Changes in the Diagnosis and Treatment of Achalasia

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Esophageal achalasia is a primary esophageal motility disorder of unknown origin characterized by the absence of esophageal peristalsis and failure of the lower esophageal sphincter (LES) to relax appropriately in response to swallowing. These abnormalities lead to impaired emptying of food from the esophagus into the stomach with consequent food stasis.

The last two decades have seen a significant change in the understanding of the clinical presentation of this disease, and in its diagnosis and treatment.

CLINICAL PRESENTATION

Dysphagia for solids and liquids is the most common symptom, and it is present in about 95% of patients. Regurgitation of undigested food is reported by 60% of patients and 40% experience chest pain. About 40% of patients also experience heartburn, due to stasis and fermentation of food in the esophagus. Unfortunately, some of these patients are often thought to have gastroesophageal reflux disease (GERD) and are treated with proton pump inhibitors or even referred for a laparoscopic antireflux operation. Today, it is also recognized that aspiration of esophageal contents can lead to respiratory symptoms.

DIAGNOSIS

Upper endoscopy is usually the first test performed to rule out cancer or a peptic stricture. Retained food is often found in the esophagus. Recently, high-resolution manometry (HRM) has replaced the conventional manometry that has been the gold standard for many years. HRM confirms the diagnosis of achalasia, and distinguishes three distinct manometric patterns (Chicago classification): type I, with minimal esophageal pressurization; type II, with pan-esophageal pressurization; and type III, with spasm, characterized by rapidly propagated esophageal pressurization attributable to spastic contractions. Patients with type II achalasia have the best prognosis, as they are more likely to respond to pneumatic dilatation (PD) or laparoscopic Heller myotomy (LHM) than patients with type I or type III achalasia. A barium swallow assesses the degree of esophageal dilatation, the axis of the esophagus (straight or sigmoid; Figure 2 a, b), and the presence of an associated epiphrenic diverticulum (Figure 3).

TREATMENT

The goal of treatment is to improve esophageal emptying and patient’s symptoms by relieving the functional obstruction at the level of the gastroesophageal junction. This is usually accomplished by decreasing the LES pressure to <10 mm Hg.

The last 20 years have witnessed a remarkable progress in the treatment of achalasia. PD has been the most common treatment for many years. This procedure has been standardized with the introduction of Rigiflex balloons (Boston Scientific Corporation, MA, USA). Usually a 30-mm balloon is initially selected, and in case of lack of response 35- and 40-mm balloons are used. In specialized Centers, the perforation rate is <5%, and post procedure reflux occurs in 30–40% of patients studied by pH monitoring. A recent multicenter and randomized trial compared PD with LHM and showed that after 2 years about 90% of patients did well with either form of treatment. It will be important to have longer follow-up to see whether these results will persist, as many retrospective studies of PD with longer follow-up have shown progressive deterioration over time, with only 50% of patients doing well after 10 years.

The endoscopic injection of Botulinum toxin in the LES aimed to decrease the sphincter pressure and it was very popular during the nineties. Unfortunately, the effect was short lasting, probably secondary to the formation of antibodies. In addition, transmural inflammation and fibrosis frequently occurred at the level of the gastroesophageal junction, making a subsequent LHM more challenging and the outcome less predictable. Therefore, botulinum toxin should be used only in patients who cannot undergo either PD or LHM.

In 1991, the first minimally invasive esophageal myotomy was performed in the United States by a left thoracoscopic approach. Subsequently, the technique evolved to a laparoscopic approach, as it allowed easier exposure of the gastroesophageal junction, a longer myotomy onto the gastric wall, and the performance of a partial fundoplication (anterior or posterior) to decrease the incidence of GERD, quite

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common after a thoracoscopic myotomy. Patients usually spend one night in the hospital, and return to their regular activities in about 10 days. Improvement of swallowing occurs in 90–95% of patients, and tends to persist in about 75% of patients 10 years after the procedure.

**Figure 1** High-resolution manometry. Achalasia type I, II, and III.

**Figure 2** Barium swallow. (a) Achalasia, straight esophageal axis; (b) sigmoid esophagus.
In 2010, Dr Inoue revolutionized the treatment of achalasia, describing for the first time the peroral endoscopic myotomy (POEM), an endoscopic procedure that allows the relief of the functional obstruction at the level of the gastroesophageal junction, combining the minimally invasive nature of an endoscopic procedure with the efficacy of a myotomy. After creation of an esophageal sub-mucosal tunnel, the circular muscular fibers of the distal esophagus and proximal stomach are selectively sectioned. As the time of the initial report, thousands of patients have been treated by POEM worldwide, creating a lot of interest and enthusiasm. However, as for any innovation, it is important to look at the evidence present in the literature. A careful review of the published studies shows some advantages of this technique but also some concerns:

**Advantages.** (1) No surgical incisions; (2) less postoperative discomfort; (3) faster return to daily activities; and (4) it has been used successfully to treat recurrent dysphagia after LHM, avoiding the need for another operation.

**Concerns.** (1) Although PD is an outpatient procedure and LHM usually requires a 24-h hospitalization, after POEM patients are often hospitalized for 2–4 days; (2) the post-POEM LES pressure is usually between 15 and 20 mm Hg, a known predictor for recurrence of symptoms; (3) although the incidence of abnormal reflux as proven by pH monitoring is around 9% after LHM and fundoplication, it is around 40–50% after POEM. But most importantly, we have only very heterogeneous short-term results from non-randomized trials: although some studies have shown excellent outcomes, others have documented a failure rate of 11% at 6 months and 18% at 1 year. Therefore, in order to establish the real value of POEM in the treatment of achalasia, long-term results of prospective and randomized trials are needed.

At the University of Chicago, patients with achalasia are seen in the Center of Esophageal Diseases, where a team of radiologists, gastroenterologists, and surgeons evaluates the patients and decides the best therapeutic approach. The LHM and Dor fundoplication are our preferred treatments based on expertise and results. In case the patient has recurrent dysphagia, we propose PD. In case of failure of PD, we use either POEM or a redo myotomy. We reserve an esophagectomy for patients who have failed all the other treatment modalities. Even though there are no available data, we feel that a multidisciplinary team probably offers the best options for every patient.

Today, we have three effective treatment modalities for patients with achalasia, which allow gastroenterologists and surgeons to tailor treatment to individual patients and individual situations based.
CONFLICT OF INTEREST

Guarantor of the article: Marco G. Patti, MD.

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1. Fisichella PM, Raz D, Palazzo F et al. Clinical, radiological, and manometric profile in 145 patients with untreated achalasia. World J Surg 2008; 32: 1974–1979.
2. Sinan H, Tatum RP, Soarez RV et al. Prevalence of respiratory symptoms in patients with achalasia. Dig. Esophagus 2011; 24: 224–228.
3. Pandolfini JE, Kwiatek MA, Nealis T et al. Achalasia: A New Clinically Relevant Classification by High-Resolution Manometry. Gastroenterology 2008; 135: 1526–1533.
4. Boeckxstaens GE, Annese V, des Varannes SB et al. Pneumatic dilatation versus laparoscopic Heller myotomy for idiopathic achalasia. N Engl J Med 2011; 12: 1807–1816.
5. Campos GM, Vittinghoff E, Rabi C et al. Endoscopic and surgical treatments for achalasia: a systematic review and meta-analysis. Ann Surg 2009; 249: 45–57.
6. 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: 572–586.
7. 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.
8. Inoue H, Minami H, Kobayashi Y et al. Peroral endoscopic myotomy (POEM) for esophageal achalasia. Endoscopy 2010; 42: 265–271.
9. Onimaru M, Inoue H, Ikeda H et al. Peroral endoscopic myotomy is a viable option for failed surgical esophagocardiomyotomy instead of redo surgical Heller myotomy: a single center prospective study. J Am Coll Surg 2013; 217: 596–605.
10. Bhayani NH, Kurian AA, Dunst CM et al. A Comparative Study on Comprehensive, Objective Outcomes of Laparoscopic Heller Myotomy With Per-Oral Endoscopic Myotomy (POEM) for Achalasia. Ann Surg 2014; 259: 1098–1103.
11. Hunneges ES, Tettelbaum EN, Santos BF et al. Comparison of perioperative outcomes between peroral esophageal myotomy (POEM) and laparoscopic Heller myotomy. J Gastrointest Surg 2013; 17: 228–236.
12. Von Renteln D, Fuchs KH, Fockens P et al. Peroral endoscopic myotomy for the treatment of achalasia: an international prospective multicenter trial. Gastroenterology 2013; 145: 309–311.
13. Familiar P, Gigante G, Marchese M et al. Peroral endoscopic myotomy for esophageal achalasia. Outcomes of the first 100 patients with short term follow-up. Ann Surg 2014; e-pub ahead of print 30 October 2014.

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