

# Achalasia

**Achalasia** is a **motility disorder** with insufficient muscle relaxation and a passage disorder of the bite from the esophagus to the stomach (*chalao* = relax), the cause of is not intraluminal obstruction or external esophageal compression. It is a **relatively rare disease** and has varying degrees of manifestation.

The esophagus is a hollow organ that connects to the pharynx and opens into the stomach. The esophageal wall is made up of, among other things, muscle. Mutually coordinated contractions of the muscle cells in the walls of the esophagus controlled local glial cells result in swallowing the correct shift of food toward the stomach, the esophagus which is separated by *the lower oesophageal sphincter* (circular muscle reinforced). As the food progresses to the stomach, the sphincter reopens thanks to the local ganglion cells and the food "falls" into the stomach.

Achalasia is based on a neuromuscular disorder (at the level of the *vagus nerve* or esophageal muscle) associated with loss of cells in the *myenteric plexus* and muscle hypertrophy. As a result of the gradual paralysis of the esophageal muscle, the movement of the bite is stuck in the stomach during swallowing. Increasing intraluminal pressure can lead to pulsed diverticula above the sphincters.

## Types

- **Cervical** (crico- pharyngeal) – a disorder of relaxation of the upper esophageal sphincter during swallowing;
- **Esophageal-cardiac** – loss of peristalsis and inability to relax the lower esophageal sphincter during swallowing.

## Causes

The causes of achalasia are not usually known. For some reason, the nerve ganglion cells in the esophageal wall and the sphincter wall begin to disappear, causing the muscle cells to lose innervation and cease to function, leading to the above problems.

## Symptoms

Manifestations of the disease arise gradually.

The main manifestation is a **swallowing disorder** or **dysphagia**. At first, the patient begins to have difficulty swallowing a large stiff bite, snorts, feels **pain** and **pressure in his chest** at mealtimes (food accumulates in the esophagus and does not progress). A sick person helps himself by drinking a lot of bites. Fluids pass better through sick esophagus and can take solid parts of food with them.

Another symptom is a **cough**, which typically occurs at night, and **recurrent lower respiratory tract infections** (recurrent pneumonia). This is due to the frequent flow of congested food into the airways. Although the airways are protected by the laryngeal valve (*epiglottis*), however, when food in the esophagus stagnates, the chances of its penetration are great, despite the existence of the valve.

## Diagnostics

The doctor will find suspicion during the patient's complaints about poorer swallowing of a solid diet and other symptoms mentioned above. From the examination it is possible to perform:

- **Gastroscopy** - eliminates other causes (such as narrowing of the esophagus by the tumor), an experienced endoscopist can recognize esophageal achalasia with just a glance.
- **Oesophageal X-ray** - during this examination the patient drinks a contrast agent and his swallowing act is clarified by X-ray. We will find out the shape of the esophagus, its filling and how the substance drains through the lower sphincter. If the substance accumulates in the esophagus and does not drain, our suspicion confirms this. According to the X-ray finding, there are 4 stages:
  1. uncoordinated lower sphincter function, muscle hypertrophy with aperistaltic contractions, without esophageal dilatation;
  2. gradual dilatation of the esophagus, loss of peristalsis with persistent lower sphincter hypertension;
  3. amotile form, lower sphincter hypertension, dilatation and elongation of the esophagus without peristalsis (esophageal atony);
  4. dolichomegaesophagus;
- The final examination is the so-called **esophageal manometry** - a thin wire is inserted into the esophagus, which measures the pressure in individual parts of the esophagus. If an increase in pressure in the lower esophageal sphincter is detected when swallowed, the diagnosis of achalasia is practically certain. This is because the pressure in the lower esophageal sphincter should drop when swallowed so that food can slip from the esophagus into the stomach.

## Prevention

It's not possible. However, if you notice any difficulty swallowing, you should see a doctor. In addition to achalasia, it could also be a tumor of the esophagus (typically men around the age of 50). A growing tumor can oppress the esophageal cavity, causing similar symptoms. In addition to treatment, a patient with established esophageal achalasia should be monitored regularly with an endoscope, as achalasia is at increased risk of developing esophageal tumors.

## Therapy

Due to the fact that we do not know the cause of achalasia, the treatment is relatively difficult.

- **Diet**
  - It is advisable to recommend that the patient eat **less solid food** (porridge, mashed and ground food) and **wash down** food with water.
- **Conservative treatment**
  - The administration of calcium channel blockers helps to relax the esophageal sphincter.
- **Endoscopic treatment**
  - Injection of botulinum toxin into the lower esophageal sphincter causes it to relax. Another option is to stretch the esophagus through balloon dilation.
- **Surgical treatment**
  - **Heller's** lower or upper sphincter **myotomy** is performed, followed by esophageal resection if the myotomy fails. At the same time, the esophageal outlet must be surgically adjusted to prevent reflux of gastric contents back into the esophagus.
  - A novelty is the so-called **oral endoscopic myotomy (POEM)**<sup>[1]</sup>

## Links

### External links

- Achalazie (<https://www.stefajir.cz/?q=achalazie>) (na stránkách stefajir.cz)

### Related articles

- Onemocnění jícnu
- Difúzní spasmus jícnu

### Source

- ŠTEFÁNEK, Jiří. *Medicína, nemoci, studium na 1. LF UK* [online]. [cit. 24.03.2010]. <<https://www.stefajir.cz/>>.
- PASTOR, Jan. *Langenbeck's medical web page* [online]. [cit. 2009]. <<https://langenbeck.webs.com/>>.

### Literature

1. Klinika hepatogastroenterologie, IKEM. *POEM dorazil do České republiky* [online]. [cit. 2012]. <<http://www.endoskopiste.cz/ruzne-1/nove-endoskopicke-techniky/poem-dorazil-do-ceske-republiky.aspx>>.



Barium swallow. Dilated esophagus with retained column of barium and "bird's beaking" suggestive of achalasia