



Surgical therapy of esophageal motility disorders

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Introduction

Primary esophageal motility disorders (PEMD) were diagnosed in the past by barium esophagram (1), but modernly they are defined by specific manometric patterns that evolved with time and are currently defined by the Chicago 4 Classification (2). The concept of 'primary' assumes that the dysmotility is not secondary to a predetermined etiology with gastroesophageal reflux disease (GERD) as the most common of them, but also including systemic diseases, medications, eosinophilic esophagitis, etc. (3). Based on this assumption, surgeons have always emphasized the need for objective GERD evaluation in patients with suspected PEMD (4), but this was only acknowledged by the last Chicago Classification (5). Also, the new classification acknowledged that manometric findings are not clinically significant unless symptoms are present and sometimes a second test (e.g., barium swallow) confirms the diagnosis (2). There is important implication in this distinction as treatment should be directed towards GERD in secondary motility disorders not on the dysmotility *per se* (4).

Surgery is frequently not the first therapeutic option in patients with PEMD, except for achalasia, but good outcomes can be expected if surgery is well indicated (6). Initial enthusiastic results and the modern appeal to technology raised endoscopic therapy for PEMD as the primary alternative for several authors. New technology

is also available to diagnostics. Functional luminal imaging probe is a catheter-based measurement of the complacency of the distal esophagus with the purpose of assessing esophagogastric junction opening dynamics and the stiffness of the esophageal wall (7). This tool has been enthusiastically used to evaluate physiology before an operation and to guide myotomy adequacy during the operation; however, high-quality data are still scarce and conflicting results are present when compared to esophageal manometry (8).

We aimed to review current evidence on the value of surgery for PEMD.

Achalasia

Achalasia is defined by abnormal peristalsis and a defective relaxation of the lower esophageal sphincter or, in manometric parameters, an abnormal median integrated relaxation pressure (IRP) and 100% failed peristalsis (2).

Achalasia (*Figure 1*) is certainly the most understood PEMD. While there are several procedures described to treat achalasia, from medications to endoscopic to surgical, pneumatic forceful dilatation of the cardia, laparoscopic Heller's myotomy (LHM) associated to a partial fundoplication and peroral endoscopic myotomy (POEM) are certainly the most used and probably the best treatments for achalasia (9). Endoscopic dilatation, once the main choice for primary treatment, lost position for LHM (10).

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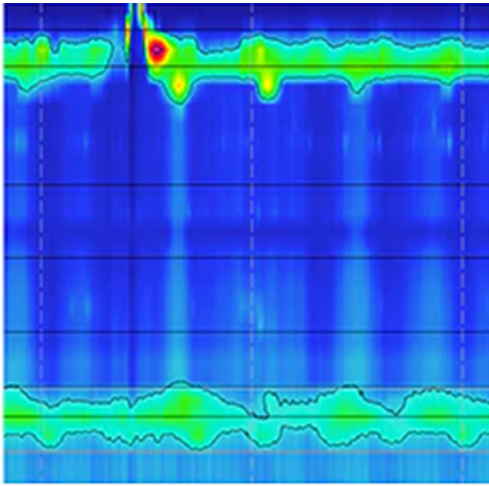


Figure 1 Manometric pattern for achalasia. Aperistalsis and defective relaxation of the lower esophageal sphincter. Example shows achalasia type I.

Even though both procedures bring excellent dysphagia relief, GERD is a critical issue for endoscopic therapy since lower esophageal sphincter defunctionalization is not followed by an antireflux procedure. Similarly, POEM relieves dysphagia, but it does not specifically prevent GERD although some argue that the privileged view of the muscles may allow preservation of the sling fibers of the sphincter (11). Nowadays, LHM and POEM are competing as the primary procedure for achalasia (12).

There are several recent meta-analyses in the last 3 years on the outcomes of different therapies for achalasia (*Table 1*). In general, POEM and LHM bring similar symptomatic relief with PD with lower efficacy. GERD is more incident in POEM.

Achalasia type III (*Figure 2*) deserves special considerations. Most authors believe that treatment should not be aimed towards the lower esophageal sphincter only but also to the esophageal body where the spastic contractions are generated. Thus, a long myotomy is necessary. Endoscopists argued that POEM should be the ideal therapy in these cases since the proximal extension of the myotomy can be easily stretched with the intraluminal approach (20). In fact, a meta-analysis showed superior outcomes for POEM versus LHM in these patients (21). However, a long myotomy can be done through laparoscopy (22). A careful analysis of the aforementioned meta-analysis show that a single study unfavorably viewed LHM for achalasia type III while the other did show superior or

comparable results to POEM. This unique study is the only one included that did not tailor myotomy's length based on the type. Thus, LHM is certainly an excellent alternative to type III as well.

Distal esophageal spasm

Distal spasm is defined by normal median IRP and $\geq 20\%$ swallows with premature/spastic contraction, i.e., short distal latency in the presence of symptoms and absence of GERD (2).

Distal esophageal spasm (*Figure 3*) comprises around 40% of the PEMD according to the most updated classification (23). It must be emphasized again that GERD must be excluded in the presence of this manometric pattern as almost half of the patients that fill criteria for distal spasm have GERD (21).

Primary treatment is usually based on medications that decrease the lower esophageal tonus or decrease peristalsis (24). Endoscopic or surgical therapy are usually left as rescue treatment.

There are no studies on LHM for distal esophageal spasm based on the new classification (25). Experience from older classifications (that probably incorporates most of the patients by the new classification) show symptomatic improvement superior to 85% (6). Some case reports and small series of POEM for distal spasm have been reported with promising results (25,26). A more challenging procedure due to reactive hyperactive spastic contractions during POEM has been reported (27). A meta-analysis of results was not possible since most studies did not report outcomes separately according to the PEMD (28).

Hypercontractile esophagus

Hypercontractile esophagus (previous nutcracker and jackhammer) is defined by normal median IRP and $\geq 20\%$ hypercontractile swallows, i.e., high distal contractility integral (DCI) in the presence of symptoms and absence of GERD (2).

Hypercontractile esophagus (*Figure 4*) comprises less than 1% of the PEMD diagnosed in a series of manometry (23).

Similar to the distal spasm, primary treatment is usually based on medications and endoscopic or surgical therapy are usually left as rescue treatment. Also, there are no studies on LHM for distal esophageal spasm based on the new classification (25). Experience from older classifications

Table 1 Meta-analysis from 2020 to 2023 comparing outcomes for different therapies for achalasia in adults.

Author, year	Number of studies included	Number of patients included	Outcomes for dysphagia relieve	Outcomes for gastroesophageal reflux disease
Gong 2023 (13)	27	2,278	POEM > LHM > PD	LHM > POEM > PD
Shiu <i>et al.</i> 2022 (14)	24	1,987	LHM = POEM > PD	LHM = POEM > PD
Dirks <i>et al.</i> 2021 (15)	28	2,368	LHM = POEM > PD	Not stated
Mundre <i>et al.</i> 2021 (16)	9	911	LHM = POEM > PD	LHM = POEM = PD
Facciorusso <i>et al.</i> 2021 (17)	6	745	LHM = POEM > PD	PD > LHM > POEM
Martins <i>et al.</i> 2020 (18)	12	893	LHM = POEM	LHM = POEM
Aiolfi <i>et al.</i> 2020 (19)	19	4,407	POEM > LHM = PD	LHM = PD > POEM

POEM, peroral endoscopic myotomy; LHM, laparoscopic Heller’s myotomy; PD, pneumatic dilatation.

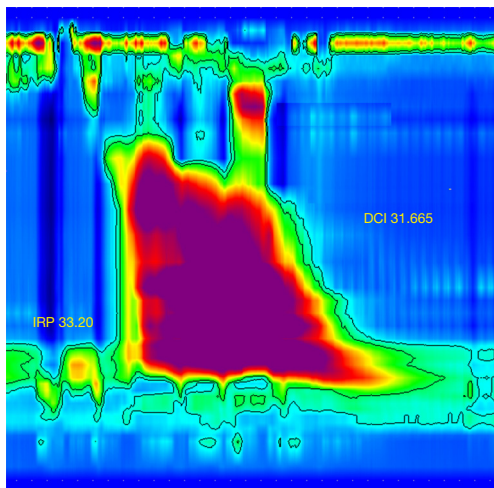


Figure 2 Manometric pattern for achalasia type III. Premature/spastic contraction and no evidence of peristalsis. DCI, distal contractile integral; IRP, integrated relaxation pressure.

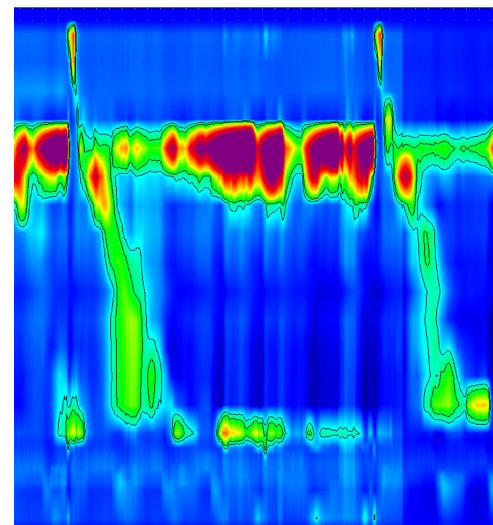


Figure 3 Manometric pattern for distal esophageal spasm. Premature waves in more than 20% of the swallows.

show symptomatic improvement superior to 80% and better outcomes in the presence of obstructive symptoms and elevated lower esophageal sphincter resting pressure or poor relaxation (6). Two recent systematic reviews compiling small series showed a clinical success of superior to 80% (28,29).

Esophagogastric junction outflow obstruction (EGJOO)

EGJOO is defined by normal peristalsis and a defective relaxation of the lower esophageal sphincter or, in

manometric parameters, an abnormal (IRP) (supine and upright) with $\geq 20\%$ elevated intrabolus pressure (supine), and not meeting criteria for achalasia (2).

Most cases of EGJOO (*Figure 5*) are asymptomatic, self-limited, or associated to mechanical obstruction especially after operations in the area (e.g., hiatal hernia, mediastinal mass, etc.). Few cases are considered PEMD.

Treatment should be cautiously indicated (30). Expectant management is recommended for patients with mild or atypical symptoms and there seems to be a limited role for medical treatment (31). LHM may be used in case of conservative treatment failure and in selected cases with

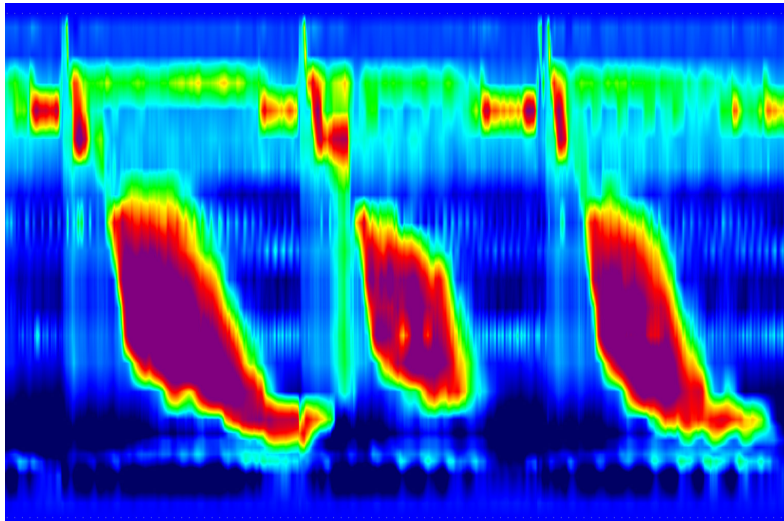


Figure 4 Manometric pattern for hypercontractile esophagus. Twenty percent or more of swallows with a DCI >8,000 mmHg-cm-s. DCI, distal contractile integral.

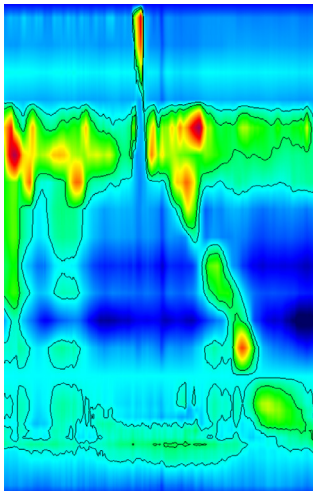


Figure 5 Manometric pattern for esophagogastric junction outflow obstruction. Normal peristalsis and a defective relaxation of the lower esophageal sphincter.

excellent results, comparable to achalasia (32). Small POEM series, including a prospective trial (n=15) and a multicenter study (n=15) show good outcomes (25,33,34).

Ineffective esophageal motility

Ineffective esophageal motility is defined by normal median IRP, with >70% ineffective swallows or $\geq 50\%$ failed

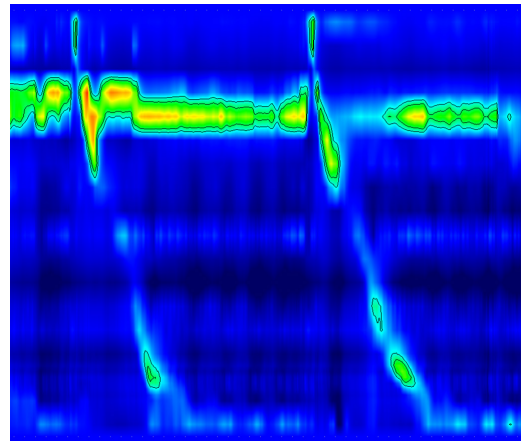


Figure 6 Manometric pattern for ineffective esophageal motility. Fifty percent or more of failed peristalsis or 70% or more of weak peristalsis.

peristalsis (2).

Ineffective esophageal motility (*Figure 6*) is the most common PEMD and the pattern most associated to GERD (21).

The treatment of hypotensive motility disorders is disappointing and relies mainly on dietary and lifestyle changes (35). Surgeons can rarely, if ever, act on this condition.

Conclusions

Surgery is frequently not the first therapeutic option in patients with PEMD, except for achalasia, but good outcomes can be expected if surgery is well indicated. We reviewed current evidence on the value of surgery for PEMD in cases of achalasia, distal esophageal spasm, hypercontractile esophagus, and EGJOO. Proper selection of patients is linked to good outcomes with low morbidity, which makes surgical therapy an adequate therapeutic option. If surgical therapy is not indicated, pharmacological therapy may be helpful even though results are not always good, side effects are common and medicine posology is usually inconvenient (36). Speech therapy rehabilitation may be helpful as well.

Achalasia, even type III, is best treated by LHM or POEM, although POEM is linked to a high incidence of GERD. LHM and POEM are competing as the primary procedure for other PMED; however, good-quality data are still elusive since classification recently changed, POEM is a relatively recent technology, and these diseases are not common.

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