

Evaluation and Treatment of Patients with Recurrent Dysphagia After Heller Myotomy

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Introduction

The introduction of minimally invasive surgery during the last two decades has led to a slow shift in the treatment algorithm of esophageal achalasia secondary to, and today a laparoscopic Heller myotomy (LHM) with partial fundoplication is considered the initial treatment modality of choice in most Centers [1–14].

The technique of the minimally invasive approach to achalasia patients has evolved over the last 20 years. Our initial experience with a myotomy performed through a left thoracoscopic approach was first reported in 1992 [15]. Using the guidance provided by intraoperative upper endoscopy, a short myotomy extending for only 5 mm onto the gastric wall, without an antireflux procedure was performed. It became soon clear that the thoracoscopic approach had several advantages when compared to the classic approach by a left thoracotomy, including a shorter hospital stay, reduced postoperative discomfort, and a faster

recovery [15]. Long-term follow-up showed that relief of dysphagia was achieved in almost 90 % of patients, but unfortunately abnormal reflux was documented in 60 % of patients [1]. The laparoscopic approach was then chosen as it provided a better exposure of the gastroesophageal junction (GEJ), the ability to easily extend the myotomy for 1–1.5 cm onto the gastric wall, and the performance of a partial fundoplication [1]. Over time, the length of the myotomy onto the gastric wall was increased, as studies showed that better relief of dysphagia was obtained with a longer myotomy [3, 6]. For instance, Oelschlager et al. compared the outcomes of a conventional myotomy (which extended 1.5 cm onto the gastric wall) to those obtained with an “extended” myotomy (which extended 3 cm below the GEJ) [3] showing long-term relief of dysphagia in 83 and 97 % of patients respectively [6]. Today, our standard technique for patients with achalasia includes an extended myotomy of this sort. As the first branch of the left gastric artery is used as a landmark to gauge the extent of the myotomy onto the gastric wall, we feel that in most cases intraoperative upper endoscopy is not necessary to assess the distal extension of the myotomy in relationship to the GEJ.

Overall, about 90 % of patients undergoing LHM have a major improvement in esophageal emptying and symptom relief [4, 6, 7, 10]. Some patients however experience recurrent dysphagia over time [16]. This chapter will focus on the technical elements that are important for a successful

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and long-lasting operation, and our approach to the diagnosis and treatment of patients with recurrent dysphagia after a LHM.

Recurrent Dysphagia

These are patients who experience substantial relief of symptoms for months or years after the initial LHM and then eventually experience again progressive dysphagia [16]. It is not always easy to elucidate the specific cause of recurrent dysphagia. These are the most common causes:

1. *Scarring of the distal edge of the myotomy.* The most common cause in patients who experience recurrent symptoms after a long symptom free interval is the scarring at the distal edge of the myotomy [2, 17, 18]. While no predictive factors have been identified, we believe that a longer myotomy and a wider separation of the edges of the myotomy at the time of initial LHM should decrease the occurrence of this problem [3, 6].
2. *Wrong fundoplication.* In 2004, Richards et al. reported the outcomes of a prospective randomized trial comparing LHM alone and LHM with Dor fundoplication [5]. While similar improvement of dysphagia was reported in the two groups, abnormal reflux was found at post-operative pH monitoring in 48 % of patients after LHM alone and in only 9 % of patients when a Dor was added, suggesting that the addition of a Dor fundoplication prevented reflux in most patients without impairing esophageal emptying [5]. The use of a total fundoplication has been proposed as a more effective antireflux procedure [19]. This approach however is associated with poor long-term results [20, 21]. For instance, Rebecchi et al. compared 71 patients who underwent a LHM and Dor fundoplication to 67 patients who had a LHM and a Nissen fundoplication [21]. With a mean follow-up of 125 months, the incidence of pathologic reflux was similar in the two groups but dysphagia was present in 2.8 and 15 % of patients respectively, suggesting that a 360° fundoplication
- causes too much resistance at the level of the GEJ, thus impairing esophageal emptying. To date a partial fundoplication is the recommended antireflux procedure in addition to a LHM as it takes into consideration the lack of esophageal peristalsis [9, 11, 12]. There is evidence suggesting that the anterior (Dor) posterior (Toupet) fundoplication are equally effective in preventing reflux [14].
3. *Gastroesophageal reflux disease.* Pathologic reflux is present postoperatively in 50–60 % of patients when a LHM alone is performed, and in 20–40 % when a partial fundoplication is added. Abnormal reflux is considered a common cause of recurrent dysphagia. For instance, Csendes et al. documented a progressive clinical deterioration of the initially good results after a Heller myotomy mainly due to an increase in pathologic reflux and the development of short or long-segment Barrett's esophagus [16]. Unfortunately most patients who develop pathologic reflux are asymptomatic [1]. Therefore, an ambulatory 24-h pH monitoring after the operation is recommended to rule out the presence of reflux, particularly in young patients [22]. If abnormal reflux is demonstrated, acid-reducing medications should be prescribed, and closer endoscopic follow-up obtained.
4. *Effect of previous treatment.* This may occur due to the presence of scar tissue at the level of the GEJ secondary to prior endoscopic treatments [7, 17, 23–25]. Both pneumatic dilatation and intra-sphincteric injection of Botulinum toxin can cause scarring at the level of the GEJ, fibrosis and loss of the normal anatomic planes. In these cases the myotomy performed after endoscopic treatment is more challenging, is associated with higher risk of mucosal perforation, and the outcomes are worse. For instance, Smith et al. compared 154 patients who had undergone endoscopic therapy before surgery to 55 patients who were referred directly to surgery [25]. A higher failure rate of the myotomy was found in the endoscopically treated group (19.5 % versus 10.1 %).
5. *Esophageal cancer.* In achalasia patients the risk of developing squamous cell carcinoma is

increased. In addition, Barrett's esophagus and adenocarcinoma can develop in the presence of pathologic reflux after the myotomy, causing recurrent dysphagia [26]. Even though there are no specific recommendations about endoscopic follow-up of achalasia patients, an upper endoscopy should be routinely performed every 3–5 years.

Diagnostic Evaluation

When patients complain of persistent or recurrent dysphagia, a thorough work-up is critical to identify the cause and site of obstruction in order to formulate a tailored treatment plan.

The first step should always include the revision of the entire history. It is very useful to review when available the diagnostic tests performed before the initial operation as sometimes a wrong diagnosis of achalasia is made. It is also very important to review the report of the original operation. Often there are clues that explain the recurrent dysphagia, such as the description of scar tissue at the level of the GEJ due to prior treatment, failure of identifying the anatomic planes, a short myotomy, or something related to the fundoplication, including a wrong configuration of the wrap.

The symptomatic evaluation is the next step to determine which symptoms are present, and to compare them to the symptoms present before the first operation.

A barium swallow is probably the most useful diagnostic test to evaluate the cause of recurrent dysphagia. It assesses the emptying of the barium from the esophagus into the stomach and shows the diameter and shape (straight versus sigmoid) of the esophagus. Loviscek et al. recently reported a series of patients with recurrent dysphagia after Heller myotomy who underwent redo surgery. They correlated the preoperative radiologic findings on barium swallow to the postoperative symptom improvement. All patients with a straight esophagus (normal or dilated caliber) experienced improvement of dysphagia postoperatively, whereas poorer results were obtained in patients with a sigmoidesophagus [27].

An upper endoscopy should be obtained in every patient. It shows if there is mucosal damage secondary to reflux, or *Candida* esophagitis due to slow emptying, and rules out the presence of cancer. When pseudo-achalasia secondary to the presence of a sub-mucosal tumor or a tumor outside the esophagus is suspected, endoscopic ultrasound and computed tomography can help establish the diagnosis [28].

Esophageal manometry is the key test to confirm the diagnosis of achalasia and to measure the pressure and length of the lower esophageal sphincter. When compared to the preoperative test, the postoperative manometry can show if the extension of the myotomy onto the gastric wall has been appropriate, or if a residual high-pressure zone is still present.

Ambulatory 24-h pH monitoring should also be obtained. The analysis of the pH tracing besides the reflux score is critical to distinguish between real reflux and false reflux due to stasis and fermentation of esophageal contents [29].

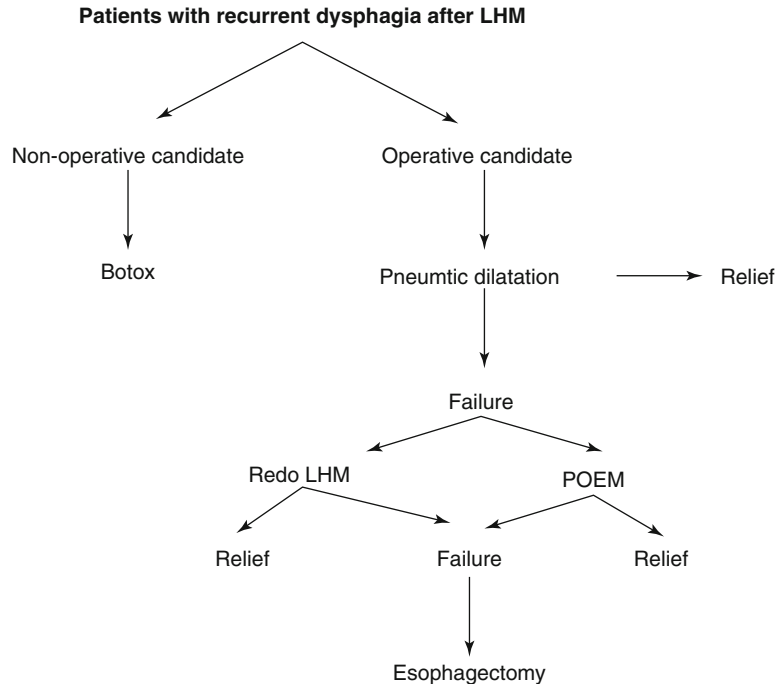
Treatment

Figure 16.1 summarizes our treatment algorithm for patients with recurrent dysphagia after Heller myotomy.

Pneumatic Balloon Dilatation

The initial treatment of these patients should always include a pneumatic balloon dilatation in these patients. Contrary to common belief, the risk of esophageal perforation is very low since the stomach if a Dor fundoplication was performed or the left lateral segment of the liver if a Toupet was added to the myotomy cover the myotomy, or by Zaninotto et al. reported recurrent dysphagia in 9 of 113 patients (8 %) after LHM and Dor fundoplication [17]. Seven of the nine patients were effectively treated by balloon dilatation (median two dilatations, range 1–4), while a second operation was necessary in two. Similar outcomes were described by Sweet et al. who reported on the effectiveness of dilatation for the treatment of both persistent and recurrent dysphagia [7].

Fig. 16.1 Treatment algorithm of recurrent dysphagia after Heller myotomy. *LHM* laparoscopic Heller myotomy, *POEM* peroral endoscopic myotomy



Revisional Surgery

If dysphagia is not relieved by dilations, a reoperation must be considered. When discussing with the patient the risks and benefits, it is important to stress that even though the laparoscopic approach is feasible in most cases, a laparotomy might be needed. In addition, patients must be aware that in case of severe damage to the mucosa during the course of the operation, an esophagectomy may be necessary.

The first step of the operation consists in separating the liver from the stomach and the esophagus. The fundoplication must be then taken down and the fundus brought to the left in order to expose the esophageal wall. Adequate and complete exposure of the esophageal wall, including a thorough dissection of the previous myotomy is the next step. Once this has been accomplished, it is easier to perform a new myotomy rather than trying to extend the prior myotomy. The new myotomy is performed on the opposite side on an unscarred part of the esophageal wall (Fig. 16.2). The myotomy should be extended for about 3 cm below the GEJ, and intra-operative endoscopy should be performed to evaluate for inadvertent

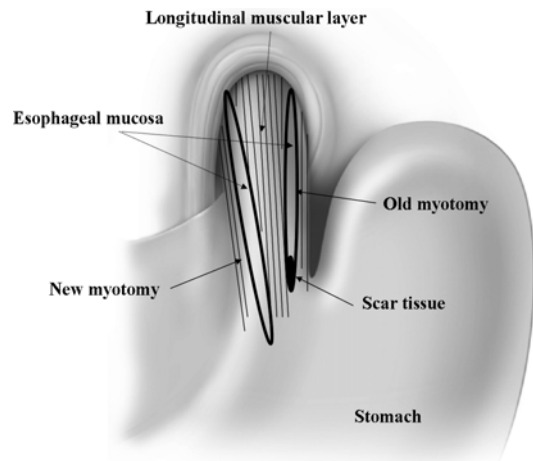


Fig. 16.2 New myotomy performed on the opposite side of the esophagus

esophageal or gastric mucosal injury. After the myotomy is completed, consideration should be given whether or not to add a fundoplication. Certainly, if a mucosal injury has occurred, a Dor fundoplication may decrease the risk of a leak and prevent reflux in most patients. Otherwise it is important to make it sure that a fundoplication

will not cause any added resistance at the level of the GEJ. In cases when the esophagus is dilated, or when part of the fundus of the stomach has been damaged during the dissection, is better to avoid performing a fundoplication. If the patient develops abnormal reflux, it can be treated with proton pump inhibitors. Loviscek et al. recently showed excellent results using this approach [27]. They analyzed the outcome in 43 achalasia patients who had re-do Heller myotomy for recurrent dysphagia between 1994 and 2011. The only take down of the previous fundoplication was performed in 3 patients, while a redo myotomy extending for 3 cm onto the gastric wall was also performed in the remaining 40 patients. A fundoplication was recreated in only about one quarter of these patients. All patients were followed for at least 1 year after the operation. At a median follow-up of 63 months in 24 patients, improvement of dysphagia, with median overall satisfaction rating of 7 (range 3–10) was reported in 19 patients (79 %). An esophagectomy was necessary in four patients for persistent dysphagia. Other authors have reported similar results [30–32].

Sometimes patients present with recurrent dysphagia after a Heller myotomy performed through either a left thoracotomy or a left thoracoscopic approach. Because there are no adhesions in the abdomen and the right side of the esophagus is free of scar tissue created by the first operation, a LHM can be safely performed on the right side of the esophagus with excellent outcomes [33]. Depending on the esophageal size, a partial fundoplication can be added to the myotomy.

Esophagectomy

Esophagectomy should be avoided whenever possible as it is associated with a mortality rate ranging between 2 and 4 % and high morbidity even in expert hands and high volume Centers [34, 35]. For instance, Devaney et al. reported a 10 % rate of anastomotic leak, 5 % rate of hoarseness, and 2 % rate of bleeding and chylothorax requiring thoracotomy among 93 patients who had an esophagectomy for achalasia [35]. In

addition, dysphagia secondary to an anastomotic stricture requiring dilatation occurred in 46 % of patients, regurgitation was complaint by 42 % of patients, and dumping syndrome was demonstrated in 39 % of patients. The average hospital stay was 12.5 days. Despite these shortcomings, esophagectomy is sometimes the only option in patients with end-stage achalasia, dilated and sigmoid shaped esophagus who have already had a failed Heller myotomy and sometimes a re-do Heller myotomy. When performing an esophagectomy, we prefer to use the stomach as an esophageal substitute. Because the esophagus is frequently dilated and fed by large blood vessels, the dissection of the thoracic esophagus is safer under direct vision, either thoracoscopically or by a right thoracotomy. The esophago-gastric anastomosis can be placed either in the neck or at the apex of the right chest.

Alternative Treatment Modalities

A peroral endoscopic myotomy (POEM) is a new treatment modality proposed in achalasia patients, with short term relief of dysphagia in most patients [36, 37]. Because LHM is performed on the anterior wall of the esophagus, POEM could be used instead of a redo Heller myotomy in patients with persistent or recurrent dysphagia by performing a myotomy on the posterior wall of the esophagus [38, 39]. For instance, Onimaru et al. reported excellent short-term results in ten patients undergoing POEM for recurrent dysphagia after Heller myotomy [38]. At 3 months after POEM, the lower esophageal sphincter pressure decreased from 22.1 ± 6.6 to 10.9 ± 4.5 mmHg and the Eckardt score decreased from 6.5 ± 1.3 to 1.1 ± 1.3 . Long term follow-up will be needed to confirm the validity of these short term results.

Conclusions

A LHM with partial fundoplication is today the recommended treatment modality for achalasia patients. The technical steps have been clearly established, and failure to follow them is the main cause of persistent or recurrent dysphagia.

Even though the success rate of LHM is very high, recurrence of symptoms eventually occurs in some patients, with the need for further treatment, particularly if the first operation was done at an early age. When this occurs, a thorough work-up is important for the identification of the cause and to plan a tailored treatment. The best outcomes are obtained in high volume Centers where radiologists, gastroenterologists and surgeons with experience in the diagnosis and treatment of this disease work as a team.

Conflict of Interest The authors have no conflicts of interest to declare.

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