Achalasia of the Esophagus: A Surgical Disease

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Achalasia is a primary esophageal motility disorder of unclear etiology. As treatment has evolved during the past 10 to 15 years, it has become primarily a surgical disease. It is uncommon, but not rare, affecting approximately 1 in 100,000 individuals per year.1 Occurring equally in men and women, it is an acquired condition usually diagnosed between 20 and 50 years of age, but can occur at any age. In 1672, Sir Thomas William first described the disease as “cardiospasmy” and treated the problem with dilation using a whale sponge attached to a whale bone. It was not until 1927, when AF Hurst determined that the problem was the result of an inability of the lower esophageal sphincter (LES) to relax, and named the disease achalasia, a Greek term meaning failure to relax.2 Normal individuals have a lower esophageal high-pressure zone, which completely relaxes with initiation of a swallow. In achalasia, residual pressures in the LES remain well above normal after swallowing, resulting in a functional outflow obstruction at the gastroesophageal junction. In addition, the LES can be hypertensive, resulting in higher than normal resting pressures, and the esophageal body is aperistaltic, all leading to failure of bolus transport.3 When untreated, intraesophageal pressures rise and the esophagus slowly dilates, often to the point of gross deformity.4

ETIOLOGY

Achalasia can be primary (idiopathic) or secondary. Primary achalasia is thought to result from a complete loss or relative absence of inhibitory ganglion cells in the myenteric (Auerbach’s) plexus of the esophagus. This loss creates an imbalance between excitatory and inhibitory neurons causing failure of the LES to relax.5 Several studies have documented diminished numbers of myenteric ganglion in diseased esophageal specimens and demonstrated the presence of lymphocytic infiltrate and collagen deposition within ganglion.6-8 Given these findings, an infectious agent, such as a virus, and its subsequent immune response, is thought to be the cause of the ganglion loss, but the exact etiology remains unknown. Neurotransmission and cell signaling as it relates to the distal esophagus and LES in achalasia has been a subject of growing investigation. Nitric oxide is thought to be the major inhibitory neurotransmitter controlling relaxation of esophageal smooth muscle.9,10 It has been suggested that nitroergic inhibitory neurons are lost in greater proportion than cholinergic neurons during the achalasia disease process.11

Supporting this thesis, studies in knockout mice lacking nitric oxide synthase (NOS) have found high LES resting pressures and absent LES relaxation.12 In humans, specimens from achalasia patients have shown complete loss of NOS enteric neurons and NOS activity as compared with nonachalasia specimens.13 Bruley des Varannes and colleagues14 recently reported a study suggesting a serum-borne substance, supporting an immunologic mechanism. Gastric fundic muscle from normal humans was cultured with serum from achalasia patients, gastroesophageal reflux disease patients, or normal subjects. The normal fundus incubated with serum from achalasia patients had a decrease in NOS neurons as compared with controls. In addition, relaxation of electrically stimulated circular muscle contractions was substantially decreased after incubation with serum from patients with achalasia.

The most common secondary cause of achalasia is Chagas’ disease15,16 a systemic disease caused by infection by the protozoan Trypanosoma cruzi. Chagas is transmitted to humans by the reduvid or “kissing bug” and infects intramural neurons, causing autonomic dysfunction. Chagas’ disease is common in South and Central America, and worldwide is considered to be the most common cause of achalasia.15,16 Pseudoachalasia occurs through infiltration of the LES by cancer or by iatrogenic condition, such as a tight fundoplication or external appliance, such as a band.17,18

PRESENTATION AND DIAGNOSIS

Achalasia is a slowly progressive disease. As a consequence, patients often present late in the course of this disease, when their symptoms and anatomic abnormalities have become prominent. Diagnosis earlier in the disease process is possible, given conscientious attention to patients’ esophageal reports and a high degree of suspicion. Clinical symptoms include slowly progressive dysphagia for solids and liquids, regurgitation of bland undigested food, chest pain, and weight loss. Patients can also have a history suggestive of aspiration, including recurrent pneumonia or chronic cough.
Of important note, patients with achalasia can present with heartburn and, as such, mimic the presentation of gastroesophageal reflux disease. Reviewing the symptoms of 67 patients with achalasia, Spechler and colleagues found that 32 (48%) reported heartburn. This fact, coupled with diagnostic challenges in patients with early achalasia, including presence of a normal-caliber esophagus and artifactual LES relaxation on manometry (caused by relative movement of the catheter and sphincter), likely leads to a small but real number of patients with achalasia erroneously diagnosed with gastroesophageal reflux disease and undergoing Nissen fundoplication. In fact, it is not uncommon for patients with achalasia to be treated with proton pump inhibitors. The clinician should be aware of this possibility; a careful review of both the manometric study and video barium examination will almost always reveal the characteristics of achalasia. This occurs for several potential reasons. Physiologically, heartburn has been shown to be precipitated by esophageal distention. Different neural stimuli (luminal acid versus distention) can be interpreted by the brain as the same symptom. It has also been suggested that achalasia can develop in the setting of underlying gastroesophageal reflux disease. The retrospective symptom review referenced here showed that of those with heartburn, 47% (n = 15) had heartburn before development of dysphagia, which persisted once dysphagia developed, and an additional 28% (n = 9) had heartburn before development of dysphagia, which stopped after onset of dysphagia. Heartburn symptoms developed in the remaining patients (25%) after onset of dysphagia. Interestingly, those patients reporting heartburn had considerably less LES resting pressures as compared with those without heartburn symptoms. Both mechanisms are likely at play.

A simple chest radiograph can suggest the diagnosis in advanced cases by the absence of a gastric bubble and presence of a dilated fluid-filled esophagus, generally seen as a right-sided posterior mediastinal shadow. Upper endoscopy should be performed in all patients to exclude obstruction from tumor or stricture. Findings will commonly include retention of saliva and undigested food associated with a dilated esophagus, but can be normal. A barium swallow is often diagnostic, showing a dilated aperistaltic esophagus, presence of an air-fluid level, or a characteristic “bird’s beak” appearance created by a nonrelaxing LES. Barium studies can be interpreted as “normal,” particularly in early achalasia, with the radiologists’ focus on anatomic characteristics, such as dilation and emptying. Reviewing the symptoms and barium studies of 38 patients with confirmed achalasia, Blam and colleagues found a poor correlation between symptoms and barium study findings, which led to a diagnostic delay, particularly in patients who presented with atypical symptoms (i.e., heartburn, weight loss, cough, asthma). Atypical symptoms were reported in 34% of patients and resulted in a median delay of 30 months before correct diagnosis, as compared with 11 months in those who reported typical symptoms (dysphagia, chest pain, regurgitation). Careful examination of fluoroscopic or video images should be routinely done, which will almost always reveal a flaccid nonperistaltic esophagus devoid of the usual “stripping” waves.

The gold standard for diagnosis of achalasia is esophageal manometry, which is the most sensitive diagnostic test...
Manometry is especially helpful in establishing the diagnosis in patients who present in an early disease state when other diagnostic evaluations appear normal. There are four classic manometric characteristics (Table 1): hypertensive LES, present in approximately 50% of patients; nonrelaxing LES; esophageal aperistalsis; and elevated lower esophageal baseline pressure. A subset of achalasia patients can have simultaneous contraction waves of variable amplitudes, consistent with preserved muscle function, this is termed vigorous achalasia. Examining the manometric tracings of 26 patients with achalasia, Shi and colleagues found residual LES relaxation pressure to be the most accurate diagnostic manometric criteria. Specifically, a residual pressure of $>12\text{ mmHg}$ had 92% sensitivity for the diagnosis of achalasia. Coupled with aperistalsis, a residual pressure of $>10\text{ mmHg}$ had 100% sensitivity and 100% positive predictive value for achalasia. Similarly, in a later study by the same group, examination of 58 achalasic manometric studies revealed only one variant patient who had a residual pressure after swallow-induced LES relaxation that was $<10\text{ mmHg}$.

Although traditionally, a nonrelaxing sphincter and complete aperistalsis were thought to be universally required for diagnosis of achalasia (it is debated which of these might be the primary and most important feature), it is likely that some heterogeneity exists in the manometric findings in patients with the clinical characteristics of achalasia. Hirano and colleagues have recently emphasized this fact. They carefully analyzed the clinical history and manometric recordings of 58 patients with achalasia and 43 control subjects. Endoscopy, barium swallow, histopathology, and response to treatment were used to help establish the diagnosis in patients without classic manometric findings. Although it was uncommon, four manometrically distinct variants were seen, ie, high-amplitude nonperistaltic esophageal body contractions, short segments of aperistalsis, retained complete LES relaxation on swallowing, and nonswallow transient relaxation. These findings were seen in only 4 of 58 patients, the vast majority had classic manometric findings of achalasia. This study provides a strong argument that variations truly exist. The clinician should be aware of these possibilities when evaluating patients with dysphagia.

The introduction of high-resolution manometry (HRM) and multichannel intraluminal impedance (MII) studies can improve the diagnostic manometric assessment of achalasia and assessment of treatment efficacy. HRM is readily evident on HRM and the technique facilitates assessment of sphincter relaxation, one of the most difficult aspects of conventional manometry (Fig. 3). HRM is made possible by a solid-state catheter with 36 circumferential sensors spaced at 1-cm intervals. Pressure measurements obtained from the sensors are plotted in reference to both space and time. The closely spaced pressure transducers and interpolation of the data eliminate the movement artifacts, which confound standard manometric techniques. Data have shown that, in the absence of previous hiatal surgery, failure of the LES high-pressure zone to relax below 15 mmHg is highly specific and sensitive for achalasia. The ability of MII to quantitate bolus transport can prove to be useful in the postoperative assessment of patients with achalasia after laparoscopic myotomy, although few studies to date have investigated its benefit.

**TREATMENT**

Achalasia is an incurable disease and treatment is focused on relief of symptoms. The goal of both surgical and nonsurgical treatment is to eliminate the outflow obstruction afforded by a nonrelaxing sphincter, relieve dysphagia, and maintain a barrier against gastroesophageal reflux when possible. Nonsurgical treatment includes calcium channel blockers and nitrates, injection of botulinum toxin (Botox;
Allergan) into the LES, and large-caliber (30 to 40 mm) pneumatic dilation of the LES. Surgical treatment includes division of the muscle fibers of the LES and proximal stomach. Although pharmacologic treatments such as calcium channel blockers, nitrates, and sildenafil have been shown to have physiologic effects on esophageal function, they rarely result in meaningful clinical improvement. With the advent of minimally invasive surgery, laparoscopic myotomy has slowly shifted treatment of achalasia toward the greater use of surgical therapy.

Pharmacologic treatment
Pharmacologic treatments of achalasia, such as oral nitrates and calcium channel blockers, act to inhibit intramural neurons. These drugs attempt to recreate a balance between stimulatory and inhibitory nerve fibers to decrease the resting and residual pressure of the LES. Their effect is sporadic and has shown only short-term success. In light of their poor outcomes and potentially harmful systemic effects, these agents are largely unreliable and unrealistic modalities for long-term symptom relief.

Flexible endoscopic treatment
Endoscopic injection of Botox into the LES aims to block release of acetylcholine from cholinergic neurons in an effort to lower both basal and residual LES pressures. This treatment is short-lived, rarely results in substantial reduction in LES pressure, and, although dysphagia can be improved, often requires repeat injections for continued relief. In a prospective randomized controlled trial comparing Botox injection (n = 40) with laparoscopic myotomy and partial fundoplication (n = 40), Zaninotto and colleagues found that although patients in both groups showed initial improvement in symptoms, 6 months after treatment dysphagia and regurgitation recurred in nearly half (45%) of those treated with Botox. In addition, they determined that the probability of being symptom-free at 2 years was considerably higher after myotomy (87.5%) than after Botox injection (34%). It is now recognized that Botox injection creates an inflammatory reaction in the distal esophagus with consequent submucosal fibrosis, which can make subsequent surgical myotomy more difficult. Although commonly tried as a short-term treatment option, the authors believe Botox should be reserved for those who are not surgical candidates or abandoned altogether.

Pneumatic balloon dilation decreases esophageal outflow resistance by forceful “tearing” of the LES muscle fibers. Protocols vary among clinicians when performing pneumatic dilation. In general, graded balloon dilation is begun using a 30-mm balloon as the initial dilator. The balloon dilator is passed into the esophagus and placed at the gastroesophageal junction by using fluoroscopy or direct endoscopic vision. A Gastrografin esophagram followed by a barium swallow is performed after pneumatic dilation to exclude esophageal perforation. Repeat balloon dilation can be performed using progressively larger dilator sizes to 35 mm and 40 mm. The most common balloon dilators in current use are the Rigiflex and Witzel polyethylene balloons. Rigiflex is a balloon on a catheter, which is placed using fluoroscopy, and the Witzel balloon is inserted while attached to the endoscope. Risk of perforation using the Rigiflex is about 3% and that of the Witzel is 6%.

Comparing pneumatic balloon dilation with Botox injection in a randomized trial, Cleveland Clinic investigators found that 70% (14 of 20) of patients who underwent dilation were in symptomatic remission at 1 year as compared with 32% (7 of 22) of patients who underwent Botox treatment. In addition, they found that balloon dilation resulted in substantial improvements in LES pressure, esophageal barium column height, and esophageal diameter, and Botox injection failed to show any change in these parameters. Similarly, when comparing pneumatic balloon
dilation with Botox injection in a double-blinded randomized study, Bansal and colleagues found that 89% (16 of 18) of patients who underwent balloon dilation had a decrease in their symptom grade as compared with only 38% (6 of 16) of patients who underwent Botox injection. Most recently, a meta-analysis evaluating pneumatic balloon dilation and Botox injection for treatment of achalasia reported that, although short-term failure rates were similar between the two techniques, long-term (>6 months) results showed considerably less failures after dilation as compared with Botox.

In an analysis evaluating the long-term efficacy of pneumatic dilation for achalasia, Katz and colleagues retrospectively evaluated 72 patients at a mean of 6.5 years after dilation. Defined as no additional therapy needed for achalasia, success was seen in 85% of patients. Four patients required additional pneumatic dilation.

More recently, Zerbib and colleagues retrospectively evaluated long-term remission rates of pneumatic dilation for 150 patients with achalasia, using a fastidious dilation protocol that included several dilation sessions until development of remission and close follow-up with fairly liberal “on-demand” repeat dilation sessions, if needed. They found initial symptom remission to be 91.3% after a median of 2.6 dilations. Recurrent symptoms developed in 35% of patients. At 5 and 10 years, the calculated probability of being in remission was 67% and 50%, respectively. Of those patients with recurrent symptoms who required repeat dilation treatment, 96.4% were in remission at a mean of 3.5 years. The probability of being in remission after repeat dilation was 96.8% and 93.4% at 5 and 10 years, respectively.

Although pneumatic dilation has been found to have a more lasting effect on dysphagia than Botox injection, outside of a rigorous dilation protocol, most studies have not demonstrated the long-term outcomes seen after surgical myotomy. Until recently, the only prospective randomized trial comparing pneumatic dilation to surgical myotomy was reported by Csendes and colleagues from Chile. They found that 100% of patients treated with myotomy (n = 19) had only mild or no dysphagia at a mean of 3.5 years, as compared with only 61% of those treated with balloon dilation (n = 18). Very late follow-up of this trial has also been published.

Two more recent evaluations are also worth noting. West and colleagues retrospectively evaluated long-term outcomes of 125 achalasia patients followed prospectively for >5 years after pneumatic dilation. In this experience, only 50% of patients had no or occasional (less than once per week) dysphagia at 5 years, and only 40% at 15 years. Kostec and colleagues recently reported a multicenter Swedish prospective randomized study of 51 patients comparing pneumatic dilation (n = 26) with laparoscopic myotomy (n = 25). A predefined dilation protocol was used, beginning with a 30-mm (women) or 35-mm (men) Rigiflex balloon and progressing as needed to 40 mm in those with persistent dysphagia at 7- to 10-day intervals. The lack of such a protocol was one of the major criticisms of the study performed by Csendes and colleagues. Laparoscopic myotomy was performed by experienced surgeons using intraoperative endoscopy and Toupet posterior hemifundoplication. Outcomes were assessed at 1 year after treatment. Failure was defined as incomplete symptom control or symptom relapse requiring more than three treatments during the 12-month period, serious complications or side effects, or patient request for alternative treatment. Esophageal perforation occurred in two patients in the dilation group (7.6%). All surgical procedures were completed laparoscopically with no substantial perioperative morbidity. Treatment failure was significantly more common in the dilation group (1 of 25 [4%] versus 6 of 26 [23%]; p = 0.04). Mean symptom score after operation was 11.3 and after dilation was 17.7 (p > 0.05). The authors concluded that laparoscopic myotomy and partial fundoplication was superior to pneumatic dilation in newly diagnosed patients with achalasia.

**Surgical treatment**

Esophageal myotomy for achalasia was first described by Ernest Heller in 1913. In this operation, both the anterior and posterior lower esophageal sphincter muscle fibers were disrupted. A modified version of this procedure, referred to today as the Heller myotomy, consists of a single anterior longitudinal myotomy and has become the standard operative technique. Once performed through either a thoracotomy or laparotomy, esophageal myotomy is now performed almost invariably through a laparoscopic approach.

Outcomes of thoracoscopic myotomy have been shown to be inferior to the laparoscopic approach. In a retrospective evaluation of patients undergoing either thoracoscopic Heller myotomy (n = 30) or laparoscopic Heller myotomy (n = 30), Patti and colleagues found laparoscopic myotomy to be superior with regard to complete dysphagia relief (70% versus 77%) and development of postoperative reflux (60% versus 10%). Similarly, Stewart and colleagues comparing the outcomes of 24 achalasia patients who had undergone myotomy through a thoracoscopic approach with 63 patients who had a laparoscopic approach, found that the laparoscopic approach was not only associated with decreased operative time, conversion rate, and length of stay, but was also better able to relieve dysphagia and prevent heartburn than the thoracoscopic approach. A re-
cent review of studies evaluating thoracoscopic myotomy from 1993 to 2003 (n = 10 studies; 204 patients) and laparoscopic myotomy from 1995 to 2002 (n = 15 studies; 499 patients) by Abir and colleagues found symptom relief to occur in 76% versus 94% of patients, respectively. Development of gastroesophageal reflux disease occurred in 35% of patients after thoracoscopic myotomy and 13% of patients after laparoscopic myotomy. The inferior symptomatic outcomes seen after a thoracoscopic myotomy reflect the difficulty in extending the myotomy adequately onto the stomach from the chest and the inability to create a fundoplication to counteract development of reflux disease.

Technique: laparoscopic esophageal myotomy and partial fundoplication

The patient is placed on the table in the modified lithotomy position with the lower extremities abducted; positioning identical to that of a laparoscopic fundoplication. Pneumoperitoneum is established and four additional ports are placed under video laparoscopic control. The dissection begins by freeing the upper third of the gastric fundus in preparation for later Dor anterior hemifundoplication. The short gastric vessels are sequentially dissected and divided with the aid of ultrasonic shears. In contrast to a Nissen fundoplication for gastroesophageal reflux disease, the posterior attachments and pancreaticogastric branches should not be divided, in an effort to preserve as much of the normal posterior gastroesophageal anatomy as possible. If a Toupet posterior hemifundoplication is planned, a posterior dissection is necessary. This would be the preferred approach in the small (4% to 5%) subset of patients who present with a concomitant hiatal hernia.

The gastroesophageal junction is then exposed by dissecting the gastroesophageal fat pad and retracting it laterally. The gastroesophageal junction is cleared for 6 to 8 cm in preparation for the myotomy. A vessel loop can be placed around the gastroesophageal junction, including the anterior vagus nerve, and brought out through the subxiphoid trocar if necessary. Once the esophagus is mobilized and gastroesophageal junction exposed, the myotomy is performed with a combination of scissors and hook-type electrosurgery (Fig. 4). The extent of the myotomy should be limited proximally by the anterior crural confluence and to that which can be covered by a fundoplication. It should then be carried down across the gastroesophageal junction, to ensure that the clasp fibers are divided, and onto the anterior wall of the fundus of the stomach for 2 to 3 cm. The edges of the myotomy are carefully separated from the underlying mucosa for 40% to 50% of the esophageal circumference. The procedure is completed by fashioning an anterior (Dor) or posterior (Toupet) partial fundoplication.

An anterior partial fundoplication of the Dor type is the most commonly performed antireflux barrier created after myotomy for achalasia. After fundic mobilization the anterior fundus is laid across the myotomy site and the left fundic edge is sewn to the left cut edge of the esophageal myotomy with three or four interrupted sutures of 2-0 silk. The highest stitch is taken through the left crural pillar as an anchor to prevent torsion of the fundus. The fundus is then sewn to the right edge of the cut esophageal muscle similarly; again using three or four interrupted sutures of 2-0 silk and taking the highest stitch through the right crural pillar to prevent torsion.

To create a posterior partial fundoplication (Toupet), the anterior fundus is brought to either side of the myotomy, and two columns of sutures are placed on either side, leaving the myotomy site bare and open.

DATA SUPPORTING TECHNICAL COMPONENTS

Extent of myotomy

Recent data show that both the proximal and distal extent of esophageal myotomy are important. Chen and colleagues have reported that epiphrenic pseudodiverticulum developed in as many as 67% of patients followed 7 to 16 years post myotomy and fundoplication. This is likely caused by an absence of coverage by the fundoplication over the proximal extent of the myotomy. This increased recognition of the potential for diverticulum formation has led many to limit the proximal extent of the myotomy to that which can be covered by a fundoplication.

Relief of dysphagia might be better when lengthening the distal extent of myotomy. Oelschlager and colleagues reported better symptomatic improvement and a lower in-
cidence of recurrent dysphagia by increasing the distal extent of myotomy onto the proximal stomach from 1.5 cm to 3 cm. They compared outcomes of 52 patients treated with a standard myotomy of 1.5 cm onto the proximal stomach with outcomes of 58 patients treated with an extended myotomy of 3 cm. Postoperative LES pressures were substantially lower (9.5 mmHg with extended versus 15.8 mmHg with standard myotomy) and postoperative dysphagia improved after longer myotomy. Importantly, postoperative 24-hour pH data showed that extension of the distal aspect of the myotomy did not result in a higher prevalence of gastroesophageal reflux.

**Addition of fundoplication**

Although myotomy effectively lowers esophageal outflow resistance and improves esophageal emptying, it also increases the propensity for development of gastroesophageal reflux. The importance of the addition of a fundoplication to the myotomy has, until recently, been debated.

A recent prospective randomized study has put this issue to rest. Richards and colleagues reported on 43 achalasia patients randomized to laparoscopic Heller myotomy with and without Dor fundoplication. Gastroesophageal reflux, defined by 24-hour distal esophageal acid exposure time >4.2%, was present in 47.5% of patients undergoing Heller myotomy alone compared with 9.1% of patients undergoing Heller myotomy with partial fundoplication.

Importantly, there was no substantial difference in postoperative LES pressures or dysphagia scores between the two groups. Similarly, a retrospective evaluation of 146 patients who underwent laparoscopic Heller myotomy with (n = 88) or without (n = 61) Dor fundoplication by Rice and colleagues, found that pH proved gastroesophageal reflux occurred considerably less often after addition of a partial fundoplication. The addition of Dor fundoplication to Heller myotomy did not decrease esophageal emptying when assessed by barium esophagography.

In the presence of an aperistaltic, often dilated and tortuous esophagus, most agree that a complete fundoplication should be avoided. On the other hand, the precise degree (ie, 180 degrees, 270 degrees) and location (anterior, posterior) of a partial fundoplication is unclear. Current data suggests a minimal difference between a posterior 270-degree (Toupet) fundoplication and an anterior 90- to 180-degree (Dor) fundoplication. For example, Arain and colleagues found no difference in relief of symptoms, including dysphagia, heartburn, or chest pain, or the need for proton-pump inhibitors when comparing myotomy with Dor to myotomy with Toupet. Because of the preservation of natural posterior attachments and its easier technical construction, most centers have migrated toward routine use of a Dor fundoplication.

**SURGICAL OUTCOMES**

Outcomes of laparoscopic Heller myotomy have now been studied for well more than a decade. These data show that surgical myotomy is more effective than nonsurgical treatments at providing longterm relief of dysphagia, regurgitation, and chest pain, while promoting weight gain and patient satisfaction. Many surgeons and gastroenterologists alike believe that surgical myotomy should be recommended as primary therapy for virtually all patients with achalasia.

Medium-term outcomes of laparoscopic myotomy have been reported from centers around the world (Table 2). Assessed 2 to 5 years after operation, >90% of patients have either complete relief of, or only mild persistent, dysphagia. Investigators from Padua, Italy, have provided some of the longest-term data. Portale and colleagues reported 113 patients who under-
went laparoscopic Heller myotomy from 1992 to 1999. More than 90% (91.2%) were symptom-free at a median of 2 years followup, with a Kaplan-Meier analysis showing a 90% probability of remaining asymptomatic at 5 years. Most published results report similar improvement. Degree of improvement might not be the same with all symptoms. Arain and colleagues\textsuperscript{54} found that the most marked improvement occurred for the symptom of regurgitation (74% complete relief). Moderate relief was seen for dysphagia (33% complete relief) and the least relief was observed for chest pain (18% complete relief). Difficulty in relieving chest pain has also been reported after pneumatic dilation.\textsuperscript{64}

Complications average 10% to 15%, and commonly include pneumothorax, wound infection, and esophageal leak.\textsuperscript{47,55-63} Mortality is uncommon, zero in most large published series. Inadvertent violation of the esophageal mucosa occurs in 0 to 14% of procedures and, when recognized and repaired, is rarely of clinical consequence. Unrecognized mucosal perforation will present as acute abdominal or chest pain in the early postoperative period and has been treated both operatively and nonoperatively. Conversion from a laparoscopic operation to open laparotomy is infrequent in experienced laparoscopic centers and is caused by esophageal perforation, uncontrolled hemorrhage, or adhesions.

Postoperative assessment through contrast esophagography, manometry, and pH monitoring has shown that esophageal outflow obstruction improves and esophageal diameter decreases, as does resting and residual LES pressure. Zaninotto and colleagues\textsuperscript{62} reported pH data on 84 of 113 patients who underwent postoperative 24-hour pH monitoring. Only 5 of 84 (6%) had tracings consistent with gastroesophageal reflux, three of these reported mild heartburn and one had erosive esophagitis at endoscopy. Longterm followup data from the era of open myotomy suggest that gastroesophageal reflux is a common cause of recurrent symptoms, with a substantial minority developing progressive regurgitation, heartburn/chest pain, erosive esophagitis, and Barrett’s esophagus. Whether this will also be true of the laparoscopic era remains to be determined.

Providing the longest followup of surgical myotomy ever reported, Csendes and colleagues\textsuperscript{42} published the clinical, endoscopic, histologic, manometric, and acid reflux studies of 67 patients enrolled in a prospective randomized study carried out in the 1970s. Patients were divided into three groups based on the length of followup: 6 to 10 years, 10 to 20 years, and 20 to 30 years. Most patients (73%) had long-lasting symptomatic response, although substantial recurrent symptoms were found in 7% of patients <10 years, 23% of those 10 to 20 years, and 35% of patients >30 years postmyotomy. Similarly, there was a slowly progressive increase in patients found to have pathologic esophageal acid exposure 15%, 28%, and 53% respectively (Fig. 5). Barrett’s esophagus developed in nine patients and esophageal squamous cell carcinoma developed in three.

Figure 5. Proportion of patients with normal and abnormal esophageal acid exposure after myotomy and partial fundoplication for achalasia. Group 1: <10 years, group 2: 10 to 20 years, group 3: >30 years followup. Gray bar, normal acid reflux test; black bar, abnormal acid reflux test. Reprinted from: Csendes A, Braghetto I, Burdiles P, et al. Very late results of esophagomyotomy for patients with achalasia; clinical endoscopic, histologic, manometric and acid reflux studies in 67 patients for a mean follow-up of 190 months. Ann Surg 2006;243:196–203, with permission.

Outcomes predictors
Several factors have been proposed as predictors of outcomes after treatment for achalasia. Degree of improvement in the resting LES pressure has been reported by several authors to substantially affect longterm success of pneumatic dilation. Patients in whom the LES pressure was reduced to ≤10 mmHg had long-lasting (5 years or more) relief of dysphagia, and recurrent dysphagia developed within the first 12 to 24 months in those with persistent LES pressures >20 mmHg (Fig. 6).\textsuperscript{65} This principle is likely as true for surgical myotomy as it is for pneumatic dilation, although lowering resting LES pressure to near 10 mmHg is much more reproducible with myotomy. Young patients (younger than 40 years) have also been shown to respond less well to pneumatic dilation and are good candidates for primary myotomy. In a longterm prospective study comparing outcomes of patients who required myotomy after failed pneumatic dilation with patients who responded well to dilation therapy, Gockel and colleagues\textsuperscript{66} found that the younger the patient age at diagnosis the greater the probability of needing surgical myotomy for
symptomatic relief. Using odds ratios to determine risk for eventual myotomy, the authors found that a patient diagnosed with achalasia at age 15 has a 70% probability of needing myotomy, a patient diagnosed at age 40 has a 35% probability and a patient at diagnosed at age 70 has an 8% probability (Fig. 7).

Several factors have also been suggested to affect outcomes of laparoscopic myotomy. These include the magnitude of preoperative LES resting pressure, the degree of preoperative esophageal dilation or tortuosity, or both, and the presence or absence of earlier nonoperative interventions.

Evaluating outcomes of 78 consecutive achalasia patients treated with laparoscopic myotomy, Arain and colleagues\(^54\) found that a high preoperative resting LES pressure was an independent predictor of relief of dysphagia. This was confirmed in a recent series of 200 consecutively treated achalasia patients by investigators at Vanderbilt University.\(^67\) They found that patients with a preoperative LES resting pressure of \( \geq 35 \text{ mmHg} \) were 21.3 times more likely to have relief of their dysphagia than those with preoperative LES pressures \( < 35 \text{ mmHg} \). These authors also reported that the greater the decrease in LES pressure after operation, the greater the improvement in postoperative dysphagia (Fig. 8).

The benefit of surgical myotomy in patients with a dilated and tortuous esophagus is unclear. In this setting, in which the esophagus is not only massively dilated but sigmoid-shaped, bolus flow is impaired by the shape of the esophagus in addition to LES spasm. Even in these circumstances, patients can perceive benefits. Patti and colleagues\(^68\), evaluating 66 patients with varying degrees of dilation, found that all 7 patients with esophageal diameters of \( > 6 \text{ cm} \) and tortuosity had satisfactory outcomes. In contrast, a case-controlled study by Pechlivanides and colleagues\(^69\) found that esophageal diameter decreased in all patients with esophageal dilation, but only patients with a postoperative diameter of \( < 4 \text{ cm} \) experienced excellent results.\(^69\) Given this data and the low morbidity and mortality associated with laparoscopic Heller myotomy as com-
pared with esophagectomy, myotomy is often chosen as the first line of treatment regardless of the extent of dilation. Endoscopic interventions performed in achalasia patients before surgical intervention likely influence the success of surgical myotomy. Smith and colleagues,33 recently reported that previously given treatment can effect symptomatic outcomes. Patients with \((n = 154)\) or without \((n = 55)\) a history of earlier balloon dilation or Botox injection were compared. Defined as persistent or recurrent symptoms or the need for additional treatment, the failure rate of those with earlier interventions was found to be almost twice that of those who had no earlier intervention (19.5% versus 10.1%). In addition, patients with previous endoscopic therapies were found to have more difficult dissection planes, increased mediastinal scarring, which resulted in a higher incidence of intraoperative mucosal perforations, and longer postoperative recovery than those without earlier treatment. These findings support use of surgical myotomy as the initial treatment method for achalasia and put forth that endoscopic interventions should be reserved for the rare patient who is not a surgical candidate.

**Persistent or recurrent symptoms**

Long-term outcomes data suggest that a properly performed laparoscopic myotomy and partial fundoplication will be the only therapeutic intervention required in up to 80% of patients with achalasia.70 Mild to moderate symptoms of dysphagia, regurgitation, heartburn, and chest pain recur with time in as many as 40% to 50% of patients, but few of these require additional intervention beyond lifestyle and behavioral changes or proton-pump inhibitor therapy. Mechanisms of failure include inadequate myotomy, scarring or anatomic distortion of the gastroesophageal junction, complications of gastroesophageal reflux disease, and progressive esophageal dilation or distortion, usually secondary to persistent outflow resistance. Early failures are usually caused by failure to extend the myotomy far enough onto the stomach or anatomic distortion, or both. Late failures occur secondary to development of complicated gastroesophageal reflux with strictureing or Barrett’s changes and disease progression. Development of a distal esophageal diverticulum, thought secondary to proximal extension of the myotomy into the lower posterior mediastinum, is also an increasingly recognized occurrence. The predisposing factors and symptomatic importance of this are as yet unclear.

Recurrent dysphagia after surgical myotomy should be investigated by upper endoscopy to exclude an obstructing lesion, by contrast, esophagography to assess for esophageal dilation or tortuosity, and by repeat manometry to assess for high persistent LES resting pressure (≥10 to 15 mmHg). Management options include pneumatic dilation or remyotomy for suspected incomplete or scarred myotomy, and esophagectomy for those with end-stage dilation and tortuosity, stricture, or cancer. Pneumatic dilation, once believed to be contraindicated after myotomy, can be an attractive option for early persistent dysphagia. In a retrospective analysis of 113 patients treated with myotomy for achalasia, Zaninotto and colleagues62 found that 78% of 10 patients with recurrent dysphagia could be effectively treated with pneumatic dilation. They concluded that pneumatic dilation should be considered as the first-line treatment for patients with persistent dysphagia and that reoperation should be reserved for those who do not respond.

Both medical and surgical therapies exist as treatment options for patients with achalasia. Administration of pharmacologic agents have had very little clinical effect. Botox injection has shown symptomatic relief in some, but is often transient, requires multiple treatment sessions, and can make later surgical treatment more difficult. Although pneumatic dilation has been found to have a more lasting effect on dysphagia than Botox injection, it has not demonstrated the longer-lasting outcomes seen with surgical myotomy. Laparoscopic Heller myotomy has become the initial primary treatment and standard surgical approach for patients with achalasia.

Several studies have shown surgical myotomy to provide superior long-term symptom relief as compared with nonsurgical interventions. An extended distal myotomy, along with a partial fundoplication, has been found to provide greater dysphagia relief with minimal development of gastroesophageal reflux. A high preoperative LES pressure portends better symptomatic outcomes after operation, as does the absence of earlier nonoperative interventions. Persistent or recurrent symptoms after surgical myotomy that do not respond to lifestyle changes can be treated effectively with pneumatic dilation.

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