postoperative GERD. Majority of the studies have utilized 24 hour pH study for assessing GER but some have made use of heartburn grading system (60) or health related QOL assessment scale (61) but all the studies have provided data on the number of patients affected by severe GER. Patients undergoing Toupet, with more fuller wrap, interestingly showed a trend towards experiencing higher rates of GER in the first ever comparative study by Richardson et al. (38) (20% Dor vs. 33% Toupet). Torres-Villalobos et al. (58) in their latest RCT have similarly shown at 6 months a significant better GER control in Dor group compared to Toupet (7% Dor vs. 34% Toupet). However, at 24 months there was no statistical difference was noted (10.5% Dor vs. 31.5% Toupet). Nonetheless other studies have shown
contradictory results. Rawling et al. in the first RCT comparing Dor vs. Toupet after a HM (52) reported 41.7% of Dor patients were suffering from GER compared to 21% in the Toupet group, while Kiudelis et al. reported a similar result (35% Dor vs. 11% Toupet). So what is the clinical relevance of the GER rates for these two procedures in terms of further treatment? The answer is none because no difference in intake of proton pump inhibitors or H2 blockers or antacids has been reported between the two groups. It is entirely possible that with the long term follow-up, the GER rates may achieve parity between these two types of partial fundoplications. Only objective analysis with ambulatory pH studies, will be able to differentiate the long term effects of these antireflux procedures post HM.

Conclusions

Primary esophageal motility disorders (PEMD) are uncommon conditions and present a challenge in terms of their surgical treatment and long term benefits except for achalasia. Although these motility disorders are diagnosed on HRM, on one hand, they may not have any significant functional or symptomatic consequences but on the other hand they may progress to something more sinister such as achalasia. What is challenging is how to detect those PEMD which will eventually require a surgical treatment in the long term? A surgical treatment therefore should be based on symptomatology and HRM and some of these conditions may require sequential HRM to see their progress. However, this strategy is expensive, carries complications albeit small and disliked by a vast majority of patients who have already experience HRM in the past. What is clear from the surgical literature e.g., in achalasia where dysphagia is the dominant symptoms, that surgery directed at obstructive symptoms has a tremendous health benefit and one would expect the same sort of results with other PEMD especially DES. However, the literature is sparse and unclear due to the rarity of some of these conditions and unpredictable surgical results. As far as relief of chest symptoms such as non-cardiac chest pain is concerned, does surgical therapy play any role? It seems a HM has an inconsistent effect based on a very limited surgical literature and the beneficial effects seems to be temporary. Furthermore, the myotomy may lead to weakened peristalsis leading to dysphagia that had not been present preoperatively. Nonetheless, minimally invasive surgical therapy for PEMD such as a HM with or without partial fundoplication has brought a shift in the treatment algorithm and is an attractive option but the results seem to be less satisfactory except for achalasia. As far as the obstructive symptom of dysphagia is concerned irrespective of type of PEMD, the extent of myotomy plays a key role in not only relieving the primary dysphagia but the recurrence of dysphagia. The length of myotomy should be at least 1 cm beyond the affected segment on the esophageal side and about 2 cm beyond the GOJ on the gastric side. Total wrap e.g., Nissen fundoplication to control GER produces unacceptable degree of dysphagia after a HM and therefore a partial wrap either anterior Dor or posterior Toupet is advocated which provide a satisfactory antireflux barrier without causing dysphagia in a vast majority of patients. This type of minimally invasive therapy has a proven track record in achalasia and considered to be a gold standard. Other minimally invasive modalities such as POEM are gaining acceptance for, not only achalasia, but other PMED, however no long-term results or any RCTs between POEM vs. HM are available and therefore presently the role POEM is being cautiously evaluated.

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Footnote

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