



Expert opinion on the management of esophageal achalasia from the Society for Surgery of the Alimentary Tract (SSAT) Global Outreach Committee

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Abstract: Achalasia is the best studied primary esophageal motility disorder that we know most about its pathophysiology (though still evolving) and has appropriate treatments to relieve its most common symptom presentation (dysphagia). The disease diagnosis, however, demands suspicion and a complete workup. There are currently several options to treat the disease with pneumatic dilatation, peroral endoscopic myotomy (POEM), Heller myotomy, and esophagectomy as the main options. This review aims to compile experts' recommendations from the Society for Surgery of the Alimentary Tract (SSAT) Global Outreach Committee. This expert opinion paper summarizes the current evidence-based data and personal experience of a panel of experts for achalasia diagnoses and treatment. Upper endoscopy is essential to rule out malignancy. Barium swallow helps confirm the diagnosis and provides a baseline for esophageal diameter. Esophageal manometry is the gold standard for diagnosis and aids in prognosis and treatment selection. The routine use of endoluminal functional lumen imaging probes remains unclear. First-line treatment options include pneumatic dilatation, POEM, and laparoscopic Heller myotomy (LHM), which offer similar dysphagia relief but varying postoperative reflux outcomes. Botulinum toxin injection is a conduct of exception at present. End-stage disease may be best treated by LHM, while a dilated sigmoid esophagus poses challenges for POEM. Esophagectomy should be an option for patients who fail less aggressive treatments. In conclusion, diagnosing achalasia requires suspicion and a thorough workup. Several treatment options are available for non-advanced achalasia, including pneumatic dilatation, POEM, and LHM. These treatments offer similar outcomes in terms of dysphagia relief but vary in postoperative reflux results. End-stage disease is likely best managed with LHM + Dor (LHD).

Keywords: Achalasia; esophageal manometry; laparoscopic Heller myotomy (LHM); peroral endoscopic myotomy (POEM); end-stage achalasia

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Introduction

Background

Achalasia is often considered a rare disease, but its incidence varies globally. One US study reported 10–26 cases per 100,000 people annually (1), compared to 4.7 per 100,000 for esophageal cancer in the same country (2). Achalasia may be as common as esophageal cancer, yet it is frequently misdiagnosed as gastroesophageal reflux disease (GERD) and other benign and malignant diseases of the esophagogastric junction (EGJ), leading to treatment delays. Nearly half of achalasia patients in one series were on antacid medication, wrongly diagnosed with GERD (3). Misdiagnosis among surgeons during laparoscopic antireflux surgery may reach up to 4% of cases (4).

Rationale and knowledge gap

To address these issues, the Society for Surgery of the Alimentary Tract (SSAT) Global Outreach Committee organized a panel of experts at their 65th annual meeting to discuss achalasia management.

Objective

This review compiles their opinions and recommendations.

Methods

This expert opinion paper summarizes the current evidence-based data and personal experience of a panel of experts for achalasia.

The experts were lecturers invited by the SSAT Global Outreach Committee during the 65th annual meeting in Washington, DC, USA in May 2024. Topics were selected by the members of the Committee to include diagnoses and treatment. All presenters were requested to send their lectures that were compiled together and revised by the whole group.

Diagnoses

Symptoms

Symptoms can occur due to the functional obstruction of the esophagus and EGJ (dysphagia, regurgitation), its consequences (weight loss, halitosis, heartburn due to food fermentation and acid production in the esophagus), spastic

uncoordinated contractions (chest pain) or aspiration (cough). Dysphagia, regurgitation, and weight loss are, however, the common triad of symptoms in patients with achalasia. It is noteworthy that symptoms are common to other esophageal disorders including GERD. The Eckardt score is certainly the most used grading system (5) (*Table 1*). Eckardt scores range from 0 to 12, with a score of 3 or less considered ‘normal’ or minimally symptomatic.

Upper digestive endoscopy

Upper digestive endoscopy is essential when achalasia is suspected but does not diagnose the disease. It appears normal in half of the cases, while the other half may show indirect signs like food residue (despite adequate fasting), leukoplakia, or puckering at the gastroesophageal junction. The endoscopist may notice the esophageal dilatation in cases with marked dilatation. Resistance to passing of the scope may be noticed, even though the lower esophageal sphincter (LES) opens even in normal individuals only in response to swallowing what does not occur during sedation. This may raise the suspicion of pseudoachalasia due to a hidden malignancy at the EGJ.

Endoscopy mainly aims to rule out other conditions, especially esophageal cancer, since achalasia increases the risk of squamous cell cancers (7). The risk is, apparently, not decreased by therapy. Thus, endoscopic surveillance is advisable. The ideal interval between tests is, however, unclear. We believe that an endoscopy at least every 4 or 5 years is recommended.

Posttreatment, endoscopy may be necessary to evaluate dysphagia recurrence but especially to evaluate GERD that has a variable incidence after therapy as it will be discussed along the paper.

Barium esophagogram

A barium esophagogram is a straightforward test for diagnosing and grading the disease. A massively dilated esophagus with tapering of the distal portion (bird’s beak sign) usually does not have significant differential diagnoses. Minor dilatation without the bird’s beak sign may be observed in connective tissue disorders with aperistalsis (8) or in advanced GERD with aperistalsis. Less pronounced esophageal dilatation with tapering can result from tumoral obstruction and pseudoachalasia (4). Even when diagnosis is achieved through other methods, an esophagogram can serve as a baseline for comparison following therapy in

cases of treatment failure. Additionally, aside from aiding in diagnosis, the extent of esophageal dilatation may help grade the disease and identify end-stage conditions.

Timed barium swallow measures the contrast column after a set volume and time to evaluate esophageal emptying. It can help in unclear cases and is often used post-treatment to check if there is persistent outflow obstruction at the EGJ in case of dysphagia recurrence (9).

Esophageal manometry

Achalasia was defined during the conventional manometry era as a failure of complete relaxation of the LES and aperistalsis as identified as 100% of simultaneous or absent waves. High-resolution manometry did not change this definition even though LES relaxation is measured by a more complex and supposedly better parameter—integrated relaxation pressure (IRP)—and aperistalsis is measured by the presence of failed waves (10).

Table 1 Eckardt score for achalasia symptoms

Score	Dysphagia	Regurgitation	Chest pain	Weight loss (kg)
0	None	None	None	None
1	Occasional	Occasional	Occasional	<5
2	Daily	Daily	Daily	5–10
3	Each meal	Each meal	Each meal	>10

Each item is graded on a score of 0 to 3, with a maximum score of 12. Symptoms are frequently considered relevant with a final score >3. Based on reference (6).

Some Latin American authors believe in what they call an “undetermined” stage of achalasia. This stage is assumed to occur in patients with Chagas’ disease but depicting manometric findings other than the classical definition (unrelaxing LES and aperistalsis) assuming that this is an initial stage presupposing that they would eventually evolve to aperistalsis (11). In our experience, we have never seen such cases (12). Patients with Chagas’ disease may never develop esophagopathy and the manometric findings that do not characterize achalasia may be secondary to GERD (13).

A widely adopted classification is the Chicago Classification that defines three types of achalasia based on esophageal pressurization during swallows (*Figure 1*) (14). This classification gained acceptance due to the fact that it is linked to prognosis (15) and may guide therapy (16). Several variants have been identified, but it is unclear if they need the same or different treatments. Moreover, lack of dedicated studies leads to a very low level of recommendation (17). The Chicago Classification 4.0 presented 32 recommendations: 18 (56%) were strong recommendations while the other 14 (44%) were conditional recommendations or accepted clinical observations. The level of supportive evidence, using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) process, was moderate in 2 (9%) instances, low in 11 (52%), and very low in 8 (38%).

The group from the University of Rochester first described variants that challenged the concept of “all or nothing at all” to aperistalsis as a prerequisite for the disease (18). The group advocated that the disease may start in the LES and aperistalsis may be a secondary, metachronous finding.

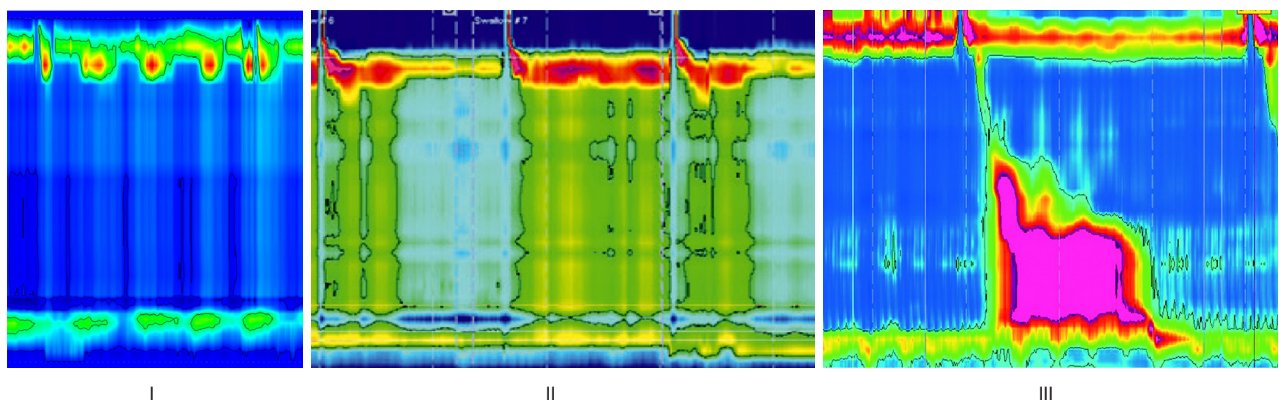


Figure 1 Chicago classification for manometric subtyping for achalasia. Type I achalasia: 100% failed peristalsis without PEP. Type II achalasia: PEP in at least 20% of swallows. Type III achalasia: at least 20% of swallows premature with no appreciable peristalsis. PEP, panesophageal pressurization.

They described the following variants: subtype 1, abnormal LES relaxation with normal/hypertensive peristalsis; subtype 2, abnormal/borderline LES relaxation with mixed peristalsis or simultaneous waves; and subtype 3, abnormal/borderline LES relaxation and aperistalsis with occasional short segment peristalsis. This classification, however, never gained popularity.

Chicago Classification (19) described variants that denote an inconclusive diagnosis and demand further investigation with other tests. They are: (I) absent contractility with no appreciable peristalsis in the setting of IRP values at the upper limit of normal in both positions, with or without panesophageal pressurization in 20% or more swallows; (II) appreciable peristalsis with changing position in the setting of a type I or II achalasia pattern in the primary position; and (III) abnormal IRP with evidence of spasm and evidence of peristalsis. Again, the evidence provided is low (17). Furthermore, normal IRP is frequently observed, occurring in over half of the cases in our series (20). This is compounded by the fact that surgeons might receive patients after an endoscopic treatment that disrupts the LES, thereby compromising its manometric analysis.

Provocative tests may be added to the test. This complementation is based on very low levels of evidence (17), with discrepant results (21), makes the test longer, losing one of the advantages of high-resolution manometry and leads to the battle between the findings of conflicting results—before and after provocation (22).

This increase in complexity from the simple aperistalsis to numerous variants leads to an interobserver agreement for the manometric diagnosis of achalasia ranging between 75% and 84% even including experienced interpreters (23–26). It is too soon to determine if artificial intelligence will improve these numbers (27).

Functional lumen imaging probe

Endolumenal functional lumen imaging probe (EndoFLIP) measures EGJ compliance. It is described by the Chicago Classification as a supportive test in cases of inconclusive diagnosis (19). It may identify abnormal EGJ distensibility in patients with typical achalasia symptoms but without classic manometric features of achalasia (28).

pH monitoring

GERD is paradoxical in treatment naïve cases since the lack of LES relaxation forbids flux and reflux. Pseudoreflux,

however, may occur due to intraluminal food fermentation and intrinsic acid production (7).

Other tests

Other tests to assess extraesophageal disease may be used if pseudoachalasia is suspected, such as endoscopic ultrasound and tomography (4).

Treatment

Peroral endoscopic myotomy (POEM)

POEM is the newest of the most common procedures on the continuum of achalasia treatment. Like other surgical interventions, it involves cutting the LES. Surgical division of the LES is the gold standard treatment of achalasia since Heller's open myotomy was first performed in 1913. This was transitioned to a thoracoscopic myotomy in 1992 and a laparoscopic myotomy in 1993. In 2010, Inoue presented his first series of POEM for achalasia (29).

POEM is indicated for disorders of outflow including all subtypes of achalasia. It can also be used for sole treatment in the setting of epiphrenic diverticulum (30). It has also seen success in disorders of esophageal peristalsis including distal esophageal spasm (DES) and hypercontractile esophagus (31). It is important to exercise caution in the use of POEM in these instances, as some cases of DES and hypercontractile esophagus are actually secondary effects of GERD (32). There are some cases where POEM has a distinct advantage over traditional approaches to esophageal myotomy. In cases where patients have had a prior myotomy, doing a peroral myotomy on the opposite side of the esophagus can be a relatively scar-free plane with acceptable success and lower morbidity than an attempt at redo laparoscopic or thoracoscopic myotomy (33).

In type 3 achalasia, some argue that there is a superior rate of symptom improvement for POEM as compared to traditional myotomy (34). A meta-analysis concludes for this premise (16); however, the evidence underpinning this is quite weak—cohort studies with short-term follow-up are often compared with longer-term outcomes following myotomy and an outlier in the meta-analysis did not use an extended Heller myotomy affecting final result. Finally, in cases where patients have had prior foregut surgery, an endoscopic approach to myotomy can be more straightforward.

In comparison to surgical myotomy, POEM has

comparable success rates (35). Thus, most gastroenterology and gastrointestinal surgical societies advocate that POEM is a reasonable option for patients in a setting with appropriate local expertise and if patient preference dictates. Overall, there has been persistent use of surgical myotomy for achalasia over time, but there has been a notable increase in POEM usage.

In comparison to nonsurgical treatments like pneumatic dilation (PD), randomized controlled trials have shown higher success rates with POEM over PD (92% as compared to 54%) with a lower rate of adverse events (0% *vs.* 7%). This is likely due to the more controlled nature of the circular muscle division in POEM. The GERD rate after POEM, and consequent need to long-term medication usage, is somewhat higher than after PD, unsurprising as it is more effective in relaxing the esophagogastric barrier for symptom relief in achalasia (36).

In comparison to Heller myotomy, POEM is equally successful in randomized controlled trials. In a study by Werner *et al.* clinical success rate for POEM and Heller plus fundoplication were comparable (83% *vs.* 81.7%) with a similar rate of adverse events (2.7% for POEM and 7.3% for Heller) (35). Again, reflux rate was somewhat higher after POEM at 44% compared to Heller's 29%, but this was not statistically significant. This randomized controlled data cemented POEM as a mainstay in the armamentarium of esophageal surgeons. More recent studies have also concluded that GERD rate over time decreases with POEM (37).

Inadequate myotomy has long been one potential risk of surgery for achalasia. Often, this is inadequate extension of myotomy onto the stomach. In the case of POEM, adjunct intraoperative testing like use of a second scope within the lumen to verify tunnel length can decrease the likelihood of incomplete tunneling and incomplete myotomy (38). Additionally, use of EndoFLIP during POEM can help target an esophageal diameter and distensibility index (DI) that is more likely to lead to clinical success (39). Current EndoFLIP parameters during POEM are guided based off of cohort data as well as studies evaluating these parameters in normal subjects (40). Early EndoFLIP data during POEM was collected using EndoFLIP 1.0. This found that DI of 4.5–8.5 was optimal (39). However, more recent studies using the updated EndoFLIP 2.0 system have shown that a DI less than 3.1 correlates with treatment failure (41). The same research group found that a DI of over 2.7 and/or a diameter over 11 mm is more likely to develop esophagitis (42). The conclusion for this early EndoFLIP data in POEM is

that there is an overall lack of standardization of parameters to determine adequacy of myotomy with EndoFLIP. However, in general there is evidence that the use of EndoFLIP is associated with better outcomes. Over time, the use of EndoFLIP may allow the length of myotomy to become shorter with comparable symptom improvement. There is evidence that a shorter endoscopic myotomy can effectively treat achalasia with equivalent rates of success, but with a lower risk of GERD and esophagitis (43,44). This provides an exciting opportunity to continue to optimize POEM for achalasia.

One of the challenges of POEM is the steep learning curve some experience. In general, endoscopists who have robust interventional practice are able to adopt POEM with a learning curve that varies between 13 and 20 cases (45-47). Longitudinal training with an expert operator can also lead to successful adoption, and fellowships in minimally invasive surgery as well as third space endoscopy have the potential to train surgeons and gastroenterologists to perform POEM. Current formal training pathways include a general surgery residency plus a minimally invasive fellowship with an endoscopic focus, a medicine residency plus a GI fellowship followed by an advanced endoscopy fellowship, and a thoracic surgery residency plus advanced endoscopy fellowship. Informal training pathways outside of fellowship are numerous and include a practice of interventional endoscopy plus ideally ESD, plus didactic training and hands-on skill sessions through surgical and gastroenterology societies, plus proctored early cases. These training pathways are in evolution as the technical worlds of GI surgery and interventional endoscopy continue to develop. What is likely to continue to be true is that POEM is a mainstay for achalasia treatment, is equivalently effective when compared to Heller myotomy and is largely superior to PD. Reflux after POEM is a continuous debate and lacks good study including pH monitoring. Finally, tailoring POEM with EndoFLIP is likely to improve outcomes.

Laparoscopic Heller myotomy (LHM)

Surgical treatment for esophageal achalasia was introduced by Heller in 1913, consisting of an anterior and posterior myotomy across the LES (48), but the current technique was modified in 1923 by Zaaijer to simply an anterior myotomy (49). LHM was first described by Shimi *et al.* in 1991 (50). With the minimally invasive surgery revolution, the Padova group was the first to propose the laparoscopic myotomy with the addition of a Dor fundoplication

[LHM + Dor (LHD)] (51). In a few years, LHD seems to have completely changed the achalasia management algorithm and it has rapidly become the procedure of choice for treating this disease. So far, a 6–7 cm anterior myotomy is performed, extending it 2–3 cm onto the proximal stomach. Early middle success rates with LHD have been high, around 90% (52,53) and the first 20 years follow-up study on a large cohort showed that >80% of patients were still symptom-free (54).

Looking at the available randomized trials and knowing that the definition of failure and when to recommend re-treatment remains ill-defined (55). One can clearly see that the outcome of LHD is similar than POEM and PD but with a lower incidence of intraoperative and long-term complications (35,56).

According to recent studies examining large cohorts of achalasia patients, the presence or absence of a sigmoid esophagus and the pre-operative manometric patterns represent the strongest predictors of outcome in terms of dysphagia and food-regurgitation relief (35,54–56). These data confirm the importance of a detailed pre-treatment work-up to define the patient's radiological grade (57,58) and manometric subtype (59–61).

A recent study has demonstrated that the LHD myotomy with the “pull-down” technique is an effective treatment in more than 90% of patients with end-stage diseases and it should be the first surgical option offered to this difficult group of patients before considering esophagectomy (62).

Patients with manometric types I and II have better clinical outcomes after LHD than those with type III (55,59,60). Moreover, manometric studies showed that the LES is longer in type III achalasia, and that extending the myotomy both downwards and upwards could improve the outcome in this cohort of patients (63,64).

It is also not clear if age should therefore be considered a factor in the choice of treatment for achalasia. Curiously, most specialized centers do not tailor treatment based on advanced age (65). While two recent studies showed that LHD can be used as the first (and often only) therapeutic approach to achalasia in elderly patients with an acceptable surgical risk (65,66), endoscopic dilatation is also a possibility (65), but botulinum toxin injection offers unacceptable results even though it looks attractive in the elderly.

Moving to the point of view of children, recently the Padova group published the 25-year experience of LHD in pediatric patients and they concluded LHD is a safe and long-term effective treatment even in the pediatric age,

with a success rate comparable to that usually obtained in the adult population (67).

LHD-related morbidity is very low. The main complications associated are mucosal perforations, which occur in 2–7% of patients (52,54). Most of them are detected during the procedure, however, and repaired intraoperatively, with no further clinical consequences or influence on the outcome (68). The procedure carries a really small mortality risk and is not correlated with the surgical procedure, so a careful patient selection needs to be done during the pre-treatment assessment.

The main long-term complication reported after LHD is GERD. In a recent report on a large series of 1,001 patients treated with LHD and assessed with an objective evaluation after surgery (24-h pH monitoring) only 9.1% of patients showed a pathological distal esophageal acid exposure (54). Comparing this complication between the other available treatments, in a recent Italian propensity score match trial, the incidence of GERD was statistically lower after LHD (17%) than after POEM (38.4%) (69). While the European trial showed no difference in distal esophageal acid exposure at 10 years follow-up between LHD and pneumatic dilatation (56).

In conclusion, LHD can durably relieve achalasia symptoms in more than 80% of patients at 20 years after treatment, comparable or superior to other treatment options (*Table 2*). Complications of surgery are rare, and postoperative reflux occurs in less than 10% of patients. The pre-operative manometric pattern and the presence of a sigmoid esophagus need to be assessed to appropriately modify the LHD: a longer myotomy (in pattern III) or the pull-down technique (in end-stage disease).

Diagnosis and treatment of end-stage achalasia

The definition of end-stage achalasia is still a matter of debate. Most of the authors rely on the assessment of radiographic features. In 2018, the International Society for Diseases of the Esophagus (ISDE) published a consensus guideline on achalasia, and specifically recommended barium esophagogram as the defining study in characterizing end-stage achalasia (57). The diagnostic radiographic feature is a massive dilation and tortuosity, which is termed sigmoid esophagus (*Figure 2*) (74). Manometry may be difficult to perform in the setting of end-stage achalasia due to dilation and tortuosity of the esophagus and retained debris that may lead to catheter looping or kinking without crossing the LES (57,75). Thus,

Table 2 Treatment outcome of randomized controlled trials in achalasia patients

Authors	Year	Patient, n	Follow-up (months)	Treatments (A vs. B)	Outcome A (%)	Outcome B (%)	P value
Novais (70)	2010	94	3	PD vs. LHD	73.8	88.3	0.08
Persson (71)	2015	39	78	PD vs. LHT	61	88	0.025
Werner (35)	2019	221	24	POEM vs. LHD	83	81.7	n.s.
Sediqi (72)	2021	43	120	PD vs. LHT	64	92	0.016
Kuipers (73)	2022	125	60	PD vs. POEM	40	81	<0.01
European trial (56)	2024	76	120	PD vs. LHD	74	74	n.s.

LHD, laparoscopic Heller myotomy + Dor; LHT, laparoscopic Heller myotomy + Toupet; n.s., not significant; PD, pneumatic dilatation; POEM, peroral endoscopic myotomy.

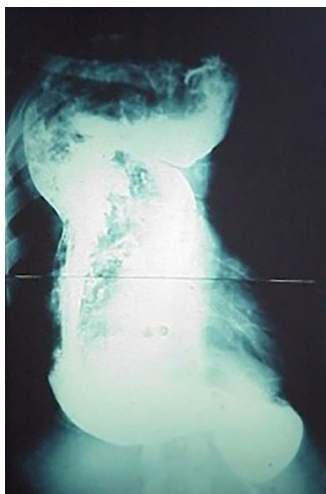


Figure 2 Barium esophagogram shows a massive dilation and tortuosity (sigmoid esophagus).

a megaesophagus with sigmoid shape and distal esophageal sump would widely be considered representative of the end-stage achalasia.

A descriptive summary of current treatment and the disadvantage of each approach is presented in *Table 3* (76).

The primary role of treatment in achalasia patients is to palliate symptoms, most commonly through disruption of the LES and allow gravity-dependent esophageal emptying. Moreover, the treatment for end-stage achalasia includes also reducing risk of aspiration and improvements of nutritional deficiencies related to the non-functional esophagus (76).

Endoscopic approaches such as PD or temporizing paralysis of the LES with botulinum toxin may be employed, but more definitive treatment with LES myotomy is preferred in candidates suitable for general

anesthesia. The two most common approaches currently are minimally invasive Heller myotomy (LHM), which is typically performed transabdominally with concurrent partial fundoplication, and POEM. Megaesophagus and sigmoid esophagus are also atypical initial presentations of achalasia, and patients with end-stage disease have often failed one or more of these initial treatment methods (74,76,77).

There is no consensus on which surgical therapeutic approach should be used for the end-stage achalasia treatment. However, most experts agree that the most definitive treatment for achalasia is esophagectomy, and esophageal resection should be considered in patients with end-stage disease or progression that has failed a less aggressive approach, including LHM or POEM (74,76,78). Much of the literature is retrospective, relying on expert opinion, case series, and individual discussions with patients, based on treatment successes in high-volume esophageal centers.

Heller myotomy

LHM is suitable for treating achalasia, even with sigmoid or megaesophagus. The minimally invasive transabdominal approach focuses on the distal esophagus, LES, and cardia while avoiding most of the redundant intrathoracic esophagus (76,79,80).

A pull-down technique was introduced for certain patients with sigmoid esophagus to align the esophageal axis. By encircling the gastro-esophageal junction with a string (easy flow/Penrose drain), approximately 10 cm of the lower mediastinal esophagus is isolated. Multiple stitches are placed on each side and then tied to secure the esophageal wall to the diaphragmatic pillars. Once the

Table 3 Current treatment options for end-stage achalasia

Intervention	Disadvantages
PD	Risk of perforation, sepsis, and bleeding
Botulinum toxin	Temporary effects (3–6 months): fibrosis
POEM	Higher risk of GERD; technically challenging in a sigmoid esophagus
Minimally invasive Heller myotomy	Risk of complications; improperly constructed fundoplication, fibrosis
Stapled cardioplasty	Intractable GERD; persistent regurgitation
Esophagectomy	Significant morbidity and mortality; high-volume centers; acceptable QoL
POPE	Risk of intraluminal injury; novel approach without long-term follow-up

GERD, gastroesophageal reflux disease; PD, pneumatic dilation; POEM, peroral endoscopic myotomy; POPE, peroral plication of the esophagus; QoL, quality of life.

esophageal axis is verticalized, the LHD is performed as previously described (63,81).

Salvador *et al.* [2023] (81) studied 73 patients diagnosed with end-stage achalasia with a sigmoid esophagus treated with LHM. Of the 73, 23 had previous endoscopic treatment. They showed satisfactory results in 71.2%. Regarding the failures (21 patients), five patients refused additional treatment, while all the others underwent endoscopic PD as the first retreatment, with a success of 56% (9/16). No dilation-related complications were recorded. Further treatments for persistent failures included: a redo myotomy in four patients, a Botox injection in one, POEM in one, and esophagectomy in one.

A recent systematic review also showed good outcomes for LHM in end-stage achalasia (82).

POEM

POEM is a widely accepted initial approach to achalasia; however, there is some discussion involving its efficacy in the setting of end-stage achalasia (64–73% clinical success rate) (83,84). The loss of the organ's axis and dilation significantly increase the complexity of the technique and the likelihood of complications.

A major concern following myotomy is the development of reflux disease. In particular, the rates of reflux after POEM have been reported to be nearly double those after LHD at 44% (35).

Esophagectomy

Esophagectomy for benign disease is a topic of discussion over the years. However, esophageal resection is considered

definitive management for a non-functional esophagus. The restoration of alimentary continuity using gastric or colonic interposition aims to improve emptying and subsequently reduce the risk of weight loss or aspiration (78,85). The goal of esophagectomy for end-stage achalasia is to remove esophageal obstruction and the inert reservoir creating a direct path for food and liquid to enter the stomach.

When performing an esophagectomy, there is significant variability in the approach (minimally invasive *vs.* open, transabdominal, transthoracic, or a hybrid approach) as well as in the choice of interposition conduit to restore alimentary continuity. One common option for interposition is the use of the stomach as a conduit, relying on the blood supply of the gastroepiploic artery to perfuse a surgically narrowed stomach that is positioned through the mediastinum or in a substernal position and anastomosed to the proximal esophagus. This choice of conduit requires a single anastomosis between the stomach and esophagus, and the intra-abdominal position of the stomach allows for relatively straightforward transposition into the thoracic cavity (12,13,76–78). While colon interposition is an option, achalasia is non-malignant and typically does not affect life expectancy. Over time, interposed colons may dilate and cause food and liquid stasis, replicating the issues of end-stage achalasia.

This surgical procedure presents several challenges, including: sigmoid deviation of the esophagus into the right chest; transmural inflammation causing adhesions to the pleura, trachea, and crura; neovascularization of hypertrophied esophageal muscle leading to bleeding and potential reoperation; and prior fundoplication resulting in abdominal adhesions that shorten the gastric conduit (4–6,85,86). In high-volume surgical centers, the overall

rate of morbidity ranges 30–60%, with operative mortality of 3–5% (74,77).

Esophagectomy and gastric transposition are safe procedures with acceptable morbidity, mortality, and reasonable postoperative clinical results in benign diseases, representing the definitive and last option in the management of end-stage achalasia. However, this type of reconstruction after esophageal resection causes modifications in anatomy and physiology on the upper gastrointestinal tract. Resection or disruption of natural antireflux mechanisms, esophageal-gastric direct anastomosis, pyloric drainage, impairment of gastric motility, recovery of acid secretion from gastric conduit, impaired motility of esophageal remnant all contribute to esophageal stump mucosal damage (13). Gastric and duodenal refluxate have been documented in 60–80% of esophageal resected patients. Long-term follow-up has revealed some late complications: (I) increasing incidence of esophagitis and Barrett's esophagus in the esophageal stump; (II) diffuse gastritis of the transposed stomach; and (III) development of carcinoma in the esophageal stump (87,88).

The pathogenesis of these three late complications can be attributed to the following factors: (I) resection of the EGJ; (II) recovery of gastric acid secretion over time after surgery; and (III) development of Barrett's epithelium following esophagectomy and gastric tube reconstruction (86,88).

da Rocha *et al.* (87) have studied 101 patients who underwent esophagectomy and cervical gastropasty and were followed-up prospectively for a mean of 10.5 ± 8.8 years. All patients underwent clinical, endoscopic, and histopathological evaluation every 2 years. Gastric acid secretion was also assessed. The incidence of esophagitis in the esophageal stump (45.9% at 1 year, 71.9% at 5 years, and 70.0% at 10 years follow-up); gastritis in the transposed stomach (20.4% at 1 year, 31.0% at 5 years, and 40.0% at 10 or more years follow-up), and the occurrence of ectopic columnar metaplasia and Barrett's esophagus in the esophageal stump (none until 1 year, 10.9% between 1 and 5 years, 29.5% between 5 and 10 years, and 57.5% at 10 or more years follow-up), all rose over time. Gastric acid secretion returns to its preoperative values 4 years postoperatively. Esophageal stump cancer was detected in the setting of chronic esophagitis in five patients: three squamous cell carcinomas and two adenocarcinomas. Barrett's esophagus is also a common finding (89).

Peroral plication of the esophagus (POPE)

POPE is a technique that uses an endoscopic suturing device to plicate the esophagus in a distal to proximal manner to eliminate the distal esophageal “sump” and facilitates emptying. This procedure is still experimental and should be performed by a skilled endoscopist in a high-volume center under clinical research scrutiny. POPE may offer some advantages for refractory sigmoid esophagus. It is important to note that full-thickness bites may be taken with the endoscopic suturing device, and injury to adjacent mediastinal structures can be fatal (90).

Current recommendations comprise myotomy with either LHD or POEM as initial surgical treatment of end-stage achalasia. A dilated sigmoid esophagus may denote significant technical challenges for POEM compared to a minimally invasive transabdominal surgical myotomy. Esophagectomy should be considered as a treatment option for end-stage achalasia, particularly in patients who have failed less aggressive treatment modalities (58,76).

Conclusions

This is essentially an opinion paper. The review was not intended to include new findings to direct future research but to be a practical clinical guide based on academic experts from high-volume centers. This may introduce bias in the selection of reviewed papers but, nonetheless, they underpin the concepts exposed.

Diagnosing achalasia requires suspicion and a thorough workup. Upper endoscopy is essential to rule out malignancy or premalignant conditions. Barium swallow helps confirm achalasia and provides a baseline for esophageal diameter. Esophageal manometry is the gold standard for diagnosis and aids in prognosis and therapy selection. The routine use of endoluminal functional lumen imaging probe remains uncertain.

Several treatment options are available for non-advanced achalasia, including pneumatic dilatation, POEM, and LHM. Botulinum toxin injection is a conduct of exception at present. These first-line treatments offer similar outcomes in terms of dysphagia relief but vary in postoperative reflux results. End-stage disease is likely best managed with LHD. A dilated sigmoid esophagus may present significant technical challenges for POEM. Esophagectomy should be considered as a viable treatment option for patients who have not responded to less aggressive treatments.

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Footnote

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