

Treatment of esophageal achalasia with Heller myotomy: retrospective evaluation of patient satisfaction and disease-specific quality of life

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Background: Prospective randomized studies have suggested that surgery palliates esophageal achalasia more effectively than pneumatic dilatation, but for some dilatation is still the procedure of choice for initial treatment. We decided to compare achalasia symptoms before and after Heller myotomy by means of postoperative questionnaires. **Methods:** The study included 22 patients who underwent Heller myotomy for achalasia at the Hotel Dieu Hospital, Queen's University, Kingston, Ont., since July 1990; 5 of them required repeat myotomy for symptom recurrence, for a total of 9 open and 18 laparoscopic procedures. Median follow-up was 43 (range 6–109) months. Preoperative and postoperative data regarding dysphagia, regurgitation, chest pain and overall patient satisfaction were gathered. Symptom scores were calculated by adding severity (0 = none, 2 = mild, 4 = moderate, 6 = severe) to frequency (0 = never, 1 = occasionally, 2 = once a month, 3 = every week, 4 = twice a week, 5 = daily). Patients having a repeat procedure were instructed to evaluate symptoms with respect to their initial myotomy. **Results:** Seventeen (77%) patients were successfully contacted, 4 of them had subsequent repeat myotomy for symptom recurrence. Initially, overall symptom scores decreased for all but 1 patient, with mean preoperative and postoperative values of 23.1 and 7.3 respectively ($p < 0.001$). The patient in whom symptoms did not improve is a candidate for a repeat procedure. Repeat myotomy was performed after a median of 38 (range 23–75) months, corresponding to an overall 3-year positive outcome in 13 (76%) of the 17 patients. Fifteen (88%) patients considered their myotomies a success and 16 (94%) would choose to have this procedure again given the outcome. **Conclusion:** Heller myotomy appears to be effective in alleviating the symptoms of achalasia. Repeat myotomy is occasionally required.

Contexte : Des études randomisées prospectives ont indiqué que la chirurgie est plus efficace comme traitement palliatif contre l'achalasia de l'œsophage que la dilatation pneumatique, mais que dans certains cas, la dilatation demeure l'intervention de choix comme traitement initial. Nous avons décidé de comparer les symptômes de l'achalasia avant et après une myotomie de Heller en utilisant des questionnaires postopératoires. **Méthodes :** L'étude a porté sur 22 patients qui ont subi une myotomie de Heller pour une achalasia à l'Hôpital Hôtel Dieu de l'Université Queen's, à Kingston (Ontario) depuis juillet 1990 : il a fallu répéter la myotomie dans 5 cas parce que les symptômes sont réapparus, ce qui donne un total de 19 interventions sanglantes et 18 laparoscopies. Le suivi médian s'est établi à 43 (fourchette de 6 à 109) mois. On a réuni des données préopératoires et postopératoires sur la dysphagie, la régurgitation, les douleurs à la poitrine et la satisfaction globale des patients. On a calculé les scores des symptômes en additionnant la gravité (0 = aucun, 2 = bénin, 4 = moyen, 6 = sévère) à la fréquence (0 = jamais, 1 = à l'occasion, 2 = une fois par mois, 3 = toutes les semaines, 4 = deux fois par semaine, 5 = tous les jours). On a demandé aux patients qui ont subi une nouvelle intervention d'évaluer les symptômes par rapport à leur myotomie initiale. **Résultats :** On a réussi à communiquer avec 17 (77 %) des patients, dont quatre ont subi par la suite une deuxième myotomie parce que les symptômes sont réapparus. Au début, les scores globaux des symptômes ont diminué chez tous les patients sauf un, et les valeurs préopératoires et postopératoires moyennes se sont établies à 23,1 et 7,3 respectivement ($p < 0,001$). Le patient chez lequel les symptômes ne se sont pas améliorés est candidat à une deuxième

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intervention. On a procédé à une deuxième myotomie après une médiane de 38 (fourchette de 23 à 75) mois, ce qui correspond à un résultat positif global à trois ans chez 13 (76 %) des 17 patients. Quinze (88 %) des patients ont considéré leur myotomie comme une réussite et 16 (94 %) choisiraient de subir de nouveau l'intervention compte tenu du résultat. **Conclusions :** La myotomie de Heller semble atténuer efficacement les symptômes de l'achalasia. Il faut à l'occasion répéter l'intervention.

Achalasia is a relatively uncommon esophageal motor disorder with a reported annual incidence of 1 in 100 000 people.¹ Characteristic manometric findings include a hypoperistaltic esophageal body and absence of lower esophageal sphincter (LES) relaxation. Dilated esophagi with retained food, as well as an air-fluid level with a characteristic bird-beak appearance of a constricted LES, are late radiographic findings. Although the cause of the disease remains unclear it has been suggested that the disorder may be secondary to abnormal innervation of the myenteric plexus of the esophagus.² In conjunction with radiographic and manometric studies for the diagnosis of achalasia, endoscopy serves an important role in evaluating potentially obstructive causes of achalasia.²

The characteristic presentation of achalasia is that of dysphagia and regurgitation; less frequent symptoms include chest pain, weight loss and heartburn.^{3,4} Dysphagia is progressive and contributes to both weight loss and esophageal dilatation. Long-term retention of food in the esophagus worsens the symptom of regurgitation, which can lead to aspiration pneumonia and pulmonary abscesses. Thus, early therapy is important in halting and potentially reversing the progressive esophageal dilatation.

Therapeutic methods aim to palliate the symptoms of achalasia, as no current therapy exists to treat the underlying neuropathology. The basic treatment principle is disruption of the LES to reduce sphincter pressure. This can be achieved by the 2 mainstays in achalasia therapy: pneumatic dilatation and LES (Heller) myotomy.² Other therapies, including botulinum toxin injection and pharmacotherapy, are not effective in

the long term.² Prospective randomized studies suggest that surgery palliates dysphagia more effectively than pneumatic dilatation.^{5,6} Despite these studies some still consider that pneumatic dilatation is the initial treatment of choice.^{7,8} The advent of minimally invasive laparoscopic surgery, with its high success rate and minimal morbidity, has brought about a shift in surgical practice; more patients are being referred for myotomy as primary treatment.⁹

This study aims to compare the main symptoms of achalasia (dysphagia, regurgitation and chest pain) by means of retrospectively mailed questionnaires previously used by Zaninotto and colleagues.^{10,11} Pre- and postoperative symptom scores were gathered from patients treated with esophageal Heller myotomy, both open and laparoscopic. Overall patient satisfaction was also assessed.

Methods

Starting in July 1990, 22 patients underwent Heller myotomy for esophageal achalasia at the Hotel Dieu Hospital, Queen's University, Kingston, Ont. Five patients required repeat Heller myotomy after a median of 38 (range 16–75) months due to symptom recurrence, for a total of 9 open and 18 laparoscopic procedures. There were 12 males and 10 females with a mean age of 42 (range 19–73) years. Eighteen (82%) patients had failed previous nonoperative treatment. They included 10 (46%) patients who had pneumatic dilatations; 5 patients had 1 dilatation, 3 patients had 2 dilatations, and 2 patients had 3 failed dilatations. One patient had botulinum toxin injections, and 13 patients had previously been treated with calcium channel antagonists. One patient had

undergone remote surgery performed transthoracically; however, further investigations revealed significant residual constriction at the LES and as such he was a candidate for Heller myotomy.

Standard preoperative work-up included upper gastrointestinal series, endoscopy and esophageal manometry to confirm the diagnosis of achalasia. Transabdominal laparoscopic Heller myotomy was performed by one surgeon (C.D.M.), according to the method described by Hunter and associates.¹² The gastroesophageal junction was exposed through division of the phrenicosophageal ligament, and the esophageal fat pad was excised. The myotomy was initiated on the esophagus by hook cautery 6 cm proximal to the LES and extended 3 cm onto the gastric cardia. Longitudinal and circular muscle fibres were carefully divided until the submucosal plane was adequately identified. Length and adequacy of the myotomy were monitored by intraoperative esophagogastrosopy. A partial anterior (Dor) fundoplication was used to complete the procedure. Open Heller myotomy was performed through an upper, midline incision except in 2 patients who underwent transthoracic Heller myotomy. Of the 17 patients who had a single Heller myotomy, 5 were open (2 transthoracic, 3 transabdominal) and the remaining 12 were laparoscopic. The laparoscopic approach was preferred as it allowed excellent visualization of the LES, a more complete myotomy and decreased morbidity and hospitalization times. Of the 5 patients who had repeat Heller myotomy, all were initially treated laparoscopically; however, 4 of the 5 procedures were performed through a transabdominal laparotomy with the remaining 1 per-

formed laparoscopically. Myotomy alone was used in 10 patients, and an anterior Dor fundoplication was added in the remaining 12 patients.

Clinical data regarding the pre- and postoperative symptoms of dysphagia, regurgitation and chest pain were retrospectively gathered by means of a mailed, unvalidated, questionnaire, which was developed by Zaninotto and colleagues.^{10,11} Dysphagia was described as “difficulty swallowing,” and regurgitation was described as the sensation of stomach contents going up the esophagus. The authors sent identical symptom questionnaires in a single mailing, labelled as either “preoperative” or “postoperative” to prospective patients in 2004, along with a letter of research intention and an invitation to participate. Patients having had repeat myotomy were instructed to complete the preoperative and postoperative symptom questionnaires to reflect their initial myotomy. Patients were asked to assign values to each of the 3 symptoms with respect to severity (0 = none, 2 = mild, 4 = moderate, 6 = severe) and frequency (0 = never, 1 = occasionally, 2 = once a month, 3 = every week, 4 = twice a week, 5 = daily). Symptom scores were calculated, with the highest possible symptom score being 11 and the highest possible total score being 33. Overall patient satisfaction was assessed by asking the following yes/no questions included in the questionnaire: Would you consider your surgery to be a success? and If given the opportunity, would you choose to have this procedure again? Paired-samples *t*-tests were used to compare pre- and postoperative symptom scores. A *p*-value of 0.05 was considered significant.

Results

Six (27%) patients had complications, 3 of them intraoperatively; 2 were esophageal perforations that were repaired immediately, the third was a

gastric perforation diagnosed 1 week postoperatively which was repaired by laparotomy (converted from laparoscopy). Three patients had postoperative complications, consisting of 1 case of pulmonary edema after transthoracic open myotomy and 2 complications at the trocar site. One patient had mild cellulitis, which responded well to antibiotics taken orally. The other patient had a hernia at the trocar site. Conversions to an open procedure were required in 3 (14%) patients due to scarring and adhesions from a previous failed myotomy that made adequate laparoscopic myotomy impossible. There were no operative deaths. The median length of hospital stay was 7 days for open myotomy and 3 days for laparoscopic myotomy. Two patients required postoperative pneumatic dilatations; 1 reported a good outcome.

Four patients noted a return of achalasia symptoms after a median time of 38 (range 23–75) months, constituting a 3-year successful outcome in 13 (76%) of the 17 patients.

Symptom questionnaire

Symptom questionnaires were collected from 17 (77%) patients (4 of whom required a repeat operation). Of the nonrespondents, 1 patient had died, and 4 were lost to follow-up. Median follow-up was 43 (range 6–109) months. Symptom scores for dysphagia, regurgitation and chest pain, as well as total symptom scores are shown in Table 1. Symptom scores for the 4 patients who required repeat operation reflect their initial Heller myotomy. The decrease in individual symptom scores for all 3 symptoms was significant. Overall, total symptom scores decreased from

Table 1

Pre- and postoperative mean symptom scores for patients with achalasia treated by Heller myotomy

Symptom	Mean score (median) (and SD)		Change, %	<i>p</i> value
	Preoperative (n = 17)	Postoperative (n = 17)		
Dysphagia	10.18 (11) (2.01)	3.88 (3) (4.04)	61.9	< 0.001
Regurgitation	7.94 (10) (3.91)	2.06 (0) (2.49)	74.1	< 0.001
Chest pain	4.94 (5) (4.23)	1.35 (0) (2.45)	72.7	0.002
Total score	23.06 (21) (6.8)	7.29 (6) (6.7)	68.4	< 0.001

*Change scores represent % improvement over preoperative scores.

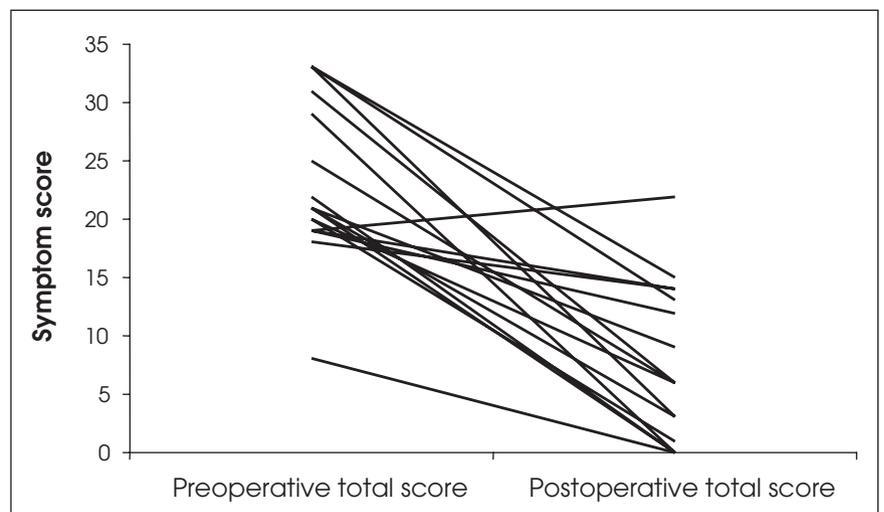


FIG. 1. Preoperative versus postoperative total symptom scores in 17 patients with achalasia treated by Heller myotomy.

a mean preoperative value of 23.1 (standard deviation [SD] 6.8) to a mean postoperative value of 7.3 (SD 6.7) ($p < 0.001$) (Table 1). Total symptom scores decreased in 16 (94%) of the 17 patients (Fig. 1). The one with a positive variation reported no change in preoperative versus postoperative symptom scores for dysphagia and regurgitation, with constant values of 9 and 5 respectively and worsening chest pain, with preoperative versus postoperative values of 5 and 8 respectively. Median time to repeat operation was 38 (range 23–75) months, corresponding to an overall 3-year positive outcome in 13 (76%) patients.

Of the 17 respondents, 15 (88%) considered that their operation was a success. The 2 (12%) who did not were the ones who reported no improvement in total symptom scores at time of questionnaire follow-up. Sixteen (94%) respondents stated they would choose to have this procedure performed again if given the opportunity; the 1 who would not was the patient in whom a repeat Heller myotomy failed.

Discussion

The management of choice for achalasia remains controversial. Pharmacotherapy provides symptomatic short-term relief, with some studies reporting success; however, in terms of long-term symptomatic improvement, pharmacotherapy plays a minimal role.^{13,14} An alternative medical management for achalasia is botulinum toxin injections into the LES. Botulinum toxin inhibits acetylcholine release from presynaptic nerve terminals, thereby inhibiting muscle contraction and decreasing LES pressure.¹⁵ Initial results were promising, but patients frequently relapsed within a year.¹⁶ In addition, patients who had unsuccessful botulinum toxin injections demonstrated an increase in fibrosis at the LES, making future surgical intervention more difficult and adding to its inher-

ent risks.^{17,18} As a result, some suggest that botulinum toxin injection should be reserved for elderly or frail people who are poor candidates for the more definitive treatments.²

Pneumatic dilatation has been the mainstay in achalasia management for a number of years because it is cheaper than surgical myotomy and because its long-term benefits surpass all other nonsurgical treatments. However, the only prospective randomized trial comparing pneumatic dilatation to surgical myotomy found that dysphagia was palliated in 95% of myotomized patients whereas only 65% of patients were relieved after dilatation.⁶ These results were criticized by several studies, which cited a suboptimal dilatation technique and a greater baseline of disease severity among patients in the dilatation group.^{19,20} Although these results remain the best source for comparative data,² the best treatment for achalasia remains controversial as both endoscopic and surgical techniques have evolved since this randomized study. More recently, satisfactory results using graded pneumatic dilatations have been reported, with 90% of patients noting symptom improvement, albeit with a 74% failure rate at 4-year follow-up.²¹ Complications of dilatation include esophageal perforation, which occurs in approximately 5% of patients.² Surgical myotomy, while proving more efficacious, once involved serious morbidity as access to the site was initially gained through thoracotomy or laparotomy. However, minimally invasive surgery through laparoscopic techniques has been introduced with success rates equal to open cardiomyotomy but with significantly reduced morbidity.²² An antireflux procedure is often performed simultaneously to prevent postoperative reflux, and controversy remains over what type of fundoplication, if any, should be used. In our patients, a shortened myotomy alone was performed earlier in our series to avoid the need for an antireflux procedure.

However, because some patients had symptoms of recurrent dysphagia, we changed our surgical practice to adopt an extended myotomy with a Dor fundoplication, resulting in superior outcomes. Anterior Dor fundoplication, in comparison to partial posterior (Toupet) fundoplication, has the advantage of requiring less extensive mobilization of the gastroesophageal junction as well as creating a wrap that may be less prone to the development of dysphagia.²³ The advent of minimally invasive surgery has brought a shift in the treatment of achalasia, as surgical myotomy has become the preferred treatment of most gastroenterologists and other referring physicians.⁹

Our study has a number of limitations. First, 17 patients is admittedly a modest sample size; however, it was sufficient to achieve adequate statistical power for illustrating the effect of surgical intervention on symptom outcome, as scores had, on average, a 15-point change. Postoperative symptom trends were evaluated over a median of 43 months, allowing for adequate clinical assessment of success. Second, esophagomyotomy was achieved by a heterogeneous mix of procedures; 2 patients had open thoracotomy, 3 patients had open laparotomy and the remainder were approached by laparoscopy. In addition, 10 patients early in the laparoscopic series were treated with only a short myotomy (1 cm onto the gastric wall), while the remainder were treated with a longer myotomy (3 cm onto the gastric wall), with the addition of an anterior Dor fundoplication. In our opinion this reflects in part the evolution of the surgical management of achalasia, with the more recent laparoscopic approach proving superior in terms of visualization of the LES as well as a decrease in associated morbidity and postoperative length of hospital stay.

Finally, we recognize that the symptom questionnaire developed by Zaninotto and colleagues^{10,11} has yet to be validated. Although there exist

numerous validated quality-of-life assessment scales and gastrointestinal symptom scores, our rationale for using this particular questionnaire was based on its concise and pertinent content. We acknowledge the limitations of a retrospective study, particularly recall bias, and to limit this bias as much as possible, a short questionnaire focusing on the most common symptoms of achalasia was chosen. It is our belief that such a questionnaire evaluating dysphagia, chest pain and regurgitation, based solely on the criteria of frequency and severity, best reflects the degree of symptoms in our particular study, where some patients were asked to recall remote esophageal symptoms. By omitting emotional, physical and psychosocial symptoms contained in other questionnaires, and by keeping requested outcome measures brief, recall bias would be minimized.

Our study reports that Heller myotomy affords significant relief for the symptoms of dysphagia, regurgitation and chest pain. Total symptom scores improved in 16 (94%) patients, a success rate in concordance with published literature. Further investigation of the failure demonstrated persistently elevated LES pressure on manometry, and that this patient is a candidate for a repeat procedure. The 4 patients required another myotomy when repeat endoscopy and esophageal motility studies confirmed a persistently elevated LES pressure. Repeat procedures have been less successful in alleviating achalasia symptoms.

Conclusions

Esophageal Heller myotomy is effective in alleviating the predominant symptoms of achalasia. Repeat myotomy is occasionally required.

Competing interests: None declared.

References

1. Sonnenberg A, Massey BT, McCarty DJ, et al. Epidemiology of hospitalization for achalasia in the United States. *Dig Dis Sci* 1993;38:233-44.
2. Spiess AE, Kahrilas PJ. Treating achalasia. From whalebone to laparoscope. *JAMA* 1998;280:638-42.
3. Peracchia A, Bonavina L. Achalasia: Dilation, injection or surgery? *Can J Gastroenterol* 2000;14:441-3.
4. Katilius M, Velanovich V. Heller myotomy for achalasia: quality of life comparison of laparoscopic and open approaches. *JLS* 2001;5:227-31.
5. Csendes A, Velasco N, Braghetto I, et al. A prospective randomized study comparing forceful dilation and esophagomyotomy in patients with achalasia of the esophagus. *Gastroenterology* 1981;80:789-95.
6. Csendes A, Braghetto I, Henriquez A, et al. Late results of a prospective randomized study comparing forceful dilation and oesophagomyotomy in patients with achalasia. *Gut* 1989;30:299-304.
7. Da Silveira EB, Rogers AI. Achalasia: a review of therapeutic options and outcomes. *Compr Ther* 2002;28:15-22.
8. Richter JE. Comparison and cost analysis of different treatment strategies in achalasia. *Gastrointest Endosc Clin N Am* 2001;11:359-70.
9. Patti MG, Fisichella PM, Perretta S, et al. Impact of minimally invasive surgery on the treatment of esophageal achalasia: a decade of change. *J Am Coll Surg* 2003;196:698-705.
10. Zaninotto G, Costantini M, Molena D, et al. Treatment of esophageal achalasia with laparoscopic Heller myotomy and Dor partial anterior fundoplication: prospective evaluation of 100 consecutive patients. *J Gastrointest Surg* 2000;4:282-9.
11. Zaninotto G, Costantini M, Portale G, et al. Etiology, diagnosis, and treatment of failures after laparoscopic Heller myotomy for achalasia. *Ann Surg* 2002;235:186-92.
12. Hunter JG, Trus TL, Branum GD, et al. Laparoscopic Heller myotomy and fundoplication for achalasia. *Ann Surg* 1997;225:655-65.
13. Gelfond M, Rozen P, Keren S, et al. Effect of nitrates on LOS pressure in achalasia: a potential therapeutic aid. *Gut* 1981;22:312-8.
14. Gelfond M, Rozen P, Gilat T. Isosorbide dinitrate and nifedipine treatment of achalasia: a clinical, manometric and radionuclide evaluation. *Gastroenterology* 1982;83:963-9.
15. Pasricha PJ, Ravich WJ, Hendrix TR, et al. Treatment of achalasia with intrasphincteric injection of botulinum toxin: a pilot trial. *Ann Intern Med* 1994;121:590-1.
16. Annese V, Basciani M, Perri F, et al. Controlled trial of botulinum toxin injection versus placebo and pneumatic dilation in achalasia. *Gastroenterology* 1996;111:1418-24.
17. Horgan S, Hudda K, Eubanks T, et al. Does botulinum toxin injection make esophagomyotomy a more difficult operation? *Surg Endosc* 1999;13:576-9.
18. Patti MG, Feo CV, Arcerito M, et al. Effects of previous treatment on results of laparoscopic Heller myotomy for achalasia. *Dig Dis Sci* 1999;44:2270-6.
19. Katz P. Achalasia: two effective treatment options — let the patient decide. *Am J Gastroenterol* 1994;89:969-70.
20. Robinson RJ, Mayberry JF. Achalasia: the debate continues. *Am J Gastroenterol* 1995;90:1204-5.
21. Vaezi MF, Richter JE. Current therapies for achalasia: comparison and efficacy. *J Clin Gastroenterol* 1998;27:21-35.
22. Dempsey DT, Kalan MM, Gerson RS, et al. Comparison of outcomes following open and laparoscopic esophagomyotomy for achalasia. *Surg Endosc* 1999;13:747-50.
23. Ali A, Pellegrini CA. Laparoscopic myotomy: technique and efficacy in treating achalasia. *Gastrointest Endosc Clin N Am* 2001;11:347-58.