Achalasia
What it is and how it is treated

Achalasia is a rare disorder that keeps food and liquid from easily entering the stomach. This handout describes symptoms and treatments of achalasia.

Contents

What is achalasia? ............................................................... 2
What causes achalasia? ....................................................... 2
What are the symptoms of achalasia? .................................. 3
What can I do to ease the symptoms of achalasia? ............... 3
How is achalasia diagnosed?................................................ 3
  Barium Swallow Study, Esophageal Manometry,
  Flexible Upper Endoscopy
What are the complications from having achalasia? ............ 5
How is achalasia treated? .................................................... 6
  Medicines, Botox Injection, Endoscopic Dilation, Surgery
What happens during a myotomy? ...................................... 8
Acid Reflux ........................................................................ 8
How to Prepare for Surgery ................................................. 9
Before Your Surgery ........................................................... 9
  Medicines, Liquid Diet, Shaving, Reminder Call, Fasting
Day of Surgery ................................................................. 10
Arriving at the Hospital ..................................................... 10
After Surgery ..................................................................... 11
  In the Recovery Room, In Your Hospital Room
Going Home....................................................................... 12
  Diet and Nutrition, Medicines, Incision Care, Showering,
  Activity, Sexual Activity, Driving
When to Call Your Surgeon................................................ 13
Follow-up Care ................................................................. 14
  Short-term Follow-up Care, Long-term Follow-up Care
What is achalasia?

In normal digestion, swallowed food goes down the *esophagus* (the tube that goes from the throat to the stomach) and into the stomach. The food is moved by *peristalsis*, a series of wave-like muscle contractions.

A muscle called the *lower esophageal sphincter (LES)* is at the bottom of the esophagus. The LES acts as a valve between the esophagus and stomach. When food reaches the stomach, the LES relaxes (opens) to allow the food to enter.

In a rare disorder called *achalasia*, 2 things do not work properly. They are:

- Peristalsis in the esophagus
- Relaxation of the LES

In someone with achalasia, food and liquid collect in the esophagus until their mass creates enough pressure to push through the LES. Achalasia tends to get worse over time.

Achalasia is very rare. Only 10 to 20 people out of 1 million have the condition. It affects about 3,000 people in the United States.

What causes achalasia?

We do not know the exact cause of achalasia. We know that it is linked to a lack of certain nerve cells called *ganglion*, inside the muscles, that work during peristalsis. This lack of cells may also cause the esophagus to become narrower closer to the stomach.
In people with achalasia, nerve cells produce less nitric oxide, a substance that helps the LES relax. Achalasia sometimes occurs as a symptom of another condition, such as infection or damage to the nerves, or cancer.

Achalasia is most common in middle-aged and older adults. The average age of people with achalasia is 49. But, achalasia can occur at any age. It occurs equally among men and women of different races.

What are the symptoms of achalasia?

The main symptom of achalasia is difficulty or pain with swallowing. This is called dysphagia. This often means a person eats less, which can cause weight loss and malnutrition.

Other symptoms of achalasia are:
- Regurgitation of food (food backs up into the esophagus or mouth from the stomach)
- Chest pain
- Heartburn
- Aspiration of food into the lungs, when food “goes down the wrong pipe” and enters the lungs when breathing (occurs in advanced stages of achalasia)

What can I do to ease the symptoms of achalasia?

- Eat slowly.
- Lessen stress, which can make achalasia worse.
- To help ease chest pain or spasms, try these tips:
  - Drink warm or room-temperature water or seltzer water
  - Chew crackers, bread, ice, or hard candy
  - Gulp milk
  - Drink warm milk
  - Take antacid medicine (such as Tums, Mylanta, Maalox, and Gaviscon)

How is achalasia diagnosed?

To diagnose achalasia, your health care provider will do a full health exam. This will include a detailed review of your medical history and these tests:
- Barium swallow study
- Esophageal manometry
- Flexible upper endoscopy of your esophagus and stomach
Barium Swallow Study

A barium swallow study is often used to diagnose achalasia. In this study, you will swallow a contrast solution that contains barium. This liquid will look white on X-ray images of your esophagus (see “Barium X-ray image” in the drawing below).

If you have achalasia, the X-ray images will likely show that your esophagus gets very narrow where it enters the stomach. This narrowing makes your esophagus look like a bird’s beak.

The barium liquid helps your doctor see the inside of your esophagus.

Esophageal Manometry

A manometry test is an important step in diagnosing achalasia. A device will be inserted through your nose or mouth. The device measures the peristaltic waves (contractions) at different points inside your esophagus.

Flexible Upper Endoscopy

In a flexible upper endoscopy, a specialist will check your esophagus and stomach with a thin, flexible tube called an endoscope. This tube will be directed down your throat.

The endoscope has a light and a tiny camera on one end. The camera will send pictures of the inside of your esophagus to a monitor for your doctor to see.
During an endoscopy, a thin, flexible tube called an endoscope is put down your throat.

An endoscopy is needed to make sure that a tumor in the esophagus is not causing your achalasia symptoms. This condition, called pseudoachalasia (“false” achalasia) would require different treatment.

People with pseudoachalasia most often:
- Have had symptoms less than 6 months
- Have lost more than 15 pounds
- Are older than 55

**What are the complications from having achalasia?**

Complications of achalasia include:
- Weight loss
- Malnutrition

A person with advanced achalasia can inhale food into their lungs when they breathe. This can cause:
- Lung infection
- Pneumonia
- Lung abscesses (inflamed, pus-filled areas)

Achalasia also is linked with a higher risk of cancer of the esophagus.
**How is achalasia treated?**

The goal of treatment is to relax the LES. You may be treated with:

- Medicine
- Botox (*botulinum*) injection of the LES
- *Endoscopic dilation* (stretching) of the LES
- Surgery

Of these treatments, surgery provides the best chance for long-term relief of symptoms. The surgery can be done in a *minimally invasive* way for almost all people (see page 8).

**Medicines**

These medicines are most often used to treat achalasia:

- Calcium-channel blockers
- Nitrates
- Phosphodiesterase inhibitors

These medicines decrease blood flow to the LES. This relaxes the LES. But, these medicines do not work well for all people, and we do not know how well they will keep working over a long period.

If you have severe achalasia symptoms and you are waiting to have surgery, your doctor may prescribe medicines to relieve your symptoms while you wait.

**Botox Injection**

*Botulinum toxin*, or Botox, paralyzes the nerve cells that tell the LES valve to contract. Botox injections give short-term relief (6 months to 1 year) for about 70% of patients (70 out of 100). If you choose to get a Botox injection, you will likely need more injections for long-term relief.

Before your injection, you will receive medicine to make you sleepy and relaxed. This is called *conscious sedation*. You will be awake, but you will not feel pain. The procedure will be done using an endoscope (see drawing on page 5).

Botox injections can increase the rate of complications in patients who have *myotomy* surgery (see pages 7 and 8). Botox injections to treat achalasia should not be given to people who will be having a myotomy. These injections are usually given to people who are too ill for anesthesia or who are expected to live less than 3 years.
Endoscopic Dilation

In endoscopic dilation, a balloon catheter (a flexible tube with a balloon on one end) is used to weaken the muscles that make the LES contract. In this procedure, your doctor uses an endoscope and a guide wire to direct a catheter into your esophagus. The catheter is placed inside the LES and then the balloon is inflated for up to 1 minute.

This procedure is usually done in the clinic using conscious sedation. It is successful for about 70% of patients (70 out of 100) 1 year after dilation and for about 50% of patients (50 out of 100) 5 years after dilation. We do not know much about longer-term results.

A rare complication of endoscopic dilation is a tear in the wall of the esophagus. This happens in less than 5% of people (less than 5 out of 100) who have this procedure. This tear may heal on its own, but surgery may be needed to repair it.

Surgery

Many people with achalasia have surgery to open their LES. This surgery is called a myotomy. During surgery, the muscle around the LES is cut. The cut is about 2 to 2.5 cm (¾ to 1 inch) long.

Cutting this muscle helps the LES relax. This helps food move more easily from the esophagus to the stomach.

Myotomy eases achalasia symptoms in about 95% of patients (95 out of 100). Of all the treatments for achalasia, a myotomy gives the longest lasting relief of achalasia symptoms.

The surgery also has a risk of complications, such as tearing or leaks in the esophagus. It is important to find an esophageal surgical specialist to do your procedure.

What happens during a myotomy?

You will have general anesthesia for your myotomy. This means you will be asleep during the surgery.
Myotomy is done using laparoscopy, a method that lets your surgeon work through very small incisions. Instead of doing open surgery through 1 large incision in your abdomen, about 5 small incisions will be made (see drawings below). Your surgeon will insert tiny instruments and a tiny camera through these incisions.

With laparoscopy, you will have a shorter hospital stay and less pain than with open surgery. Laparoscopy is known as minimally invasive surgery.

Acid Reflux

One of the side effects of myotomy surgery is acid reflux. Acid reflux is when normal stomach acids flow up into the esophagus past the LES. These acids can irritate the esophagus. Acid reflux may occur after a myotomy because the LES is opened up during surgery.

To reduce acid reflux, your surgeon may also do a partial fundoplication procedure after doing your myotomy. In this procedure, your surgeon will wrap the top part of your stomach partway around the base of your esophagus. This will create a 1-way valve to help keep stomach acid from entering your esophagus.
How to Prepare for Surgery

- **Exercise and eat well in the weeks before surgery to be in the best possible health.** As with any surgery, a myotomy puts stress on your heart and lungs (your *cardiopulmonary* system). This is why it is so important that you are in good physical shape before surgery.

- **Increase your protein intake with nutritional shakes and supplements.** Good nutrition will help you recover after surgery. This can be a challenge for people who have been losing weight and muscle mass as result of achalasia. If you are not able to eat enough on your own, your doctor may advise you to get nutrition through a feeding tube.

**Before Your Surgery**

- **Medicines:** For 1 week before your surgery, do **NOT** take aspirin, ibuprofen (Advil, Motrin) or naproxen (Aleve, Naprosyn) **unless** you are taking it for a specific health condition. If you are unsure whether you should stop taking it, please call the clinic.

- **Liquid Diet:** For 3 days before surgery, do **NOT** eat any solid foods. One of the risks when you are under anesthesia is *aspiration* (inhaling food into your lungs). This may occur if there is food left in your esophagus when you receive general anesthesia for your surgery. When anesthesia is given, this food may travel back to the area at the back of your mouth and enter your airways. If this food reaches your lungs, it can cause pneumonia and even lung injury.

- **Shaving:** For 2 days before your surgery, do **not** shave any part of your body that you do not usually shave every day. If you usually shave near your surgical site, **stop shaving that area 2 days before your surgery.**

- **Reminder call:** A staff member from the Pre-Anesthesia Clinic will call you between 2 and 5 p.m. the day before your surgery. If your surgery is on a Monday, you will receive this call Friday afternoon.

  The Pre-Anesthesia staff will remind you:
  - What time to arrive at the hospital
  - Where to check in
  - What your pre-surgery instructions are, including what medicines to take and not take the day of surgery

- **Fasting:** You will already be on liquid diet before surgery. Do **not drink** anything after midnight the night before your surgery. The Pre-Anesthesia nurse who calls you may change this instruction based on what time your surgery will start.
• **Pre-surgery showers:** Both the night before and the morning of surgery, you should shower or bathe using Chlorhexidine Gluconate soap.
  - Do **not** use this special soap on your face or hair. Use your regular soap and shampoo for these areas.
  - Wash thoroughly from your neck down, especially around the area of your surgery.

### Day of Surgery

- **Do not** put on makeup, deodorant, lotions, hair products, or fragrances.
- **Do not** wear contact lenses. Wear your glasses instead.
- **Remove** all jewelry.
- Wear loose clothing that will be easy to take off and comfortable to wear home.

**Arrive early.** Please leave home early and plan to arrive ahead of your scheduled check-in time. Allow for traffic and the chance that operations scheduled before yours may end early.

- Bring these items with you on the day of surgery:
  - **List of your current medicines,** including their exact doses and when you last took them
  - **Photo ID**
  - Your **insurance** and **pharmacy cards**
  - **Co-payments** for discharge medicines

- If you have these items, please also bring:
  - A copy of your **advance health care directive** and/or **durable power of attorney for health care.** They will be placed in your medical record.
  - Your **CPAP machine,** if you use one for sleep apnea.

### Arriving at the Hospital

When you arrive at the hospital for your surgery, you will:

- Check in and sign admission forms.
- Be asked your name, date of birth, and what procedure you will be having. It is normal for many staff members who are caring for you to ask you these same questions. This is for your own safety.
• Be covered with a heating blanket to keep your body warm. This will help reduce your risk of infection.

• Have an intravenous (IV) line placed.

• Receive an injection in your abdomen to help prevent blood clots.

Once these steps are done and your operating room is ready, your anesthesia provider will take you back to the operating room.

Your surgery will take about 2 to 3 hours.

**After Surgery**

**In the Recovery Room**

• You will spend about 2 hours in the recovery room waking up after surgery.

• Nurses in the recovery room will monitor your pain level and give you medicine to make you comfortable.

• Your family may be able to visit you in the recovery room. This depends on how you are doing and the care of other patients in the recovery room.

When you wake up, you will have:

• An oxygen mask over your face to give you extra oxygen. You will be switched to nasal prongs (oxygen into your nose) when your lungs are ready.

• Inflatable stockings called sequential compression devices (SCDs) on your legs. These stockings squeeze your legs off and on to improve blood flow. This helps keep blood clots from forming.

• A urinary catheter in your bladder. This lets us monitor your urine output during and after your surgery. The catheter will be removed early the next morning.

**In Your Hospital Room**

• **Medicines:** All your medicines will be crushed or in liquid form.

• **Exercise your lungs:** Your nurse will show you how to use an incentive spirometer to help you exercise your lungs. It is important to exercise your lungs to prevent complications such as pneumonia.

• **Activity:** It is important for you to get up and try to walk, even in the evening after your surgery. Your nurse will help you the first few times to make sure you are steady on your feet. Please ask your nurse to help you walk. Do not try to walk on your own at first.
• **Diet:** After your surgery, you will be started on clear liquids and advanced to an *esophageal diet*. A dietitian will visit you the day after your surgery to talk about your diet when you leave the hospital.

• **Family and friends:** Family and friends can be important to your recovery. They can do things to help make you more comfortable, such as fluff your pillow, get you a glass of water, or find your remote control.

### Going Home

You will be in the hospital overnight. We recommend that patients who live more than a 2-hour drive from the hospital stay in the Seattle area 1 to 2 nights after their surgery. This rest time will help your recovery. You will also be close by in case any problems develop.

### Diet and Nutrition

Right after surgery, you will be started on a clear liquid diet. Your diet will be advanced to a full liquid diet by the time you leave the hospital. You will continue to slowly advance your diet at home.

This diet will help keep food from getting stuck in the area where the surgery was done. If you have any questions about your diet, read the handout “Esophageal Diet After Surgery” that your dietitian gave you before you left the hospital. Or, call your dietitian or your surgery team.

### Medicines

**You cannot swallow whole pills for 4 weeks after your surgery.** You will go home with liquid medicines or pills that can be crushed. This includes your pain medicine and anti-nausea medicine.

### Incision Care

- You may remove your outer bandage in 48 hours.

- Leave the white tapes (Steri-Strips) in place. You may remove them in 1 week, or they may fall off on their own in about 1 week.

### Showering

- You may shower the day after surgery. The dressings on your incision will repel the water.

- **Do not** take a bath, sit in a hot tub, or go swimming for 4 weeks after your surgery.

### Physical Activity

- Do not lift anything that weighs more than 10 pounds for 6 weeks after your operation. A gallon of milk weighs 8 pounds.
• Avoid doing activities that make you contract your abdominal muscles.

• It is important to walk. You should walk 3 to 4 times every day. Slowly increase how far you go.

**Sexual Activity**

• You may resume sexual activity 2 weeks after your surgery.

• Once you resume it, continue to follow the other activity restrictions listed above in “Physical Activity.”

**Driving**

• Do **not** drive for at least 2 weeks after your surgery.

• When you start driving again, you should be off all of your pain medicine. You should be able to move easily and quickly apply brakes if needed.

**When to Call Your Surgeon**

Call your surgeon if you have:

• A fever higher than 100.5°F (38°C)

• Shaking or chills

• A hard time getting food or liquids down

• Nausea or vomiting that will not go away or keeps getting worse

• Abdominal or chest pain that keeps getting worse

• Any signs of infection in your incision:
  - Redness
  - Swelling
  - Foul-smelling drainage

**If you are calling weekdays between 8 a.m. and 4 p.m.:**

• Call the Surgical Specialties Clinic Nurse Advice Line at **206-598-4477**.

**If you are calling after hours or on a weekend or holiday:**

• Call **206-598-6190** and ask for the resident on call for Surgery to be paged.
Follow-up Care

Short-term Follow-up
We would like to see you back in the clinic 2 to 3 weeks after you leave the hospital. When you get home, please call our Patient Care Coordinator at 206-598-4547 to schedule your appointment if one is not already scheduled.

If you live more than 2 hours away, please ask your surgeon if you need to have this follow-up appointment.

Long-term Follow-up
Your long-term follow-up appointment should be about 6 months after your surgery. This visit is important to assess the success of your surgery and to see if you need any other treatment.

At this visit, you may have these tests:
- 24-hour pH monitoring
- Upper gastrointestinal barium X-rays

Questions?
Your questions are important. Call your doctor or health care provider if you have questions or concerns.

Weekdays from 8 a.m. to 4 p.m., call the Nurse Line for the Surgical Specialties Clinic at 206-598-4477.

After hours and on weekends and holidays, call 206-598-6190 and ask for the Resident on call for Surgery to be paged.