Introduction

Achalasia is the most recognized motor disorder of the esophagus and the only primary motility disorder with an established pathology. The term means “failure to relax” and describes the predominant feature of this disorder—a poorly relaxing lower esophageal sphincter (LES). The first case of achalasia was reported more than 300 years ago by Sir Thomas Willis [1]. The patient’s esophageal obstruction responded to dilation with a whale’s bone.

Achalasia occurs with equal frequency among men and women. Case studies show an age distribution between birth and the ninth decade of life, with the peak incidence between 30 and 60 years. There is no racial predilection [1]. Achalasia is an uncommon disease, but it occurs frequently enough to be encountered at least yearly by most gastroenterologists. Esophageal specialists, both gastroenterologists and surgeons, can see six to 10 cases of achalasia a year. The disease prevalence is approximately 10 cases per 100,000 population. Its incidence has been fairly stable over the last 50 years at about 0.5 cases per 100,000 population per year [2].

The cause of achalasia is unknown. Available data suggest that hereditary, degenerative, autoimmune, and infectious diseases are possible causes, the latter two being the most commonly accepted [1]. Pathologic changes identified at necropsy or from myotomy specimens are seen in the esophageal myenteric (Auerbach’s) plexus and include a prominent but patchy inflammatory response consisting of T lymphocytes and variable numbers of

Opinion statement

The goals in the treatment of achalasia are threefold: 1) relieving the symptoms, particularly dysphagia and bland regurgitation; 2) improving esophageal emptying by disrupting the poorly relaxing lower esophageal sphincter (LES); and 3) preventing the development of megaesophagus. Although achalasia cannot be permanently cured, excellent palliation is available in over 90% of patients, especially those with pneumatic dilation and laparoscopic Heller myotomy. The efficacy for short- and long-term therapy seems to be similar when performed by experts. Pneumatic dilation done as an outpatient surgery disrupts the LES muscle from within by using balloons of progressively larger diameter (3.0, 3.5, and 4.0 cm). Repeat dilations may be required; secondary severe gastroesophageal reflux disease (GERD) is rare, but approximately 2% of patients will have an esophageal perforation. A surgical Heller myotomy is now being done laparoscopically through the abdomen that cuts the LES and extends the myotomy 2 to 3 cm onto the stomach. Usually 2 days of hospitalization is required, and patients can normally return to work in 1 to 2 weeks. Severe GERD with esophagitis and peptic stricture is a common complication; therefore, most surgeons combine the myotomy with an incomplete fundoplication. Medical therapy is much less effective than these invasive procedures. Smooth muscle relaxants (nitrates and calcium channel blockers) taken immediately before meals improve dysphagia, but side effects and drug tolerance are common. The injection of botulinum toxin (100 to 200 units) endoscopically into the LES gives short-term relief of symptoms and improves esophageal emptying. This treatment is most effective in the elderly, as symptom relief can last up to 1 to 2 years with a single injection. Several studies suggest the most cost-effective management of achalasia is initial treatment with pneumatic dilation.
esosisophils and mast cells, loss of ganglion cells and some degree of myenteric neural fibrosis \[3\cdot 4\]. The end result of the inflammatory process is a selective loss of postganglionic inhibitory neurons containing nitric oxide and vasoactive intestinal polypeptide. Because postganglionic excitatory neurons are spared, cholinergic stimulation continues unopposed \[5\], leading sometimes to high resting LES pressure. The loss of inhibitory input results in abnormal and usually incomplete LES relaxation, and aperistalsis is caused by loss of the latency gradient that permits sequential contractions along the esophageal body, a process medicated by nitric oxide.

The diagnosis of achalasia should be suspected in patients complaining of dysphagia for solids and liquids with regurgitation of bland food and saliva \[1\]. Some patients initially have only solid food dysphagia but by the time of clinical presentation, nearly all complain of solid food dysphagia while eating and drinking, especially cold beverages. Regurgitation of bland, nonacidic, undigested food, or accumulated saliva occurs postprandially and at night while recumbent, often waking the patient from sleep because of coughing and choking. Chest pain occurs in some patients but is more common in milder disease. Heartburn is a frequent complaint in achalasia, despite the fact that achalasia is not associated with increased episodes of acid reflux by esophageal pH monitoring. The symptom cause is speculative but probably related to the production of lactic acid from retained food or ingested acid foods and beverages such as carbonated drinks. Most achalasia patients have some degree of weight loss at presentation; however, the loss is usually only 5 to 20 lbs., and some patients are obese.

When achalasia is suspected, a barium esophagram with fluoroscopy is the best initial test \[6\]. The esophagus may be of normal caliber, is usually dilated and sometimes tortuous, does not empty, and retained food and saliva produces an air-fluid level at the top of the barium column. The distal esophagus is characterized by a smooth tapering leading to the closed LES that resembles a bird’s beak. Fluoroscopy always shows a lack of peristalsis replaced by to-and-fro movement in the supine position. Sometimes an epiphrenic diverticulum is seen.

Esophageal manometry is required to establish the diagnosis of achalasia and must be done in any patient where invasive therapies are planned \[1\]. Because achalasia only involves the smooth muscle of the esophagus, the manometry abnormalities are confined to the distal two-thirds of the esophagus. All patients have at least two manometric abnormalities; they are aperistalsis and abnormal LES relaxation. In the esophageal body, all swallows are followed by low amplitude (10 to 40 mmHg), simultaneous waves that are identical to each other (isobaric or mirror images). The term “vigorous achalasia” is used to describe cases with aperistalsis and normal or even high amplitude contractions in the esophageal body \[7\] and may represent an early histologic form of the disease \[4\]. Abnormal LES relaxation is seen in all achalasia patients, with 70% to 80% having incomplete or absent relaxation. In the remaining group, the relaxations are complete to gastric baseline but are of short duration (usually < 6 seconds) and functionally inadequate, as shown by barium and nuclear studies of emptying \[8\]. Other abnormalities seen include elevated LES pressure in up to 50% of patients and an increase in esophageal baseline pressure, often greater than gastric pressure due to retention of food and saliva.

Some tumors, especially adenocarcinoma of the stomach, can produce a pseudoachalasia picture. Therefore, all patients with suspected achalasia will need a careful upper endoscopy with close examination of the cardia and gastroesophageal junction. Endoscopic ultrasound and computed tomography scan are sometimes needed to help make the diagnosis of pseudoachalasia.

No treatment can restore muscular activity to the denervated esophagus in achalasia. Esophageal aperistalsis and impaired LES relaxation are rarely, if ever, reversed by any mode of therapy. Therefore, every treatment for achalasia is directed to reducing the pressure gradient across the LES, thus facilitating esophageal emptying by gravity and hopefully preventing the future development of megaeosophagus \[6\]. This disruption of the LES gradient is best accomplished with pneumatic dilation or surgical myotomy and less effectively by pharmacologic agents injected endoscopically into the sphincter (eg, botulinum toxin) or taken orally (eg, calcium channel blockers or nitrates). The symptoms of regurgitation and dysphagia are the easiest to treat, but chest pain can be problematic in some patients, even in those with successful esophageal decompression. Overall, using single or multiple modalities of treatment, over 90% of achalasia patients will do well \[1\].

### Treatment

#### Diet and lifestyle

- Hard, dry foods should be avoided. Substitute softer, well-chewed foods.
- Cold liquids worsen symptoms in some patients.
- Carbonated beverages with their expanding gases may augment esophageal emptying.
- Lifting the neck and arms while swallowing hard increases intraesophageal pressure and may promote esophageal emptying.