Treatment of Achalasia

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Opinion statement
Achalasia is a primary motility disorder of the esophagus that causes dysphagia. Normal esophageal motility and lower esophageal sphincter (LES) function cannot be restored; thus treatment is directed at decreasing the pressure or disrupting the muscle fibers of the LES to allow passage of ingested material. Effective therapy for achalasia can be broadly characterized as surgery based or endoscopy based. Medications (calcium channel blockers and nitrate derivatives) do not provide adequate relief of dysphagia and have substantial side effects, and thus are rarely used as long-term therapy. Botulinum toxin injection, a recently introduced endoscopic therapy, enjoyed much enthusiasm initially but was shown to have only transient effect and is now recommended only for poor operative candidates. The mainstay of therapy remains endoscopic dilation or laparoscopic esophagomyotomy (LEM) combined with an antireflux procedure. We have found that patients who can tolerate a laparoscopic abdominal surgery are best served with an LEM and Toupet (270°) posterior fundoplication. This provides good or excellent relief of dysphagia in 90% to 95% of patients with very little morbidity.

Introduction
The human esophagus is composed of striated muscle proximally and smooth muscle distally that act in a coordinated fashion with the pharynx to facilitate oral progression of ingested materials. Two sphincters function in a highly coordinated manner with the esophageal propulsive wave to allow deglutition. The upper esophageal sphincter (UES), formed by the cricopharyngeus muscle, is found around 18 to 24 cm from the nares. The lower esophageal sphincter (LES), assisted by the diaphragmatic pinchcock and the transition from negative thoracic pressure to positive abdominal pressure, is usually found between 36 and 48 cm from the nares. Changes in any part of the esophageal mechanism can cause dysphagia.

Achalasia is a primary motility disorder of the esophageal body and lack of lower esophageal sphincter (LES) relaxation. Although it is the most common primary esophageal motility disorder, achalasia is still uncommon with reported incidence of 0.5 to 1.0 per 100,000 in North America. The first description likely occurred more than 300 years ago, though it was referred to as cardiospasm [1]. Coined by Lendrum in 1937, achalasia means “failure to relax” [2]. The disease manifests without gender predilection, usually in those between the ages of 20 and 50 (but can occur much earlier and later), as progressive dysphagia, first to solids, then liquids. Symptoms may also include regurgitation, chest pain, and weight loss. Heartburn may also occur but is usually caused by fermentation of stagnant food in the esophagus and not by gastroesophageal reflux [3]. However, misdiagnosis as gastroesophageal reflux disease is a common reason for delay in treatment that occurs commonly with patients with achalasia. Patients may also complain of cough, aspiration, choking, recurrent pneumonia, and delayed regurgitation of undigested food.

Manometry is essential to make the diagnosis. However, endoscopy and contrast fluoroscopy are used to fully evaluate the upper gastrointestinal tract and to rule out other abnormalities. Pseudoachalasia, usually caused by a malignancy, can mimic achalasia [3]. Patients over 50 years of age with short symptom duration (<6 months) or weight loss (>10 lbs.) should be carefully evaluated, even pursuing endoscopic ultrasound and/or computed tomography (CT) scan to rule out pseudoachalasia [3-5].

The etiology of achalasia is unknown, but is thought to be caused by progressive loss of post-ganglionic excitatory
and inhibitory neurons of the esophageal myenteric plexus and by lack of nitrous oxide synthase in the LES fibers resulting in poor propagation of the food bolus and lack of LES relaxation [6]. Reduction of nitric oxide and vasoactive intestinal peptide, both important mediators of LES relaxation, combined with ganglion-cell loss of the myenteric plexus in the body of the esophagus result in symptoms and dilation of the esophagus. Histologically, myenteric inflammation, depletion of ganglion cells and neural fibrosis, are found in the muscle tissue [7].

Achalasia treatment has undergone substantial changes over the last 20 years. Balloon dilation and esophageal myotomy are the mainstays of treatment. The advent of the minimally invasive approach removes the majority of surgical morbidity. Cost effectiveness of surgical intervention must be weighed fairly against cost of repeated dilations, as are usually required [8,9]. Laparoscopic esophageal myotomy and Toupet fundoplication should be considered primary treatment for achalasia for most patients.

**Treatment**

**Diet and lifestyle**

- Most patients adapt their eating behavior before the diagnosis is made. Because the dysphagia is usually slowly progressive, first for solids and then liquids, symptoms are often quite severe upon presentation. Patients often augment food passage by gulping large quantities of liquids after swallowing food. They describe standing after meals, raising their hands over their heads, and jumping repeatedly in an attempt to pass food into the stomach with gravity’s assistance. If these maneuvers fail, spontaneous or forced regurgitation is common. Stress and cold liquids may exacerbate the dysphagia. In addition, due to impaired esophageal emptying, 10% to 39% of patients suffer bronchopulmonary complications due to repeated regurgitation and aspiration [10]. Twenty-four percent of patients also report heartburn [11]. Though 24-hour pH monitoring rarely demonstrates gastroesophageal reflux, it can be falsely positive due to lactic acid produced by fermentation of retained food in the esophagus [3,12,13].

- Patients with achalasia report low quality of life scores, approximately 31% lower than normal cohorts. Scores have been shown to improve to near normal with successful treatment of dysphagia, regurgitation, and chest pain [10].

**Pharmacologic treatment**

- Medical management of achalasia has modest results at best. Although a certain population of patients will have limited benefit, most clinicians agree pharmacologic treatment should be reserved for those who refuse further intervention or are very poor surgical or dilation candidates [14–17]. A wide variety of medications have been tested to reduce the LES pressure, these include anticholinergics, nitrates, calcium channel blockers, theophylline, β2 agonists, and amyl nitrate. Calcium channel blockers (diltiazem, nifedipine) decrease lower esophageal sphincter pressure (LESP) by 13% to 49% for 30 to 120 minutes, with maximum effect (nifedipine) at 20 to 45 minutes. Improvement in patient symptoms is reported in up to 75% [1,14]. Iosorbide dinitrate reduces LESP from 30% to 66% for 30 to 90 minutes, with maximum effect (nifedipine) at 20 to 45 minutes. Improvement in patient symptoms is reported in up to 75% [1,14]. Iosorbide dinitrate reduces LESP from 30% to 66% for 30 to 90 minutes, with maximum effect (nifedipine) at 20 to 45 minutes. Improvement in patient symptoms is reported in up to 75% [1,14]. Nitrates are more potent LES relaxants whereas calcium channel blockers tend to be better tolerated [18]. Anticholinergics, loperamide, dicyclomine, verapamil, butylscopolamine, aminophylline, and others have not shown a significant decrease in LESP and no significant improvement in the symptoms [17]. Therefore, discussion here is limited to medications most often utilized. Medical therapy seems to work in patients with a transverse diameter of esophagus less than 5 cm, at least partial LES relaxation (30%), and LESP decrease of greater than or equal to 30% in at least one period following medication test dose [16].