Surgical Management of Achalasia: Recurrent Dysphagia

39

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Introduction

Advances in minimally invasive surgery have led to a significant shift in the surgical management of esophageal achalasia, and today, laparoscopic Heller myotomy and partial fundoplication are considered the procedures of choice [1–12]. The laparoscopic approach is the preferred approach because it provides excellent exposure of the gastroesophageal junction (GEJ) and allows for performance of a partial fundoplication [2, 13]. In addition, as a result of studies showing that a longer gastric myotomy results in improved relief of dysphagia, the length of the myotomy performed on the gastric wall has increased [4, 14]. This is facilitated by the laparoscopic approach.

This approach can achieve significant improvement in esophageal clearance in upward of 90–95% of patients [1, 4, 6, 12]. However, patients can experience persistence of their symptoms or recurrence over time with recurrent (late) dysphagia being more common than persistent (early) dysphagia. This chapter will focus on the diagnostic and therapeutic approach to patients with persistent or recurrent dysphagia following a Heller myotomy.

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Persistent Dysphagia

Persistent dysphagia can be defined as typically presenting immediately following a Heller myotomy or after a temporary relief of symptoms (i.e., less than 6 months). We believe there are several factors, primarily attributable to surgical technique, that are responsible for this occurrence (Table 39.1).

Inadequate Gastric Myotomy

The most common cause of persistent dysphagia is an incomplete myotomy, primarily on the gastric side of the GEJ. In the early 1990s, as minimally invasive surgery began to be applied to the treatment of achalasia, the Heller myotomy was performed through a left thoracoscopic approach that was first published by Pellegrini and colleagues [13]. A 7 cm esophageal myotomy was performed and extended for only 5 mm onto the gastric wall, without an antireflux procedure based on the conventional technique described by Ellis et al. [15]. Early experience resulted in a high incidence of dysphagia that was attributed to inadequate extension of the myotomy onto the gastric wall [13]. Because exposure of the GEJ and gastric wall was difficult through the thoracoscopic approach, the decision was made to switch to the laparoscopic approach and extend the myotomy 1–1.5 cm below the GEJ [2]. Our group had initial success with this approach with resolution of dysphagia in patients with a prior thoracoscopic myotomy and lower rates of dysphagia for primary operations, but we still had occasional patients who required reoperation. We observed that our outcomes appeared to be directly related to the length of the gastric myotomy and decided to extend the myotomy even further to 3 cm below the GEJ. We published our outcomes in a 2003 comparative study that looked at the results of a conventional myotomy (1.5 cm on the gastric wall) versus those with an extended myotomy (3 cm on the gastric wall) [14]. Those patients with an extended myotomy had significantly reduced lower esophageal sphincter pressure and incidence of dysphagia postoperatively. These results persisted in a long-term follow-up study by our group and as a result have become our standard practice [4]. Intraoperatively, surgeons can use laparoscopic instruments and/or endoscopy to help ensure that an adequate gastric myotomy length has been achieved. Laparoscopic graspers can be used to estimate the length of the gastric myotomy since an open grasper measures approximately 3 cm (Fig. 39.1). Endoscopy can also assist with

Table 39.1 Most common causes of persistent dysphagia

1. Incomplete myotomy (a) Inadequate gastric myotomy (most common) (b) Inadequate muscle fiber division
2. Lack of separation of myotomy muscle edges
3. Misconfiguration of the fundoplication
4. Tight closure of the hiatus

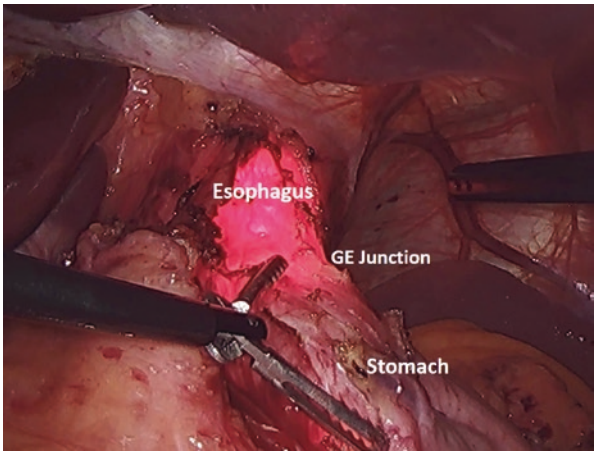

Fig. 39.1 Intraoperative picture of a Heller myotomy. An open laparoscopic grasper roughly measures 3 cm and can be used as a guide for the target length of the gastric myotomy

assessment of the entire myotomy, allows for easy identification of the squamocolumnar junction, and can confirm adequate distal extension of the myotomy onto the gastric wall.

Inadequate Division of Muscle Fibers

Incomplete division of esophageal muscle fibers can also, in theory, cause persistent dysphagia. Several studies have shown that this may occur because of scar tissue that develops at the level of the GEJ secondary to prior endoscopic treatment [16–18]. Pneumatic dilatation and intrasphincteric injection of botulinum toxin are endoscopic treatments that historically were selected as first- and second-line therapies to avoid the morbidity of open surgery. While the success of minimally invasive approaches to surgical myotomy has been well documented [2, 3, 7, 12, 13, 15, 19], pneumatic dilatation and botulinum toxin injection still continue to be offered and performed by gastroenterologists [16]. Though some studies suggest these preoperative procedures do not influence the outcome of surgical myotomy [20, 21], several studies and our own experience support the belief that these procedures, designed to cause disruption of the LES, can lead to scarring which can make the subsequent myotomy more difficult, incomplete, and prone to mucosal perforation with less predictable success rates [16, 17].

Lack of Separation of Myotomy Edges

Persistent dysphagia can also result when the myotomy edges re-approximate postoperatively and form a new scar that can result in esophageal narrowing. To decrease this occurrence, we recommend that the edges of the muscle layers are separated so that approximately 30–40% of the mucosa is uncovered [22]

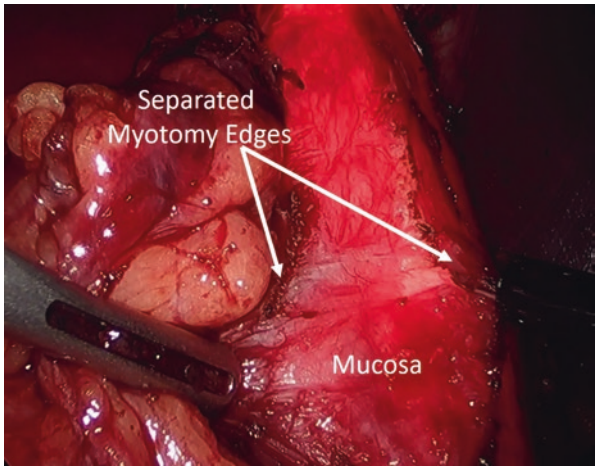

Fig. 39.2 Wide separation of the myotomy edges can prevent the postoperative re-approximation that can lead to recurrent dysphagia

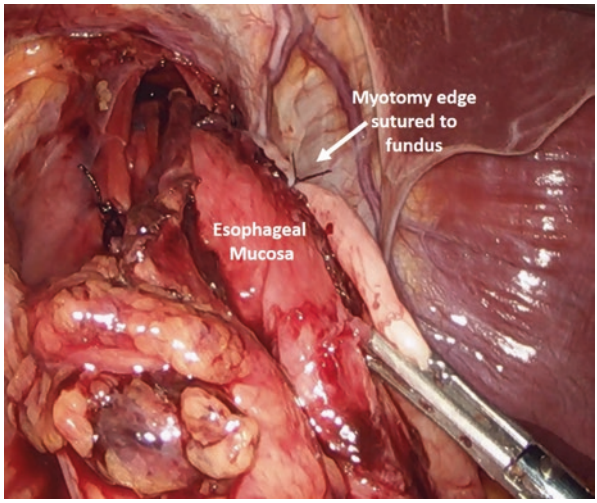

Fig. 39.3 Toupet fundoplication performed with Heller myotomy. The fundus of the stomach is sutured to the myotomy. This splays open the myotomy edges limiting re-approximation

(see Fig. 39.2). Our practice is to perform a Toupet fundoplication at the time of myotomy. The fundus of the stomach is sutured to the myotomy which splays open the myotomy edges and limits re-approximation and scarring (Fig. 39.3).

Fundoplication Misconfiguration

Our practice advocates performing a partial fundoplication following a Heller myotomy to prevent reflux. A 360-degree (Nissen) fundoplication may create a mechanical obstruction due to the lack of esophageal peristalsis that is characterized by achalasia [22]. While the type of partial fundoplication performed (anterior or

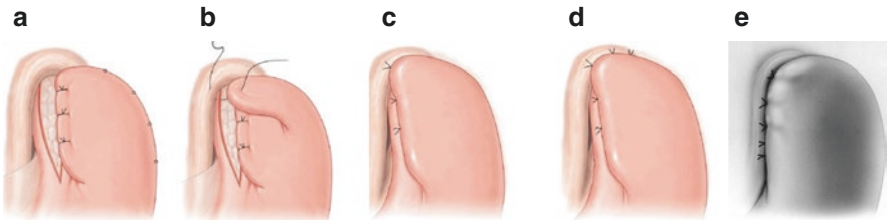

Fig. 39.4 A Dor fundoplication is created by two rows of sutures. (**a**) The first row is created by suturing the left myotomy edge to the fundus. (**b**) The fundus is then folded over the exposed mucosa, and the second row is created with three additional sutures. The first suture incorporates the fundus of the stomach, the esophagus, and the right crus. (**c**) The second and third sutures incorporate just the esophageal wall and the fundus. (**d**) Apical sutures seen in the 12 o'clock position are often placed to reduce tension. (**e**) This picture illustrates *improper* formation of the Dor fundoplication as the second row of sutures incorporates the right crus. This may lead to angulation at the GEJ and contribute to early re-approximation of the myotomy edges

posterior) may vary from center to center, poor construction of either fundoplication may lead to persistent or recurrent dysphagia, just as they can in patients who have a primary fundoplication for GERD. A Dor fundoplication is a 180-degree anterior fundoplication that is constructed with two rows of sutures. Once the first row is created on the left side (Fig. 39.4), the fundus is folded over the exposed mucosa, and three additional sutures are placed. The first suture incorporates the fundus of the stomach, the esophagus, and the right crus. The second and third sutures incorporate just the esophageal wall and the fundus, as incorporating the right crus as well may lead to angulation at the GEJ. Too many stitches at this level may cause constriction at the level of the GEJ and contribute to early re-approximation of the myotomy edges (Fig. 39.4) [7, 22, 23]. We choose a Dor fundoplication to provide coverage of a primary mucosal repair when there is an esophageal perforation. A Toupet fundoplication, our preferred partial fundoplication, is a 270-degree posterior fundoplication that may also lead to postoperative dysphagia when the fundoplication is constructed incorrectly. For example, this can occur if the posterior aspect of the fundoplication is placed under too much tension, creating an anterior angulation of the esophagus that may result in dysphagia [22].

Tight Closure of the Hiatus

Closure of the hiatal opening may impair esophageal emptying by causing constriction or creating a similar anterior angulation of the esophagus that can occur with a poorly constructed Toupet fundoplication. In general, we do not close the hiatus of the average patient with achalasia like we would with an antireflux procedure. If the patient has an associated large hiatal hernia (rare) or if the esophagus is particularly dilated or sigmoid in configuration, we recommend partial closure to avoid dysphagia.

Recurrent Dysphagia

Recurrent dysphagia is more common than persistent dysphagia and typically occurs greater than 6 months after the initial Heller myotomy. It is reported to occur in approximately 3–13 percent of patients [4, 12, 23–26]. When a patient presents for consultation, it is important to determine if the patient was asymptomatic for a period of time and was able to tolerate food. This helps to establish that the initial surgical procedure was successful and may help clarify the etiology of the recurrence. Similar to our discussion on persistent (early) dysphagia, incomplete myotomy on the gastric wall and along the esophagus is also the most common reason for recurrent (late) dysphagia. While causes of recurrence can be multifactorial, we will highlight the most common causes of recurrence that follow incomplete myotomy (Table 39.2).

Myotomy Scarring

Scarring/fibrosis at the distal edge of a properly formed myotomy can be a common cause of recurrent dysphagia following a symptom-free interval [7, 12, 22, 23, 27]. A meta-analysis performed by Campos and colleagues described an incidence of 27% [28]. We believe that the creation of a longer myotomy with a wider separation of myotomy edges should decrease the frequency of this occurrence [4, 14].

Gastroesophageal Reflux Disease

The disruption of the LES, as a result of the myotomy, places patients at risk for the development of postoperative reflux (POR). POR has an incidence of 50–60% when a myotomy is performed without a fundoplication and 2–26% when a partial fundoplication is added [4, 8, 22, 28–31]. Complications of GERD such as esophagitis, Barrett's esophagus, and peptic stricture could lead to dysphagia. Patients who develop pathologic reflux following Heller myotomy may often be asymptomatic, so it is important to perform ambulatory pH monitoring postoperatively [32]. Our practice is to perform this test at our 6-month postoperative visit. If abnormal reflux is documented, typically this is treated with acid-reducing medications if there is no abnormality with the fundoplication. In our experience, this is rare but the most common reason for severe, recurrent dysphagia.

Table 39.2 Most common causes of recurrent dysphagia

1. Incomplete myotomy (a) Inadequate gastric myotomy (most common) (b) Inadequate muscle fiber division
2. Myotomy scarring
3. Gastroesophageal reflux disease
4. Fundoplication abnormality
5. Esophageal cancer

Fundoplication Abnormality

Fundoplication herniation or movement is a less common cause of postoperative dysphagia but has a reported incidence of 10–13% [28]. Misconfiguration of the fundoplication at the time of the initial operation could make this occurrence more likely. If the fundoplication is the cause of dysphagia and the patient requires reoperation, our preference is to take down the fundoplication without reconstructing it. If it is apparent at the time of reoperation that the fundoplication was misconfigured initially (i.e., a fatal flaw such as too much fundus included in the wrap and misplaced sutures), it may be reasonable to redo both the myotomy and the fundoplication.

Esophageal Cancer

Patients with untreated achalasia are at an increased risk of developing squamous cell carcinoma. Long-term studies looking at the development of esophageal cancer following myotomy are scarce. Zaninotto and colleagues performed a retrospective review of 226 patients who received a Heller myotomy for achalasia [31]. Two percent (four) of these patients developed squamous cell carcinoma at 2, 8, 13, and 18 years following myotomy. Pathologic reflux following myotomy can result in Barrett's esophagus and adenocarcinoma which can also cause recurrent dysphagia [33]. Currently there are no precise guidelines about endoscopic follow-up in achalasia patients.

Diagnostic Evaluation

A meticulous and systematic approach to patients with dysphagia is necessary to aid the physician in choosing the most effective treatment modality. Though the etiologies of persistent and recurrent dysphagia somewhat differ, the diagnostic workup is the same (Table 39.3).

As is true for all patients, obtaining a thorough history is a crucial step in the diagnostic workup. Along with the patient's current symptoms, a review of symptoms *before* the initial operation should be elucidated as it helps establish that the initial operation was performed for the correct indication.

Table 39.3 Diagnostic workup for patients presenting with persistent/recurrent dysphagia

1. Upper gastrointestinal series
2. Upper endoscopy
3. Esophageal manometry
4. Ambulatory 24-h pH monitoring
5. Computed tomography[a]
6. Endoscopic ultrasound[a]

[a] Indicates studies that can help rule out pseudoachalasia when other studies are inconclusive

Prior manometry, endoscopy, and upper gastrointestinal (UGI) series images should be obtained along with the operative report. The operative report can provide important clues that may explain the patient's current symptoms, such as a description of scar tissue due to prior treatment, the extent of the myotomy, difficulty identifying anatomic planes, or an enlarged left lobe of the liver that precluded adequate superior extension of the myotomy [22]. In terms of imaging studies, we believe that an UGI is the most important initial study, followed by endoscopy, esophageal manometry, and then pH monitoring.

Upper GI Series

An UGI is most useful for determining if there is a persistent obstruction at the GEJ. Additionally, it can clarify the etiology of the obstruction such as a misconfigured fundoplication, incomplete gastric myotomy, scarring due to GEJ narrowing, hiatal hernia, or an overly tight hiatal closure [22, 23] (Fig. 39.5).

Independent of etiology, the grade of esophageal dilatation determined by the UGI can also help predict the success of revisional surgery. Our center looked at patients requiring a redo myotomy for recurrent dysphagia, and we found that patients with an UGI demonstrating a straight esophagus (normal or dilated, Grades 1–3) all had improved dysphagia following revisional surgery [34]. Dysphagia improvement was less consistent if the esophagus was in a sigmoid configuration (Grade 4).

Endoscopy

An upper endoscopy is the next useful diagnostic study that we recommend performing even if the etiology of dysphagia is apparent on the UGI. Endoscopy serves to identify reflux esophagitis, candida esophagitis secondary to delayed emptying, and strictures and to rule out cancer [22]. It can also reveal a malformed fundoplication and may be useful in characterizing fibrosis [23]. Additionally, at the time of endoscopy, one should consider performing a 20 mm (60 French) balloon or Savary dilatation. When performed properly it is of little risk but may provide substantial relief without proceeding to more aggressive procedures (e.g., pneumatic dilatation $\geq$30 mm).

Esophageal Manometry

Esophageal manometry can confirm the diagnosis of achalasia, especially if the previous two studies are inconclusive and can help identify those patients who may or may not benefit from reoperation. Chapman and colleagues showed that dysphagia is much less common in patients with a hypotensive LES pressure (less than 10 mmHg) following myotomy [35]. If postoperative manometry reveals

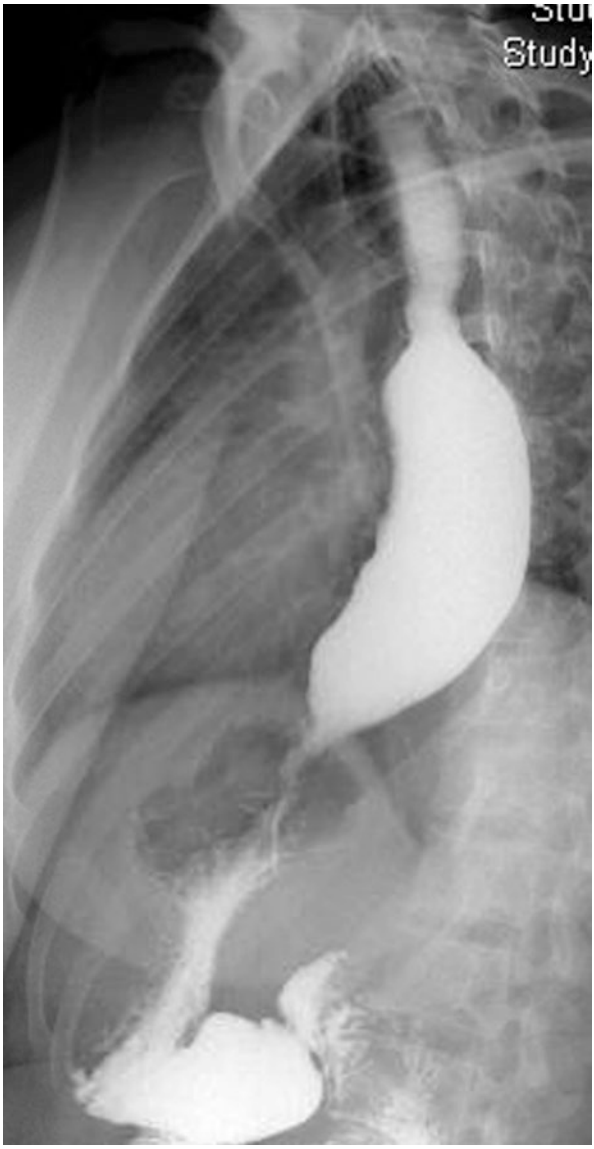

Fig. 39.5 Upper GI study in a patient presenting with recurrent dysphagia following a previous Heller myotomy with Dor fundoplication for achalasia. The patient's operative report noted a 2 cm gastric myotomy. The X-ray at presentation suggests persistent obstruction at the GE junction. This could be due to the Dor fundoplication or an inadequate gastric myotomy

persistently elevated LES pressures, this may indicate an incomplete or fibrotic myotomy that may benefit from a redo myotomy (Fig. 39.6).

Ambulatory 24-Hour pH Monitoring

GERD can lead to dysphagia and occurs frequently after myotomy, even when a fundoplication is included. The best way to confirm GERD is 24-h pH monitoring,

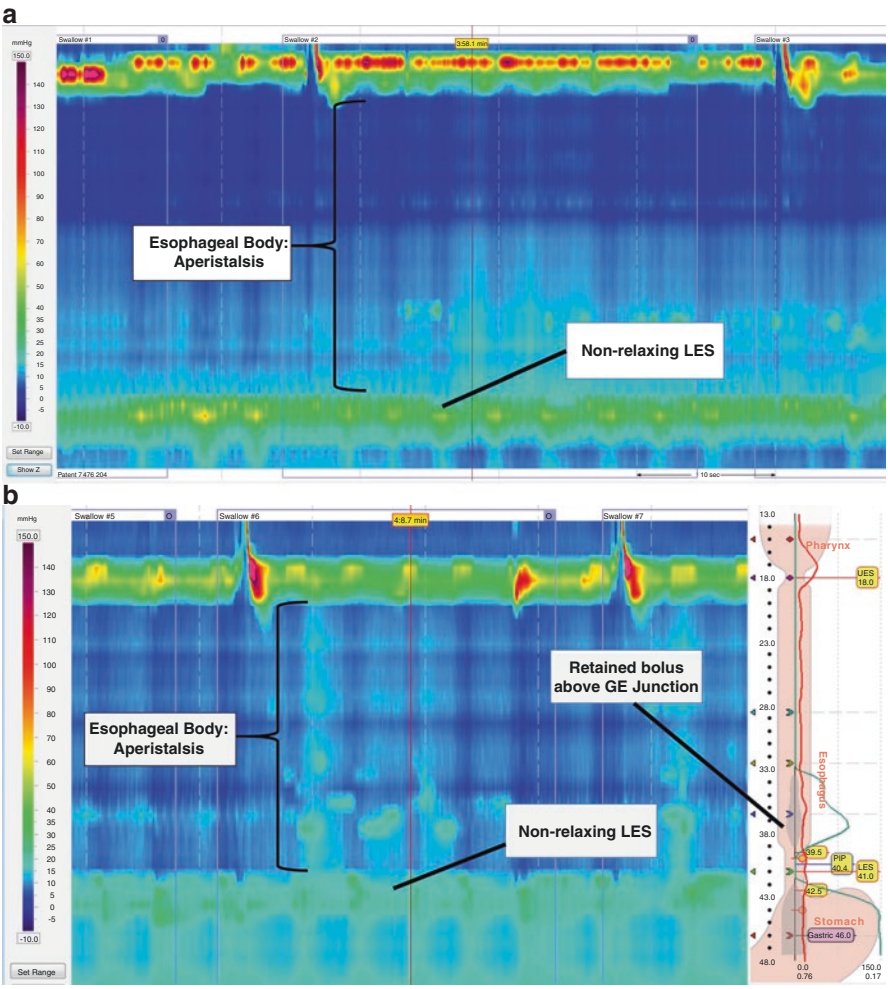

Fig. 39.6 (**a**) Classic high-resolution manometry (HRM) in a patient with type 1 achalasia. (**b**) HRM in a patient presenting with recurrent dysphagia following a Heller myotomy with Dor fundoplication. The patient still has evidence of abnormal relaxation of the LES with a LES pressure of 17 mmHg (normal post myotomy <10 mmHg) and retained bolus above the GE junction

and it should be performed in this patient population. It is important to critically review the pH tracings as there can be false positive results from stasis and fermentation. Simple review of the final composite reflux score will not reveal this, but the tracings are key in differentiating true reflux from false reflux. False reflux is secondary to the stasis of food that can occur if there is abnormal relaxation of the LES. The stasis causes "acidification" of the food as it decomposes. This is depicted on a pH tracing as a slow decrease in pH rather than the abrupt change that is seen with true reflux [23] (Fig. 39.7). There will also be long periods of the pH remaining

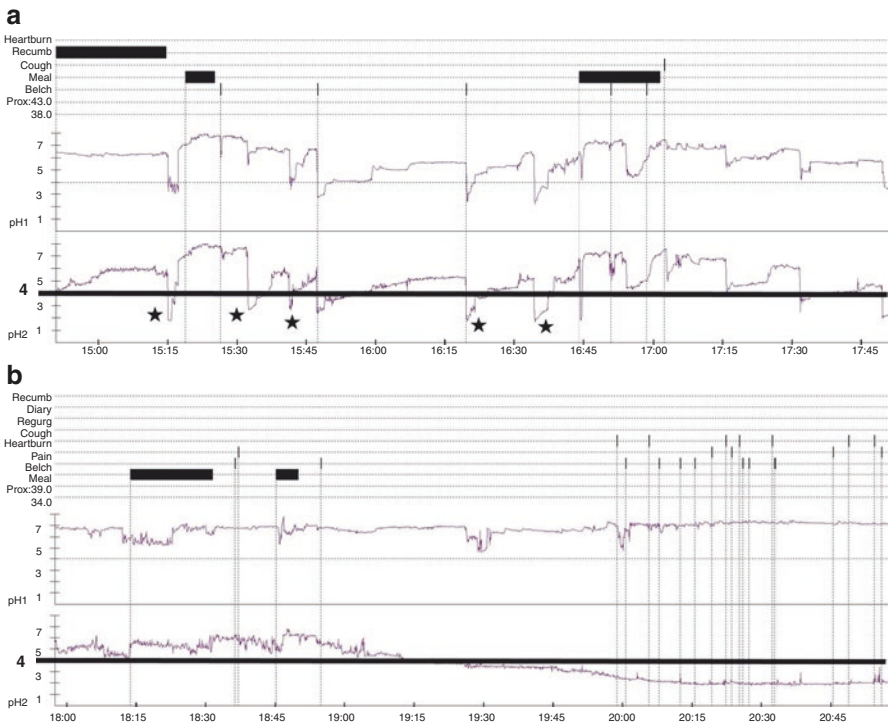

Fig. 39.7 (**a**) 24-h ambulatory pH tracing demonstrating "true" reflux with characteristic abrupt drops below a pH of 4 (stars). (**b**) A false-positive pH tracing demonstrating a slow decrease in the pH that is characteristic of food stasis and fermentation

below 4 and a very abnormal score. The key point here is that false reflux is a manifestation of poor esophageal emptying, not actual reflux. Additionally, patients with achalasia will always have an element of poor emptying as the intrinsic function of the esophagus (peristalsis) is abnormal and unable to be restored post myotomy. Those patients with true reflux may benefit from the addition of acid-reducing medications or revision/addition of a fundoplication [23]. As stated earlier in our section on GERD, reflux can often be silent, so this test should be performed routinely even on asymptomatic patients. Our center typically performs this at the time of our 6-month follow-up and if recurrent symptoms occur.

Other Studies

The workup of these patients can essentially be completed with the above studies, but rarely patients may present with postoperative dysphagia even when manometry is consistent with achalasia and when an adequate myotomy has been performed. Should this happen, computed tomography and/or endoscopic ultrasound may be

useful adjuncts in identifying the unfortunate scenario of a previously missed pseudoachalasia secondary to a submucosal tumor or a tumor outside of the esophagus [36, 37].

Treatment

Endoscopic therapy, revisional surgery, esophagectomy, and newer endoscopic approaches to myotomy are all considerations for the treatment of persistent and recurrent dysphagia.

Pneumatic Balloon Dilatation

Balloon dilatation is a relatively noninvasive procedure that uses increasing intraballoon pressure to disrupt the fibers of the LES. The balloon dilatations used in these cases are larger (30–40 mm) than the 20 mm balloon dilatations mentioned earlier in the endoscopy section so as to adequately disrupt the LES fibers. The procedure carries a low risk of perforation because the previous myotomy site is covered by the stomach if a Dor fundoplication is performed and the left lateral segment of the liver if a Toupet is performed. Dilatation to treat recurrent dysphagia following Heller myotomy has reported early success rates ranging from 30% to 80% [12, 38]. Zaninotto and colleagues performed a retrospective review of 113 patients, 9 of these patients (8%) had recurrent dysphagia following laparoscopic Heller myotomy and Dor fundoplication [25]. Seven of the nine patients were effectively treated with balloon dilatation (median, two dilatations; range, 1–4), and two patients required reoperation. Sweet et al. described similar results in patients with both persistent and recurrent dysphagia [6]. These considerations make balloon dilatation a good treatment option for patients presenting with dysphagia following myotomy.

Revisional Surgery

Revisional surgery should be considered if there is evidence of esophageal obstruction that can be improved with myotomy extension or takedown of the fundoplication, and symptoms fail to respond to pneumatic dilatation. If the patient has already undergone an extended myotomy, the need for reoperation should be rare unless there is an abnormality with the fundoplication [23]. In addition, reoperation carries a risk of irreparable damage to the esophageal mucosa that may require an esophagectomy, a high-risk procedure, even at high-volume centers.

In those patients whose diagnostic workup suggests that the original myotomy was not extended well enough onto the stomach (or the esophagus), a redo myotomy is reasonable to consider. It is also reasonable to consider a redo myotomy for those in whom we believe the main problem is fibrosis/scarring that is

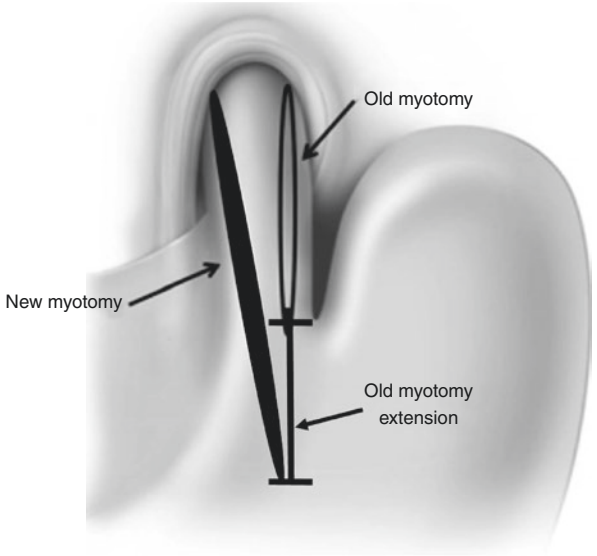

Fig. 39.8 Revisional surgery for recurrent achalasia. The surgeon can extend the prior scarred myotomy (right) or perform a new myotomy (solid oval) to the side of the old myotomy

nonresponsive to pneumatic dilatation. Intraoperatively, we either extend the prior scarred myotomy or perform the second myotomy to the side of the previous myotomy (Fig. 39.8). This allows the surgeon to take advantage of the uncut muscularis layer of the esophagus and the stomach [22, 23]. The myotomy should be extended for 3–4 cm below the GEJ and superiorly on the esophagus as far as the prior effective myotomy. Once the myotomy is completed, the surgeon should determine whether or not to perform a fundoplication. If the mucosa is inadvertently injured, performing a Dor fundoplication to buttress the repair may decrease the chance of immediate complications and prevent further reflux. In the absence of perforation, our tendency is to, in most cases, avoid performing a fundoplication in the setting of a redo operation, particularly in the presence of a dilated esophagus [22]. Our rationale is based on three concepts: (1) dysphagia, not GERD, is the primary indication for reoperation; (2) should the fundoplication fail or exacerbate dysphagia, a third operation would be increasingly difficult; and (3) abnormal reflux can be more easily treated with medications than dysphagia. This approach has been supported by several studies in the literature [34, 38–40]. However, if the esophagus is normal and there is a clear abnormality corrected (i.e., inadequate myotomy, misconfigured fundoplication), one can then consider a fundoplication if reasonably confident that it will not result in recurrent obstruction.

Peroral Endoscopic Myotomy (POEM)

Peroral endoscopic myotomy is a newer endoscopic technique for performing a LES myotomy that was developed in 2008 (Fig. 39.9). Early studies have been associated with excellent relief of dysphagia, and a recent long-term study by

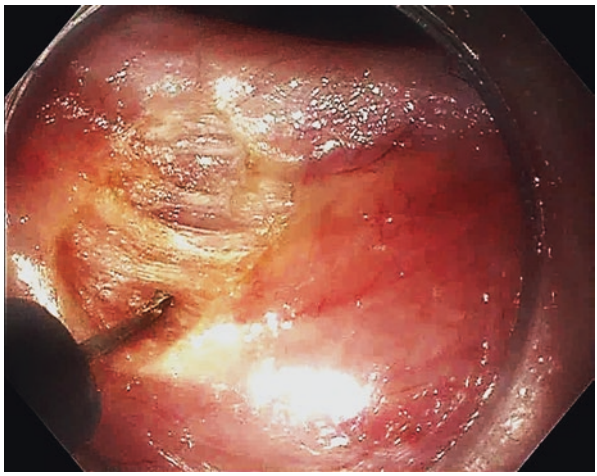

Fig. 39.9 Intraoperative photo of a POEM. This technique allows for endoscopic division of the esophageal muscle fibers, avoiding the need for laparoscopy

Hungness et al. reported symptomatic relief in 92% of patients with a low rate of complications (follow-up range 12–52 months) [41–43]. Forty percent of these patients had objective evidence of reflux. While a laparoscopic myotomy is performed on the anterior wall of the esophagus, the POEM procedure can be performed on the posterior wall of the esophagus. This makes POEM a reasonable consideration in patients presenting with persistent or recurrent dysphagia after a failed myotomy or redo operation and may allow patients to avoid an esophagectomy. One limitation of the POEM in cases of recurrent dysphagia is that it does not address the fundoplication as an etiology for the patient's dysphagia. If a fundoplication was performed at the time of initial surgery, this approach does not allow for the fundoplication to be taken down. Because of this, POEM is an ideal option in those patients without a prior fundoplication. If a POEM is performed in the setting of a prior fundoplication, and dysphagia persists, one should consider taking it down in a subsequent setting. Use of the POEM procedure is not widespread and is only available at a limited number of centers with highly trained surgical endoscopists and gastroenterologists. While the initial studies are promising, further long-term studies are needed, particularly related to recurrent dysphagia before widespread use recommendations can be made.

Esophagectomy

Esophagectomy is associated with a mortality rate between 1% and 4%, even in expert hands [44, 45]. Devaney and colleagues reported a 10% rate of anastomotic leak, 5% rate of hoarseness, and 2% rate of bleeding among 93 patients who had an esophagectomy for achalasia [45]. As a result, it should only be undertaken when all other less invasive options are exhausted. At that point, esophagectomy should be considered in patients with end-stage achalasia, characterized by a sigmoid-shaped esophagus, and those who have already failed a myotomy or redo myotomy [34].

Given the risks of an esophagectomy, the lower morbidity of a laparoscopic redo myotomy, and some success with a redo myotomy in end-stage patients [34], we tend to offer esophagectomy only to those patients who do not improve after a redo myotomy. In terms of surgical approach, the esophagus is frequently dilated with large collateral veins on the surface that make the transhiatal approach risky. Our approach in these patients is to dissect the esophagus under direct vision either thoracoscopically or via thoracotomy [22, 23].

Conclusion

Laparoscopic Heller myotomy with partial fundoplication is the procedure of choice for patients with achalasia. Persistent or recurrent dysphagia can develop over time secondary to surgical technique, fibrosis, fundoplication configuration, reflux, or esophageal cancer. A systematic and thorough approach is necessary for the diagnostic evaluation of these patients to help determine the most appropriate treatment modality. Endoscopic therapy and revisional surgery are the cornerstones of treatment, and newer endoscopic therapies may provide a more minimally invasive approach to the management of this disease. Esophagectomy, however, should be considered as a surgical option in cases that are refractory to these less invasive treatment therapies to open the LES.

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