

Minimally invasive surgery for esophageal achalasia

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Abstract

Esophageal achalasia is the most commonly diagnosed primary esophageal motor disorder and the second most common functional esophageal disorder. Current therapy of achalasia is directed toward elimination of the outflow resistance caused by failure of the lower esophageal sphincter to relax completely upon swallowing. The advent of minimally invasive surgery has nearly replaced endoscopic pneumatic dilation as the first-line therapeutic approach. In this editorial, the rationale and the evidence supporting the use of laparoscopic Heller myotomy combined with fundoplication as a primary treatment of achalasia are reviewed.

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INTRODUCTION

Idiopathic achalasia is a primary motor disorder characterized by incomplete relaxation of the lower esophageal sphincter and aperistalsis of the esophageal body secondary to loss of inhibitory ganglion cells in the myenteric plexus. It is unclear whether the primary event occurs in the brain or whether the neurologic changes are the result of a direct injury of the myenteric plexus. The etiology of the disease is unknown, with genetic, autoimmune, infectious, and environmental factors being implicated. Achalasia, usually diagnosed between 20 and 40 years or after 60 years of age, is the most common primary esophageal motor disorder; it is second only to gastroesophageal reflux disease as the most common

functional esophageal disorder to require surgical intervention. The first account of a successfully treated case of achalasia was described by Thomas Willis in 1674, in which a whale bone was used to forcibly dilate the cardia. The first surgical myotomy was performed by Ernst Heller in 1913.

PATHOPHYSIOLOGY AND ASSESSMENT OF ACHALASIA

Failure of the lower esophageal sphincter to completely relax upon swallowing results in a functional obstruction and pressurization of the esophageal body. Defective esophageal emptying progressively leads to dilatation and tortuosity of the esophagus with loss of the peristaltic waveform. Overall deterioration of esophageal function and structure with time, and the fact that peristalsis can return after surgical myotomy, suggest that the motor abnormalities secondary to esophageal outflow obstruction may be reversible and that early definitive treatment of achalasia is essential to preserve esophageal function^[1].

Achalasia has an insidious onset. Dysphagia and food regurgitation are the two major presenting symptoms of the disease. Nocturnal regurgitation often leads to recurrent episodes of aspiration pneumonia. In about 40% of patients the diagnosis of achalasia is delayed by the reported symptoms of chest pain and heartburn simulating gastroesophageal reflux disease. As the disease progresses, inability to swallow causes malnutrition and weight loss. Squamous-cell carcinoma of the esophagus appears to develop with greater frequency in patients with long-standing achalasia than in the normal population^[2].

The most sensitive tests for detecting achalasia are esophageal manometry and barium swallow videofluoroscopy. Aperistalsis and incomplete lower esophageal sphincter relaxation are the typical manometric features. Radiological abnormalities include aperistalsis, esophageal dilatation, and minimal lower esophageal sphincter opening with a bird-beak appearance. Endoscopic assessment is important to exclude the diagnosis of malignancy-induced secondary achalasia, often referred to as pseudoachalasia, before invasive therapies are implemented. Clinical features suggesting a tumor of the gastroesophageal junction are a short duration of dysphagia, a significant weight loss, and an elderly patient. Since adenocarcinoma of the cardia may present endoscopically as an infiltrating lesion with apparently normal mucosa, CT scan, endoscopic ultrasonography, and even exploratory laparoscopy should be used liberally in this subgroup of patients^[3].

MANAGEMENT OF ACHALASIA

Endoscopic dilation versus surgery

Treatment of achalasia is palliative and is directed toward elimination of the outflow resistance caused by the abnormal lower esophageal sphincter function without creating excessive gastroesophageal reflux. Sustained symptomatic relief of dysphagia can be achieved by endoscopic pneumatic dilation or by surgical myotomy. Retrospective studies have shown better results with myotomy performed by an experienced surgeon, and in the only prospective randomised trial myotomy gave better long-term results compared with pneumatic dilatation^[4]. A recent study of 217 patients who received a median of 4 pneumatic dilations over a 12-year period reported a long-term therapeutic success of only 50%^[5]. Over the past 15 years, the advent of minimally invasive technology has made surgery a more attractive option as a first-line therapy to both patients and referring physicians^[6]. It has been shown that pneumatic dilation is 72% effective versus 92% for the laparoscopic Heller myotomy. In addition, laparoscopic techniques have sharply reduced surgical morbidity^[7] (Table 1).

Principles of surgical therapy

The basic goal of surgery in the treatment of achalasia is the extramucosal division of both layers of the esophageal muscularis propria and of the oblique gastric fibers, the so-called Heller myotomy. For many years this operation has been performed through a laparotomy, often with the addition of a fundoplication, or through a left thoracotomy. The extent of the myotomy in the stomach, which in part depends on the type of surgical access, has long been a matter of controversy. Ellis, the pioneer of the transthoracic approach, advocated a limited (< 1 cm) gastric myotomy without an antireflux procedure. However, at a very late follow-up the level of symptomatic improvement markedly deteriorated over time with this approach, and the rate of excellent results (i.e., asymptomatic patients) progressively decreased from 54% at 10 years to 32% at 20 years^[8]. The majority of European surgeons have instead favored the transabdominal approach which includes a longer gastric myotomy (1 to 2 cm) and a fundoplication to protect the esophagus from iatrogenic gastroesophageal reflux. Another debated issue has been the direction of the myotomy over the stomach, which may be closer to the lesser curve or to the greater curve, and, as a consequence, can divide the semicircular (clasp) fibers and the oblique (sling) fibers in distinct proportions^[9].

A third major issue is the opportunity to add an antireflux repair to the myotomy. The rationale for considering a concomitant fundoplication is the assumption that a well performed surgical myotomy renders the lower esophageal sphincter incompetent. Furthermore, gastric contents propelled retrograde into an aperistaltic esophagus are not effectively cleared, thus magnifying the damage caused by postoperative reflux. The majority of surgeons worldwide presently favor a transabdominal approach and add to the myotomy a partial fundoplication, either a 180 degree anterior (Dor) or a 270 degree posterior fundoplication

Table 1 Pooled response rate of achalasia treatments^[7]

Therapy	n	Weighted response rate (%)	Weighted follow-up (yr)
Botulinum toxin	149	32	1.1
Pneumatic dilation	1276	72	4.9
Heller myotomy			
Thoracotomy	1221	84	5
Laparotomy	732	85	7.6
Laparoscopy	171	92	1.2

(Toupet). The partial fundoplication, as opposed to the 360 degree Nissen repair, does not cause significant resistance to esophageal emptying, therefore reducing the risk of postoperative dysphagia. The main advantage of an anterior Dor fundoplication is that this technique does not require mobilization of the distal esophagus and cardia, and provides a natural patch over the denuded esophageal mucosa. Overall, there has been a greater than 90% symptomatic relief and a less than 10% incidence of pH proven gastroesophageal reflux with this approach^[10]. This operation should be proposed as the first therapeutic option also in patients with sigmoid-shaped esophagus, although the chances of symptomatic relief are reduced in such circumstances and in some of these individuals an esophagectomy can eventually be required.

Minimally invasive surgery for achalasia

During the early times of the minimally invasive surgical approach, both laparoscopy and thoracoscopy have been used to perform a Heller myotomy. However, it soon became clear that laparoscopy offers several inherent advantages, including superior visualization of the gastroesophageal junction, a single lumen endotracheal intubation, the ability to add an antireflux procedure, and a shorter hospital stay. In addition, laparoscopy showed a better symptomatic outcome and a lesser incidence of postoperative gastroesophageal reflux^[11].

The first laparoscopic Heller procedures were performed in England and in Italy during the early 1990s^[12,13]. Compared to the open abdominal approach, a technical innovation introduced in the laparoscopic era has been the use of intraoperative endoscopy as a means of precisely identifying the gastroesophageal junction, checking for the completeness of the myotomy and testing for the presence of occult perforations. During the early phase of the learning curve, a Rigiflex balloon dilator was also used to distend the cardia in order to facilitate division of all residual muscle fibers^[14]. Subsequently, transillumination and air inflation provided by the endoscope were considered appropriate to assist the myotomy^[15]. Further demonstration of the effectiveness of intraoperative endoscopy came from a study showing that endoscopic and laparoscopic criteria were discordant in the identification of the esophagogastric junction in 58% of the cases, the cardia being in all these cases at a more distal site with endoscopic criteria. As a consequence, based on the laparoscopic appearance, the surgeon may err by underestimating the caudal extent of the myotomy and

can perform a too short myotomy on the gastric side^[16]. Although we are still convinced of the effectiveness of intraoperative endoscopy during the learning curve of this operation and during reoperative surgery, we have now discontinued this practice in our high-volume referral center where between 15 and 20 patients per year undergo Heller myotomy as a primary treatment for achalasia.

It should also be noted that a common reason for an incomplete myotomy on the gastric side is the fear of producing a mucosal injury, which typically occurs just at the esophagogastric junction. The mean reported rate of mucosal perforation is about 5%, but the frequency is largely dependent on surgeon's experience. There are no consequences if the mucosal injury is detected intraoperatively and repaired with interrupted stitches^[15].

The principle of limited surgical dissection of the cardia, already advocated by some surgeons in the open era^[10], has been successfully reproduced in many centers with the minimally invasive laparoscopic approach. Dissection is limited to the anterior and lateral attachments of the phrenoesophageal membrane as this allows enough space to perform the myotomy and helps to prevent the occurrence of postoperative gastroesophageal reflux. Only in patients with an associated hiatal hernia or epiphrenic diverticulum the distal esophagus is encircled and a posterior crural repair is performed. By grasping the cardia and pulling it in a caudal direction, the myotomy is started on the distal esophagus using a L-shaped electrocautery hook until the submucosal plane is identified. The myotomy is carried out for about 6 cm on the esophagus, toward the left of the anterior vagus nerve, and up to 2 cm on the gastric side to include the oblique fibers.

Despite the minimal surgical dissection herein described, an antireflux repair added to the Heller myotomy appears to be beneficial and is nowadays supported by the majority of esophageal surgeons. The anterior Dor fundoplication is a technically simple procedure, quick and safe to perform laparoscopically. It does not require mobilization of the cardia, and places a gastric fundic patch over the myotomy site to protect the mucosa and prevent re-healing of the myotomy. Reconstruction of the angle of His is first performed by suturing the medial wall of the fundus to the left edge of the myotomy and to the left crus of the diaphragm. The Dor fundoplication is fashioned by folding the anterior fundic wall over the myotomized surface and securing the stomach to the right crus of the diaphragm and to the right edge of the myotomy with interrupted stitches. It is important to pay attention to the geometry of the fundoplication in order to avoid undue tension and to provide a uniform patch over the myotomized esophagus, but usually there is no need to divide the short gastric vessels. The Heller-Dor operation is carried out in about one hour by an expert surgeon. A gastrographin swallow study is performed the following day to check esophageal transit and to rule out leaks. A soft diet is then permitted, and the patient is usually discharged home on postoperative d 2 or 3.

Studies including large number of patients have shown that an extramucosal myotomy of the esophagus and cardia combined with an anterior fundoplication can be

Table 2 Twenty-four h pH data of patients submitted to Heller myotomy and Heller myotomy plus Dor fundoplication in a randomized study^[24]

Parameter	Heller (n = 18)	Heller-Dor (n = 21)	P value
Time (%) pH < 4 upright	8.1 + 10.4	0.8 + 1.1	0.015
Time (%) pH < 4 supine	9.1 + 18.3	2.0 + 6.9	0.002
No. episodes pH < 4	113 + 128	25 + 6.9	0.001
No. episodes pH < 4 for ≤ 5 min	3.4 + 4.7	0.5 + 1.6	0.001

performed safely and effectively through laparoscopy, with clinical and functional results similar to that obtained with the open transabdominal approach and relief of dysphagia in more than 90% of patients^[17-22]. Although previous endoscopic treatments, such as balloon dilatation or intrasphincteric botulinum toxin injection, may cause submucosal scarring at the esophagogastric junction resulting in a more difficult surgical procedure and an increased operative morbidity, no statistically significant differences as far as concern the clinical outcome have been reported^[15].

Radiologic and manometric studies after the laparoscopic Heller-Dor operation have shown a significant decrease of the internal diameter of the thoracic esophagus and of the resting lower esophageal sphincter pressures^[19]. In a recent report, at a minimum follow-up of 6 years after the operation, about 82% of patients were satisfied with the treatment and were able to eat normally; more than a half of the symptomatic recurrences occurred during the first year of follow-up and were effectively treated with pneumatic dilations. Nine (12.7%) patients either had abnormal acid exposure on postoperative 24-h pH study or were on treatment with proton-pump inhibitors for reflux symptoms; however, in none of these individuals did endoscopy reveal more than grade A esophagitis^[23].

An important preoperative factor affecting the outcome of the laparoscopic operation is the magnitude of the resting pressure of the lower esophageal sphincter. On a multivariate analysis, only a high resting pressure prior to surgery was a predictor of resolution of dysphagia. Interestingly, all patients with a preoperative sphincter pressure greater than 36 mmHg had their dysphagia resolved after surgery^[24].

The issue of whether an antireflux procedure should be added to the Heller myotomy has long been controversial and supported only by personal feelings and retrospective studies. Recently, a randomised double-blind clinical trial comparing the outcome of myotomy plus Dor fundoplication versus myotomy alone has shown that the former operation is superior in terms of reflux control (Table 2). The addition of a Dor fundoplication reduces the risk of pathologic gastroesophageal reflux by 9-fold as tested by 24-hour esophageal pH monitoring. Even the median esophageal acid exposure was lower in the Heller-Dor group, indicating that the few unfortunate individuals in whom reflux might occur are easily managed with

medical therapy^[25].

The predominant mechanism of failure after the Heller operation is an incomplete distal myotomy. In such circumstances, pneumatic dilatation can represent a reasonable therapeutic option when dysphagia is mild. In patients with major symptoms, revisional laparoscopic surgery with repeat myotomy and fundoplication is feasible and effective with a low morbidity rate^[26]. Intraoperative endoscopy is helpful as a guide during dissection to identify the cleavage plans and to clearly recognize the gastroesophageal junction. When a properly performed myotomy has failed in a patient with sigmoid esophagus, and the redundant supradiaphragmatic esophageal loop still interferes with emptying, a transthoracic or a transhiatal esophagectomy is the treatment of choice^[27].

It has recently been suggested that a laparoscopic myotomy extended for 3 cm on the gastric side can result in better dysphagia scores and in fewer additional procedures to treat recurrent dysphagia, provided that a Toupet fundoplication is added to the procedure^[28]. However, this improved outcome should be balanced against the risk of severe gastroesophageal reflux disease induced by the longer gastric myotomy and the necessity to fully mobilize the cardia to perform a posterior fundoplication. Interestingly, a very long-term study of 67 patients who underwent an open Heller-Dor operation and were followed for more than 20 years has shown a progressive clinical deterioration of results and a 22.4% failure rate due to an increased esophageal acid exposure and development of Barrett's esophagus^[29], suggesting once again that surgical therapy for achalasia involves a very delicate balance between relief of outflow obstruction and destruction of the antireflux barrier.

Robot-assisted Heller operation has been performed with satisfactory outcomes and no mucosal perforation^[30]. Operative times were similar to those of standard laparoscopic operation after the first 30 cases in a multicenter study^[31]. No data from randomised clinical trials have been reported yet. At present time, despite the advantage of the three-dimensional view, elimination of tremor, and 360 degree of freedom of movement of the robotic arms, the role of this technology in the management of esophageal achalasia remains to be determined.

CONCLUSIONS

The management of achalasia has changed significantly over the past 15 years. Minimally invasive surgery has influenced the management of esophageal disease more than any other gastrointestinal disorder and in a manner similar as in the therapy of cholelithiasis. In several institutions around the world, the laparoscopic surgical myotomy has replaced pneumatic dilation as the initial therapy of choice for achalasia. Current evidence from the literature suggests that it is conceivable to extend the Heller myotomy on the gastric side for about 2 cm to encompass the oblique fibers without fear of inducing significant gastroesophageal reflux, provided that a minimal antero-lateral dissection of the phrenoesophageal membrane is performed and a Dor fundoplication is routinely added to the myotomy.

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