UW Medicine
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Achalasia

What it is and how it is treated

Contents

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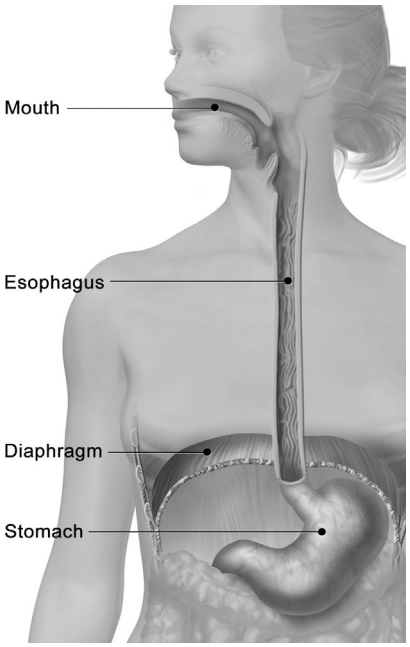
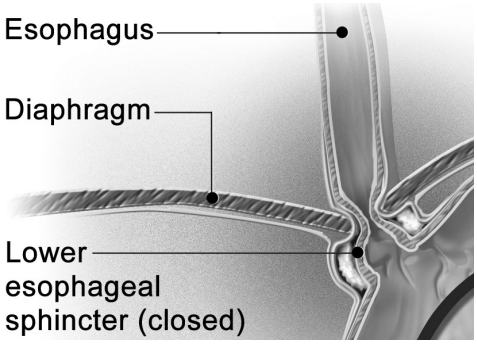


In normal digestion, swallowed food goes down the esophagus and into the stomach.

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*, wave-like muscle contractions.

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

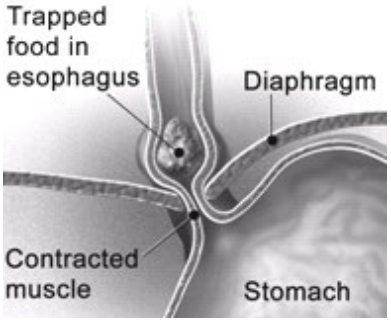


Most times, the LES is closed to keep stomach acid from rising up into the esophagus.

In achalasia, 2 things happen:

- Peristalsis in the esophagus does not work properly
- The LES does not relax as it should

When someone has achalasia, food and liquid collect in their esophagus, right above the LES. It stays there until it creates enough pressure to push through the LES.



With achalasia, food and liquid collect above the LES.

What causes achalasia?

We do not know the exact cause of achalasia. It sometimes occurs as a symptom of another condition, such as an infection, damage to the nerves, or cancer.

We do know that achalasia is linked to a lack of nerve cells inside the muscles that help with peristalsis. These nerve cells, called *ganglia,* produce *nitric oxide*, a substance that helps the LES relax.

People with achalasia produce less nitric oxide, so their LES does not relax. And, the lower part of their esophagus may become narrower.

Who gets achalasia?

Achalasia is very rare. Only 10 to 20 people out of 1 million have it. It is most common in middle-aged and older adults. The average age of people with achalasia is 49. But, it can occur at any age.

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Achalasia affects about 3,000 people in the United States. It occurs equally among men and women of different races. It tends to get worse over time.

What are the symptoms of achalasia?

The main symptom of achalasia is trouble swallowing, 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*, when 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

Aspiration tends to occur in advanced stages of achalasia.

What can I do to ease the symptoms of achalasia?

- Eat slowly.
- Lessen stress. Tension can make achalasia worse.
- To help ease chest pain or spasms, try:
  - Drinking warm or room-temperature water or seltzer water
  - Chewing crackers, bread, ice, or hard candy
  - Gulping milk
  - Drinking warm milk
  - Taking antacid medicine such as Tums, Mylanta, Maalox, or Gaviscon

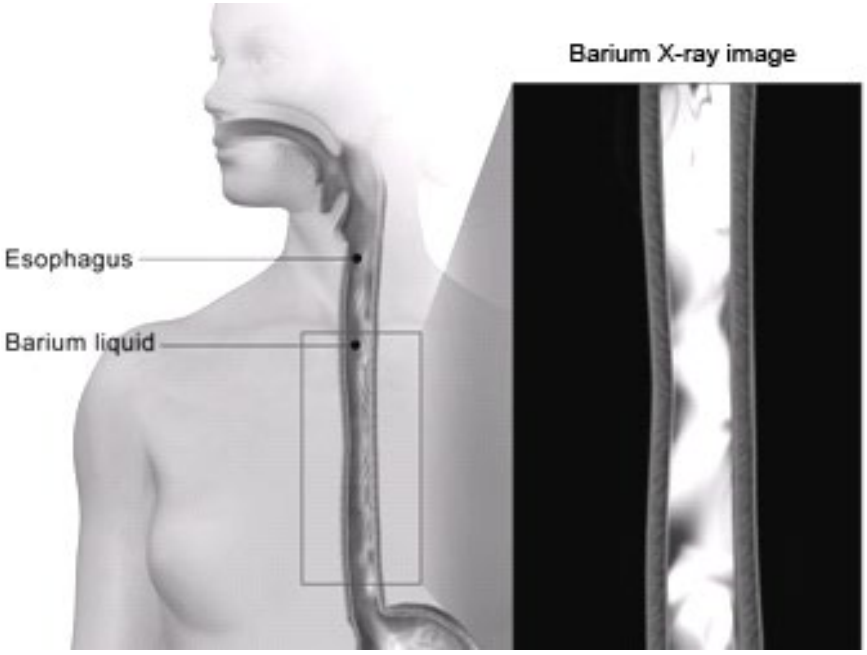
How is achalasia diagnosed?

To diagnose achalasia, your healthcare provider will do a full health exam. This will include a detailed review of your medical history. You will also have 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" below.) If you have achalasia, the images will likely show that your esophagus gets very narrow where it enters your stomach.



Barium liquid helps your doctor see the inside of your esophagus.

Esophageal Manometry

A manometry test is an important step in diagnosing achalasia. For this test, a device will be inserted through your nose or mouth. The device measures the peristaltic waves at different places inside your esophagus.

Flexible Upper Endoscopy

In a flexible upper endoscopy, a specialist checks your esophagus and stomach with a thin, flexible tube called an *endoscope*. This tube is put down your throat.

The endoscope has a light and a tiny camera on one end. The camera sends pictures of the inside of your esophagus to a monitor for your doctor to see.

An endoscopy is needed to make sure that you do not have a tumor in your esophagus. This condition is called *pseudoachalasia* ("false" achalasia). It requires different treatment.

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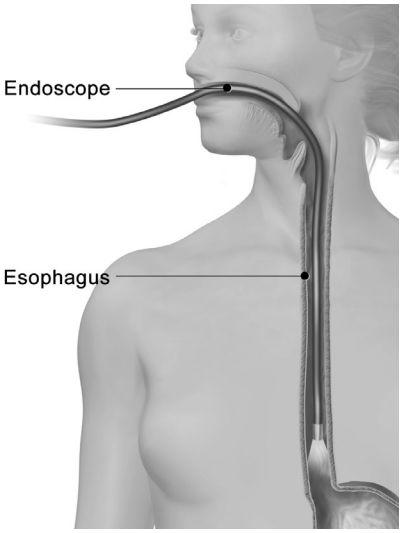

During an endoscopy, a thin, flexible tube called an endoscope is put down your throat.

People with pseudoachalasia most often:

- Have had symptoms less than 6 months
- Have lost more than 15 pounds
- Are older than 55

What other problems can occur with achalasia?

Other problems that can occur with achalasia include:

- Weight loss
- Malnutrition

A person with advanced achalasia can *aspirate* (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 7).

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 ease your symptoms while you wait.

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Botox Injection

Botulinum toxin (Botox) paralyzes the nerve cells that tell the LES valve to contract. Botox injections:

- Give short-term relief (6 months to 1 year)
- Work for about 70% of patients (70 out of 100 patients)

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 to treat achalasia should **not** be given to people who will be having *myotomy* surgery (see below). Botox can increase problems for these patients. Botox is usually given to people who are too ill for anesthesia or who are expected to live less than 3 years.

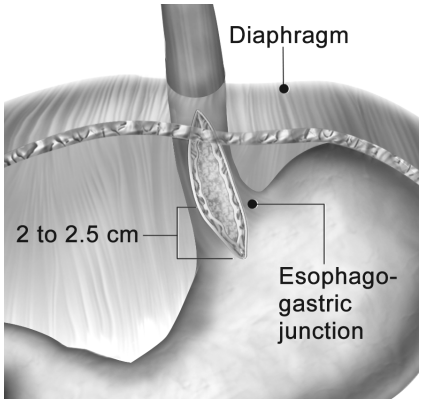
Endoscopic Dilation

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 move a catheter into your esophagus. The catheter is placed inside the LES and the balloon is inflated for up to 1 minute.

This procedure is:

- Most often done in the clinic using conscious sedation.
- 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 problem that can occur in endoscopic dilation is a tear in the wall of the esophagus. This happens in less than 5% of people (fewer than 5 out of 100) who have this procedure. This tear may heal on its own, but surgery may be needed to repair it.



In a myotomy, the surgeon makes a small cut in the muscle around the LES where the esophagus meets the stomach (the esophagogastric junction).

Myotomy 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.

During laparoscopic surgery, a tiny camera will take images of the inside of your belly and project them onto a monitor. This helps your surgeons see the areas being worked on.

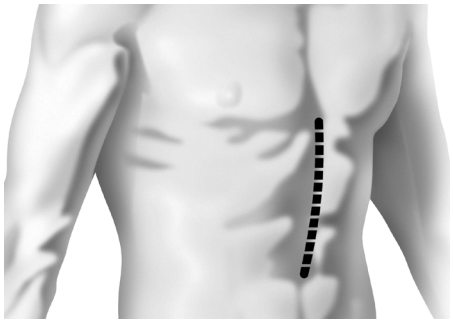
Myotomy eases achalasia symptoms in about 95% of patients (95 out of 100 patients). This surgery gives longer lasting relief of achalasia symptoms than other treatments.

Myotomy also involves risks, such as tearing or leaks in the esophagus. Make sure an *esophageal surgical specialist* does your procedure.

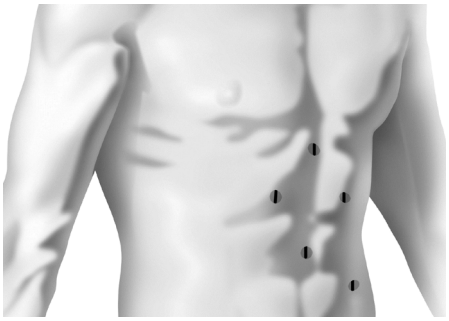
What happens during a myotomy?

Myotomy is done using *laparoscopy*. This means that instead of doing *open surgery* through 1 large incision in your belly, your surgeon will make about 5 small incisions (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.



Open incision



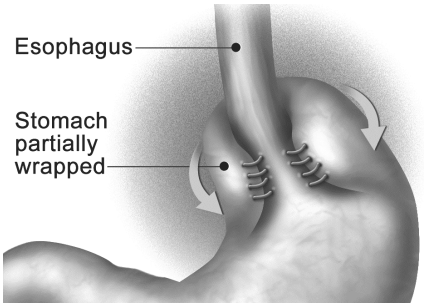
Laparoscopic incisions

You will have *general anesthesia* for your myotomy. This means you will be asleep during the 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 creates a 1-way valve to help keep stomach acid from entering your esophagus.



Partial fundoplication

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It is important to be in the best possible health for your surgery. This includes eating a healthy diet and getting exercise.

Preparing for Surgery

For best outcomes, it is important to be in the best possible health for your surgery. Please eat a healthy diet and exercise every day from now until your surgery.

Liquid Diet to Prevent Aspiration

For **3 days** before your 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.

When anesthesia is given, any food that is in your esophagus may travel to the back of your mouth and enter your airways. If this food reaches your lungs, it can cause pneumonia and even lung damage.

Surgery Day

To learn what happens on the day of your surgery, please read the purple packet we gave you.

After Surgery

In the Recovery Room

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

- Nurses in the recovery room will monitor your pain level. They will give you medicine to make you comfortable.
- Your family may be able to visit you in the recovery room after you are awake. This depends on how you are doing and the care needed by 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 under your nose) when your lungs are ready.
- An **intravenous catheter (IV)** to give fluids and medicines.
- **Sequential compression devices (SCDs) on your legs.** These leg wraps help keep blood from pooling in your calves. They inflate from time to time to improve blood flow and help prevent blood clots. You will feel pressure on your legs when SCDs inflate.

You may or may not have:

- A **urinary catheter** (tube) in your bladder. This lets us monitor your urine output. The catheter will be removed at midnight.

In Your Hospital Room

When your doctor says you are ready, you will be moved from the recovery room to a room in one of the units in the hospital.

During your hospital stay:

- **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 problems such as pneumonia.
- **Activity:** It is important for you to get up and try to walk, even the evening after your surgery. Do **not** try to walk on your own at first. 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.
- **Diet:** After surgery, you will be started on clear liquids. When your doctor says you are ready, you will begin an esophageal diet. A dietitian will visit you the day after your surgery to talk about your diet when you leave the hospital.
- **Visitors:** Family and friends can do things to help make you more comfortable. They can fluff your pillow, get you a glass of water, or find your remote control. If one of your visitors is sick, we may ask them to put on a mask or gown. For the safety of patients and staff, we may ask them to leave the hospital and return when they are well.

Going Home

Most patients are discharged by 11 a.m. the day after surgery. If you live more than a 2-hour drive from the hospital, we advise you to stay in the Seattle area an extra 1 or 2 nights after you leave the hospital. This rest time will help your recovery. You will also be nearby in case any problems occur.

Self-care at Home

For Your Safety

The medicines you receive during surgery can affect your ability to think clearly. For 24 hours after surgery **and** while you are taking opioids:

- Do **not** drive
- Do **not** travel alone
- Do **not** drink alcohol
- Do **not** be home alone

- Do **not** be responsible for children, pets, or an adult who needs care
- Do **not** use machines
- Do **not** sign any legal papers or make important decisions

Driving

Do **not** drive:

- For at least 2 weeks after surgery.
- As long as you are taking prescription pain medicine (opioids).

You may begin driving when you feel that your reaction time is normal.

Pain Control

- You will have some pain at your incision sites. We advise you to take acetaminophen (Tylenol) or ibuprofen (Advil, Motrin) for pain relief. Follow the dosing schedule in your discharge instructions.
- Ice packs on your incisions can help with pain.
  - Do **not** place ice directly on your skin. Put a clean cloth between the ice pack and your skin.
  - Use the ice pack for 20 minutes at a time, and wait 20 minutes before using it again.
- You will receive a prescription medicine (opioids) to help with moderate to severe pain. **Only** use opioids if you have severe pain that acetaminophen and ibuprofen do not relieve.
- If you want to request an opioid refill, you must either:
  - Come to the hospital pharmacy to pick up a prescription; or
  - Call us before you run out of your opioids. We cannot send opioid prescriptions directly to your pharmacy. It will take a few days for your prescription to arrive in the mail.
- You may also have shoulder pain for 4 to 5 days after your surgery. This is caused by the gas (*carbon dioxide*) that we used to inflate your belly during surgery. If this pain bothers you, try walking, massage, or heating pads. Opioids do not have much effect on this pain.
- Some pain medicines can make you dizzy. Ask for help when you get out of bed so that you do not fall.
- Some pain medicines can cause constipation. Take the laxative as prescribed. Stop taking it if you start having loose stools or diarrhea.

Medicines

- **For 4 to 6 weeks after surgery:** All of your medicines must be crushed or in a liquid form. Do **not** swallow whole pills during this time. We will give you a pill crusher before you go home. Call your pharmacy if you have questions about crushing any of your pills.
- Stop taking all antacids. If your symptoms return, call your surgeon's office.
- Take all of the medicines you received at discharge, as prescribed. One of these medicines will help prevent nausea and vomiting. It is important **not** to vomit in the first few weeks after your surgery. Follow the written instructions that come with your medicines.
- You may resume all of your other usual medicines, unless your provider tells you not to.

Activity

- For 6 weeks after surgery:
  - Do **not** lift anything that weighs more than 15 pounds (a gallon of water weighs almost 9 pounds).
  - Avoid activities that use your stomach muscles, increase your heart rate, or make you breathe hard.
- It is important to walk. Start walking as soon as you can after surgery. Walk 3 to 4 times a day, for a total of at least 1 mile a day. Walk farther as you recover.
- Slowly increase your activity as you heal.
- You may resume sexual activity 2 weeks after your surgery, as long as you follow all activity precautions.
- Let pain be your guide! If an activity causes pain, stop doing it. Try it again on another day.

Dressing and Skin Care

- Remove your dressings (gauze and Tegaderm) 48 hours after your surgery.
- You will have white tape called Steri-Strips under your dressings. Do **not** peel them off. They will fall off by themselves in 1 or 2 weeks.

Showering

- You may shower the day after surgery. The Tegaderm is plastic and repels water.

- Once you remove your dressings (48 hours after surgery), it is OK to shower and get the Steri-Strips wet.
- Gently pat the Steri-Strips dry after showering. Do **not** rub them dry.
- Do **not** take a bath, go swimming, sit in a hot tub, or soak your incisions until they are fully healed.

Diet and Nutrition

In the hospital, you will be on a liquid diet after your surgery. When you leave the hospital, you will start a soft esophageal diet. This diet helps keep food from getting stuck in the area where your surgery was done.

- You will stay on this soft diet for 4 to 6 weeks.
- Follow your dietitian's instructions on what foods you can eat at home after your surgery. Call your dietitian if you have questions.
- Try soft foods like mashed potatoes, eggs, cottage cheese, and thick soups.
- Instead of eating 3 large meals, eat 5 to 6 small meals a day. Take small bites, chew them well, and eat slowly. Stop when you feel full.
- Do **not** drink carbonated liquids or use a straw to drink.
- Most patients lose about 10 pounds after this surgery. You will gain this weight back unless you try not to.
- You will shift to a regular diet in 4 to 6 weeks.

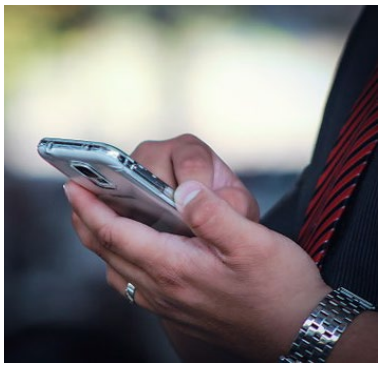
Bowel Movements

- You may have diarrhea (loose stools) after surgery. This is mostly due to the change in your diet. This usually goes away in a few days.
- Call the clinic if you have diarrhea for more than 3 days.
- Do **not** take any medicines for diarrhea unless your surgeon's team says it is OK.

When to Call Your Doctor

During the 7 days after surgery, call your primary care provider (PCP) if you:

- Cannot swallow foods, or you can only drink liquids
- Cannot keep fluids down
- Have trouble swallowing



Call your surgeon if you have a fever higher than 100.5 ℉ (38 ℃).

- Vomit a lot, even if you are taking medicines to prevent nausea
- Have vomit that is green, bloody, or looks like coffee grounds
- Have chest pain or shortness of breath
- Have severe, constant pain that is not eased by pain medicines and rest
- Have back or shoulder pain that does not go away
- Feel very full and your belly is swollen (*distended*)
- Cannot have a bowel movement or have diarrhea
- Have black or tarry stools
- Feel dizzy or faint when you stand up
- Have new or increased weakness, numbness, or tingling
- Have these symptoms in one of your legs or arms: warmth, tenderness, pain, swelling, or redness
- Have more bleeding from your incisions
- Have any sign of infection around your incisions:
  - Fever higher than 100.5°F (38°C)
  - Shaking or chills
  - Increase in drainage, or drainage is thick or smelly
  - Redness or swelling
  - Increasing pain or tenderness at or spreading away from the incision sites

Notes

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Questions?

Your questions are important. Call your doctor or healthcare provider if you have questions or concerns.

Weekdays from 8 a.m. to 5 p.m., call 206.598.4477, and press 2 when you hear the recording.

After hours and on weekends and holidays, call 206.598.6190 and ask to page the Surgery "O" team.

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