ACHALASIA

INTRODUCTION

Achalasia is a rare swallowing disorder that affects only 1 in every 100,000 people. Patients typically sense increasing difficulty swallowing solid food and also liquids. Most people are diagnosed between the ages of 25 and 60 years. It is usually a chronic condition that worsens over time and does not resolve.

WHAT CAUSES ACHALASIA?

The specific cause of achalasia is unknown. However, patients with achalasia have two problems in the esophagus (the tube which carries food from the mouth to the stomach). The first is that the esophagus stops working, it essentially becomes a tube without muscular contractions that would normally propel food and liquids. The second problem is in the lower esophageal sphincter (LES), the valve between the esophagus into the stomach that normally helps prevent food from flowing backwards, from the stomach into the esophagus. The LES should relax in response to swallowing to allow food to enter the stomach. In patients with achalasia, the LES fails to relax, creating a barrier that prevents food and liquids from passing into the stomach. These changes cause food, liquid and saliva to accumulate in the esophagus.
SYMPTOMS OF ACHALASIA

The major symptom is difficulty swallowing (liquids or solids). 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. These symptoms often are slowly progressive and many people do not seek medical attention until symptoms are advanced.

MAKING THE DIAGNOSIS OF ACHALASIA

Achalasia is usually suspected based upon the presence of the symptoms described above, but tests are needed to confirm the diagnosis. In addition, it is important 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.

X-rays Testing — A barium test involves swallowing a chalky-tasting mixture of barium while x-rays are taken of the esophagus. The barium shows the shape and contour of the esophagus and sphincter. In Achalasia, it reveals an absence of contractions in the esophagus after swallowing and narrowing with hang up of barium at the lower esophageal sphincter.

Manometry— Manometry refers to the measurement of pressure within the esophagus and the LES. Pressures are measured by advancing a thin tube through the nose into the esophagus while patients are awake. Patients will be asked to swallow sips of water while the tube is in place while the equipment measures the esophageal contractions. 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 allows for direct visualization of the inside of the esophagus, LES, and stomach using a thin, lighted, flexible tube. Endoscopy is done while a patient is sedated. In people with achalasia, endoscopy often reveals that the esophagus appears dilated and there may be residual food in it.; it may also reveal inflammation, small ulcers caused by residual food or pills, and candida (yeast) infection. It is important to evaluate for other problems at that time such as reflux esophagitis and cancer.

**TREATMENT CHOICES FOR ACHALASIA**

There is no cure or definitive treatment for achalasia. Since the primary problem is poor emptying of the esophagus, available treatments reduce symptoms by improving emptying by opening up the spastic lower esophageal sphincter either mechanically/physically (pneumatic balloon dilation and surgery) or chemically/medically (Botox). Most studies have shown essentially equal effectiveness of pneumatic dilation and surgery in at least 75% of patients. Botox provides temporary relief.

**Balloon dilatation (pneumatic balloon dilatation)**

Balloon dilatation mechanically/physically stretches out the spastic lower esophageal sphincter muscles to then improve emptying from the esophagus. It is performed during endoscopy, while sedated and also using fluoroscopic (X-ray) guidance to visualize and monitor the treatment. A balloon (3cm or larger) is positioned within the LES and inflated to fracture/open up the sphincter. After the patients wakes up, an X-ray of the esophagus is performed to assess and assure there are no leaks or tears.

The most significant complication of balloon dilatation 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, and it is most likely to occur during the first dilatation session. Symptoms of persistent or worsening pain in the hours
after the procedure may indicate a perforation. The development of gastroesophageal reflux disease (GERD) can occur after dilation because the LES is the principal barrier which prevents the reflux of stomach contents into the esophagus, its disruption can lead 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)

Myotomy can be used to directly cut the muscle fibers of the LES. The surgical technique used most often is called the Heller myotomy and is usually done laparoscopically through a tiny incisions through which surgery is performed using a thin lighted tubes inserted internally.

Like balloon dilatation, there is a risk of reflux following myotomy. Surgeons generally perform a fundoplication (wrapping a portion of the stomach around the esophagus to prevent regurgitation of stomach contents) at the time of surgery; however this does not always prevent reflux. Patients should be regularly monitored for this complication, and may require acid suppressing medications.

Botulinum toxin (Botox) injection

Botulinum toxin temporarily paralyzes the nerve cells that signal the LES to contract thereby relaxing the spastic muscles and helping to relieve the obstruction. The injection procedure is performed during routine endoscopy, while patients are sedated. The botulinum toxin is injected directly into the LES.

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). Since the effect of Botox wears off over time, additional periodic injections will be needed to control symptoms. This treatment may or may not halt progressive esophageal damage and dilation of it.