



Achalasia

Patient and family information, brought to you by the Education Committee of APSA

Overview - “What is it?”

Achalasia is an abnormality of the esophagus that may explain problems in swallowing. The esophagus is the tube that connects the mouth to the stomach and is made up of muscle layers. The region between the esophagus and the stomach is defined by a slight tightness in the muscle called the lower esophageal sphincter. In achalasia, there are two problems: (1) the muscles of the esophagus that move food from the mouth to the stomach do not work, and (2) the lower esophageal sphincter (LES) does not relax completely. The combination of non-working esophageal muscles and a tight LES cause the food to get stuck in the esophagus.

Most cases of achalasia have no defined cause (idiopathic).

Achalasia occurs in 1-2 people in 200,000. It is most common in adults mostly between ages 30 to 50. About 10% of all patients with achalasia are children and teenagers

Signs and Symptoms - “What symptoms will my child have?”

Early signs: Patients with achalasia may present with coughing at night. When lying flat, food and spit may collect in the esophagus. Food and spit may spill in the airways and lungs. Symptoms of achalasia include problems swallowing, vomiting undigested food and chest pain behind the sternum. Because undigested food remains in the esophagus for a long time, bad breath and foul-smelling burps can be observed.

Later signs/symptoms: If the child has continued vomiting, he or she may exhibit weight loss.

Conditions that mimic this condition:

- Because patients present with vomiting and aspiration, patients can be thought to have gastroesophageal reflux.
- In South America, a parasitic disease called Chagas’ disease causes injury to the nerves of the esophagus and therefore cause the similar symptoms.

- Diffuse esophageal spasm is even less common than achalasia. Symptoms of this disorder are similar to symptoms seen with achalasia. Patients present with a long-standing history of swallowing problems, regurgitation of undigested food, bad breath and weight loss.

Diagnosis - “What tests are done to find out what my child has?”

Achalasia is usually diagnosed with a combination of an esophagram and measurements of esophageal manometry and motility.

- An esophagram is a test where the patient drinks barium and X-rays are taken during the swallowing process (See Figure 1). Findings of this study show a large, non-moving esophagus with a closed off (beaked) esophagus (See Figure 2).



Figure 1: Chest x-ray shows an air-fluid level in the middle of the chest (arrow) in a patient with achalasia. (Image credits: R. Ignacio)



Figure 2: Contrast study with a “bird’s beak” deformity (arrow) in a patient with achalasia. (Image credit: R. Ignacio)

- Esophageal manometry measures the pressure within the LES and esophageal motility measures movement of the esophagus. A physician places a small tube into the esophagus to measure these characteristics. In achalasia, the LES pressure is high and there is no effective movement of the esophagus that pushes food from the stomach. These two main problems cause food to stay in the esophagus instead of going in to the stomach.
- Endoscopy is when a flexible telescope is placed in the mouth and is gently pushed through the esophagus, stomach and part of the small intestine. The doctor can look at evidence of inflammation directly. This study is used to make sure that there are no other reasons for the child’s symptoms.

Treatment - “What will be done to make my child better?”

Medicine: Medications that decrease the pressure of the LES have unpredictable effects on symptoms.

Endoscopic dilation: The LES can be opened forcefully by inflating a balloon within the esophagus during endoscopy. This maneuver stretches the circular fibers of the LES. It can achieve some resolution of symptoms over weeks or months. Dilation has about a 2-6% risk of perforation.

Endoscopic injection of botulinum toxin (Botox): Botox is a substance that has been used in several conditions to achieve muscle relaxation (most commonly to decrease the appearance of facial wrinkles). Botox can be injected into the esophageal sphincter muscles in achalasia to effect relaxation of the muscles. The effect lasts about three months.

Surgery: Surgical treatment of achalasia is the only modality that results in lasting results. There are different approaches to surgery:

- Esophageal myotomy (also called the Heller myotomy – named after the surgeon who first described the procedure). The surgeon creates an incision through the LES effectively releasing the circle of the sphincter and loosens up this tight region. The opening in this region allows food to pass from the esophagus to the stomach, but it also increases the likelihood of having food pass from the stomach to the esophagus (gastroesophageal reflux). In order to avoid reflux of food and acid to go from the stomach to the esophagus, surgeons create a partial fundoplication, where part of the stomach is wrapped around the lower esophagus. At the time of the operation, some surgeons may require having a specialist (gastroenterologist) put a telescope in the esophagus to make sure that the surgery is successful. The operation can be done with a large vertical cut through the middle of the upper abdomen from the bottom of the breastbone to the belly button.
- Another way of doing the surgery is through laparoscopy, where small incisions that allow a scope and instruments to be inserted into the belly. This second approach results in a faster recovery and smaller scars. It is important to know that the surgery opens up LES, but does not do anything to correct the abnormal movement of the esophagus. There is no surgery to help the movement of the esophagus.
- Another surgical option for treatment of achalasia is a technique called POEM which stands for Peroral (through the mouth) Endoscopic Myotomy. This is a form of a newer, even less invasive, type of surgery called natural orifice surgery, where a procedure on an internal organ is done through one of the body’s orifices, such as the mouth. In this procedure, a surgeon or gastroenterologist (often the two working together) introduces a special camera through the mouth. Under camera vision and using special instruments, the lining of the esophagus (called the mucosa) is opened and the inner muscle layer of the esophagus is cut. The mucosa is then closed with special staples.

There has been increasing experience with this operation in adults but only a few cases have been performed in children. If considering this option, it is very important to have your child treated at a center with experience in this new technique.

Preoperative preparation: Patients are usually asked to shower or bathe on the night before surgery. Patients are asked to stop eating or drinking for a few hours before surgery.

Informed consent: A consent form is a legal document that states the tests, treatments or procedures that your child may need and the doctor or practitioner that will perform them. Before surgery, your doctor should tell you what the operation is, the goal of the surgery and other possible treatment options that are available. Your doctor should explain the risks and benefits of the surgery. You give your permission when you sign the consent form. You can have someone sign this form for you if you are not able to sign it. You have the right to understand your child's medical care in words you know. Before you sign the consent form, make sure all of your questions are answered. It is important to know that during surgery, there are things that can happen that your doctor may have not predicted before going in. He or she will explain these to you after the surgery.

Emotional support: Stay with your child for comfort and support as often as possible while he or she is in the hospital. Ask another family member or someone close to the family to stay with your child when you cannot be there. Bring items from home that will comfort your child, such as a favorite blanket or toy.

Postoperative care: After surgery, some surgeons may require the patient to have an X-ray where the patient swallows contrast to make sure that there are no small leaks in the esophagus. In addition, this study may show that contrast travels from the esophagus to the stomach, showing that the surgery was successful. The surgeon may want a nasogastric (NG) tube inserted through your child's nose and down into his stomach. This tube keeps air and fluid out of the stomach during surgery and immediately after

Risks/Benefits: In this procedure, the surgeon makes an incision through the muscle layer only of the LES and not the inside lining of the esophagus. In about 2% of cases, the internal layer of the esophagus is opened (perforation). It is usually recognized at the time of the operation and repaired. If this were to happen, the surgeon may delay feeding the patient for some time to allow the repair to heal.

Home Care - "What do I need to do once my child goes home?"

Diet: Most patients are instructed to eat a soft diet for several days after the surgery. When they are advanced to a general diet, they are to continue eat with small bites and chew thoroughly.

Activity: If the patient had the procedure with small incisions (laparoscopic), he or she can be back to normal activity in 1-2 weeks. If the surgery is done through a big incision, then he or she can be back to normal activities in six weeks, with a weight restriction of 10 pounds up until that time.

Wound care: The patient can shower in three days but may want to wait 5-7 days after surgery before soaking the wound.

Medicines: Medication for pain such as acetaminophen (Tylenol) or ibuprofen (Motrin or Advil) or something stronger like a narcotic may be needed to help with pain for a few days after surgery. Sometimes, if the patient has heartburn symptoms, medications to decrease the acidity of the stomach (antihistamine blockers such as ranitidine and proton pump inhibitors such as omeprazole) may help. Stool softeners and laxatives are needed to help regular stooling after surgery, especially if narcotics are still needed for pain.

What to call the doctor for: After discharge from surgery, problems that may indicate infection such as fevers, wound redness and discharge should be addressed. If there is a lot vomiting, chest pain or food getting stuck in the esophagus, the surgeon should be contacted.

Follow-up care: The patient should be seen by a surgeon at least once to check the surgical wound. The patient's gastroenterologist may require multiple visits for months to years to make sure that the patient remains symptom free.

Long Term Outcomes - "Are there future conditions to worry about?"

After surgery, patients may have bad heartburn symptoms. Heartburn usually means that there is acid that is going up the esophagus. When acid from the stomach contacts the esophagus for a long time, the esophagus is harmed. The esophagus can get narrow, inflamed or even have a higher risk of cancer. Patients with achalasia have a higher chance of getting esophageal cancer compared to the general population. It is important that the esophagus is examined by endoscopy to make sure that inflammation or cancer is not happening. However, there are no guidelines as to how often or for how long these endoscopies should be performed.

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