SSAT Patient Care Guidelines

Esophageal Achalasia

Introduction
Esophageal achalasia is a primary esophageal motility disorder of unknown etiology, characterized by absence of esophageal peristalsis and increased or normal resting pressure of the lower esophageal sphincter (LES), which fails to relax completely in response to swallowing.

Clinical Presentation
Dysphagia is the most common symptom, experienced by virtually all patients. Regurgitation is the second most common symptom, and is present in about 60% of patients. It occurs more often in the supine position, and exposes the patients to the risk of aspiration of undigested food. Chest pain occurs in about 40% of patients, and is usually experienced at the time of a meal. Heartburn is experienced by about 40% of patients. In untreated patients this symptom is usually due to stasis and fermentation of food or esophageal distension.

Diagnosis
In addition to careful symptomatic evaluation, the following tests should be routinely performed: Barium swallow usually shows narrowing at the level of the gastroesophageal junction ("bird beak"), and various degrees of esophageal dilatation. Endoscopy is important to rule out the presence of a peptic stricture or cancer, and gastroduodenal pathology. In patients older than 60 years of age, with recent onset of dysphagia and excessive weight loss, secondary or pseudo-achalasia (obstruction due to a submucosal neoplasm in the distal esophagus) should be ruled out. Because a cancer of the gastroesophageal junction is the most common cause of pseudo-achalasia, an endoscopic ultrasound or a CT scan of the gastroesophageal junction can help to establish the diagnosis. Esophageal manometry is the key test for establishing the diagnosis. The classic manometric findings are: (a) absence of esophageal peristalsis, and (b) hypertensive or normotensive LES which fails to relax completely in response to swallowing.

Treatment
Treatment is directed toward elimination of the outflow resistance at the level of the gastroesophageal junction. The following treatment modalities are available to achieve this goal:

Traditionally, pneumatic dilatation has been the first line of treatment for esophageal achalasia, while surgery was reserved for patients who had persistent dysphagia after multiple dilatations or who had suffered a perforation during dilatation. Today, minimally invasive surgery has completely changed this treatment algorithm and a laparoscopic Heller myotomy and partial fundoplication is preferred by most gastroenterologists and surgeons as the primary treatment modality. When properly performed, a Heller myotomy can be expected to result in permanent relief of dysphagia in 85-100% of patients. Critical details of the operation include a generous myotomy of the lower esophagus, extending well onto the gastric wall. Because of the lack of esophageal peristalsis, a partial (Dor or Toupet), rather than a total fundoplication is frequently added to prevent reflux. A recent prospective, randomized study demonstrated that Heller myotomy plus a partial fundoplication is superior to Heller myotomy alone in regard to the incidence of postoperative reflux as measured by 24 hour pH testing. Patients can usually eat the morning of the first postoperative day, and can be discharged home after one or two days. In the only prospective, randomized trial performed comparing balloon dilation with surgery, myotomy outperformed balloon dilation 95% to 65%.

Historically, the most popular treatment for achalasia has been by forceful pneumatic dilation. The success rate of this procedure is 55-70% with a single dilation but can be increased to nearly 90% with multiple dilations. However, the risk of perforation with each dilation is at least 3-5% and has been reported as high as 12% in some series. These patients may require open surgery to close the perforation and perform a myotomy. Furthermore, when stratified by age, balloon dilation is less than 50% effective in patients younger than 40 years old and is rarely effective in adolescents.

Intrasphincteric injection of botulinum toxin (BOTOX) injection is less effective than balloon dilation and requires re-treatment to maintain an efficacy rate of 65%. Of greater concern is the fact that BOTOX injection leads to scar formation in the submucosal plane which results in a more difficult myotomy and higher mucosal perforation rate (up to 30%) during dissection. Thus, BOTOX should be reserved for the treatment of patients who are poor candidates for surgery and poor candidates for balloon dilation (dilated sigmoid esophagus) or as a bridge to surgery. An additional utility for BOTOX is in aiding in the diagnosis of patients who have equivocal findings on initial evaluation. A good response to BOTOX is usually an indication that the patient will have long-term relief following surgical myotomy.
In selected patients such as a hostile, multiply operated abdomen or following a failed abdominal myotomy, the thoracic or thoracoscopic approach may be preferred. The thoracic approach is also appropriate in managing patients with proximal esophageal motility abnormalities.

Occasionally the degree of esophageal aperistalsis is so advanced that myotomy alone will not relieve the dysphagia and the patient is better served with esophagectomy. Esophagectomy should be considered in a patient who has had a previous myotomy, with a resting LES pressure of less than 10 mmHg, and a dilated sigmoid esophagus. The need for esophagectomy for achalasia is very uncommon, even in the presence of a dilated esophagus, and should be reserved for failures after myotomy.

All patients undergoing treatment for achalasia should be followed by surveillance endoscopy, because they are at increased risk for development of both squamous and adenocarcinoma.

**Risks**

Aspiration of retained food in the esophagus at the time of induction of anesthesia and perforation of the esophageal mucosa are the most common operative complications. Persistent or recurrent dysphagia occurs in 5% to 10% of patients. The combination of intraoperative manometry and endoscopy can better guide the extent of the myotomy and can improve the adequacy of myotomy and are useful tools in decreasing the incidence of significant dysphagia after antireflux surgery. A complete work-up is necessary to evaluate the cause of the dysphagia in these patients, and either pneumatic dilatation or a second operation can often correct the problem. Up to 15% of patients may experience gastroesophageal reflux after myotomy, as measured by pH monitoring. In patients undergoing elective myotomy the mortality rate is less than 1%.

**Expected Outcomes**

About 90% of patients have long-term relief of dysphagia after a myotomy, with a low incidence of symptomatic acid reflux. There is often a poor correlation between symptoms of reflux and measurable reflux as demonstrated by pH study. All patients should be studied by postoperative pH study. Patients with demonstrated reflux by pH study or with reflux symptoms after surgery should be treated long-term with proton pump inhibitors.

**Qualifications for Performing Operations for Achalasia**

The qualifications of a surgeon performing any operative procedure should be based on training (education), experience, and outcomes. At a minimum, surgeons who are certified or eligible for certification by the American Board of Surgery, the Royal College of Physicians and Surgeons of Canada, or their equivalent should perform operations for achalasia. Achalasia surgery should preferably be performed by surgeons with special knowledge, training and experience in the management of gastroesophageal swallowing disorders. These surgeons have successfully completed at least 5 years of surgical training after medical school graduation and are qualified to perform operations on the esophagus and stomach. When performing laparoscopic or thoracoscopic operations, it is highly desirable that the surgeon has advanced videoscopic skills. The level of training in advanced videoscopic techniques necessary to conduct minimally invasive surgery is important to assess.

**Suggested Readings**


