Review Article

Managing recurrent symptoms after treatment of achalasia

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Abstract: Recurrent dysphagia and/or regurgitation occurs in some patients after all treatments for achalasia. Further treatment following botulinum toxin or pneumatic dilatation is generally not difficult, and surgical or transoral endoscopic myotomy are feasible and can generally be undertaken as the next step. Following a failed myotomy, the author's preference is for pneumatic dilatation, with revision myotomy considered if this is not successful. Revision myotomy is technically easier if undertaken via a different body cavity or route to the original myotomy. Symptom improvement can be achieved in 80–90% of individuals after revision treatments. However, a small group continue to experience troublesome symptoms, and if fit should be considered for esophagectomy.

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Introduction

Whilst effective treatment can be offered to patients with achalasia, a good outcome is not always achieved for some individuals. Ideally treatment will permanently relieve dysphagia and regurgitation, without any trade-offs or side effects. Other papers in this series discuss specific outcomes in more depth. It is clear, however, that all treatments can be followed by failure, and a strategy to deal with these issues will be addressed.

When considering outcomes and the risk of failure, it is important to be realistic about the potential for success with each treatment modality. Treatment with botulinum toxin is always temporary, and recurrent symptoms generally develop 6–12 months after treatment (1). Pneumatic dilatation achieves a good outcome in 75–90% of patients, but often requires a series of repeated treatments, and an acceptable outcome is not achieved in a subset of patients (2,3). Laparoscopic cardiomyotomy is claimed to be successful in 85–95% of patients, but with the reported success rates dependent on the length of follow-up (4). Outcomes from series reporting longer term follow-up are generally less optimistic than series reporting short term follow-up. Peroral endoscopic myotomy (POEM) appears to achieve a similar outcome to laparoscopic cardiomyotomy, although with a potentially higher risk of gastroesophageal reflux as a concurrent partial fundoplication cannot be added following POEM (5).

It is important to remember that achalasia is not curable; the underlying motility disorder is not correctable. Hence, treatment aims to palliate dysphagia, and symptom outcomes may deteriorate over time. Poorer, but more realistic outcomes are generally reported in series that follow patients for longer periods of time, and these contrast with the early enthusiasm and optimistic short term outcomes which have been consistently reported by proponents of new therapies developed over the last 50 years; e.g., pneumatic dilatation, Botulinum toxin, thoracoscopic myotomy, laparoscopic myotomy and POEM!

Recurrent dysphagia and its evaluation

The recurrent symptoms which develop most commonly after treatment of achalasia are dysphagia and regurgitation (6). These can arise following inadequate disruption of the lower esophageal sphincter due to an incomplete myotomy (surgical or POEM), or incomplete sphincter disruption

following pneumatic dilatation. Following surgical cardiomyotomy or POEM, this problem is most commonly associated with inadequate distal extension of the myotomy resulting in failure to completely divide the non-relaxing lower esophageal sphincter muscle. To avoid this, surgeons aim to extend the myotomy onto the cardia, usually by 2–3 cm. Further treatment to address failure or recurrent symptoms needs to ensure a complete myotomy is achieved.

Occasionally, however, recurrent dysphagia is associated with post-treatment gastroesophageal reflux which can sometimes lead to the development of a peptic esophageal stricture. This problem is addressed by ensuring reflux control (generally with a proton pump inhibitor) and dilating the stricture endoscopically. Another cause of dysphagia following myotomy is progressive dilatation of the esophagus resulting in the development of a sump which displaces the esophageal outlet more proximally onto the side of the dilated distal esophagus. A sump can also arise following the development of diverticulum at the site of a previous myotomy.

In general, if dysphagia persists following treatment, this is due to either an inadequate myotomy, or treatment was attempted in a patient who presented with an already significantly dilated esophagus. Later recurrence of dysphagia after an initially good outcome, generally indicates the development of a diverticulum at the myotomy site, development of a peptic stricture, or progressive dilatation of the esophagus associated with the underlying disease process. Whilst patients presenting with a dilated esophagus are at higher risk of treatment failure, and should be counselled accordingly, simpler treatment options for recurrent symptoms can still be effective and should always be tried first.

Before considering treatment for recurrent symptoms, the clinician should to try to understand the reason why failure has occurred. Endoscopy examination of the esophagus and stomach is the preferred initial investigation. This should clarify whether recurrent dysphagia is due to an inadequate initial myotomy, esophageal dilatation, or a reflux related peptic stricture. Rarely, late onset dysphagia follows the development of esophageal cancer, which obviously follows a different treatment pathway.

If the gastroesophageal junction appears visibly "closed" at endoscopy, then an inadequate previous myotomy is likely. If the junction is patulous and open, then lower risk treatments that address an inadequate myotomy can sometimes still work, and should still be considered before escalating to higher risk or more complex options. In general, if achalasia was convincingly demonstrated at the original preoperative esophageal manometry, the author sees little value in repeating an esophageal manometry study, as outcomes do not change the recommended treatment options. A barium contrast swallow does, however, sometimes provide complementary information; it can better demonstrate the extent of any esophageal dilatation, the presence of a post-myotomy diverticulum, and demonstrate how well the gastroesophageal junction opens with swallowing. It might not change initial decision making, but it can inform any discussion about the likelihood of success of any offered treatment.

Recurrence after treatment with botulinum toxin

Recurrence after treatment with botulinum toxin is inevitable! Repeat treatment is an option, but will again be followed by recurrence. Hence, escalation to a more definitive option is preferred for most patients. After botulinum toxin, all treatment options are feasible, and the debate about which option is preferred is similar to that in patients who have not undergone any previous treatment. Perhaps the only issue to consider following botulinum toxin injection is that botulinum toxin generates a local inflammatory response, and this can cause fibrosis within the critical submucosal tissue plane. As easy separation of the mucosa from the deeper muscularis propria is required during myotomy or pneumatic dilatation, fibrosis after multiple botulinum toxin treatments makes separation of the mucosa from the muscularis propria more difficult, and increases the risk of mucosal perforation during these treatments (7).

Recurrence after pneumatic dilatation

Pneumatic dilatation often entails a series of dilatations, and may require escalation of the balloon diameter, starting initially at 30 mm, and sometimes escalating to 35 mm, and rarely to 40 mm (3). Recurrent symptoms after initially successful treatment can be managed by repeat pneumatic dilatation or a myotomy. Repeat pneumatic dilatation generally proceeds as per previous dilations, and is successful in many instances (3). If unsuccessful, or patient preference is for an alternative, esophageal myotomy (either laparoscopic or POEM) becomes the obvious next step. When performing a laparoscopic myotomy, the procedure is usually no more difficult than a myotomy in an untreated patient. Occasionally an area of scarring and disruption

is seen at the gastroesophageal junction, consistent with the previous dilatation, although in most instances the site of the previous dilatation is not identified when the previous muscle rupture occurred laterally or posteriorly, and not anteriorly into the new operation field. Clinicians undertaking POEM in this situation also generally report few difficulties, and similarly to surgical myotomy, additional difficulties only arise when myotomy is attempted at the same site as the previous muscle disruption (8).

Recurrence after cardiomyotomy

After a previous surgical myotomy, adhesions and some scarring will be present. Where these are encountered depends on the previous surgical approach (6). Significant abdominal or pleural adhesions will be encountered following open abdominal or thoracic surgery respectively. Fortunately, these scenarios are uncommon, with most previous surgery undertaken laparoscopically or thoracoscopically. Some adhesions to adjacent structures will be encountered after these approaches, but in general the difficulties are limited to the immediate area of the old operation site and can be dissected more easily than after open surgery. After a previous thoracoscopic myotomy, the lung can adhere to the old myotomy site. After laparoscopic surgery, there will be adhesions between the under-surface of the left liver and the operation site, and some fibrosis of the anterior hiatus. If an anterior partial fundoplication was added at the first procedure, the anterior surface is general adherent to the liver and the posterior surface is adherent to the myotomy site, whereas after a posterior partial fundoplication, the myotomised mucosa can be directly adherent to the liver. Care should be taken when re-operating to avoid damage to the mucosa at the old myotomy site. Intra-operative endoscopy can help guide the revision procedure, and a skilled assistant, preferably another surgeon, is also important.

For symptom recurrence or failure after a previous surgical myotomy, the options include revision surgical myotomy, POEM or pneumatic dilatation. If pneumatic dilatation is chosen, this generally proceeds in the same manner as a primary dilatation procedure. In this situation, the previous laparoscopic dissection and partial fundoplication actually provide some additional protection from esophageal rupture by surrounding the distal esophagus with adhesions and partially covering it with the gastric fundus. In the author's experience, this has been a safe and easy initial approach to recurrent symptoms after failed surgery, improving dysphagia in most patients. When pneumatic dilatation fails, other causes of dysphagia should be considered, and these will often require a surgical solution. For example, an improperly positioned or constructed fundoplication can cause obstruction by creating a constriction band across the front of myotomy. Inappropriate or tight closure of the hiatus can also create a ring of scar tissue which obstructs and narrows the diaphragmatic hiatus.

If revision surgery is to be undertaken, any revision via the same surgical cavity (i.e., laparoscopic revision of previous laparoscopic myotomy, or thoracoscopic revision of previous thoracoscopic myotomy) will be more difficult than a primary procedure. The author's group has described an experience with revision surgery, and identified that when performing revision surgery, the simplest approach is generally to change body cavities (6). If the original procedure was thoracoscopic, then a revision laparoscopic myotomy usually proceeds with no more difficulty than a primary laparoscopic myotomy. Similarly, a thoracoscopic myotomy via the left chest is a generally straightforward after a previous laparoscopic myotomy. The author's experience is that the change in cavity also repositions the revision myotomy onto a different part of the esophageal wall, as laparoscopic myotomies are usually undertaken between the 12–2 o'clock positions (when the esophagus is viewed from below), whereas a left thoracoscopic myotomy is usually undertaken at the 4–5 o'clock positions. Symptom improvement was achieved in 89% of patients at mean 3.6 years follow-up (6).

Similar to the experience with changing body cavities for revision surgery, POEM proceduralists generally report few difficulties with performing this procedure after a previous laparoscopic cardiomyotomy, and usually can identify and cut the lower esophageal sphincter muscle at a different site (9).

When encountering a patient with recurrent esophageal obstruction after a previous myotomy, the author's preference is to proceed to pneumatic dilatation in the first instance as this is generally simpler and safe after previous surgery. If a further myotomy is to be considered, myotomy via a different body cavity is preferred, or POEM could be considered if local expertise is available. Either of these approaches is likely to be easier than re-operating via the original body cavity.

Recurrence after POEM

Conceptually, POEM is a thoracoscopic myotomy,

but performed via a trans-oral route and with different equipment. It divides the esophageal muscle via a transthoracic approach, and does not allow the addition of a partial fundoplication. Following a failed POEM, the approaches are similar to those outlined for failed surgical myotomy; i.e., pneumatic dilatation, repeat POEM or surgical myotomy. For this scenario, the author's preference would be to undertake a laparoscopic cardiomyotomy and anterior partial fundoplication, with the expectation that this approach will be no more difficult than in an untreated patient.

When all other options fail!

It should be remembered that the approaches described above do not cure achalasia. They aim to relieve symptoms, and they achieve this by disrupting the non-relaxing lower esophageal sphincter and improving drainage of the esophagus. Perhaps surprisingly, this restores what most patients perceive to be normal swallowing, even though objective manometric testing will suggest otherwise. However, these treatment options, as well as revision procedures, do fail in some individuals, with the risk of failure increasing as follow-up lengthens. In a small subset, dysphagia, regurgitation and sometimes aspiration are particularly troublesome. The risk of a poor outcome is greatest in patients with a significantly dilated esophagus, and food and fluid can be retained in the esophagus despite a demonstrably adequate myotomy. If these patients remain reasonably fit, and their esophagus is demonstrably "failing", the best option is generally esophagectomy (10). In appropriately selected patients, removal of the majority of the esophagus, and restoration of continuity with a gastric conduit can be life changing.

Quality of life after esophagectomy is not normal, but it is generally substantially better than the situation encountered by these individuals. Post-esophagectomy eating disturbances usually subside with time, and post-esophagectomy reflux is manageable and an acceptable trade-off for most patients (11). It should also be recognised that patients with achalasia considering esophagectomy are generally fitter than those considered for cancer resection, they are not deconditioned by neoadjuvant therapies, and they do not require an extensive lymph node dissection. For these reasons, morbidity and mortality risks are lower than that for esophagectomy for cancer, and surgical mortality should be less than 1%. In the author's group, approximately 2% of patients presenting with achalasia have progressed to esophagectomy.

Conclusions

All treatments for achalasia are palliative and failures are inevitable. Optimistic reports of good short outcomes are generally tempered by time, and failure inevitably occurs in a proportion of patients, perhaps in the order of 15–20%, across longer term follow-up. Following initial treatment with botulinum toxin or pneumatic dilatation, myotomy can generally be undertaken as the next step. Following a failed myotomy, the author's preference is to try pneumatic dilatation first. If this fails, another myotomy is feasible, but will be easier if undertaken via a different body cavity/route to the original myotomy. Good outcomes are expected for approximately 80–90% of individuals after revision treatments. However, a small group continue to experience troublesome symptoms, and if fit, should be considered for esophagectomy.

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Footnote

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References

1. Bansal R, Nostrant TT, Scheiman JM, et al. Intrasphincteric botulinum toxin versus pneumatic balloon dilation for treatment of primary achalasia. J Clin Gastroenterol 2003;36:209-14.
2. Lopushinsky SR, Urbach DR. Pneumatic dilatation and surgical myotomy for achalasia. JAMA 2006;296:2227-33.
3. Moonen A, Annese V, Belmans A, et al. Long-term results of the European achalasia trial: a multicentre randomised controlled trial comparing pneumatic dilation versus laparoscopic Heller myotomy. Gut 2016;65:732-9.
4. Chen Z, Bessell JR, Chew A, et al. Laparoscopic cardiomyotomy for achalasia: clinical outcomes beyond 5 years. J Gastrointest Surg 2010;14:594-600.
5. Werner YB, Hakanson B, Martinek J, et al. Endoscopic or Surgical Myotomy in Patients with Idiopathic Achalasia. N Engl J Med 2019;381:2219-29.
6. Grotenhuis BA, Wijnhoven BP, Myers JC, et al. Reoperation for dysphagia after cardiomyotomy for achalasia. Am J Surg 2007;194:678-82.
7. Wu QN, Xu XY, Zhang XC, et al. Submucosal fibrosis in achalasia patients is a rare cause of aborted peroral endoscopic myotomy procedures. Endoscopy 2017;49:736-44.
8. Ling T, Guo H, Zou X. Effect of peroral endoscopic myotomy in achalasia patients with failure of prior pneumatic dilation: a prospective case-control study. J Gastroenterol Hepatol 2014;29:1609-13.
9. Ngamruengphong S, Inoue H, Ujiki MB, et al. Efficacy and Safety of Peroral Endoscopic Myotomy for Treatment of Achalasia After Failed Heller Myotomy. Clin Gastroenterol Hepatol 2017;15:1531-1537.e3.
10. Glatz SM, Richardson JD. Esophagectomy for end stage achalasia. J Gastrointest Surg 2007;11:1134-7.
11. Leibman S, Smithers BM, Gotley DC, et al. Minimally invasive esophagectomy: short- and long-term outcomes. Surg Endosc 2006;20:428-33.

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