

Achalasia

Achalasia ("no relaxation") is a failure of esophageal smooth muscle fibers to relax, which can cause two problems in the esophagus:

- The muscles in the tube of the esophagus are not able to contract in a coordinated way (or at all) to propel food down the esophagus.
- The lower esophageal sphincter fails to remain closed and fails to open when food is swallowed and moves down the esophagus. This creates a functional blockage preventing food from easily moving into the stomach for digestion.

The most common form is primary achalasia, which has no known underlying cause. It is due to the failure of distal esophageal inhibitory neurons. However, a small proportion occurs secondary to other conditions, such as esophageal cancer or Chagas disease (an infectious disease common in South America). Achalasia affects about one person in 100,000 per year. There is no gender predominance for the occurrence of disease.

Symptoms

The main symptoms of achalasia are:

- Difficulty in swallowing (dysphagia)
- Regurgitation of undigested food,
- Chest pain behind the sternum,
- and sometimes weight loss

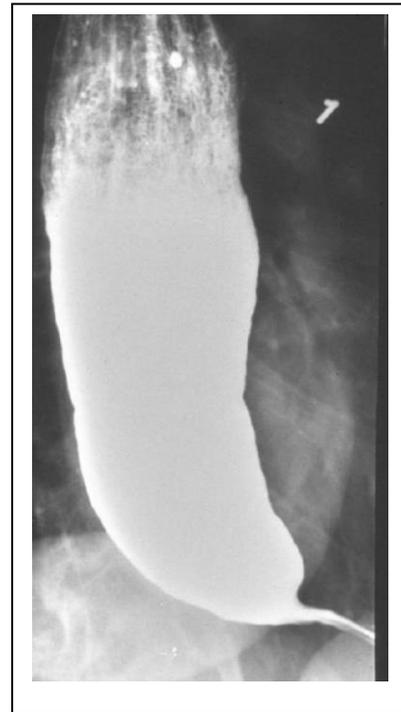
Dysphagia tends to become progressively worse over time and to involve both fluids and solids. Some people may also experience coughing when lying in a horizontal position. The chest pain experienced, also known as cardiospasm and non-cardiac chest pain can often be mistaken for a heart attack. It can be extremely painful in some sufferers. Food and liquid, including saliva, are retained in the esophagus and may be inhaled into the lungs (aspiration).

Diagnosis

Due to the similarity of symptoms, achalasia can be mistaken for more common disorders such as gastroesophageal reflux disease (GERD), and hiatal hernia. Specific tests for achalasia are barium swallow and esophageal manometry. In addition, endoscopy of the esophagus, stomach, and duodenum (esophagogastroduodenoscopy or EGD) is typically performed to rule out the possibility of cancer. The internal tissue of the esophagus generally appears normal in endoscopy, although a "pop" may be observed as the scope is passed through the non-relaxing lower esophageal sphincter with some difficulty, and food debris may be found above the LES.

Barium swallow

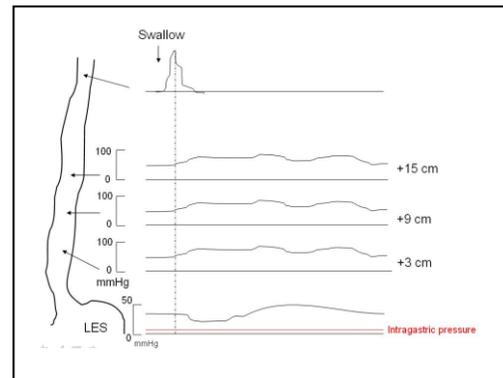
The patient swallows a barium solution, with an X-ray recording to observe the flow of the fluid through the esophagus. Normal movement of the esophagus is not seen. There is a tapering at the lower esophageal sphincter and narrowing at the gastro-esophageal junction, producing a "bird's beak" appearance. The esophagus above the narrowing is often dilated (enlarged) to varying degrees as the esophagus is gradually stretched over time.^[5] An air-fluid margin is often seen over the barium column due to the lack of peristalsis. A five-minutes timed barium swallow can provide a useful benchmark to measure the effectiveness of treatment.



"Bird's beak" appearance and "megaesophagus," typical in achalasia.

Esophageal manometry

Manometry is a test that measures the pressures inside the esophagus and the contraction and relaxation of the muscle using a small tube placed in the nose. It is an essential test for establishing the diagnosis of achalasia. The catheter (thin tube) is inserted through the nose, and the patient is instructed to swallow several times. The probe measures muscle contractions in different parts of the esophagus during the act of swallowing. Manometry reveals failure of the LES to relax with swallowing and lack of functional peristalsis in the smooth muscle esophagus.



Schematic of manometry in achalasia showing **aperistaltic** contractions, increased intraesophageal pressure, and failure of relaxation of the lower esophageal sphincter.

Characteristic manometric findings are:

- Lower esophageal sphincter (LES) fails to relax upon wet swallow (<75% relaxation)
- Pressure of LES <26 mm Hg is normal, >100 is considered achalasia, > 200 is **nutcracker achalasia**.
- Limited to no movement of the esophageal body called aperistalsis.
- Relative increase in intra-esophageal pressure as compared with intra-gastric pressure

Upper Endoscopy (EGD)

An upper endoscopy is performed to make sure there is not a cancer in the lower esophagus causing the obstruction. Biopsies can be performed of the lining of the esophagus to make sure.

Treatment

Sublingual nifedipine significantly improves outcomes in 75% of people with mild or moderate disease. It was classically considered that surgical myotomy provided greater benefit than either botulinum toxin or dilation in those who fail medical management. However, a recent randomized controlled trial found pneumatic dilation to be non-inferior to laparoscopic Heller myotomy.

Lifestyle changes

Both before and after treatment, achalasia patients may need to eat slowly, chew very well, drink plenty of water with meals, and avoid eating near bedtime. Raising the head off the bed or sleeping with a wedge pillow promotes emptying of the esophagus by gravity. After surgery or pneumatic dilatation, proton pump inhibitors are required to prevent reflux damage by inhibiting gastric acid secretion, and foods that can aggravate reflux, including ketchup, citrus, chocolate, alcohol, and caffeine, may need to be avoided.

Medication

Drugs that reduce LES pressure are useful. These include calcium channel blockers such as nifedipine and nitrates such as isosorbide dinitrate and nitroglycerin. However, many patients experience unpleasant side effects such as headache and swollen feet, and these drugs often stop helping after several months.

Botulinum toxin (Botox) may be injected into the lower esophageal sphincter to paralyze the muscles holding it shut. As in the case of cosmetic Botox, the effect is only temporary and lasts about 6 months. Botox injections cause scarring in the sphincter which may increase the difficulty of later Heller myotomy. This therapy is recommended only for patients who cannot risk surgery, such as elderly people in poor health. Pneumatic dilatation has a better long term effectiveness than botox.

Pneumatic dilatation

In balloon (pneumatic) dilation or dilatation, the muscle fibers are stretched and slightly torn by forceful inflation of a balloon placed inside the lower esophageal sphincter. Gastroenterologists who specialize in achalasia have performed many of these forceful balloon dilatations and achieve better results and fewer perforations. There is always a small risk of a perforation which requires immediate surgical repair. A perforation is a serious complication that can lead to death from infection. Pneumatic dilatation causes some scarring which may increase the difficulty of Heller myotomy if the surgery is needed later. Gastroesophageal reflux (GERD) occurs after pneumatic dilatation in some patients. Pneumatic dilatation is most effective in the long-term on patients over the age of 40; the benefits tend to be shorter-lived in younger patients. It may need to be repeated with larger balloons for maximum effectiveness.

Surgery

Robotic Heller myotomy is the “gold standard” for treating achalasia. When performed by an experienced surgeon it provides the best long term outcome with the least risk. The myotomy is a lengthwise 8 cm cut along the esophagus, starting above the LES by the base of the heart and extending down onto the stomach a little way. The esophagus is made of several layers, and the myotomy cuts only through the outside muscle layers which are squeezing it shut, leaving the inner muscosal layer intact. A partial fundoplication or "wrap" is generally added in order to prevent excessive reflux, which can cause serious damage to the esophagus over time. After surgery, patients should keep to a soft diet for several weeks to a month, avoiding foods that can aggravate reflux.

The most recommended fundoplication to complement Heller myotomy is Dor fundoplication, which consists of a 180- to 200-degree anterior wrap around the esophagus.

Endoscopic myotomy

A new endoscopic therapy for achalasia management was developed in 2008 in Japan. It is still under investigation and experimental. Per-oral endoscopic myotomy or POEM is a minimally invasive type of natural orifice transluminal endoscopic surgery that follows the same principle as the Heller myotomy. A tiny incision is made on the esophageal mucosa through which an endoscope is inserted. The innermost circular muscle layer of the esophagus is divided and extended through the LES until about 2 cm into the gastric muscle. Since this procedure is performed entirely through the patient's mouth, there are no visible scars on the patient's body.

Patients usually spend about 1–4 days in the hospital and are discharged after satisfactory examinations. Patients are discharged on full diet and generally able to return to work and full activity immediately upon discharge. Major complications can occur after POEM including a leak from the esophagus. This can cause serious infection and even death. Long term patient satisfaction is similar following successful POEM compared to standard laparoscopic Heller myotomy.