Achalasia (Cardiospasm) — Diagnosis and Surgery

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Achalasia (cardiospasm) is a functional disorder caused by the narrowing of the lower esophageal muscles. It is marked by the reduced or absent peristaltic movement of the esophagus and failure of the lower esophageal sphincter to relax during swallowing. This abnormality results in functional obstruction at the junction of the esophagus and stomach with symptoms such as dysphagia, regurgitation, heartburn, stomach pain, and weight loss. It is diagnosed with a barium swallow test, esophageal monitoring, and endoscopy. Pharmacological and non-surgical interventions are used to treat achalasia.

Definition of Achalasia

Achalasia as a functional disorder of the esophagus
Achalasia is caused by the narrowing of the lower esophageal muscles. It is characterized by decreased peristalsis and the absence of physiological slackening of the lower esophageal sphincter during swallowing. If not treated, achalasia progresses from stage I (hypermotile form) to stage II (hypomotile form) and stage III (amotile form).

**Epidemiology of Achalasia**

**Achalasia as an exceptional disease**

With an incidence of 1/100,000 people, achalasia is a rather rare disease. It affects men and women with the same frequency, mostly in the third to fifth decade of life. Achalasia is even rarer in the context of the autosomal-recessive congenital disease triple A syndrome (achalasia, alacrimia, and adrenal insufficiency).

**Etiopathogenesis of Achalasia**

**Features of achalasia**

To date, the causes of primary achalasia are unknown. Degeneration of the inhibitory ganglia cells of the myenteric plexus (Auerbach’s plexus) occurs in the lower esophagus. Normally, these neurons cause the esophagus to relax, especially during swallowing. Their absence results in deficient slackening during the swallowing reflex and increased resting pressure of the lower esophageal sphincter. Achalasia is a type of esophageal motility disorder.

With achalasia, food piles up in front of the narrowed lower esophageal sphincter, and later dilatation of the esophagus directly in front of the narrow passage develops. In sporadic findings, megaesophagus can form.
If achalasia occurs in the context of another disease, this is referred to as secondary achalasia. **Chagas disease** (*Trypanosoma cruzi infection*) and tumor diseases can be the cause of secondary achalasia.

**Symptoms of Achalasia**

**Dysphagia and regurgitation at achalasia**

Patients primarily complain about having dysphagia for several years. Often, they have to drink a lot of fluids after food intake to be able to swallow solid food completely. However, even swallowing fluids can be hard in the later stages.

Regurgitation of food occurs, especially when lying down, which is associated with the complications of aspiration pneumonia and esophagitis. This inflammation of the esophagus called retention esophagitis increases the carcinoma risk 30-fold compared to the normal population. With increased **regurgitation**, **foetor ex ore** can often be detected.

Furthermore, patients often describe bloating. Very rarely, convulsive pain (**hypermotile achalasia**) is also present, especially at the beginning of the disease. This decreases later in the disease.
Diagnosis of Achalasia

Anamnesis of achalasia

Concerning anamnesis, one should ask if the complaints occurred suddenly and if they are rapidly progressing. This would suggest a diagnosis of carcinoma. However, in the case of achalasia, complaints often exist for many years, and the patient presents with a long medical history.

Manometry of achalasia

Manometry (physical pressure measurement) is the gold standard for detection of achalasia. This test measures pressure in different parts of the esophagus (the normal resting pressure is 20 mm Hg).

![Manometric schematic of achalasia demonstrating aperistaltic contractions, increased intraesophageal pressure and failure of relaxation of the lower esophageal sphincter.](Image: "Manometric schematic of achalasia demonstrating aperistaltic contractions, increased intraesophageal pressure and failure of relaxation of the lower esophageal sphincter," by JC Petit. License: CC-BY 3.0)

Three characteristic features can be detected, including a lack of slackening during the swallowing reflex, pathological peristalsis, and increased resting pressure. Classification of the severity of achalasia is mostly made during manometry.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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<tr>
<td>Stage I</td>
<td><strong>Hypermotile:</strong> The muscles of the esophagus actively contract to pass the narrowed lower esophageal sphincter.</td>
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<tr>
<td>Stage II</td>
<td><strong>Hypomotile:</strong> The distal esophagus dilates at a constant contraction of the lower esophageal sphincter.</td>
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<tr>
<td>Stage III</td>
<td><strong>Amotile:</strong> The esophageal muscles do not contract anymore. Only the esophageal sphincter is still narrow; increasing dilatation of the preceded esophageal part.</td>
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Esophagus barium swallow at achalasia

With an esophagus barium swallow test, the so-called “champagne glass shape” with prestenotic dilatation (megasophagus) can be seen in an x-ray. Peristalsis is disturbed. After a delay, the contrast agent drains from the esophagus into the stomach.

Note: In exams, x-ray barium swallow images are often shown. You should recognize the “champagne glass shape” and be able to associate it with achalasia.

Endoscopy with biopsy at achalasia

Esophagoscopy with biopsy is needed to exclude the presence of carcinoma. Due to the risk of developing carcinoma later in the disease, the procedure should be repeated every one to two years. In the context of achalasia, endoscopy typically shows a widened esophagus and a lower esophageal sphincter that can easily be passed through by the endoscope.

Note: Due to the increased risk of carcinoma, regular endoscopic check-ups are necessary.

Differential Diagnosis

Alleged achalasia

The differential diagnosis of esophagus achalasia mainly includes esophageal carcinoma, which is characterized by a distinctly more rapid progression. You should perform a biopsy to make a differential diagnosis. Further motility disorders, which are a differential diagnosis of esophagus achalasia, include:

- **Esophagus spasm:** Esophagus spasm is a rare, neuromuscular disorder of the esophagus of unknown genesis. Paroxysmal retrosternal pain and dysphagia occur. 24-hour manometry is the leading diagnostic and shows primarily reinforced and continuous esophagus contractions. In milder cases, calcium antagonists are administered. In severe cases, botulinum toxin is injected, or surgical intervention is used.

- **Hypercontractile esophagus (nutcracker esophagus):** This motility disorder is characterized by non-physiologically long-lasting contractions (> 5 seconds) of the esophageal muscles and particularly high-pressure amplitudes (individual amplitudes are partial > 200 mm Hg). This can be measured in manometry. The function of the lower esophageal sphincter is not impaired in this motility disorder.

Therapy of Achalasia

Pharmacological therapy of achalasia

Unfortunately, the long-term results are bad. However, therapy with a calcium antagonist such as nifedipine should be attempted, especially in the hypermotile stage. Nifedipine decreases the pressure of the lower esophageal sphincter. It should be taken half an hour before meals.
Interventional therapy at achalasia

**Pneumatic balloon dilation:** During esophagoscopy, the cardiac muscles are dilated or torn during this procedure. Often, one session is not enough, and the patients have to be treated several times. The success rate is 60%. Due to the risk of perforation (2–5%), an esophagus barium swallow test should always be performed after the intervention.

If there is a suspicion of perforation, barium must not be used for this procedure (risk of mediastinitis or peritonitis), but a water-soluble contrast agent should be used.

Another alternative is the endoscopic injection of botulinum toxin. It is injected directly into the lower esophageal sphincter. However, the procedure still has to be repeated after half a year. Due to the short duration of the effect, this method is for patients who cannot undergo other interventions due to distinct comorbidities.

Surgical interventions at achalasia

Surgical interventions are used as alternative therapies. The success rate is greater than 90%. However, one has to consider the complication of post-surgical reflux disease. Primarily, two surgery techniques are used:

1. **Extramucosal cardiomyotomy, according to Gottstein/Heller (laparoscopic or transabdominal):** The muscles of the stomach entry (cardia) are split longitudinally in a 6 cm long line. The mucosa is not affected. To avoid reflux disease, Nissen fundoplication is subsequently performed in most cases.

2. **Transgastric esophagus fundostomy, according to Prager:** During this procedure, a left-sided side-to-side esophagus fundostomy (TSE) is performed transgastrically, during which the gastrostomy is sealed. In addition, partial fundoplication can also be performed.

In the event of all treatment approaches failing, esophagectomy with a small intestine interpolation has to be considered as a last-resort measure.

**Note:** Balloon dilatation is first-resort therapy.

**Review Questions**

The answers are below the references.

1. **Which answer most likely describes achalasia?**
   
   A. A congenital malformation of the esophagus.
   
   B. The disturbed slackening of the lower esophageal sphincter.
   
   C. An overshoot in ganglia cells of the esophageal Auerbach’s plexus.
   
   D. A decreased resting tone of the lower esophageal sphincter.
   
   E. None of the mentioned answers are correct.

2. **Which treatment measure is most likely indicated in a 32-year-old patient with achalasia in the amotile stage?**
   
   A. Nissen’s fundoplication
   
   B. Esophagectomy with small intestine interpolation
   
   C. Pharmacological therapy with nifedipine
   
   D. Pneumatic balloon dilatation
   
   E. Botulinum injection into the lower esophageal sphincter
A 40-year-old patient presents with long-existing dysphagia in your general medicine practice. He describes that he now has even troubles with swallowing fluids. Especially at night, chronic cough and regurgitation agonize him. Which of the following diagnostic measures proves the suspected diagnosis best?

A. Long-term pH-metry
B. MRI of the neck and thorax
C. H2-breathing test
D. Chest x-ray
E. Esophagus manometry

References


**Correct answers:** 1B, 2D, 3E

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