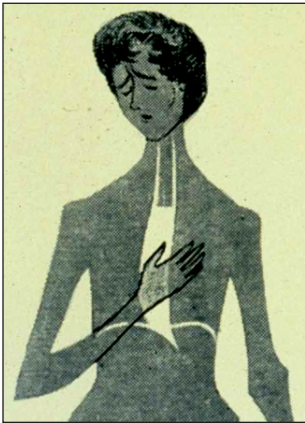


Achalasia

An educational handout for patients



What is achalasia?

Achalasia (ak" ah-la' ze-ah) is a rare disorder of the esophagus (food pipe). Achalasia results from a malfunction of the nerves that control the movement of food through the esophagus. It makes it difficult to swallow food and liquid.

What are the symptoms of achalasia?

- Trouble swallowing, both liquid and solid food; food seems to hang up in your chest
- Food or liquid comes back up into your throat (regurgitation), especially when you are lying down; it is often mixed with saliva and mucous.
- Chest pain or discomfort
- Weight loss

How does the esophagus work?

The esophagus is a hollow muscular tube that is about 10 inches long. It has two valve-like muscles, one at each end. These muscles are called the upper and the lower esophageal sphincters. These sphincters control the passage of food. In achalasia, there is a problem with the esophageal muscles and the lower esophageal sphincter.

Normally, after you swallow food, rhythmic contractions of the esophagus push food downward into the stomach. At the lower end of the esophagus, where it meets the stomach, the lower esophageal sphincter is normally closed to prevent stomach contents from coming back up into the esophagus. However, when you swallow, the lower esophageal sphincter opens to let the swallowed food into the stomach. This pattern of swallowing is called peristalsis. It is controlled by nerves in the wall of the esophagus.

What causes achalasia?

The exact cause of achalasia is not known. However, there is evidence that your own immune system destroys the nerves in the wall of the esophagus. As a result, the normal pattern of swallowing (peristalsis) does not occur, and the lower esophageal sphincter muscle does not relax properly. These defects prevent the passage of swallowed food into the stomach. As achalasia gets worse, the esophagus gradually enlarges as food collects within it. This food may be regurgitated hours or even days after being eaten.

Does achalasia affect other parts of the body?

Achalasia affects only the esophagus and typically does not involve other organs, such as the stomach or intestine. Rarely, other conditions mimic achalasia; for example, some forms of cancer and infectious disease (Chagas disease). These conditions do involve other organs. Depending on a person's risk factors, it may be necessary to test for these conditions.

What are the complications of achalasia?

In the early stages of achalasia, the main symptoms are trouble swallowing or chest pain. If achalasia is not treated, the food that collects in the esophagus can cause complications. This material can sometimes spill into the lungs causing bronchitis, pneumonia, or chronic lung disease. The retained food can also cause chronic irritation of the esophageal lining, sometimes with fungal infections. Poor nutrition may lead to weight loss or malnutrition. There is also a small increased risk for esophageal cancer.

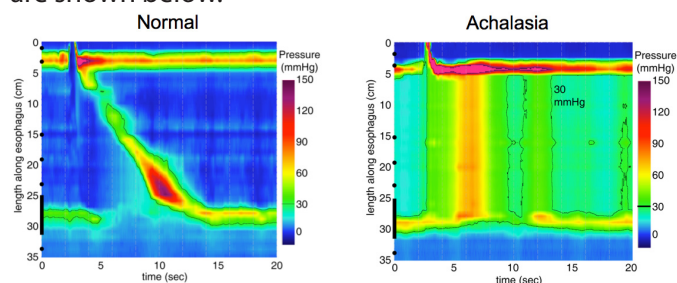
What tests are used to diagnose achalasia?

Achalasia is a rare condition. It can be confused with more common esophageal problems, such as gastroesophageal reflux disease (GERD). Because of this, patients can be misdiagnosed for many months or years. If you have constant trouble swallowing, you should see your doctor or be referred to a specialist in gastroenterology. Several tests are used to diagnose achalasia. Some tests verify the diagnosis of achalasia, while other tests rule out diseases that mimic achalasia.

Upper endoscopy: This test, also referred to as esophagogastroduodenoscopy (EGD), allows the

doctor to examine the lining of the esophagus, stomach, and duodenum. A thin, flexible instrument with a built-in camera is passed through your mouth into your esophagus while you are lightly sedated. It is the best way to detect ulcers, growths, or infections.

Esophageal manometry: This test can find out if peristalsis is working and if the lower esophageal sphincter is relaxing. A thin tube with pressure sensors is passed through your nose into the esophagus and stomach. You are asked to swallow small amounts of water while your esophageal muscle contractions are recorded on a computer. Manometry is one of the best ways to diagnose achalasia. Recent improvements in this technology include high-resolution manometry (HRM) or high-resolution esophageal pressure topography (EPT). Examples of images taken by these instruments are shown below.



Esophagram or upper GI: This is an X-ray study of the esophagus and stomach. X-rays are taken after you drink a barium solution. The X-ray shows the movement of barium through the esophagus and stomach.



What are the initial ways to treat achalasia?

- Currently, the nerve damage in the esophagus cannot be repaired. Fortunately, there are several medical and surgical treatments that improve swallowing function. However, esophageal function never completely returns to normal. You will always have some trouble swallowing. You can try the following techniques to improve your swallowing function:
- Eat slowly and drink plenty of liquids while you eat.
- Chew food thoroughly.
- Stay upright while you eat and for at least 1 hour after eating.
- Drink a full glass of water with pills!

Medical treatments

Medicines that relax the lower esophageal sphincter are helpful. Examples include nitrates such as isosorbide dinitrate or calcium channel blockers such as nifedipine; these medicines are commonly used to treat heart conditions and high blood pressure.

Alternatively, a Botox® injection into the lower esophageal sphincter may make it easier to swallow for 6 to 12 months. Continued relief is usually possible only after one of the definitive treatments: pneumatic balloon dilation or Heller myotomy.

Endoscopic treatment

Pneumatic balloon dilation is an outpatient procedure that is done along with an endoscopy. A balloon dilator is positioned across the lower esophageal sphincter and expanded to stretch the sphincter and reduce its strength. This can provide long-lasting relief, but it may need to be repeated from time to time.

Surgical treatment

Surgery is an important treatment option for achalasia. The preferred operation is a laparoscopic **Heller myotomy**. This involves a keyhole surgical technique, similar to the way gallbladders are now removed. The lower esophageal sphincter muscle is cut so that it no longer obstructs the passage of food into the stomach. The Heller myotomy is usually accompanied by another procedure called fundoplication, in which the upper portion of the stomach (fundus) is wrapped around the lower end of the esophagus. This procedure helps to reduce the risk of too much acid reflux from the stomach. After surgical procedures, patients may need to take medicine (usually a proton pump inhibitor) to reduce the secretion of stomach acid. In very advanced cases of achalasia, or if traditional treatments fail, it may be necessary to remove the diseased part of the esophagus.

Are there any new or experimental therapies?

New endoscopic treatments for achalasia are being investigated. For more information, visit the ANMS web site at www.motilitysociety.org and the National Institutes of Health web site at www.nih.gov.

For a list of doctors in your area who specialize in gastrointestinal motility problems, go to our web site:

www.motilitysociety.org

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