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Patient information: Achalasia (Beyond the Basics)

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Disclosures

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Literature review current through: Mar 2013. | **This topic last updated:** Feb 1, 2012.

ACHALASIA OVERVIEW — Achalasia is an uncommon swallowing disorder that affects about 1 in every 100,000 people. The major symptom of achalasia is usually difficulty with swallowing. Most people are diagnosed between the ages of 25 and 60 years. Although the condition cannot be cured, the symptoms can usually be controlled with treatment.

ACHALASIA CAUSE — In achalasia, nerve cells in the esophagus (the tube that carries food from the mouth to the stomach) degenerate for reasons that are not known. The loss of nerve cells in the esophagus causes two major problems that interfere with swallowing ([figure 1](#)):

- The muscles that line the esophagus do not contract normally, so that swallowed food is not propelled through the esophagus and into the stomach properly.
- The lower esophageal sphincter (LES), a band of muscle that encircles the lower portion of the esophagus, does not function correctly.

Normally, the LES relaxes when we swallow to allow swallowed food to enter the stomach. When the food has moved through the esophagus into the stomach, the LES muscle contracts to squeeze the end of the esophagus closed, thus preventing the stomach contents from flowing backwards (refluxing) into the esophagus.

In people with achalasia, the LES fails to relax normally with swallowing. Instead, the LES muscle continues to squeeze the end of the esophagus, creating a barrier that prevents food and liquids from passing into the stomach ([figure 2](#)). Over time, the esophagus above the persistently contracted LES dilates, and large volumes of food and saliva can accumulate in the dilated esophagus.

ACHALASIA SYMPTOMS — The most common symptom of achalasia is difficulty swallowing. Patients often experience the sensation that swallowed material, both solids and liquids, gets stuck in the chest. This problem often begins slowly and progresses gradually. Many people do not seek help until symptoms are advanced. Some people compensate by eating more slowly and by using maneuvers, such as lifting the neck or throwing the shoulders back, to improve emptying of the esophagus.

Other symptoms can include chest pain, regurgitation of swallowed food and liquid, heartburn, difficulty burping, a sensation of fullness or a lump in the throat, hiccups, and weight loss.

ACHALASIA DIAGNOSIS — Achalasia may be suspected based upon symptoms, but tests are needed to confirm the diagnosis.

Chest x-rays — A chest x-ray may reveal a dilated esophagus and absence of air in the stomach. However, a chest x-ray is not adequate for a diagnosis of achalasia and further testing is required.

Barium swallow test — The [barium](#) swallow test is a common screening test for achalasia. The test involves swallowing a chalky-tasting, thick mixture of barium while x-rays are taken. The barium shows the outline of the esophagus and lower esophageal sphincter (LES) ([figure 2](#)). Characteristic findings of achalasia on barium swallow include a persistently narrowed region at the end of the esophagus (the LES), with a dilated esophagus above the narrowed region. The barium swallow may also show spastic contractions in the esophagus above the LES, a condition called “vigorous achalasia”.

Manometry — Manometry is a test that measures changes in pressures within the esophagus that are caused by the contraction of the muscles that line the esophagus. The test involves the passage of a thin tube through the mouth or nose into the esophagus. The tube is lined by numerous pressure sensors that convey pressures within the esophagus to a device that records those pressures. Patients are usually instructed to have nothing to eat or drink for eight hours before the test, and they are given sips of water to swallow while the tube is in place.

Manometry is almost always used to confirm the diagnosis of achalasia. The test typically reveals three abnormalities in people with achalasia: high pressure in the LES at rest, failure of the LES to relax after swallowing, and an absence of useful (peristaltic) contractions in the lower esophagus. The last two features are the most important and are required to make the diagnosis of achalasia.

Endoscopy — Endoscopy allows the physician to see the inside of the esophagus, LES, and stomach using a thin, lighted, flexible tube. Most patients are given sedatives during the endoscopy procedure. This test is usually recommended for people with suspected achalasia and is especially useful for detecting other conditions that can mimic achalasia such as cancer of the upper portion of the stomach. (See "[Patient information: Upper endoscopy \(Beyond the Basics\)](#)".)

In people with achalasia, endoscopy often reveals a dilated esophagus that contains retained food; it may also reveal inflammation, small ulcers caused by residual food or pills, and candida (yeast) infection.

The endoscope can be advanced through the LES and into the stomach to check for stomach cancer. Cancer in the upper part of the stomach can produce symptoms and abnormal manometry results that are virtually identical to those of achalasia. This condition is called pseudoachalasia (meaning “false” achalasia) or secondary achalasia. Biopsies (small samples of tissue) are often obtained in the lower portion of the esophagus to look for cancer cells. Having a biopsy of the esophagus taken during endoscopy is not painful and is generally a safe procedure.

ACHALASIA TREATMENT — Several options are available for the treatment of achalasia. Unfortunately, none can stop or reverse the underlying loss of nerve cells in the esophagus of patients with achalasia. However, the treatments are usually effective for improving symptoms.

None of the available treatments are expected to restore normal (peristaltic) contractions in the esophagus of patients with achalasia. Rather, the treatments aim to weaken the lower esophageal sphincter (LES) muscle to the point that it no longer poses a barrier to the passage of food. The LES can be weakened by drugs, or mechanically by procedures that tear or cut the LES muscle.

Drug therapy — Two classes of drugs, nitrates and calcium channel blockers, have LES muscle-relaxing effects. These drugs can decrease symptoms in people with achalasia. The drugs are usually taken by placing a pill under the tongue 10 to 30 minutes before meals.

Drug therapy is the least invasive and safest option for treating achalasia. However, most people find that long-term drug therapy is inconvenient, ineffective, and often associated with unpleasant side effects,

such as headache and low blood pressure. Furthermore, the drugs tend to become less effective over time. For these reasons, medications are recommended primarily for patients who are not interested in or not healthy enough for mechanical treatments such as balloon dilation and surgery (myotomy).

Balloon dilation (pneumatic dilatation) — For balloon dilation, the patient swallows a collapsed balloon that is positioned in the LES. An x-ray machine is often used to guide placement of the balloon. When the balloon has been positioned at the LES, it is inflated abruptly to a large size in order to tear the muscle of the LES. This procedure is effective for relieving the swallowing difficulty in patients with achalasia in approximately two-thirds of people, although chest pain persists in some. Patients frequently require more than one balloon dilation treatment for adequate relief.

Procedure — If you have balloon dilation, you will be asked to drink only liquids for 12 hours to two days in advance (a longer period is recommended if you have a great deal of food retention in the esophagus). Using endoscopy and fluoroscopy (x-ray), a physician advances a guide wire down the esophagus and positions it inside the LES. A deflated balloon is then advanced along this guide wire, positioned inside the LES, and inflated for a variable period ranging from seconds to minutes. The balloon is then deflated and withdrawn, and you are monitored in a recovery area for a number of hours to detect any complications. After the balloon dilation, some physicians routinely perform an x-ray test similar to the [barium swallow](#) described above to make sure that the balloon has not created a hole (perforation) in the esophagus. If there are no complications, you can usually resume eating when you have recovered from the procedure. If your day-to-day symptoms do not improve, additional dilations can be performed.

Success rate — A single balloon dilation session continues to relieve symptoms of achalasia in about 60 percent of people one year after the procedure and in about 25 percent of people five years after the procedure. Higher success rates have been reported in some studies. The success rate after longer periods has not been well studied, but some people have remained symptom-free for as long as 25 years.

Complications — About 15 percent of people experience severe chest pain immediately after balloon dilation and some experience fever.

The most serious complication of balloon dilation is creation of a hole (perforation) in the wall of the esophagus; this complication occurs in about 2 to 6 percent of people undergoing the procedure. Symptoms of persistent or worsening pain in the hours after the procedure may indicate a perforation.

Perforations of the esophagus after balloon dilation are usually small. For the treatment of small perforations, your doctor will probably admit you to the hospital for intravenous feeding and antibiotic treatment. This usually results in healing of the perforation within one week without surgery. Large perforations will require emergency surgery for repair. In some cases, your doctor might recommend surgery even for a small perforation. A key factor in the successful treatment of perforation of the esophagus is rapid identification of the perforation and rapid implementation of treatment. You should call your physician immediately if you experience increasing pain after balloon dilation, especially if you develop a fever or chills.

Bleeding is another important, but infrequent complication of balloon dilation. This complication usually occurs immediately after the dilation. Symptoms of bleeding include dizziness or fainting, especially on standing up, vomiting of blood or material that looks like coffee grounds, and the passage of black or bloody stools. You should notify your doctor immediately if you experience these symptoms.

Patients also can develop gastroesophageal reflux disease (GERD) after balloon dilation. Because the LES is the principal barrier that prevents stomach contents from refluxing (backwashing) into the esophagus, LES disruption by balloon dilation can lead to acid reflux. GERD occurs in about 2 percent of people after balloon dilation, but is usually easily controlled with acid-reducing medications. (See "[Patient information: Acid reflux \(gastroesophageal reflux disease\) in adults \(Beyond the Basics\)](#)".)

Surgery (myotomy) — Myotomy is an operation that is used to weaken the LES by cutting its muscle fibers. The most common surgical technique used to treat achalasia is called the Heller myotomy, in which the surgeon cuts the muscles at the end of the esophagus and at the top of the stomach. In the past, this surgery was performed through a large (open) incision in the chest or abdomen. Today, this surgery is usually performed laparoscopically, using instruments and a television camera that are passed into the abdomen through small abdominal incisions. People who undergo laparoscopic myotomy are given general anesthesia, and generally stay in the hospital for one to two days.

Success rate — Surgery relieves symptoms in 70 to 90 percent of people. Symptom relief is sustained in about 85 percent of people 10 years after surgery and in about 65 percent of people 20 years after the surgery. Thus, some consider surgery to be a more definitive treatment for achalasia than balloon dilation or botulinum toxin injection (see below).

Complications — Postoperative pain is expected, and is treated with pain medications. Like balloon dilation, there is a risk of acid reflux following myotomy, which can cause damage to the esophagus over time.

During the operation for myotomy, surgeons often perform an additional procedure called a fundoplication in which a portion of the stomach is wrapped around the esophagus to prevent the reflux of stomach contents ([figure 3](#)). However, the fundoplication does not always prevent reflux, and it can cause additional complications such as difficulty swallowing, bloating, flatulence, and diarrhea. (See "[Patient information: Acid reflux \(gastroesophageal reflux disease\) in adults \(Beyond the Basics\)](#)".)

Botulinum toxin injection — Botulinum toxin injections temporarily paralyze the nerves that signal the LES to contract, thereby helping to relieve the obstruction. Botulinum toxin injection also is used occasionally as a diagnostic test for people who appear to have achalasia but who have inconclusive test results.

Procedure — The injection procedure is performed during endoscopy, while the patient is sedated. The botulinum toxin is injected through the lining of the esophagus directly into the LES muscle.

Success rate — A single botulinum toxin injection session relieves symptoms in 65 to 90 percent of people in the short term (three months to approximately one year). Additional injections can relieve symptoms in patients whose symptoms return. Botulinum toxin injection is more likely to be effective in people over the age of 50 years and in people who have the vigorous form of achalasia.

When compared with balloon dilation, botulinum toxin has a similar effectiveness for relieving symptoms in the first one to two years after the procedure; however, prolonged effectiveness requires multiple botulinum toxin injections because the paralyzing effect of the toxin is temporary. The long-term safety and effectiveness of botulinum toxin injection are unknown.

Complications — About 25 percent of people have chest pain for a few hours after the procedure and about 5 percent develop heartburn. Damage to the esophageal wall and lining are rare. The short-term safety of botulinum toxin injection is greater than the short-term safety of both balloon dilation and surgery; this greater short-term safety may make botulinum toxin injection a better choice for patients with other serious medical conditions (eg, advanced age, severe heart or lung problems) who cannot tolerate a balloon dilation or myotomy.

LONG-TERM RISK OF ESOPHAGEAL CANCER — People with achalasia have an increased risk of developing esophageal cancer. Your doctor might recommend endoscopy for early detection of this cancer. (See "[Patient information: Upper endoscopy \(Beyond the Basics\)](#)".)

ACHALASIA FOLLOW-UP — Since none of the treatments for achalasia cure the underlying disease, regular follow-up is needed. The goal is to recognize and treat recurrent symptoms or complications of treatment (eg, acid reflux) early. Recognizing and treating these problems can help to prevent the

development of severe enlargement of the esophagus (mega-esophagus) and cancer, which could require surgical removal of the entire esophagus.

WHERE TO GET MORE INFORMATION — Your healthcare provider is the best source of information for questions and concerns related to your medical problem.

This article will be updated as needed on our web site (www.uptodate.com/patients). Related topics for patients, as well as selected articles written for healthcare professionals, are also available. Some of the most relevant are listed below.

Patient level information — UpToDate offers two types of patient education materials.

The Basics — The Basics patient education pieces answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials.

[Patient information: Achalasia \(The Basics\)](#)

[Patient information: Esophageal cancer \(The Basics\)](#)

Beyond the Basics — Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are best for patients who want in-depth information and are comfortable with some medical jargon.

[Patient information: Upper endoscopy \(Beyond the Basics\)](#)

[Patient information: Acid reflux \(gastroesophageal reflux disease\) in adults \(Beyond the Basics\)](#)

Professional level information — Professional level articles are designed to keep doctors and other health professionals up-to-date on the latest medical findings. These articles are thorough, long, and complex, and they contain multiple references to the research on which they are based. Professional level articles are best for people who are comfortable with a lot of medical terminology and who want to read the same materials their doctors are reading.

[Clinical manifestations and diagnosis of achalasia](#)

[Evaluation of dysphagia in adults](#)

[Overview of the treatment of achalasia](#)

[Pathophysiology and etiology of achalasia](#)

[Pneumatic dilation and botulinum toxin injection for achalasia](#)

The following organizations also provide reliable health information.

- National Library of Medicine

(www.nlm.nih.gov/medlineplus/ency/article/000267.htm, available in Spanish)

- The Society of Surgery of the Alimentary Tract

(www.ssat.com/cgi-bin/achalasia.cgi)

- The Society of Thoracic Surgeons

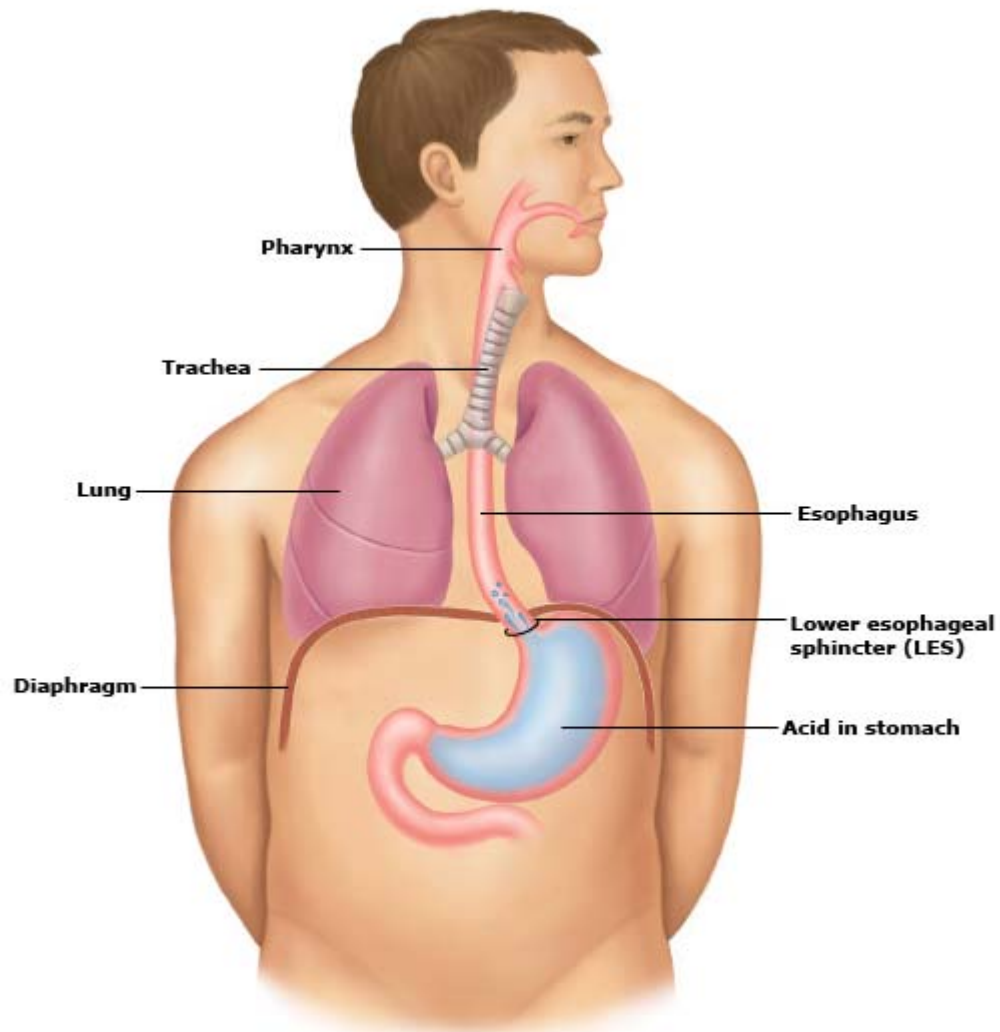
(www.sts.org)

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ACKNOWLEDGMENT — The author and editors would like to thank Dr. Pankaj J Pasricha, who contributed to earlier versions of this topic review.

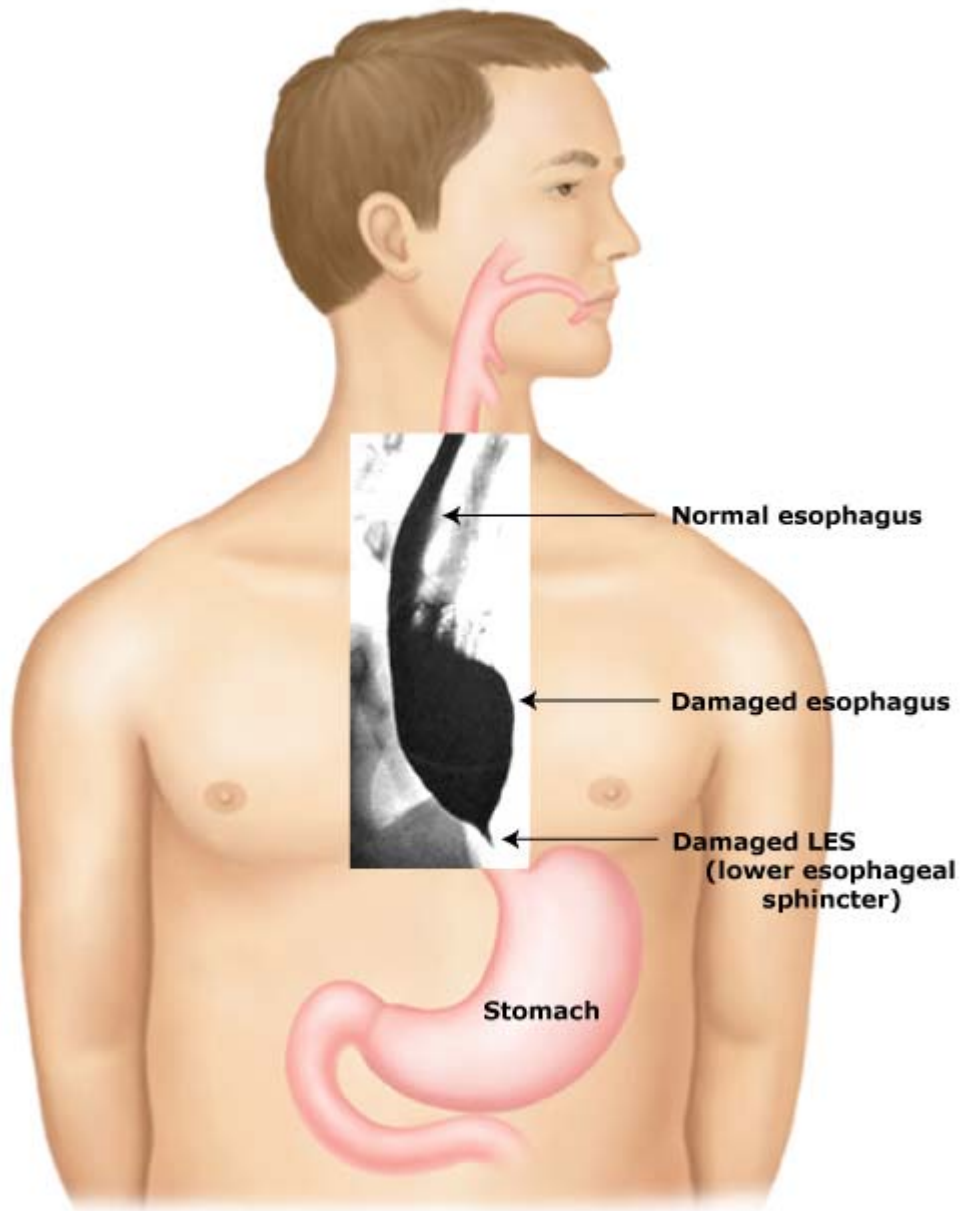
GRAPHICS

Gastroesophageal reflux disease (GERD)



When we eat, food is carried from the mouth through the esophagus, a tube-like structure that is approximately 10 inches long and 1 inch wide in adults. At the lower end of the esophagus, where it joins the stomach, there is a circular ring of muscle that relaxes and opens when food reaches that point, called the lower esophageal sphincter (LES). This allows food to enter the stomach and then closes to prevent the back-up of food and acid into the esophagus. Reflux can occur if the LES is weak or stays relaxed too long.

Barium swallow showing achalasia



This figure shows an actual x-ray of someone with achalasia. You can see that the lower esophagus is enlarged compared to the upper part, and that the lower esophageal sphincter is very narrow. The damaged lower esophageal sphincter makes it hard for food and fluid to pass into the stomach.

Nissen fundoplication



Original artwork by JP Gray, 2007.