



# The Evolution of the Treatment of Esophageal Achalasia: From the Open to the Minimally Invasive Approach

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## Abstract

**Background** Achalasia is a primary esophageal motility disorder characterized by lack of esophageal peristalsis and partial or absent relaxation of the lower esophageal sphincter in response to swallowing. This study aimed to provide an overview of the evolution of the surgical treatment for esophageal achalasia, from the open to the minimally invasive approach.

**Methods** Literature review.

**Results** No curative treatment exists for this disorder. At the beginning of the 20th century, surgical esophagoplasties and cardioplasties were mostly done to treat achalasia. The description of the esophageal myotomy by Heller changed the treatment paradigm and rapidly became the treatment of choice. For many years the esophagomyotomy was done with either an open transthoracic or transabdominal approach. With the advancements of minimally invasive surgery, thoracoscopic and laparoscopic operations became available. The ability to add a fundoplication for the prevention of reflux made the laparoscopic Heller myotomy with partial fundoplication the operation of choice.

**Conclusions** Surgical management of esophageal achalasia has significantly evolved in the last century. Currently, minimally invasive Heller myotomy with partial fundoplication is the standard surgical treatment of achalasia.

## Introduction

Achalasia is an uncommon disease characterized by lack of esophageal peristalsis and partial or absent relaxation of the lower esophageal sphincter (LES) in response to

swallowing. Due to the inadequate esophageal emptying, patients usually experience dysphagia, regurgitation, and chest pain. Weight loss is also common among patients with achalasia [1].

No curative treatment currently exists for this disorder. Management of these patients is complex and a multidisciplinary team is needed to obtain optimal outcomes (gastroenterologists, surgeons, radiologists, dieticians, etc.). The main goal of treatment is palliation of symptoms by decreasing the LES pressure and improving the emptying of the esophagus into the stomach. Pharmacologic, endoscopic, and surgical treatment modalities are available to achieve symptom relief [2]. However, as medical treatment is significantly less effective than endoscopic or surgical therapies, pharmacologic agents are mostly reserved for frail patients who are unfit for more invasive and durable treatments [3–5].

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Endoscopic treatment alternatives include as follows: botulinum toxin injection, pneumatic dilatation, and peroral endoscopic myotomy (POEM). Current surgical options include as follows: Heller myotomy and esophagectomy. The aim of this study was to review the evolution of the surgical treatment of esophageal achalasia.

### **Initial surgical procedures: from cardioplasty to myotomy**

In the early 1900's, esophageal dilatation was considered the critical contributing factor for patient's symptoms. For this reason, several operations aiming to reduce the esophageal diameter were developed such as esophagoplasties or esophago-esophageal anastomoses [6]. However, all these operations were associated with poor outcomes [7].

Soon after, attention was brought to the obstruction at the esophagogastric junction and cardioplasty became a promising surgical procedure. Wendel, inspired by the Heineke–Mikulicz pyloroplasty, reported the first cardioplasty in 1910 which consisted of a vertical incision of all layers of the anterior wall of the cardia that was then sutured transversally [8]. Heyrowsky (1913) described a different type of cardioplasty: anastomosis of the lateral wall of the distal esophagus with the gastric fundus [9]. Although the short-term results of cardioplasty were satisfactory (i.e., significant improvement of dysphagia), the onset of severe reflux esophagitis was the rule due to the lack of esophageal peristalsis and inability to clear the acid reflux from the stomach.

In 1914, Ernst Heller described the first esophageal myotomy which was a transabdominal extra-mucosal myotomy performed onto both the anterior and posterior walls of the distal esophagus and cardia [10]. The operation described by Heller showed a favorable postoperative outcome, and the surgical community started to embrace the procedure. However, the original double myotomy (i.e., anterior and posterior) was rapidly replaced by a single anterior myotomy as proposed by De Bruine Groeneveldt (1918) and Zaijjer (1923) with equivalent surgical results [11, 12].

### **Esophageal myotomy: transabdominal and transthoracic approach**

Once the esophageal myotomy was established as the ideal operation for the treatment of achalasia, many surgeons favored the transthoracic approach. Okike and colleagues analyzed their series of 468 patients undergoing standardized transthoracic esophagomyotomy between 1949 and 1976 at the Mayo Clinic and showed good to excellent outcomes in 85% of patients [13]. Although uncommon, major complications such as esophageal leak and

mediastinal sepsis were reported [13]. Gastroesophageal reflux was another concern associated with the transthoracic approach, with over 40% of patients showing objective evidence of reflux [14, 15].

The transabdominal approach gained popularity in the mid- and late twentieth century because it was felt to be associated with less risks than the transthoracic operation [16]. In addition, the laparotomy allowed to complement the myotomy with an antireflux procedure. In 1953, Lortat-Jacob proposed the fixation of the gastric fundus to the left border of the esophagus for the prevention of reflux in patients undergoing cardiomyotomy [9]. In 1962, Dor described the “technique de Heller-Nissen modifiée” for the prevention of reflux after Heller myotomy: through a laparotomy, the left side of the myotomy was secured to the anterior wall of the stomach with an initial row of sutures, which was then folded anteriorly and sutured to the right edge of the myotomy with another row of sutures [17]. In 1963, Toupet proposed a posterior partial posterior fundoplication by fixing the gastric fundus to the edges of the myotomy on either side [18]. Adding the fundoplication to the myotomy significantly improved the operation, as the dysphagia was relieved while postoperative reflux was prevented in most patients.

### **The minimally invasive era: thoracoscopic and laparoscopic myotomy**

The minimally invasive era for the surgical treatment of esophageal achalasia started at the beginning of the 1990s. In 1991, Dr. Cuschieri and his team from the University of Dundee (United Kingdom) reported the first laparoscopic Heller myotomy, which showed significant advantages such as absence of extensive abdominal scarring, early mobilization, and shorter length of hospital stay [19]. In 1992, the initial experience in the US with a thoracoscopic myotomy was described by Pellegrini and colleagues from the University of California San Francisco. The mean hospital stay was 3 days and no major complications or deaths occurred. With regard to dysphagia, symptomatic relief was excellent in 70% of the patients [20]. In 1997, Maher analyzed the outcomes of 21 patients undergoing thoracoscopic myotomy and described only one intraoperative perforation and no postoperative complications. Median length of stay was 2 days and after 22 months of follow-up, 80% of patients had excellent relief of their dysphagia [21].

Despite the favorable postoperative results, the thoracoscopic approach had still significant drawbacks: need for double lumen endotracheal intubation during the operation in order to exclude the left lung, placement of a chest tube, higher risk of respiratory complications, and inability to add a fundoplication to prevent postoperative reflux. In

1999, Patti and colleagues compared the results of left thoracoscopic myotomy ( $n = 35$ ) against laparoscopic myotomy plus a partial fundoplication ( $n = 133$ ). There were no deaths and good or excellent relief of dysphagia was obtained in 85% and 93% of patients after thoracoscopic and laparoscopic approach, respectively. The incidence of postoperative reflux, however, was significantly higher after thoracoscopic myotomy (60% vs. 17%) [22]. These results highlighted the greater benefits of the laparoscopic myotomy with fundoplication, and by the end of the 1990s, the laparoscopic Heller myotomy became the operation of choice worldwide.

### Laparoscopic Heller myotomy: standard of care in the twenty-first century

The laparoscopic Heller myotomy (LHM) is usually started about 3 cm above the gastroesophageal junction at the 11 o'clock position. Once the proper submucosal plane is reached (which can be particularly challenging in patients who underwent previous endoscopic therapeutic attempts with pneumatic dilatation and/or botulinum toxin injection) the myotomy is extended proximally for about 6 cm above the esophagogastric junction, and distally for about 2 or 3 cm onto the gastric wall. Therefore, the total length of the myotomy is usually about 8–9 cm [23].

Two types of partial fundoplication can be added to the myotomy: Dor (180° anterior) or Toupet (240° posterior). The Dor fundoplication has two main advantages: does not require posterior dissection of the esophagus and covers the exposed esophageal mucosa (which might seal an inadvertent mucosal perforation). The Toupet fundoplication, on the other hand, helps keeping the edges of the myotomy separated. Two randomized controlled trials showed that both fundoplications are associated with similar rates of symptom relief and comparable incidence of postoperative reflux [24, 25]. Thus, the choice between Dor and Toupet fundoplication is often based on surgeon's experience and preference.

Several studies have proven excellent outcomes after LHM. Zaninotto and colleagues analyzed 407 consecutive patients undergoing LHM plus Dor fundoplication and reported that the operation failed in only 10% of the patients, and the 5-year actuarial probability of being asymptomatic was 87%. In addition, pneumatic dilation improved dysphagia in 75% of patients in whom surgery was unsuccessful (overall effectiveness of 97% considering both primary operation and ancillary pneumatic dilation) [26]. Another study evaluated a consecutive series of 100 patients with LHM and partial fundoplication (94 Toupet and 6 Dor): minor postoperative complications occurred in two patients and there were no major complications or deaths. All symptom scores were significantly improved,

and at a median follow-up of 26 months, failures leading to further treatment were noted in only 4% of patients [27]. Schlottmann and colleagues studied a consecutive series of 147 patients undergoing LHM and Dor fundoplication, and at a median follow-up of 22 months, 87% of patients had symptomatic resolution and required no additional treatment [28]. The European achalasia trial (multicenter randomized controlled trial comparing pneumatic dilation with LHM) confirmed the excellent long-term results of the operation showing a success rate of 84% after 5 years [29]. Similarly, another randomized trial found that only 8% of the patients had recurrence of symptoms after a minimal follow-up of 5 years [30].

In terms of incidence of postoperative reflux, LHM has also shown favorable outcomes. A previous meta-analysis showed that the incidence of reflux after LHM plus fundoplication was 8.8% (vs. 31.5% after myotomy alone without fundoplication) [31]. Salvador and colleagues evaluated with pH monitoring 463 patients after LHM and Dor fundoplication and found that the incidence of pathologic reflux was only 8.6% [32]. Recently, a meta-analysis comprising 53 studies reporting data on LHM (5,834 patients) showed that the incidence of postoperative reflux assessed by pH monitoring was 11.1% [33].

Overall, these data suggest that a LHM with partial fundoplication is a very effective treatment modality that provides symptom relief and prevents postoperative reflux in the majority of patients.

### Robotic Heller myotomy

In 2001, Melvin and colleagues reported the first robotic LHM in the US [34], and since then the robotic platform has been slowly embraced by surgeons for the surgical treatment of achalasia. High-definition 3D vision, tremor filtration, increased degrees of freedom of surgical movements, and improved ergonomics are well-known surgical advantages of the robot. A multicenter study comparing robotic Heller myotomy ( $n = 59$ ) with LHM ( $n = 62$ ) found that intraoperative complications (i.e., esophageal perforation) were more frequent during LHM (16% vs. 0%) and both approaches had similar success rates (92% vs. 90%) [35]. Huffman et al. also compared the outcomes of both techniques: 3 (8%) esophageal perforations occurred during LHM (all repaired intraoperatively) and there were no perforations in the robotic group [36]. Another study also found that robotic myotomy was associated with a decrease mucosal injury rate, as compared to LHM (0% vs. 16%) [37]. A recent meta-analysis including 7 studies (3,214 patients) also showed that robotic Heller myotomy helps decreasing intraoperative esophageal perforation rates (OR 0.11, 95% CI 0.03–0.38) [38].

Although current evidence indicates that robotic Heller myotomy decreases the risk of intraoperative esophageal perforation, a potential learning curve bias might have influenced the results of the studies, given the robotic approach was undertaken more recently. In addition, no randomized studies comparing the laparoscopic and robotic approach are available yet. Overall, postoperative outcomes appear to be similar with both laparoscopic and robotic myotomy. Unfortunately, a robotic myotomy is associated with increased cost to patients and the health care system.

### Esophagectomy: the challenge of refractory end-stage achalasia

Management of patients with a dilated and sigmoid-shaped esophagus (i.e., end-stage achalasia) is indeed challenging. Previous studies have shown that LHM can still help these patients and should be tried before resorting to an esophagectomy. For instance, Mineo and colleagues reported that LHM improved both subjective and objective outcomes, as well as quality of life, in patients with sigmoid esophagus [39]. Other studies have also proven the effectiveness of LHM in patients with end-stage achalasia [40–42]. However, there are still some patients that present with recurrent symptoms after LHM and other endoscopic treatment modalities that might require surgical resection.

An esophagectomy is complex in patients with end-stage achalasia because the dilated esophagus occupies a major part of the posterior mediastinum, there is an altered anatomy with axial deviation, and the vessels feeding the esophagus are often significantly enlarged. Devaney and colleagues described the postoperative outcomes of 93 patients undergoing esophagectomy for achalasia (94% transhiatal esophagectomy): mean length of stay was 12.5 days, anastomotic leak occurred in 10%, and mortality rate was 2%. Anastomotic stricture occurred frequently and nearly 50% of patients required anastomotic dilatations [43].

Minimally invasive techniques for esophagectomy have also evolved dramatically in the last two decades. In fact, minimally invasive esophagectomy is associated with a significant reduction in postoperative morbidity and increased quality of life, as compared to open esophagectomy [44, 45]. Therefore, if a patient with achalasia is deemed candidate for esophagectomy because all previous therapeutic attempts have failed, a minimally invasive approach is recommended whenever possible [46].

### Conclusions

Surgical treatment for esophageal achalasia has significantly evolved in the last century. For many years, open transthoracic or transabdominal myotomy have been the available surgical options. While the emergence of minimally invasive surgery allowed for the use of either thoracoscopic or laparoscopic approach, the ability to add a fundoplication for the prevention of postoperative reflux made the LHM with partial fundoplication the operation of choice.

### Declarations

**Conflict of interest** Francisco Schlottmann, Fernando A.M. Herbella, and Marco G. Patti declare that they have no conflict of interest.

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