

Surgical Treatment of Esophageal Achalasia in the Era of Minimally Invasive Surgery

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ABSTRACT

Introduction: We have analyzed the short- and long-term results of various surgical therapies for achalasia, especially changes in postoperative esophageal function.

Patients and Methods: Between January 1, 2008 and December 31, 2017, 54 patients with esophageal achalasia were treated in our institution. Patients scheduled for surgery underwent a comprehensive gastroenterological assessment pre- and post-surgery. Forty-eight of the elective cases involved a laparoscopic cardiomyotomy with Dor's semifundoplication, while two cases entailed an esophageal resection with an intrathoracic gastric replacement for end-stage achalasia. Terek's operation was performed on two patients for iatrogenic esophageal perforation, and two others underwent primary suture repair with Heller–Dor surgery as an emergency procedure. The results of the different surgical treatments, as well as changes in the patients' pre- and post-operative complaints were evaluated.

Results: No intra-operative complications were observed, and no mortalities resulted. During the 12 to 24-month follow-up period, recurrent dysphagia was observed mostly in the spastic group (TIII: 33%; diffuse esophageal spasm: 60%), while its occurrence in the TI type did not change significantly (14.5%–20.8%). As a result of the follow-up of more than two years, good symptom control

was achieved in 93.7% of the patients, with only four patients (8.3%) developing postoperative reflux.

Conclusions: The laparoscopic Heller–Dor procedure provides satisfactory long-term results with low morbidity. In emergency and advanced cases, traditional surgical procedures are still the recommended therapy.

Key Words: Esophageal achalasia, Minimally invasive surgery, Myotomy, Megaesophagus, Iatrogenic esophageal injury.

INTRODUCTION

First described by Sir Thomas Willis in 1674, achalasia is a chronic motility disorder of the esophagus characterized by a lack of peristalsis and the inability of the lower esophageal sphincter (LES) to relax.^{1,2} Despite being rare, it is the most common primary motor disorder of the esophagus with an incidence of 1/100,000 and a prevalence of 10/100,000;³ no gender predominance can be observed. It may be developed at any age, but it occurs most commonly in the third to fifth decade of life. The etiology of achalasia is still unclear, but, ultimately, it is a selective disorder of the inhibitory neurons in the myenteric (or Auerbach's) plexus of the distal esophagus and the LES. In most cases, clinical presentation is dominated by progressive dysphagia, regurgitation, and chest/epigastric pain, with heartburn and coughing at night also common. In gastroenterological diagnostics, functional assessments play a primary role. Currently, the new high-resolution manometry (HRM), which is becoming the gold standard, facilitates an accurate diagnosis, which may consist of the following abnormal esophageal processes: aperistalsis, abnormal LES relaxation, and a dilated esophagus. HRM makes it possible to distinguish three types of esophageal motility disorder, which can be classified based on the Chicago Classification.⁴ Type I (which corresponds to the previous classic type) refers to aperistalsis of the esophageal body and a relaxation disorder of the LES. In type II (compression), the morphology of waves in the esophageal body is the same as that seen in

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type I, but the amplitudes exceed 30 mmHg. In type III (spastic), tall (> 200 mmHg), wide contractions can be seen in the distal esophagus.

The treatment for achalasia is palliative only, and all therapeutic efforts are aimed at facilitating adequate passage through the cardia and, at the same time, preventing late structural and functional esophageal complications. Non-surgical therapies for achalasia include: local administration of smooth muscle relaxants and botulinum toxin, which has a lower efficacy; endoscopic balloon dilation (EBD), which is popular for its efficacy; and the new, highly promising peroral endoscopic myotomy (POEM) used primarily in East Asia and at a few Western centers.

The safest and most effective treatment for achalasia is still surgery, which involves cutting the muscle fibers of the abnormally functioning lower esophageal sphincter (a Heller cardiomyotomy). The minimally invasive (laparoscopic and thoracoscopic) types of Heller myotomy were introduced into clinical practice in the early 1990s.^{5,6} Results from the first thoracoscopic esophagotomy performed for achalasia in Hungary were published by staff at the Department of Surgery, University of Szeged (Hungary); the procedure was well tolerated by the patients, and their swallowing functions improved.⁷

To reduce the risk of gastroesophageal reflux disease (GERD) following a cardiomyotomy, which had previously been used alone, the procedure was later completed with partial fundoplication (anterior – Dor, or posterior – Toupet). This modified laparoscopic Heller cardiomyotomy completed with semifundoplication has proved to be the most effective procedure, with minimal morbidity, both in the short and long term.⁸

Management of advanced and emergency cases forms a separate group in the treatment of achalasia. Without proper therapy, in the case of inadequate treatment or as a result of the natural progression of the disease, esophageal dilation, deformity, and at the same time, loss of function occur, leading to an end-stage disease in about 5% of cases.⁹ In these advanced cases, esophageal resection ensures the best results. The most serious complication in the endoscopic treatment of achalasia is esophageal perforation, which also requires surgical therapy. The type of surgical intervention (sutures with a myotomy and resection) and the other endoscopic therapeutic modalities are determined by the stage of the underlying disease, the general condition of the patient and the time from perforation to treatment.

In this paper, we analyze our experience with the complex surgical treatment of esophageal achalasia, emphasizing the choice of a proper treatment strategy and type of surgery, as well as the long-term changes in quality of life.

PATIENTS AND METHODS

Between January 1, 2008 and December 31, 2017, 54 patients (24 males and 30 females aged 17 to 79 years) with symptomatic esophageal achalasia were treated at the Department of Surgery, University of Szeged (Hungary). The most common complaints among the patients scheduled for surgery included dysphagia, solid food becoming stuck, epigastric pain, and less frequently, weight loss. The mean duration of the symptoms was 57.3 months (3–192). After a detailed history was taken, patients underwent a comprehensive gastroenterological assessment (a swallowing X-ray examination, upper gastrointestinal endoscopy, esophageal pH-metry, and manometry). Based on the Chicago Classification, the types of achalasia in the laparoscopic group were as follows: TI, TII, and TIII achalasia was found in 30, 3, and 9 cases, respectively; diffuse esophageal spasm (DES) was confirmed in 5 cases; and Jackhammer esophagus was observed in 1 patient. Clinical presentation of megaesophagus was diagnosed in 4 cases. Among the patients in the laparoscopic group, 18.7% (9/48) and all the patients in the acute and the reconstruction groups (4/4, 100%; 2/2, 100%) were treated with pre-operative endoscopic balloon dilation.

Surgical Treatment Elective Laparoscopic Heller–Dor Surgery

Forty-eight patients underwent a laparoscopic Heller cardiomyotomy and Dor's anterior partial fundoplication. With the supine patient in a reverse Trendelenburg position, ports were inserted into the abdominal cavity: three ports, with 15-cm intervals, along the left costal margin; one port in the epigastric region, to the right of the midline; and one port (camera port) of 10 to 12 mm in diameter directly above the umbilicus. After creating a pneumoperitoneum, the abdominal, lower mediastinal segment of the esophagus was mobilized, maintaining an intraabdominal pressure of 15 mmHg. Esophageal mucosa integrity was checked with intraoperative endoscopy in each case. A Heller esophago-cardiomyotomy was performed over a length of 8 cm on the anterior wall of the

esophagus, and over at least 2 cm on the gastric fundus, completed with Dor's partial anterior fundoplication.

Elective Esophageal Resection

Megaesophagus patients underwent surgery after bowel preparation, and ulcer, thrombosis, and antibiotic prophylaxis. Gastric replacement involved an upper midline laparotomy, widening the esophageal hiatus and then mobilizing the esophagus through the hiatus. After mobilizing the stomach and ligating the left gastric artery, a gastric conduit was created as per Akiyama using linear staplers along the lesser curvature, and then a jejunal feeding catheter was implanted. After drainage, the abdominal section was closed, the patient was turned to a left lateral decubitus position, and the esophagus was subtotally resected through a right anterolateral thoracotomy with selective intubation. The gastric conduit was pulled from the abdominal cavity through the enlarged hiatus into the thorax, where an anastomosis was performed between the esophagus and the stomach with a circular stapler.

In a patient who had previously undergone a Heller–Toupet operation and then developed recurrent symptoms and megaesophagus, the previous fundoplication was eliminated, the lower third of the esophagus and the cardia were resected, and they were replaced with an isoperistaltic jejunal segment positioned under the azygos vein (the Merendino procedure).

Emergency Surgical Interventions

Four patients underwent emergency surgery for an iatrogenic esophageal perforation due to EBD. In two cases, primary suture repair and Heller–Dor surgery were performed with traditional open surgery, using intraoperative endoscopic control, in non-septic patients with early-stage achalasia (within 8 hours). In another two emergency cases, iatrogenic perforation of megaesophagus was confirmed. More than 24 hours had passed between the injury and the surgical treatment, and the patients were in a severe septic condition at the time of surgery. In one of the patients, the perforation was caused by a diagnostic esophagoscopy performed at another institution, while the other patient developed a rupture after EBD, which was followed by two unsuccessful attempts at endoscopic clipping. Both patients underwent an esophagectomy as per Torek, a gastrotube was used for decompression purposes, and a jejunal feeding catheter was implanted.

Reconstructive Surgery

Ninety-nine and 122 days after Torek's esophageal resection, successful substernal reconstruction was performed using the right colon and the stomach, respectively.

Postoperative Care

For the postoperative period, patients in the laparoscopic group were transferred to the surgery unit after recovery from anesthesia. Parenteral fluid therapy was administered during the postoperative period. Enteral feeding was gradually introduced after the swallowing X-ray examination was conducted on postoperative day 2 if nothing abnormal was detected. Patients in the elective esophageal resection group spent three days on average (two to four days) in our department's intensive care unit for close monitoring. Once they were stable, they were transferred to our ward, where they received total parenteral feeding. A swallowing X-ray examination with a water-soluble contrast agent routinely followed on postoperative day 7. If the leak test was negative, the protocol for the gradual introduction of enteral feeding was the same as in the laparoscopic group.

Follow-up

Patients treated with a laparoscopic myotomy were given gastroenterological check-ups (a swallowing X-ray, esophageal manometry, pH-metry, and esophago-gastros-copy) an average of three months after the surgery – these assessments were carried out in 37 patients. Regular follow-up occurred with a total of 27 patients, while 10 patients did not return for the periodic follow-up visits after the initial period (months 2 and 6 post-surgery), although nine of them were complaint-free.

ETHICS

The study was registered with Regional Human Biomedical Research Ethics Committee, with the identifier 4827.

RESULTS

Postoperative Results

The average duration of the laparoscopic procedures was 72 (62–90) minutes with minimal blood loss (50 to 100 mL). No intraoperative complications were observed, and conversion was required in one case (1/48, 2%) for adhesions. The swallowing test conducted with a water-

Table 1.
Perioperative Data and Outcomes of Different Achalasia Surgeries

Indication for Surgery	Surgery (N)	Age (Mean, Years)	Type of Surgery	Timing of Surgery	Morbidity (%)	Mortality (%)	Hospital Stay (Mean, Day)
Achalasia (early stage)	48	46.2	Laparoscopic Heller-Dor	Elective	2% (1/48)	0	7.3
Achalasia (advanced stage, mega-esophagus)	2	43.3	Esophageal resection with gastric, jejunal or colonic replacement	Elective	0,0%	0	18.6
Achalasia, iatrogenic injury, early diagnosis (< 24 hours)	2	65.5	Primary suture	Emergency	50% (1/2)	0	16.5
Achalasia, iatrogenic injury, late diagnosis (> 24 hours)	2	60.5	Total esophagectomy (Torek's operation)	Emergency	50% (1/2)	0	15.5
Late reconstruction after Torek operation	2	60.5	Reconstructive surgery with gastric or colonic replacement	Elective	0,0%	0	14.5

soluble contrast agent (Gastrografin®) on postoperative day 4 (on average) revealed a leak from the site of the sutures in one case (1/48, 2%), which was treated with emergency re-operation and suture placement. The average length of stay was 7.3 (5–28) days in the elective, laparoscopic group.

There was one case of hydrothorax formation requiring a puncture and one case of atrial fibrillation in the emergency surgery group. There were no mortalities. Patients with primary suture repair were discharged after 16.5 (13–20) days, following a swallowing X-ray with normal results, while those who had undergone Torek's operation were discharged after 15.5 (14–17) days. Later, the reconstructive surgeries performed with a colon or gastric pull-up after Torek's operation were accompanied by neither intraoperative nor postoperative complications, and patients were discharged on day 14.5 after a swallowing X-ray with normal results and gradually introduced oral feeding (**Table 1**).

Long-term follow-up

At the 1 to 6-month follow-up visit, all the patients, except for one, reported unrestricted swallowing, which was also confirmed by functional assessments. Those returning for later gastroenterological check-ups were evaluated based on follow-up intervals and their swallowing function (**Table 2**).

At the 6 to 12-month follow-up visit, non-severe recurrent dysphagia was reported in eight patients. In two cases, dietary changes and medical treatment resulted in notable improvement, while persistent complaints were recorded

despite therapy in two others. EBD was required in an additional four cases (8.3%), with one of these patients requiring esophageal resection for recurrent complaints despite the EBD, considering the patient's young age and the significantly dilated esophagus. At the 12 to 24-month visit, the number of patients being followed up for dysphagia (8) had not changed, but there were three new cases. Medical therapy was successful in one, and a successful EBD was performed in another; however, one patient had persistent complaints. During the follow-up visits after 24 months, a total of seven patients were followed up for dysphagia, one of them being a new patient, who became complaint-free after conservative therapy. Those with complaints despite surgery mostly suffered from spastic motility disorders (TIII and DES).

Postoperative reflux did not occur during the 6–12-month follow-up period, while it developed in three patients in total (3/48, 60.2%) during the 12–24-month and >24-month follow-up; however, it was controlled well medically.

Our study also revealed that at the 12 to 24-month follow-up visits, symptomatic, and symptom-free patients had undergone surgery at approximately the same age (53.5 years vs. 48.1 years), and the duration of symptoms was longer in symptomatic patients (20.3 months vs. 112.8 months). However, there was no difference in pre-operative EBD (symptom-free 11% vs. 10%). During the > 2-year follow-up, there was still no difference in age (53.1 years vs. 54.8 years), and patients with satisfactory symptom control underwent surgery later than those in the symptomatic group (82.2 months vs. 40.5 months).

Table 2.
Follow-up Data after Laparoscopic Heller-Dor Procedure

Follow-up Interval	First Follow-up	6–12 Months	12–24 Months	> 24 Months
Proportion of patients with complaint	1/48 (2%)	8/48 (16.6%)	10/48 (20.8%)	9/48 (18.7%)
Type I	1/30 (3.3%)	5/30 (16.6%)	4/30 (13.3%)	4/30 (13.3%)
Type II	0/3 (0%)	0/3 (0%)	0/3 (0%)	0/3 (0%)
Type III	0/9 (0%)	3/9 (33.3%)	3/9 (33.3%)	2/9 (22%)
Diffuse esophageal spasm	0/5 (0%)	0/5 (0%)	3/5 (60%)	3/5 (60%)
Jackhammer	0/1 (0%)	0/1 (0%)	0/1 (0%)	0/1 (0%)
Postoperative reflux	0/48 (0%)	0/48 (0%)	3/48 (6.2%)	3/48 (6.2%)
Postoperative dysphagia	1/48 (2%)	8/48 (16.6%)	8/48 (16.6%)	7/48 (14.5%)

DISCUSSION

The treatment strategy for patients with achalasia in centers specializing in esophageal diseases is determined in close collaboration among gastroenterologists and surgeons. In addition to the ever growing variety of effective medical interventions, patients with persistent symptoms may undergo surgery at different stages and with different timings. The first documented surgery for achalasia was performed by Ernst Heller in 1913; it was an extramucosal myotomy at the level of the cardia, thus reducing the pressure of the LES and facilitating the passage of solid food into the stomach.¹⁰ Originally, Heller recommended a double (anterior and posterior) myotomy; however, a simple myotomy, a procedure still used today, was described by Zaaijer, a Dutch surgeon, in 1923.¹¹ The traditional open surgeries of the esophagus (via thoracotomy or laparotomy) have considerable morbidity rates. To reduce them, the minimally invasive surgical technique is currently an excellent alternative in the surgical treatment of the functional disorders of the esophagus, including esophageal achalasia. Our team has published several papers on its favorable results in the minimally invasive surgical treatment of benign esophageal disorders.^{7,12–16}

Nowadays, the laparoscopic trans-hiatal technique is one of the most accepted minimally invasive procedures in the surgical treatment of esophageal achalasia. The procedure is associated with low morbidity and ensures a long-term symptom-free condition in a considerable number of cases.¹⁷ In addition to the well-known benefits of laparoscopy (reduced postoperative pain, shorter hospital stay, and better cosmetic results), a further favorable factor is that the lower third and the abdominal segment of the esophagus can be well explored through the hiatus and

that, technically, a myotomy and antireflux surgery can be performed with precision.

Beyond an accurate diagnosis, medical treatment for achalasia is determined by the physical capacity of the patient and their response to the therapies. Smooth muscle relaxants and botulinum toxin injected into the lower esophageal sphincter may reduce dysphagia; however, their effect is only temporary, and they may make a later Heller myotomy more difficult.^{18–20} The efficacy of EBD is between 70% and 80%,²¹ and newer comprehensive studies have confirmed a perforation rate of less than 1%, which equals the rate of perforations not noticed during Heller surgery.²² Although the rate of a favorable clinical response to a surgical myotomy is better than that after EBD, serial EBD may be an appropriate alternative to surgical treatment.²³ In a prospective, randomized study conducted by Moonen et al., nearly the same success in dysphagia control was demonstrated after five years; however, this was only achieved after multiple dilations in one-quarter of the EBD group.²⁴ Persson et al. demonstrated a significantly higher five-year symptom-free rate after LHM than after EBD (95% and 65%, respectively),²⁵ and the same trend was confirmed by three different meta-analyses.^{17,26,27} When assessing the effect of different clinical parameters on therapy, it was also shown that patients below the age of 45 years benefited more from the surgical treatment than from EBD.²⁸

POEM, which requires a serious learning process and a special surgical environment, is also not a clear alternative to minimally invasive surgical treatment.^{29,30} One of the main concerns with POEM is the high rate of gastroesophageal reflux, which develops after treatment. It appears, however, that this method is useful with type III achalasia,

Table 3.
Landmark Clinical Studies Regarding Laparoscopic Heller-Dor Operation

Author	Year	Study Design	Procedure Type	Sample Size (N)	Follow up (month)	Complication Rate (%)	LOS (Day)	Success Rate (%)	Postoperative GERD (%)	Postoperative Dysphagia (%)
Ancona ³³	1995	RC	LHMD	17	6	0%	4	94.2%	0%	5.8%
			OHMD	17	6	0%	10	94.2%	5.8%	0.0%
Richards ³⁹	2004	RCT	LHMD	22	6	0%	1	NA	by pH: 9.1%	0.0%
			LHM	21	6	0%	1	NA	by pH: 47.6%	
Boeckxstaens ²³	2011	RCT	LHMD	106	24	12%	NA	90%	by pH: 23%, by EGD: 21%	6.6%
			EBD	95	24	4%	NA	86%	by pH: 15%, by EGD: 19%	
Moonen ²⁴	2016	RCT	LHMD	105	60	11%	NA	84%	by pH: 34% by EGD: 18%	NA
			EBD	96	60	5%	NA	82%	by pH: 12% by EGD: 14%	
Costantini ²⁹	2019	CCS	LHMD	140	24	2.1%	3	95.7%	by pH: 17.1%, by EGD: 15.2%	NA
			POEM	140	24	5%	2	99.3%	by pH: 38.4%, by EGD: 37.4%	
Werner ³⁰	2019	RCT	LHMD	109	24	7.3%	NA	81.7%	by pH: 30%, by EGD: 29%	NA
			POEM	112	24	2.7%	NA	83%	by pH: 30%, by EGD: 44%	
Costantini ⁴⁵	2018	RCT	LHMD	1001	62	4.7%	3	89.5%	by pH: 9.1%, by EGD: 11.6%	NA
Rawlings ⁴²	2012	RCT	LHMD	36	12	5.6%	NA	90.9%	by pH: 41.7%	8.3%
			LHMT	24	12	8.3%	NA	93.1%	by pH: 21.1%	4.1%
Torres-Villalobos ⁴¹	2018	RCT	LHMD	38	24	2.6%	2.5	100%	by pH: 10.5%	NA
			LHMT	35	24	0%	2.5	90%	by pH: 31.5%	NA
Kumaga ⁴⁰	2014	RCT	LHMD	19	12	0%	2	90.9%	by pH: 18%	18.0%
			LHMT	22	12	4.5%	2	85.7%	by pH: 38%	14.0%
Rebecchi ⁴⁴	2008	RCT	LHMD	72	60	2%	3.2	97%	symptoms: 5.6%, by pH: 2.8%	2.8%
			LHMN	72	60	1%	3.6	85%	symptoms: 0%, by pH: 0%	15.0%

RCT, randomized controlled trial; RC, retrospective cohort; CCS, case control study; OHMD, open Heller Myotomy with Dor semifundoplication; LHMD, laparoscopic Heller myotomy with Dor semifundoplication; LHMT, laparoscopic Heller myotomy with Toupet semifundoplication; LHMN, laparoscopic heller myotomy with Nissen fundoplication; EBD, endoscopic balloon dilation; POEM, per-oral endoscopic myotomy; LOS, length of stay; GERD, gastro-esophageal reflux disease; EGD, esophago-gastro-duodenoscopy; NA, not available.

where it can ensure a longer myotomy than standard LHM; at the same time, the efficacy of LHM is almost 85% here as well.³¹ Another possible future indication for this procedure is recurrent and unsuccessful cases.³²

LAPAROSCOPIC HELLER–DOR SURGERY

Laparoscopic Heller–Dor surgery was introduced at the end of the last century and since then has become the gold standard in the surgical treatment of esophageal achalasia.³³ It has excellent mortality and morbidity rates of 0.01% and 6%, respectively,¹⁷ (**Table 3**) and it provides a long-term symptom control rate of about 90%, the success of which also depends on the Chicago Classification.³¹ (**Table 3**) Mucosal injury may occur during the procedure in 6.9% (0–33%) of cases, and it may remain hidden in most cases or may be treated immediately during surgery.¹⁷ (**Table 3**)

Based on our own results, it can be established that the success rate beyond 24 months is 85.5%, which can be considered 93.7% with the supplementary conservative medical treatment of symptomatic patients. In the patient group studied, one patient in total (1/48, 2%) developed a surgery-related complication, esophageal mucosal lesion, which was discovered with the swallowing X-ray performed with a water-soluble contrast agent on postoperative day 1. The injury was supposedly caused by intraoperative thermal damage, which was not seen during the endoscopic follow-up examination after the primary surgery. No complications were observed in the other 47 patients (98%) in the group, and there was no mortality. All in all, the clinical results of this study are clearly consistent with international standards, considering both long-term symptomatic control and the morbidity rate.

As to LHM, there are two issues to be discussed: achieving a symptom-free status post-surgery (eliminating dysphagia) and the course of postoperative reflux. Both factors can basically be traced back to the proper performance of the myotomy. The 2018 International Society of Diseases of the Esophagus guidelines on achalasia state that a laparoscopic Heller myotomy is recommended over a length of at least 6 cm on the esophagus and 2 to 3 cm on the stomach for effective control of symptoms.³⁴ Two publications reported a myotomy that extended 3 cm onto the stomach, which reduced the risk of delayed dysphagia.^{35,36} Proximally, a myotomy of 6 to 8 cm in length is recommended in general, but no comparative publications are available on the length of esophageal

myotomies.³⁷ There is also a physiological basis to the proper cutting of the fibers – the high-pressure zone of the cardia is generally slightly shorter than 4 cm and extends 2 cm from the Z-line in the oral direction.

Based on our previous clinical study, it is clear that an inadequate myotomy, either in the aboral or the oral direction, may cause recurrent symptoms, which can be corrected with repeat surgery.³⁸ Our current study also demonstrates that recurrent symptoms are more common in patients with a spastic-type esophageal disorder (TIII achalasia and DES) than in those with TI or TII disease. Our results are expressive primarily after 12 months, although symptoms returned earlier among TIII cases (3/9, 33%). The trend continues in the 1 to 2-year follow-up period, since the rate of symptomatic patients is relatively high in the spastic group (TIII: 33%; DES: 60%), while in the case of the classic form, it does not change much when analyzing the time intervals (6–12 months: 14.5%; 12–24 months: 20.8%; > 24 months: 18.7%). We may thus conclude that, in certain cases, the increased tone of the esophagus may extend well above the level of the LES, where conventional and a properly performed myotomy cannot always reach.

The other myotomy-related complaint is the development of GERD. Based on observations by Campos and other authors, reflux occurs in 41.5% without an antireflux procedure and only in 14.5% with one, thus confirming that if the LES, the main barrier, is damaged, reflux may be expected.^{17,39} By completing the procedure with partial fundoplication, the occurrence of postoperative reflux can be decreased considerably, without increasing the pressure of the LES. Both anterior (Dor, 180°) and posterior (Toupet, 270°) semifundoplication are used widely after a cardiomyotomy. There is an argument between the supporters of Dor and Toupet which fundoplication is better. Those experts who are for Dor say that anterior fundoplication is easier to perform the non-dissection of the posterior part of the esophagus may help against GERD. However, the supporters of Toupet state it may keep the edges of the myotomy separated, reducing the probability of recurrent dysphagia and reducing development of GERD. Comparing these two methods there were no significant differences between the three randomized control trials and their meta-analysis regarding the postoperative dysphagia and GERD.^{40–44} (**Table 3**) Their use is determined by the preference of the surgical team. In a recent, large prospective clinical trial, laparoscopic Heller–Dor procedure has proved to be successful regarding acceptable low morbidity (4.7%) and durable symptom control (89.5%) on long-term in 1001 achalasia patients.⁴⁵

Based on our results, it is clear that the rate of GERD after a Heller–Dor procedure was minimal (6.2%, 3/48) in the medium term (24-month follow-up), and was controlled with conservative therapy.

PERSISTENT SYMPTOMS

Following surgical treatment of achalasia, there may be various mechanisms for persistent or recurrent cases, and the often unavoidable reoperations may require other surgery types (remyotomy, esophageal resection and total esophagectomy), on which our team has previously reported.³⁷ Nowadays, owing to precise diagnostic methods, misdiagnoses are rare, and inadequate surgery types that decrease patients' quality of life have also disappeared from the surgeon's repertoire. The proper performance of a myotomy is of great importance because a cardiomyotomy that is insufficient in length or depth may be the source of recurrent symptoms and, in certain cases, a secondary epiphrenal diverticulum may call attention to unsuccessful previous treatment. A myotomy without fundoplication is of historical importance, and the severe GERD and resultant esophageal stricture that develop later may require resection surgery.³⁸

MEGAESOPHAGUS

The risk of esophageal cancer is increased by long-lasting achalasia, marked esophageal dilation, and mucus congestion.^{46,47}

The best way to prevent cancer is timely treatment – primarily surgical therapy. If a cardiomyotomy is performed late, that is, after a sigmoid deformity of the esophagus has developed, effective cancer prevention can no longer be achieved; what is more, the results of radical surgery are also unsatisfactory because of the late recognition.^{48,49}

In the megaesophagus stage, the risk of aspiration pneumonia, malignant transformation, and malnutrition is markedly high. Most patients have undergone innumerable endoscopic and/or surgical procedures, and indications for surgery are continuously recurrent symptoms and the sigmoid deformity of the esophagus. The morbidity rate for radical intervention varies between 19% and 50%, with the most common complications being pneumonia and anastomotic leak. The mortality rate is between 0 and 5.4%, the length of stay is 10 to 16 days, and slightly more than one-quarter of patients will later require endoscopic balloon dilation for anastomotic stricture.⁵⁰ All our patients scheduled for elective resection

had an afunctional, significantly dilated esophagus, and their number (3/54, 5%) corresponded to the literature data. Great care was taken to individualize the type of surgery during surgical treatment, and this decision was made by the surgical team with experience of esophageal surgery after considering the patient's physical condition, the intraoperative characteristics and the long-term optimal quality of life. In the case of reconstruction, all three eligible organs were used (stomach, jejunum, and colon), neither an anastomotic leak nor pneumonia was observed after the interventions, and there was no mortality. The swallowing function of these patients is now satisfactory, and no anastomotic stricture or other complications were found during the follow-up visits.

It was long maintained that the only surgical treatment possible for megaesophagus is esophageal resection, and stomach, jejunum, or colon was used as a replacement.⁵¹ Because of the surgical burden and the relatively high rate of peri-operative morbidity, a cardiomyotomy, which may result in symptomatic improvement, could become prevalent in high-risk cases. A number of authors have reported a noticeable improvement in symptoms even in these decompensated patients with sigmoid esophagus.^{52–55} Both postoperative functional assessments of the patients and quality-of-life questionnaires confirm the justification and usability of a myotomy. Mineo's team performed a Heller myotomy in 14 achalasia patients with sigmoid deformity of the esophagus. After 85 months of follow-up, the result was excellent or good in 72% of the cases, while the postoperative dysphagia and regurgitation scores significantly decreased and matched those observed in achalasia patients operated on in the early stage.⁵² In patients undergoing surgery for megaesophagus, numerous publications have confirmed the efficacy of LHM in achieving postoperative symptom control, and resection was not necessary in any of the patients as a result of a persistently good quality of life.^{54,55}

The fact that LHM is not always effective in the treatment of sigmoid esophagus was demonstrated by Zaninotto et al. in their analysis of more than 400 cases involving a myotomy for esophageal achalasia. At the end of their long-term, prospective study, they concluded that a high preoperative LES pressure has a beneficial effect on the outcome of the surgery, while a stage IV (sigmoid) esophagus has an adverse effect. Radical esophagectomy is often unavoidable because of the persistent symptoms, but, after informing the patient properly, a minimally invasive myotomy is worth pursuing as a first step in the hope of a positive response to therapy.³¹ In our own patient population, a young woman with sigmoid esophagus underwent LHM after multiple unsuccessful

endoscopic dilations, and satisfactory swallowing function was observed for almost six months. However, since dysphagia returned after the complaint-free period, esophageal resection with jejunal interposition (the Merendino procedure) was performed. At present, after more than a decade of follow-up, the patient is completely symptom-free, her swallowing is unrestricted, and her quality of life is excellent.

POSTENDOSCOPIC ESOPHAGEAL INJURY

In large centers, a perforation rate of about 1%, as detailed above, should be expected while the patient is treated with EBD as nonsurgical treatment of achalasia. Performing the intervention on a prepared patient and timely recognition of the lesion by an experienced gastroenterologist are important factors in the patient's emergency surgery. The mortality rate for esophageal perforation ranges between 18% and 22%—even despite early recognition and treatment. If the time elapsed between injury and surgery exceeds 24 hours, the mortality rate may even reach 27 to 40%.⁵⁶ In the case of esophageal injury, a personalized treatment strategy is required in each case, and the following factors should be taken into account: etiology of the injury, existing underlying esophageal disease, time from injury to diagnosis, septic condition, comorbidities, and physical capacity.⁵⁷ Post-EBD esophageal perforations are traditionally treated surgically, but therapeutic methods also include conservative treatment and modern endoscopic techniques (over the scope clip and stent implantation).^{58–61}

Basically, early surgical treatment is required for larger lesions that cannot be treated endoscopically in the case of a contrast agent leak into the pleural and/or peritoneal space. Many authors agree that a similar decision is warranted with the involvement of a surgeon as soon as possible even for smaller lesions.⁶² Time elapsed since a perforation has a considerable effect on the success of the surgery, although there is experience with both early and later (> 24 hours) successful laparoscopic primary closures in the literature.^{62,63} In our practice, primary suture repair is used in cases that are recognized early, within 24 hours, and it is always completed with the cardiomyotomy and antireflux procedure, which has a beneficial effect on the healing of the lesion and, at the same time, may result in long-term symptom-free status.

In the case of perforations beyond 24 hours, patients usually undergo resection without reconstruction (on rare occasion, with immediate reconstruction) because of the septic condition that has developed and the reduced tendency of the esophageal wall to heal. In the case of megaesophagus, a

worse-than-average condition of the esophagus supports resection which is worth including during surgical treatment in all cases. In tertiary centers with experience of esophageal resection, mortality rates similar to that of primary suture repair can be achieved (17% [0–43%] vs. 12% [0–31%]).^{64–65} The importance of a multidisciplinary approach and a correctly chosen treatment strategy is highlighted by a study conducted at our department, which summarized cases of spontaneous esophageal perforation and demonstrated an acceptably low mortality rate (6.6%) owing to timely and well executed procedures.⁶⁶

Every patient in our acute surgery group was admitted to our unit for an esophagoscopy-associated injury. The two early cases (recognized within 24 hours), primary suture repair occurred with Heller–Dor surgery, taking the condition of the patients into account, while esophageal resection without reconstruction was performed in the two other cases with an old perforation complicated by megaesophagus. There was no mortality or notable morbidity in the emergency group.

CONCLUSION

Laparoscopic Heller–Dor surgery is a safe and effective surgical method for treating esophageal achalasia. Symptom control in patients who have undergone minimally invasive surgery is adequate even in the long term, and the rate of postoperative reflux is low. However, patients with the spastic type may develop recurrent symptoms at a higher rate. Advanced and emergency conditions are still a major challenge for surgeons, and choosing the proper therapeutic strategy depends on several factors. On the whole, surgery for achalasia is only recommended in institutions where every aspect of the condition can be managed effectively and reliably.

References:

1. Krill JT, Naik RD, Vaezi MF. Clinical management of achalasia: current state of the art. *Clin Exp Gastroenterol*. 2016;9:71–82.
2. Moonen A, Boeckxstaens G. Current diagnosis and management of achalasia. *J Clin Gastroenterol*. 2014;48(6):484–490.
3. Sadowski DC, Ackah F, Jiang B, et al. Achalasia: incidence, prevalence and survival: a population based study. *Neurogastroenterol Motil*. 2010;22(9):e256–e261.
4. Kahrilas PJ, Bredenoord AJ, Fox M, et al. International High Resolution Manometry Working Group. The Chicago Classification of esophageal motility disorders, v3.0. *Neurogastroenterol Motil*. 2015;27(2):160–174.

5. Shimi S, Nathanson LK, Cuschieri A. Laparoscopic cardiomyotomy for achalasia. *J R Coll Surg Edinb.* 1991;36(3):152–154.
6. Pellegrini C, Wetter LA, Patti M, et al. Thoracoscopic esophagomyotomy. Initial experience with a new approach for the treatment of achalasia. *Ann Surg.* 1992;216(3):291–296.
7. Olah T, Szendrényi V, Wittmann T, et al. Treatment of achalasia of the cardia using thoracoscopic esophago-myotomy. *Orv Hetil.* 1997;138(1):11–13.
8. Litle VR. Laparoscopic Heller myotomy for achalasia: a review of the controversies. *Ann Thorac Surg.* 2008;85(2):S743–S746.
9. Vela MF, Richter JE, Wachsberger D, et al. Complexities of managing achalasia at a tertiary referral center: use of pneumatic dilatation, Heller myotomy, and botulinum toxin injection. *Am J Gastroenterol.* 2004;99(6):1029–1036.
10. Heller E. Extramuköse cardioplastik beim chronischen cardiospasmus mit dilatation des oesophagus. *Mitt Grenzgeb Med Chir.* 1913;27:141–149.
11. Zaaijer JH. Cardiospasm in the aged. *Ann Surg.* 1923;77(5):615–617.
12. András L, Paszt A, Simonka Z, et al. Laparoscopic surgery for epiphrenic esophageal diverticulum. *JLS.* 2018;22(2):e2017.00093.
13. András L, Ábrahám S, Simonka Z, et al. A comparative study of short- and long-term outcomes for transcervical versus transoral surgery for Zenker diverticulum. *Orv Hetil.* 2019;160(16):629–635.
14. Simonka Z, Paszt A, Ábrahám S, et al. The effects of laparoscopic Nissen fundoplication on Barrett's esophagus: long-term results. *Scand J Gastroenterol.* 2012;47(1):13–21.
15. Lázár G, Szentpáli K, Szántó I, et al. Successful thoracoscopic surgical treatment of esophageal cyst. *Acta Chir Hung.* 1999;38(2):191–192.
16. Lázár G, Oláh T, Szendrényi V, et al. Successful videothoracoscopic surgical treatment of esophageal cyst and leiomyoma. *Magy. Seb.* 1999;52:113–116.
17. Campos GM, Vittinghoff E, Rabl C, et al. Endoscopic and surgical treatments for achalasia: a systematic review and meta-analysis. *Ann Surg.* 2009;249(1):45–57. 45e57.
18. Gelfond M, Rozen P, Gilat T. Isosorbide dinitrate and nifedipine treatment of achalasia: a clinical, manometric and radionuclide evaluation. *Gastroenterology.* 1982;83(5):963–969.
19. Pasricha PJ, Rai R, Ravich WJ, et al. Botulinum toxin for achalasia: long-term outcome and predictors of response. *Gastroenterology.* 1996;110(5):1410–1415.
20. Smith CD, Stival A, Howell DL, et al. Endoscopic therapy for achalasia before Heller myotomy results in worse outcomes than heller myotomy alone. *Ann Surg.* 2006;243(5):579–584.
21. Hulselmans M, Vanuytsel T, Degreef T, et al. Long-term outcome of pneumatic dilation in the treatment of achalasia. *Clin Gastroenterol Hepatol.* 2010;8(1):30–35. 30e35.
22. Lynch KL, Pandolfino JE, Howden CW, et al. Major complications of pneumatic dilation and Heller myotomy for achalasia: single-center experience and systematic review of the literature. *Am J Gastroenterol.* 2012;107:1817–1825.
23. Boeckxstaens GE, Annese V, Varannes S. D, et al. European Achalasia Trial Investigators. Pneumatic dilation versus laparoscopic Heller's myotomy for idiopathic achalasia. *N Engl J Med.* 2011;364(19):1807–1816.
24. Moonen A, Annese V, Belmans A, et al. Longterm results of the European achalasia trial: a multicentre randomised controlled trial comparing pneumatic dilation versus laparoscopic Heller myotomy. *Gut.* 2016;65(5):732–739.
25. Persson J, Johnsson E, Kostic S, et al. Treatment of achalasia with laparoscopic myotomy or pneumatic dilatation: long-term results of a prospective, randomized study. *World J Surg.* 2015;39(3):713–720.
26. Yaghoobi M, Mayrand S, Martel M, et al. Laparoscopic Heller's myotomy versus pneumatic dilation in the treatment of idiopathic achalasia: a meta-analysis of randomized, controlled trials. *Gastrointest Endosc.* 2013;78(3):468–475.
27. Schoenberg MB, Marx S, Kersten JF, et al. Laparoscopic Heller myotomy versus endoscopic balloon dilatation for the treatment of achalasia: a network meta-analysis. *Ann Surg.* 2013;258(6):943–952.
28. Oude Nijhuis RAB, Prins LI, Mostafavi N, et al. Factors associated with achalasia treatment outcomes: Systematic review and meta-analysis. *Clin. Gastroenterol. Hepatol.* 2020;18(7):1442–1453.
29. Costantini A, Familiari P, Costantini M, et al. Poem versus laparoscopic Heller myotomy in the treatment of esophageal achalasia: a case-control study from two high volume centers using the propensity score. *J Gastrointest Surg.* 2020;24(3):505–515.
30. Werner YB, Hakanson B, Martinek J, et al. Endoscopic or surgical myotomy in patients with idiopathic achalasia. *N Engl J Med.* 2019;381(23):2219–2229.
31. Rohof WO, Salvador R, Annese V, et al. Outcomes of treatment for achalasia depend on manometric subtype. *Gastroenterology.* 2013;144(4):718–725.
32. Ngamruengphong S, Inoue H, Chiu PW, et al. Long-term outcomes of per-oral endoscopic myotomy in patients with achalasia with a minimum follow-up of 2 years: an international multicenter study. *Gastrointest Endosc.* 2017;85(5):927–933.e2.
33. Ancona E, Anselmino M, Zaninotto G, et al. Esophageal achalasia: laparoscopic versus conventional open Heller-Dor operation. *Am J Surg.* 1995;170(3):265–270.

34. Zaninotto G, Bennett C, Boeckxstaens G, et al. The 2018 ISDE achalasia guidelines. *Dis Esophagus*. 2018 Sep 1;31(9).
35. Mattioli S, Pilotti V, Felice V, et al. Intraoperative study on the relationship between the lower esophageal sphincter pressure and the muscular components of the gastro-esophageal junction in achalasic patients. *Ann Surg*. 1993;218:635–639.
36. Oelschlager BK, Chang L, Pellegrini CA. Improved outcome after extended gastric myotomy for achalasia. *Arch Surg*. 2003; 138(5):490–495.
37. Teitelbaum EN, Soper NJ, Pandolfino JE, et al. An extended proximal esophageal myotomy is necessary to normalize EGJ distensibility during Heller myotomy for achalasia, but not POEM. *Surg Endosc*. 2014;28(10):2840–2487.
38. Horváth ÖP, Lázár G. Failed operations for achalasia. *Dis Esophagus*. 1991;4:149–153.
39. Richards WO, Torquati A, Holzman MD, et al. Heller myotomy versus Heller myotomy with Dor fundoplication for achalasia: a prospective randomized double-blind clinical trial. *Ann Surg*. 2004;240(3):405–412.
40. Kumagai K, Kjellin A, Tsai JA, et al. Toupet versus Dor as a procedure to prevent reflux after cardiomyotomy for achalasia: results of a randomised clinical trial. *Int J Surg*. 2014;12(7):673–680.
41. Torres-Villalobos G, Coss-Adame E, Furuzawa C, et al. Dor versus Toupet fundoplication after laparoscopic Heller myotomy: long-term randomized controlled trial evaluated by high-resolution manometry. *J Gastrointest Surg*. 2018;22(1):13–22.
42. Rawlings A, Soper NJ, Oelschlager B, et al. Laparoscopic Dor versus Toupet fundoplication following Heller myotomy for achalasia: results of a multicenter, prospective, randomized-controlled trial. *Surg Endosc*. 2012;26(1):18–26.
43. Siddaiah-Subramanya M, Yunus RM, Khan S, et al. Anterior Dor or posterior Toupet with Heller myotomy for achalasia cardia: A systematic review and meta-analysis. *World J Surg*. 2019; 43(6):1563–1570.
44. Rebecchi F, Giaccone C, Farinella E, et al. Randomized controlled trial of laparoscopic Heller myotomy plus Dor fundoplication versus Nissen fundoplication for achalasia: long-term results. *Ann Surg*. 2008;248(6):1023–1030.
45. Costantini M, Salvador R, Capovilla G, et al. Thousand and one laparoscopic Heller myotomies for esophageal achalasia: a 25-year experience at a single tertiary center. *J Gastrointest Surg*. 2019;23(1):23–35.
46. Leeuwenburgh I, Scholten P, Alderliesten J, et al. Long-term esophageal cancer risk in patients with primary achalasia: a prospective study. *Am J Gastroenterol*. 2010;105(10):2144–2149.
47. Tustumi F, Bernardo WM, da Rocha JRM, et al. Esophageal achalasia: a risk factor for carcinoma. A systematic review and meta-analysis. *Dis Esophagus*. 2017;30(10):1–8.
48. Orringer MB, Orringer JS. Esophagectomy: definitive treatment for esophageal neuromotor dysfunction. *Ann Thorac Surg*. 1982;34(3):237–248.
49. Za tsev VT, Dalavurak VT, Kuznetsov AV, Kudinenko AS. State of the esophageal mucosa in cardiospasm. *Klin Khir*. 1979;10:45–49.
50. Aiolfi A, Asti E, Bonitta G, et al. Esophageal resection for end-stage achalasia. *Am Surg*. 2018;84(4):506–511.
51. Watson TJ. Esophagectomy for end-stage achalasia. *World J Surg*. 2015;39(7):1634–1641.
52. Mineo TC, Pompeo E. Long-term outcome of Heller myotomy in achalasic sigmoid esophagus. *J Thorac Cardiovasc Surg*. 2004;128(3):402–407.
53. Faccani E, Mattioli S, Lugaresi ML, et al. Improving the surgery for sigmoid achalasia: long-term results of a technical detail. *Eur J Cardiothorac Surg*. 2007;32(6):827–833.
54. Sweet MP, Nipomnick I, Gasper WJ, et al. The outcome of laparoscopic Heller myotomy for achalasia is not influenced by the degree of esophageal dilatation. *J Gastrointest Surg*. 2008;12(1):159–165.
55. Panchanatheeswaran K, Parshad R, Rohila J, et al. Laparoscopic Heller's cardiomyotomy: a viable treatment option for sigmoid esophagus. *Interact Cardiovasc Thorac Surg*. 2013; 16(1):49–54.
56. Sandrasagra FA, English TAH, Milstein BB. The management and prognosis of esophageal perforation. *Br J Surg*. 1978;65(9):629–632.
57. White RK, Morris DM. Diagnosis and management of esophageal perforations. *Am Surg*. 1992;58(2):112–119.
58. Brinster CJ, Singhal S, Lee L, et al. Evolving options in the management of esophageal perforation. *Ann Thorac Surg*. 2004;77(4):1475–1483.
59. Vanuytsel T, Lerut T, Coosemans W, et al. Conservative management of esophageal perforations during pneumatic dilation for idiopathic esophageal achalasia. *Clin Gastroenterol Hepatol*. 2012;10(2):142–149.
60. Sanaka MR, Raja S, Thota PN. Esophageal perforation after pneumatic dilation for achalasia: successful closure with an over-the-scope clip. *J Clin Gastroenterol*. 2016;50(3):267–268.
61. Elhanafi S, Othman M, Sunny J, et al. Esophageal perforation post pneumatic dilatation for achalasia managed by esophageal stenting. *Am J Case Rep*. 2013;14:532–535.eCollection 2013.
62. Ghoshal UC, Karyampudi A, Verma A, et al. Perforation following pneumatic dilation of achalasia cardia in a university

hospital in northern India: a two-decade experience. *Indian J Gastroenterol.* 2018;37(4):347–352. Epub 2018 Aug 18.

63. Sánchez-Pernaute AI, Aguirre EP, Talavera P, et al. Laparoscopic approach to esophageal perforation secondary to pneumatic dilation for achalasia. *Surg Endosc.* 2009;23(5):1106–1109. Epub 2008 Sep 24.

64. Urbani M, Mathisen DJ. Repair of esophageal perforation after treatment for achalasia. *Ann Thorac Surg.* 2000;69(5):1609–1611.

65. Sudarshan M, Elharram M, Spicer J, et al. Management of esophageal perforation in the endoscopic era: Is operative repair still relevant? *Surgery.* 2016;160(4):1104–1110. Epub 2016 Aug 11.

66. Lázár G, Paszt A, Mán E. Role of endoscopic clipping in the treatment of esophageal perforations. *WJGE.* 2016;8(1):13–22.