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Patient information: Achalasia

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INTRODUCTION — Achalasia is a swallowing disorder caused by two problems in the esophagus (the tube that carries food from the mouth to the stomach). The first problem is that the lower two-thirds of the esophagus do not propel food toward the stomach properly. The second problem is in the lower esophageal sphincter (LES), a circular band of muscle that lies at the junction of the esophagus and the stomach that helps prevent food from regurgitating from the stomach to the esophagus. Normally, the LES relaxes in response to swallowing, allowing food to enter the stomach. In patients with achalasia, the LES fails to relax, creating a barrier for food and liquids to pass into the stomach.

Achalasia affects approximately 1 in every 10,000 to 100,000 people, and most people are diagnosed between the ages of 25 and 60 years. It is typically a chronic condition that initially worsens over time and does not go away on its own.

Several different treatments are available. Each treatment has specific advantages and disadvantages, and the selection among these treatments will depend on several individual factors. It is important to gather information about this condition and to discuss the various treatment options with your doctor before making a decision.

CAUSE OF ACHALASIA — The specific cause of achalasia is unknown. Normally, the LES contracts and relaxes in response to opposing signals from two groups of nerve cells located in the wall of the esophagus. For unknown reasons, in patients with achalasia, an inflammatory reaction targets nerve cells in the esophagus, particularly those that signal the LES to relax. This reaction causes these cells to gradually disappear. The end result is that the LES fails to relax and thereby creates a

blockage for swallowed material to enter the stomach. To make matters worse, nerve cells in the lower two-thirds of the esophagus are also destroyed. These cells are needed to make the esophagus contract and propel food toward the stomach, a process known as peristalsis. Thus, people with achalasia often accumulate large volumes of food and saliva in the esophagus. Food can only enter the stomach when enough pressure builds up in the esophagus to overcome the resistance posed by the non-relaxing LES.

SYMPTOMS OF ACHALASIA — Achalasia usually first occurs in mid-life, although congenital and familial cases have been described. The symptoms have a slow onset and progress gradually. As a result, many people delay seeking medical attention until symptoms are advanced. The major symptom is difficulty swallowing: about 90 percent of affected people have trouble swallowing food and 85 percent of affected people have trouble swallowing liquids. Other symptoms 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.

DIAGNOSIS OF ACHALASIA — Achalasia is usually suspected based upon the presence of the symptoms described above, but specific tests are needed to confirm the diagnosis and to rule out other conditions with similar symptoms, such as gastroesophageal reflux disease, pseudoachalasia (a rare condition in which certain tumors can mimic the features of achalasia), and an infection called Chagas' disease, which is seen almost exclusively in Central and South America.

Radiographic tests — Tests that use some form of x-ray radiation can be very helpful in confirming achalasia.

Chest x-rays — A simple chest x-ray may reveal distortion of the esophagus and absence of air in the stomach, two abnormalities that suggest achalasia.

Barium swallow test — The barium swallow test is the primary screening test for achalasia. The test involves swallowing a thick paste of barium while x-rays are taken. The barium outlines the interior and profile of the esophagus and LES.

Barium swallows are usually performed under fluoroscopy, a continuous low-grade x-ray, which is helpful for studying the motion in the esophagus. In achalasia, this test usually reveals an absence of contractions in the esophagus after swallowing. Sometimes this test reveals useless, spastic contractions of the esophagus in response to swallowing. This form of achalasia is often called vigorous achalasia and probably represents a variation of achalasia.

Manometry — Manometry refers to the measurement of pressure within the esophagus and the LES. These measurements are determined by advancing a pressure sensor into the esophagus. Manometry is always used to confirm 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.

Endoscopy — Endoscopy entails direct visualization of the inside of the esophagus, LES, and stomach using a thin, lighted tube. This test is usually recommended for people with suspected achalasia and is especially useful for detecting other

conditions that mimic achalasia. In people with achalasia, endoscopy often reveals distortion of the esophagus and the presence of residual food; it may also reveal inflammation, small ulcers caused by residual food or pills, and candida (yeast) infection. The endoscope is usually advanced through the LES and into the stomach to check for stomach cancer. Cancer in the upper part of the stomach can produce symptoms almost identical to those of achalasia, and this condition is therefore called pseudoachalasia (meaning "false" achalasia). Thus, biopsies (small samples of tissue) are often obtained in the lower portion of the esophagus.

TREATMENT OF ACHALASIA — Several different options are available for the treatment of achalasia. Most have been used for many years, but one ([botulinum toxin injection](#)) is relatively new. Unfortunately, none of these treatments can halt or reverse the loss of nerve cells in the wall of the esophagus. However, all of these treatments are effective for overcoming the blockage created by the LES and restoring normal movement of solid food and liquid down the esophagus and into the stomach.

Two of these treatments (drug therapy and [botulinum toxin injection](#)) work by pharmacologically reducing the LES pressure while two other treatments (balloon dilatation and surgery [myotomy]) work by mechanically weakening the muscle fibers of the LES. Each of these treatments has various advantages and disadvantages that you should discuss with your doctor before making a decision.

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

Medical therapy is the least invasive option for treating achalasia. However, most people find that long-term medical therapy is inconvenient, often ineffective, and sometimes associated with side effects such as headache and low blood pressure. Furthermore, these drugs tend to lose their effectiveness with repeated use.

Balloon dilatation (pneumatic dilatation) — Balloon dilatation mechanically stretches the contracted LES. This procedure is effective for relieving most symptoms of achalasia in around two-thirds of patients, although chest pain persists in some people. Up to one-half of patients may require more than one session for adequate relief. Balloon dilatation is currently considered as the standard non-surgical treatment for achalasia.

Procedure — People undergoing balloon dilatation are usually told to fast for at least 12 hours before the procedure, and some people may be placed on a liquid diet one or two days in advance. Using fluoroscopy, a doctor advances a guidewire down the esophagus and positions it inside the LES. A deflated balloon is then advanced along this guidewire, positioned inside the LES, and inflated for about 60 seconds. The balloon is then deflated and withdrawn, and the person is monitored in a recovery area for five to six hours to detect any complications of the procedure. If there are no complications, people can usually resume eating after six hours.

In most cases, only one dilatation is performed during each session; if a person's day-to-day symptoms do not improve, additional sessions can be performed. The balloons are available in three sizes; the smallest size is usually used during the first

session, and larger sizes are often used during subsequent sessions. If a person's symptoms are still present after three sessions, doctors usually recommend surgery.

Success rate — A single balloon dilatation session continues to relieve symptoms of achalasia in about 59 percent of people one year after the procedure and in about 26 percent of people five years after the procedure. However, the five-year success of this procedure may be improving with the use of newer balloon dilators. The likelihood of symptom relief at one year increases to about 89 percent if a person undergoes three dilatation sessions. The success rate at later time points has not been well studied, but some people have remained symptom-free for as long as 25 years. Prolonged relief of symptoms is more likely in people over the age of 40 years.

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

The most significant complication of balloon dilatation is creation of a tear (perforation) in the wall of the esophagus; this complication occurs in about 2 to 6 percent of people undergoing the procedure, and it is most likely to occur during the first dilatation session. Symptoms of persistent pain or discomfort in the hours after the procedure may signal that the esophagus has been torn. Some doctors routinely check x-ray tests immediately after the procedure to check for any damage of the esophagus.

Most tears are small, and some may heal on their own with antibiotics and intravenous feeding. However, many doctors routinely recommend surgery to repair these tears, regardless of their size. The risk of dying from balloon dilatation is approximately 0.2 percent. There is no way to predict perforation; however, it is sensible to choose a doctor who has a great deal of experience dilating patients with achalasia.

Other possible complications of balloon dilatation include bruising of the esophageal wall, damage of the esophageal lining, the development of small pockets (diverticula) off of the esophagus or upper stomach, and the development of gastroesophageal reflux disease (GERD). Because the LES is the principal barrier preventing the reflux of stomach contents into the esophagus, its disruption can predispose to acid reflux. GERD occurs in about 2 percent of people after balloon dilatation, but is usually easily controlled with acid-reducing medications.

Surgery (Myotomy) — Surgery can be used to directly sever the muscle fibers of the contracted LES. The surgical technique used most often is called the Heller myotomy. In the past, surgery was performed through an open incision in the chest or abdomen, but it can now be performed through a tiny incision using a thin, lighted tube (a laparoscope or a thoracoscope). This new approach is less traumatic and shortens recovery time; preliminary studies suggest that it is just as effective as open surgery for relieving symptoms of achalasia.

Success rate — Surgery relieves symptoms of achalasia in 70 to 90 percent of people. This 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. Studies suggest that surgery is a more permanent solution for achalasia than balloon dilatation or [botulinum toxin injection](#), although surgery is initially more expensive.

Complications — Successful surgery implies the virtual elimination of the LES. Therefore, there is an increased risk of gastroesophageal reflux disease, especially over time. It is vital to detect and treat this as patients with achalasia can develop severe scarring with reflux disease that can be very difficult to treat once established. This requires that patients be regularly monitored for this complication, and may require the use of acid suppressing medications. The risk of dying from this surgery is approximately 0.3 percent.

Botulinum toxin injection — [Botulinum toxin injection](#) is the newest treatment for achalasia; this procedure is still considered experimental and is often only available in academic medical centers. The botulinum toxin poisons the nerve cells that signal the LES to contract. Sometimes, botulinum toxin injection is used as a diagnostic test in people with suspected achalasia who have inconclusive test results. Because the amounts of botulinum toxin used are very small, there is virtually no risk of botulism from this procedure.

Procedure — The injection procedure is performed during routine endoscopy. The botulinum toxin is injected directly into the LES.

Success rate — A single [botulinum toxin injection](#) session alleviates symptoms in 65 to 90 percent of people in the short term (three months to approximately one year). In about half of the people who have a return of symptoms, additional injections effectively relieve symptoms. Studies suggest that 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 dilatation, botulinum toxin has a similar effectiveness for relieving symptoms in the first one to two years after the procedure. However, this prolonged effectiveness requires multiple botulinum toxin injections in 40 to 50 percent of people. Despite equivalent symptom relief, balloon dilatation decreases LES pressure and decreases food retention in the esophagus more effectively than [botulinum toxin injection](#). The long-term effectiveness of botulinum toxin injection is unknown.

Complications — About 25 percent of people experience transient (a few hours) chest pain and 5 percent of people experience heartburn immediately after [botulinum toxin injection](#). Damage of the esophageal wall and lining are rare. The short-term safety of botulinum toxin injection appears to be greater than the short-term safety of both balloon dilatation and surgery; this greater short-term safety may make botulinum toxin injection a better choice for people who must avoid more invasive procedures because of the presence of other medical conditions. The long-term safety of botulinum toxin injection is unknown.

LONG-TERM RISK OF ESOPHAGEAL CANCER — People with achalasia have an increased risk of esophageal cancer, particularly if obstruction is not adequately relieved. This risk may be as high as 16 times the risk of the general population, but the absolute risk is very low (ie, only a very small percentage of patients with achalasia will develop it). Nevertheless, some doctors recommend regular endoscopic screening for the early detection of this cancer.

WHERE TO GET MORE INFORMATION — Your doctor is the best resource for finding out important information related to your particular case. Not all patients with

achalasia are alike, and it is important that your situation is evaluated by someone who knows you as a whole person.

This discussion will be updated as needed every four months on our web site (www.uptodate.com). Additional topics as well as selected discussions written for health care professionals are also available for those who would like more detailed information.

A number of other sites on the Internet have information about achalasia. Information provided by the National Institutes of Health, national medical societies, and some other well-established organizations are often reliable sources of information, although the frequency with which their information is updated is variable.

- National Library of Medicine

(<http://www.nlm.nih.gov/medlineplus>)

- National Institute of Diabetes and Digestive and Kidney Diseases

(<http://www.niddk.nih.gov>)

- The Society of Surgery of the Alimentary Tract

(<http://www.ssat.com/>)

- The Society of Thoracic Surgeons

(<http://www.sts.org/>)

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REFERENCES

- [1.](#) West, RL, Hirsch, DP, Bartelsman, JF, et al. Long term results of pneumatic dilation in achalasia followed for more than 5 years. *Am J Gastroenterol* 2002; 97:1346.
- [2.](#) Spiess, AE, Kahrilas, PJ. Treating achalasia: From whalebone to laparoscope. *JAMA* 1998; 280:638.
- [3.](#) Eckardt, VF, Kanzler, G, Westermeier, T. Complications and their impact after pneumatic dilatation for achalasia: Prospective long-term follow-up study. *Gastrointest Endosc* 1997; 45:349.
- [4.](#) Pasricha, PJ, Ravich, WJ, Hendrix, T, et al. Intrasphincteric botulinum toxin for the treatment of achalasia. *N Engl J Med* 1995; 332:774.