

Achalasia

Achalasia is a motility disorder in which the esophagus empties slowly. The delay results from poor opening of the lower esophageal sphincter (valve) in association with the loss of normal, orderly muscle activity (peristalsis) stripping foods and liquids along the esophagus into the stomach. Achalasia results from the nerves in the esophagus and lower esophageal valve being damaged or destroyed. The cause is unknown; most cases are idiopathic and possibly viral or autoimmune in etiology; rarely it may be due to a cancer. Men and women are equally affected. The average age of presentation is approximately 50 years, but it may also be diagnosed in children or the elderly.

Symptoms of Achalasia

The symptoms of achalasia most often occur during and after a meal. All patients experience the sensation of solids, and usually liquids, hanging up and passing slowly into the stomach several times a week to every meal. Effortless regurgitation of bland, undigested food or whitish foam (saliva) is common and may be associated with coughing and choking at night. Some patients experience chest pain, and heartburn is common. The latter is not related to acid reflux into the esophagus, but rather due to retained acidic food or the fermentation of food in the esophagus. Approximately 50% of patients will lose weight.

Tests for Achalasia

The diagnosis of achalasia is suspected by barium x-rays and confirmed by esophageal manometry. Barium studies of the esophagram (esophagram) show a “bird beak” -like narrowing of the lower esophageal valve, esophageal dilation with retained barium in the upright position, poor esophageal emptying, and to-and-fro movement due to the loss of orderly peristalsis. Esophageal manometry involves placing a small tube with pressure sensors into the stomach, and slowly withdrawing while measuring lower esophageal valve pressure and peristalsis. All achalasia patients have abnormal relaxation (opening) of the lower esophageal valve combined with a lack of peristalsis (orderly wave progression) in the esophageal body. Sometimes the lower esophageal valve pressure will also be abnormally high. All achalasia patients should also have an upper GI endoscopy to exclude the unusual case of a cancer (usually adenocarcinoma of the stomach) causing a pseudo-achalasia picture.

Treatment of Achalasia

The goal of treating achalasia is to disrupt and open the lower esophageal valve to improve esophageal emptying and relieve symptoms. Unfortunately, no treatment is available to promote the return of peristalsis. The best treatments for healthy patients are pneumatic dilatation or laparoscopic Heller myotomy. Frail or elderly patients may do well with botulinum toxin (Botox) injections. Treatment markedly relieves symptoms, but are rarely curative. Retreatment may be required and alternative treatments may be needed.

Pneumatic dilatation involves upper GI endoscopy with the passage of various size balloons (3.0, 3.5, 4.0 cm) to tear the esophagus from within, opening the valve. The procedure is done with conscious sedation and takes about 30 minutes, with a loss of one day of activity. The major complication, esophageal perforation (hole in the esophagus), is rare (less than 5%), but will require major surgery.

Surgery for achalasia involves cutting the muscle (myotomy) from the outside via small laparoscopic sites on the abdomen. The procedure requires general anesthesia, hospitalization for up to two days, and limited activity for two weeks. The major side effect is cutting the muscle too much, causing bad acid reflux. Overall, the success of both of these procedures is 80%-90% and dependent on the skills of the operator.

Botulinum toxin can be injected into the esophagus and lower esophageal valve, via a needle passed through an upper endoscope. The toxin poisons the valve, relaxing the sphincter, and thereby relieving symptoms. In young patients, symptoms relief is generally for only 3-6 months, while older patients may have relief for one year or longer. Adverse events are rare.

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This article is written for the American Motility Society (AMS) and the International Foundation for Functional Gastrointestinal Disorders (IFFGD).