42 Other Esophageal Motility Disorders: Role for Laparoscopic or Endoscopic Myotomy

Francisco Schlottmann and Marco G. Patti

Clinical Presentation

Diffuse Esophageal Spasm (DES)

DES is defined by normal peristalsis intermittently interrupted by simultaneous contractions (simultaneous contraction >20% and <100%). It was first described by Osgood [1] in 1889, who reported a series of six patients with dysphagia and severe chest pain. DES is quite rare, with a prevalence of less than 10% in patients with dysphagia and/or chest pain and 3–5% in unselected patients undergoing esophageal manometry [2]. It presents with dysphagia (80%) and regurgitation (63%), followed by heartburn (51%) and chest pain (47%) [3]. Symptoms may occur during meals or physical exertion. Unlike achalasia, dysphagia is not progressive and weight loss is rare. Chest pain may mimic myocardial infarction, and it is usually described as a crushing or squeezing pain that can radiate to the jaw, arms, or back.

Nutcracker Esophagus (NE)

NE was first described by Benjamin and Castell in 1979 [4], and it is defined by peristaltic waves of very high amplitude and duration. Patients mostly complain of severe chest pain, while dysphagia, regurgitation, and heartburn are less common.

F. Schlottmann
Surgery and Center for Esophageal Diseases and Swallowing, University of North Carolina, Chapel Hill, NC, USA

M. G. Patti (✉)
Department of Surgery and Center for Esophageal Diseases and Swallowing, University of North Carolina, Chapel Hill, NC, USA
e-mail: marco_patti@med.unc.edu

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In fact, NE is the most frequent esophageal dysmotility disorder present in patients with non-cardiac chest pain [5].

Hypertensive Lower Esophageal Sphincter (HTN-LES)

HTN-LES was first described by Code et al. [6], and it is characterized by a hypertensive lower esophageal sphincter with normal peristalsis. Clinically, it is associated with dysphagia (71%) and chest pain (49%) [7].

Diagnosis

Barium Esophagogram

DES It may show indentations produced by dysfunctional muscle contractions that trap barium between contracted segments. This characteristic "corkscrew" appearance, however, is not specific for DES, and manometry remains the gold standard for the diagnosis of DES [8].

NE As all patients with NE have normal propagation of peristalsis, the barium swallow is often nondiagnostic.

HTN-LES It may show non-specific findings such as narrowing of the gastroesophageal junction with delayed esophageal emptying suggesting outflow obstruction.

Upper Gastrointestinal Tract Endoscopy

All patients with dysphagia should undergo an upper endoscopy in order to rule out malignancy. Clinical features related to malignancy-induced dysphagia are age over 60 years, presence of symptoms for less than 1 year, and weight loss over 20 pounds [9]. Findings suggestive of a primary motility disorder are found in 25% of the patients with DES, 0% of the patients with NE, and 50% of the patients with HTN-LES [3].

Conventional Manometry (Richter Classification) [8]

DES Simultaneous contractions ≥20% (but less than 100%) of wet swallows, intermittent peristalsis, and contraction amplitudes >30 mmHg.

NE Normal propagation of peristaltic waves, with a mean distal amplitude >180 mmHg, and duration >6 s.

HTN-LES Resting lower esophageal sphincter pressure >45 mmHg and normal peristalsis.

High-Resolution Manometry (HRM) (Chicago Classification) [10]

DES "distal esophageal spasm" ≥20% of wet swallows with distal latency (DL) <4.5 s and mean integrated relaxation pressure (IRP) <17 mmHg.

NE "hypertensive peristalsis" Mean distal contractile integral (DCI) >5000 mmHg/s/cm (but <8000 mmHg/s/cm which defines hypercontractile esophagus) and normal DL.

HTN-LES "EGJ outflow obstruction" Mean IRP ≥ 15 mmHg with normal or weak peristalsis (Table 42.1).

Multichannel Intraluminal Impedance

This test shows that patients with HTN-LES have outflow obstruction at the gastroesophageal junction but normal esophageal bolus clearance. Patients with NE have also normal esophageal bolus transit, while patients with DES present abnormal bolus transit [11].

Table 42.1 Manometric features of primary esophageal motility disorders

	Conventional manometry	HRM
DES	Simultaneous contractions >20% (<100%) Amplitudes >30 mmHg Intermittent peristalsis	DL < 4.5 s in ≥20% of wet swallows IRP < 17 mmHg
NE	Amplitudes >180 mmHg Duration >6 s Normal peristalsis	DCI > 5000 mmHg/s/cm in ≥20% of wet swallows Normal DL
HTN-LES	Resting LES pressure > 45 mmHg Normal peristalsis	IRP ≥ 15 mmHg Normal/weak peristalsis

HRM High-resolution manometry, *DES* Diffuse esophageal spasm, *NE* Nutcracker esophagus, *HTN-LES* Hypertensive lower esophageal sphincter, *DL* Distal latency, *IRP* Integrated relaxation pressure, *DCI* Distal contractile integral

Ambulatory pH Monitoring

When a manometric profile suggesting DES or NE is present, it is important to perform an ambulatory pH monitoring in order to exclude pathologic reflux. It is indeed known that these motility patterns can be due to abnormal reflux. If GERD is present, either medical or surgical treatment should be directed toward the control of the reflux [12].

Treatment

Since the cause of these disorders is unknown, treatment is directed toward symptom relief and improvement of esophageal emptying. Medical treatment, endoscopic treatment, and surgical intervention are the available modalities.

Medical Treatment

Treatments aimed to relax esophageal smooth muscle such as nitrates, calcium channel blockers, and antimuscarinic agents may be helpful. However, these drugs have modest effect on the resting LES pressure and do not improve LES relaxation in response to swallowing. Previous reports have shown inferior outcomes of medical treatment compared to surgery [13, 14]. Hence, pharmacologic treatment is of marginal clinical value and should be considered only in patients with mild symptoms.

Endoscopic Pneumatic Dilation and Injection of Botulinum Toxin

Symptom relief may be achieved only in patients who present with dysphagia as their main complaint and in whom manometry shows a hypertensive and nonrelaxing LES [15]. Injection of botulinum toxin in the distal esophagus acts by decreasing the release of acetylcholine by nerve endings of the myenteric plexus. In some patients it may improve dysphagia and chest pain [16].

Surgical Myotomy

Minimally invasive surgery has replaced open approaches to perform an esophagomyotomy. The operation can be done through a thoracoscopic or laparoscopic approach. While the initial experience was through a left thoracoscopic approach [17], the technique eventually switched into a laparoscopic myotomy with a partial fundoplication. Drawbacks of the thoracoscopic approach included the need for a double-lumen endotracheal tube, one-lung ventilation, right lateral decubitus, limited exposure of the gastroesophageal junction, postoperative discomfort, and a high rate of postoperative reflux. These problems were mostly eliminated with the

laparoscopic approach (single-lumen endotracheal tube, supine position, better exposure of the gastroesophageal junction, and ability to perform a fundoplication).

Laparoscopic Heller Myotomy and Dor Fundoplication Technique

The technique is similar to that of a similar operation performed in patients with esophageal achalasia [18]. In patient with NE or DES, the myotomy is usually extended more proximally on the esophageal body. The patient is in a supine position with legs placed in stirrups with knees flexed 20–30°. Five trocars are usually used for the operation.

We start by dividing the gastrohepatic ligament and identifying the right crus of the diaphragm and posterior vagus nerve.

Subsequently the peritoneum and phrenoesophageal membrane are divided, and the left crus of the diaphragm and anterior vagus nerve are identified.

The dissection should be continued into the mediastinum, lateral and anterior to the esophagus in order to expose 7–8 cm of the esophagus. No posterior dissection is needed if a Dor fundoplication is performed after the myotomy. The short gastric vessels are routinely divided.

The myotomy is performed using a hook cautery in the 11 o'clock position. In patients with the HTN-LES, the length of the myotomy is similar to that performed for patients with achalasia. For patients with NE or DES, the myotomy is extended more proximally, for about 8–9 cm proximal to the gastroesophageal junction, and then distally onto the gastric wall for 2–2.5 cm. The muscle edges are gently separated to expose the mucosa for 30–40% of the circumference.

A Dor fundoplication is then performed as previously described [19].

Outcome of Surgical Myotomy in Motility Disorders Different from Achalasia

Patti and colleagues [3] reported that in patients with DES, dysphagia and chest pain were relieved in 86% and 80%, respectively, after laparoscopic myotomy. In these patients the myotomy was usually extended more proximally than in patients with achalasia. Regurgitation was also significantly improved. Concordantly, Leconte et al. [20] reported significant improvement for dysphagia, pain, regurgitation, and heartburn in patients with DES after an extended myotomy and anterior fundoplication.

In patients with NE and chest pain, the results of surgery were disappointing with only 50% of patients experiencing symptomatic relief [3]. Dysphagia was instead improved in 80% of patients. Champion et al. [21] reported recurrence of symptoms (dysphagia or chest pain) in 75% of patients with NE submitted to myotomy and fundoplication. Overall, it seems that myotomy would be helpful only in patients with NE whose main symptom is dysphagia or when associated pathology such as an epiphrenic diverticulum is present.

Reports on myotomy for the treatment of HTN-LES have shown good results but are limited to a small number of patients [3, 22]. Tamhankar et al. [22] presented a long-term follow-up on four patients with complete relief of symptoms (dysphagia and chest pain) and complete satisfaction after the myotomy.

These data suggest that patient selection is of paramount importance. Most patients with DES and HTN-LES who complain of dysphagia improve after a myotomy. On the other hand, patients with NE whose main complaint is chest pain often do not have relief of the pain and can even develop dysphagia as a consequence of the myotomy [3].

Per-oral Endoscopic Myotomy (POEM)

In 2010 Inoue et al. reported the result of a new technique – per-oral endoscopic myotomy (POEM) in 17 consecutive patients with achalasia [23]. Since then, this endoscopic technique has been used in thousands of patients with achalasia in every continent, and most studies, albeit with a short follow-up, have documented very good results in more than 90% of patients [24–27]. Even though this technique was initially described for the treatment of achalasia, its indications have expanded to non-achalasia motility disorders such as DES, NE, and the HTN-LES [28–31].

POEM Technique

The patient is placed supine under general anesthesia. An overtube is placed, and the site for the anterior mucosotomy is selected by correlating with HRM parameters, usually 3–4 cm proximal to the upper border of the endoscopically visualized forceful esophageal contraction, in the 1 o'clock to 2 o'clock position on the ventral aspect of the esophagus. After injection of indigo carmine into the submucosal layer, a 1.5–2 cm longitudinal mucosotomy in the mid-esophagus is performed. A submucosal tunnel is then created with blunt dissection and carbon dioxide insufflation. The tunnel is extended past the esophagogastric junction for 2–3 cm onto the gastric cardia. A proximal to distal myotomy is performed with care to preserve the longitudinal muscle layers of the esophagus and stomach. Smooth endoscope passage through the esophagogastric junction, retroflexed evaluation of the valve, and a blanched gastric mucosa (distal dissection) indicate an adequate myotomy. The mucosal entry is then closed using endoscopic clips.

Outcomes of POEM in Motility Disorders Different from Achalasia

The largest series of non-achalasia motility disorders treated by POEM was described by Sharata and colleagues [32]. The authors studied the outcome of POEM in 25 non-achalasia patients with DES (5), NE (12), and HTN-LES (8) and

compared it to the outcome of POEM in 75 patients with achalasia. The study showed that dysphagia relief was better in achalasia patients (98%) than in non-achalasia patients (70%). Similarly, complete resolution of chest pain was seen in 100% of patients with achalasia but in only 75% of patients with other motility disorders. Post-POEM ambulatory pH monitoring showed abnormal reflux in 38% of patients.

Recently, Khashab et al. reported their experience with POEM for the treatment of spastic esophageal disorders refractory to medical therapy [33]. In this multicenter study (11 centers), 73 patients underwent POEM: 9 patients had DES, 10 had NE, while the remaining had type III achalasia. The mean length of stay was 3.4 days. A good clinical response was obtained in 100% of patients with DES, 96% of patients with type III achalasia, but in only 70% of patients with NE. Ambulatory pH monitoring showed pathologic reflux in 68.4% of patients. Hoppo et al. studied the utility of POEM across the spectrum of esophageal motility disorders [34]. The procedure was performed in 25 patients with achalasia and 8 patients with non-achalasia disorders. Median length of hospital stay was 3 days. At a follow-up of 7 months, dysphagia resolved in 92% of patients with achalasia and 75% of non-achalasia. Chest pain resolved in 100% of patients with achalasia and in 80% of non-achalasia.

Conclusions

Non-achalasia motility disorders are quite rare, so only a few centers have experience with their diagnosis and treatment. Few points that deserve special attention are:

- The symptoms and the manometric picture of NE and DES can be caused by GERD. Therefore, in order to have a diagnosis of "primary esophageal motility disorder," GERD must be excluded by pH monitoring. A cardiac evaluation should be routinely performed when chest pain is present.
- POEM is a relatively new procedure. As a consequence, there are no studies with long-term follow-up and no prospective and randomized trials comparing it to pneumatic dilatation or surgical myotomy.
- Many studies have shown that after POEM abnormal reflux is present in more than 50% of patients when measured objectively by pH monitoring [24, 33]. Furthermore, the multicenter study of Werner et al. of 80 patients with achalasia has shown that at a follow-up of 29 months, 3 patients had already developed Barrett's esophagus and 1 a peptic stricture [35]. The risk is that by performing POEM we might end up trading one disease process (achalasia, DES, NE, HTN-LES) with another (GERD). Contrary to what is commonly quoted by the authors of POEM studies, the incidence of GERD after myotomy and partial fundoplication is around 10% [36, 37].
- POEM has been advocated in these patients as it allows a longer myotomy onto the esophageal body. However, a long myotomy can be performed through a left thoracoscopic approach and a myotomy from the diaphragm to the thoracic inlet through a right thoracoscopic approach [3].

- Overall, we feel that the key to success is based on a complete evaluation and a careful patient selection. The best results, regardless of the technique, are in fact obtained in patients with outflow obstruction and impaired esophageal emptying, a picture similar to achalasia. In patients with NE, particularly if chest pain is the main symptom and esophageal transit is normal, the results are poor.

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