Management of Achalasia in the 21st Century: A Suggested Approach

by Donald O. Castell, Jason R. Roberts

Achalasia is the prototypical motility disorder characterized by aperistalsis of the esophageal body and incomplete relaxation of the lower esophageal sphincter. Non-motility findings that help identify this disorder include the radiographic feature of a “bird beak” on the barium esophagram, dysphagia, limited weight loss, and nighttime regurgitation. Patients with achalasia almost always complain of dysphagia and regurgitation and less commonly chest pain, nausea, hiccups, and heartburn. Prior to being diagnosed with achalasia, patients are frequently treated empirically for gastroesophageal reflux. The diagnosis is made in patients with a history suggestive of achalasia and confirmed by a combination of esophageal manometry, upper endoscopy, and barium esophagography. There are currently three different treatment modalities available—botulinum toxin injection, pneumatic dilation, and Heller myotomy. All three are focused on decreasing the LES resting pressure thereby allowing the distal esophagus to empty with the aid of gravity.

The esophageal motility disorder termed achalasia has been known for many decades and treatment approaches extend at least back to 1674 when Willis reported dilation of the cardiac sphincter with a whale bone (1). The combination of a poorly relaxing lower esophageal sphincter (producing an obstructive component) plus the absence of peristalsis in the smooth muscle portion of the esophagus (producing a lack of propulsive activity) results in the extreme dysphagia, regurgitation and weight loss found in these patients. There have been two accepted definitive therapies for many years; these include either forceful dilatation or direct myotomy in the region of the lower esophageal sphincter (LES). Much has been written regarding which technique is preferable for both immediate and long term effectiveness. The advent of a laparoscopic approach to the myotomy has markedly changed the controversy since the prior morbidity associated with thoracotomy has been eliminated. However, the basic pros and cons of the two procedures remain relatively constant: Pneumatic dilatation (PD) involves only a short outpatient encounter with the possibility of esophageal perforation being a concern. When performed by a well experienced “esophagologist”, this risk is, in fact, only 1% or less. PD carries the advantage of a lower incidence of gastroesophageal reflux. In contrast, myotomy can be expected to provide profound relief of the patient’s dysphagia resulting from the controlled destruction of the LES with the trade-off of a high likelihood of reflux. Hence, most surgeons will perform a variation of a fundoplication to control the myotomy induced reflux. As we enter the 21st century, it is appropriate to revisit the published comparisons of these two procedures.

Although Achalasia was first described and treated by physicians in the 17th century, our understanding of what constitutes best practice and the long-term effects of our interventions is lacking. At least part of the explanation for this is the low prevalence of the disease, it is difficult to accumulate experience with a disease occurring 10 in 100,000 persons. However, even at ter-
tary referral centers with the largest cohorts of patients the true answers are still not clear. The vast majority of series published in the literature are retrospective, uncontrolled reports. In our experience, it is difficult to maintain follow up with patients who respond very well to treatment. Asymptomatic patients simply do not want to undergo additional manometric or barium evaluations. This creates a recall bias that skews the collected data towards worse outcomes, with patients who have failed treatment disproportionately returning for follow-up. The use of patient symptom reporting as the primary outcome in most of these studies and not objective functional studies like timed barium swallow also remains a point of contention. Another limitation in the literature is the lack of long term data that show our interventions not only improve symptoms but also prevent the progression to an end-stage esophagus which is markedly dilated and, worse yet, tortuous. While recognizing the shortcomings of the achalasia literature, it is equally important to formulate a therapeutic approach that best utilizes the evidence that exists. Patients should be informed that botulinum toxin, pneumatic dilation, and Heller myotomy are the three acceptable interventions for achalasia, with the latter two considered to be definitive treatments.

BOTULINUM TOXIN

In the 1990s, Paschrika, et al performed a series of well devised experiments showing that 100 units of Botox divided into equal 4 quadrant injections into the LES would reduce the resting pressure and, further, that this was particularly effective in approximately 2/3 of patients with achalasia (2). In their initial report, the average duration of symptomatic remission was somewhat greater than one year although subsequent experience has shown that most patients do not achieve such long lasting effects. The need for repeated injections is the reason that we do not consider Botox as a permanent therapy for achalasia in the majority of patients. For selected patients, Botox is often the preferred approach, thus becoming definitive therapy. These are identified basically as any patient not considered a reasonable surgical risk since PD should there be a perforation, is likely to require a surgical approach. Thus, age and comorbidities are the usual indications for considering Botox as the primary therapy. We find the age question interesting in the 21st century when increasing numbers of patients with advanced age are being seen, many with minimal other medical problems. It is, therefore, difficult to recommend an age threshold at which Botox becomes the preferred treatment and the presence of other medical conditions truly becomes the decision maker.

We have found an alternative use for this therapy in our practice of treating patients with suspected Achalasia based on the observations from Katzka and Castell (3). After completing the requisite diagnostic studies, the etiology of the patient’s symptoms is not always clear. Our approach has been to use Botulinum toxin as both a diagnostic and therapeutic intervention. If the dysphagia improves, we will recommend a definitive treatment when symptoms recur. In patients whom do not respond, the diagnosis of achalasia is questioned and repeat manometry may be considered in 3–6 months or an alternative diagnosis is pursued.

PNEUMATIC DILATION

We routinely perform pneumatic dilation under fluoroscopic guidance. The patient is taken to a fluoroscopy equipped endoscopy room. Using appropriate conscious sedation, upper endoscopy is performed to observe the degree of esophageal food retention and exclude other disease in the esophagus, stomach or duodenum. A careful retroflex view of the esophagogastric junction is essential to exclude undiagnosed malignancy causing “pseudo-achalasia” and to provide qualitative assessment of tightness at the LES. As the endoscope is withdrawn, a fluoroscopic image while visualizing end of the endoscope at the EG junction will establish its relative position for subsequent centering of the dilator balloon. A guidewire is left in the antrum as the endoscope is withdrawn. The dilator is passed into the stomach over the wire and the balloon centered under fluoroscopic visualization using the prior estimated position. During fluoroscopy the balloon is inflated to 10 psi pressure and a transient “waist” should confirm proper placement in the LES. This step is crucial and should be repeated until the correct position is confirmed. Then, inflation at 10 psi is maintained for 15 seconds. The balloon is deflated and the dilator and wire removed.
Immediately following the procedure a 16 French naso-gastric tube is placed with its tip in the distal esophagus. Sixty ml of barium contrast media is instilled to fill the distal esophagus and ensure the absence of perforation. The tube is removed and the patient allowed to recover as after standard endoscopy.

This procedure is relatively short (15–20 minutes), well tolerated, and done on an outpatient basis. Gastroenterologists that do not perform or recommend this procedure to patients likely do so because they are not experienced with the technique, feel it is an inferior treatment to surgery, or do not practice in a center with a thoracic surgeon available in the unlikely event of an esophageal perforation. Evaluating the success of this procedure can be difficult as there have been many types of balloon dilators and even more techniques used over the years. The first balloon dilators were used in the 1930s. Today, the most commonly used device in the USA is the Rigiflex Balloon Dilator (Microvasive Watertown, MA). The technique employed in our lab using this device is described above; however, the variables such as inflation pressure, time of inflation, and number of inflations differ throughout the world. Using this device there are 24 uncontrolled studies with 1,256 patients followed for a mean of 20 months with 77% having a good or excellent symptom response (4–28). In the few series with follow up extending beyond 10 years the success rate drops to 55% (25,29–30). A concern among gastroenterologist and patients is the risk of esophageal perforation during pneumatic dilation. The frequency of this complication varies among studies, but in experienced hands, the rate should be ≤1%. All patients should undergo radiographic evaluation for perforation following PD.

HELLER MYOTOMY

Ernest Heller was the first to describe the surgical myotomy in 1914 through a thoracotomy (31). Since then, the technique has been refined and now the preferred approach is via laparoscopy which has reduced the morbidity and recovery times. Following the incision through the LES, a partial fundoplication (Dor or Toupet) is performed to prevent post-myotomy gastro-esophageal reflux which occurs in up to 56% of patients (23–24,32–58). The response rate was 84% in 1,420 patients from 30 uncontrolled trials (23–24,32,58). As with PD, the effectiveness of HM degrades with time. In a study by Rice et al. of 73 patients treated with HM, 89% had a good/excellent response at 6 months and 57% at 6 years (24). Because of the technical aspects of surgery, the efficacy and adverse events of HM are related to surgeon experience.

The only randomized comparison of laparoscopic HM and PD with the Rigiflex balloon was conducted by Kostic et al (23). In this study, 40 patients were randomized to either treatment and followed for 12 months. They reported 96% of patients undergoing HM and 87% of patients treated with PD had successful responses. Rice et al. reported the results of a non-randomized study including 106 patients treated with PD and 73 treated with HM (24). At 6 months the good/excellent response rates were 90% and 89% respectively, however these rates declined to 44% and 57% at 6 years.

Based on the information cited above and on our clinical experience, we believe that either PD or myotomy is an appropriate long term approach to the management of the achalasia patient. We make every attempt to discuss with the numerous patients we see in consultation that such is the case, often sending them to a surgeon to, at least, learn about the surgical option. As with any therapeutic approach that involves procedural skill and is, therefore, dependent on experience, our belief is that the treatment option primarily discussed with the patient should include referral to the individual readily available who is the most appropriately experienced. This is what is often termed the “best local expertise.”

Our preference for initial treatment of a typical achalasia patient is PD, starting with a 30 mm pneumatic balloon dilator. If the clinical response is less than satisfactory over an interval of 4–6 weeks, a repeat procedure is performed using a 35 mm diameter dilator. Failure to respond to 2 PDs is considered indication for myotomy (Figure 1).

There appears to be a trend occurring in the United States today for many gastroenterologists to ignore the PD option, in favor of a myotomy. We think it is appropriate that the gastroenterologist with limited experience in performing PD during their training years should definitely seek “local expertise”. We do not, however, think
it appropriate to consider myotomy the only, or even the preferred, option. Rather, we believe that the gastroenterologist seeing a patient with achalasia should consider a referral to an esophageal expert, for either a PD or a myotomy. Our plea is that PD be considered just as appropriate an option as myotomy for definitive treatment of achalasia; i.e. refer the patient to a gastroenterology colleague at an esophageal center.

References
1. Willis T. 
3. Katzka DA, Castell DO. Use of Botulinum Toxin as a Diagnostic/Therapeutic Trial to Help Clarify an Indication for Definitive Therapy in Patients With Achalasia. 
5. Cox J, Buckton GK, Bennett JR. Balloon dilatation in achalasia: a new dilator. 
10. Levine ML, Moskowitz GW Dorf BS et al. Pneumatic dilation in patients with achalasia with a modified Gruntzig dilator (Levine) under direct endoscopic control: results after 5 years. 
15. Lambroza A, Schuman RW Pneumatic dilation for achalasia without fluoroscopic guidance: Safety and efficacy. 

Figure 1. Algorithm for diagnosing and managing achalasia.
Management of Achalasia

A SPECIAL ARTICLE


