What is the definition of achalasia?
ACHALASIA: DEFINITION AND PATHOPHYSIOLOGY

Loss of swallow-induced relaxation of the lower esophageal sphincter (LES) and aperistalsis of the esophageal body.

Autoimmune, viral immune, or neurodegenerative degeneration of ganglion cells in the myenteric plexus of the esophageal body and the LES.

WHEN DO YOU SUSPECT ACHALASIA?
WHEN DO YOU SUSPECT ACHALASIA?

- Dysphagia for both solids and liquids
- Regurgitation of bland undigested food and saliva (mucus)
- Substernal chest pain
- Weight loss
- Heartburn (especially in early stages, inflammatory process?)
DIFFERENTIAL DIAGNOSIS
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Gastroesophageal reflux disease (PPI trial)

Chagas-Disease  (Trypanosoma cruzi-infection)
   Endemic in Middle and South America, megacolon?, heart disease?, neurologic abnormalities?

Pseudo-achalasia
   - neoplastic infiltration of the cardia (adenocarcinoma of gastroesophageal junction, pancreatic, breast, lung, or hepatocellular cancer)
   - paraneoplastic (small cell bronchial cancer)

Previous surgery (overly tight fundoplication, gastric banding)
**Endoscopy:**
- Normal findings in 1/3 of patients
- Passage of cardia against soft resistance
- Food remnants
- Dilated esophagus
- Candidiasis

Main indication: To rule out malignancy
Radiology (Timed barium swallow):

- „Bird beak“
- Dilated esophagus
- Retention of contrast media

(200 ml diluted barium suspension, height and width of contrast media column after 1, 2 and 5 minutes)

Cheap, very suitable for assessing therapeutic success
Elevated Integrated Relaxation Pressure (IRP), no normal peristalsis
Elevated LES resting pressure, esophageal pressure

Typ I: - no panesophageal pressurisation

Typ II: - panesophageal pressurisation
  >30 mmHg in > 20% of swallows

Typ III: - spastic, premature contractions
  in > 20% of swallows

RECOMMENDATIONS

1. All patients with suspected achalasia who do not have evidence of a mechanical obstruction on endoscopy or esophagram should undergo esophageal motility testing before a diagnosis of achalasia can be confirmed (strong recommendation, low-quality evidence).

2. The diagnosis of achalasia is supported by esophagram findings including dilation of the esophagus, a narrow esophagogastric junction with “bird-beak” appearance, aperistalsis, and poor emptying of barium (strong recommendation, moderate-quality evidence).

3. Barium esophagram is recommended to assess esophageal emptying and esophagogastric junction morphology in those with equivocal motility testing (strong recommendation, low-quality evidence).

4. Endoscopic assessment of the gastroesophageal junction and gastric cardia is recommended in all patients with achalasia to rule out pseudoachalasia (strong recommendation, moderate-quality evidence).

THERAPY OF ACHALASIA
THERAPY OF ACHALASIA

Goal: reduction of outflow obstruction at the level of the LES

Pharmacologic
- Nitrates, Ca++-channel-inhibitors (Nifedipine), Sildenafil,
  Botulinum-toxin

Endoscopic
- Dilatation
- Temporary stenting
  POEM = PerOral Endoscopic Myotomy

Surgical
- Laparoscopic cardiomyotomy
  ± fundoplication (Dor vs. „floppy Nissen“)
PNEUMATIC DILATATION

Performed under sedation and under fluoroscopy
Initial dilatation with 30 mm balloon, 8-15 psi for 15-60 seconds
PNEUMATIC DILATATION

Performed under sedation and under fluoroscopy
Initial dilatation with 3.0 cm balloon, 8-15 psi for 15-60 seconds
Graded dilatation approach: Cumulatively, dilation with 3.0, 3.5, and 4.0 cm balloon diameters results in good-to-excellent symptomatic relief in 74, 86, and 90 % of patients with an average follow-up of 1.6 years (range 0.1 – 6 years).

The most serious complication is esophageal perforation with an overall median rate in experienced hands of 1.9 % (range 0 – 16 %)

All patients considered for Pneumatic Dilatation must also be candidates for surgical intervention in the event of esophageal perforation needing repair.

PNEUMATIC DILATATION

1. **Achalasia dilatation**
   1.1 Perform dilatation with pneumatic balloons 30–40 mm in diameter starting at 30 mm in the first session to reduce the risk of complications (GRADE of evidence: high; strength of recommendation: strong).

   1.2 Perform a second dilatation session 2–28 days later with a larger size balloon of 35 mm (GRADE of evidence: high; strength of recommendation: strong).

   1.3 Consider repeat dilatation (after the initial series) during follow-up to maintain symptom response (GRADE of evidence: high; strength of recommendation: strong).

   1.4 Perform dilatation under endoscopic or fluoroscopic control based on clinician’s preference and local expertise (GRADE of evidence: moderate; strength of recommendation: strong).

   1.5 Consider proton pump inhibitor (PPI) therapy after dilatation as the technique has 10–40% rate of symptomatic gastro-oesophageal reflux disease (GORD) or ulcerative oesophagitis after treatment (GRADE of evidence: high; strength of recommendation: strong).

   1.6 Consider performing a water-soluble contrast swallow after dilatation to screen for perforation, but it is not essential (GRADE of evidence: moderate; strength of recommendation: weak).

# DILATATION VS. MYOTOMY

201 patients randomised, follow-up time 43 mo (95% CI 40-47 mo)

<table>
<thead>
<tr>
<th></th>
<th>Pneumatic Dilatation</th>
<th>Laparoskop. Heller Myotomy + Dor Hemipl.</th>
<th>p</th>
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</thead>
<tbody>
<tr>
<td>n</td>
<td>95</td>
<td>106</td>
<td></td>
</tr>
<tr>
<td>1 yr Eckardt Score ≤3</td>
<td>90%</td>
<td>93%</td>
<td></td>
</tr>
<tr>
<td>2 yr Eckardt Score ≤3</td>
<td>86%</td>
<td>90%</td>
<td>0.46</td>
</tr>
<tr>
<td>2 yr LES-Pressure</td>
<td>12 mmHg [95% CI 9.7-14]</td>
<td>10 mmHg [95% CI 8.7-12]</td>
<td>0.27</td>
</tr>
<tr>
<td>2 yr barium column</td>
<td>3.7 cm [95% CI 0-8.8]</td>
<td>1.9 cm [95% CI 0-6.8]</td>
<td>0.21</td>
</tr>
<tr>
<td>Perforation</td>
<td>4%</td>
<td>12%</td>
<td></td>
</tr>
<tr>
<td>Abnormal acid exposure</td>
<td>15%</td>
<td>23%</td>
<td>0.28</td>
</tr>
</tbody>
</table>

No significant differences, but:
Single surgical procedure vs. multiple pneumatic dilatations (max. 3 series with 30, 35, 40 mm)

Boeckxstans G et al. NEJM 2011
## POEM VS. LAPAROSCOPIC MYOTOMY

<table>
<thead>
<tr>
<th></th>
<th>POEM</th>
<th>Laparoskop. Heller Myotomie</th>
<th>p</th>
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</thead>
<tbody>
<tr>
<td>Reports</td>
<td>21</td>
<td>53</td>
<td></td>
</tr>
<tr>
<td>n</td>
<td>1958</td>
<td>5834</td>
<td></td>
</tr>
<tr>
<td>Follow-up time</td>
<td>16.2 mo</td>
<td>41.5 mo</td>
<td>0.001</td>
</tr>
<tr>
<td>1 yr Dysphagia improved</td>
<td>93.5%</td>
<td>91.0%</td>
<td>0.01</td>
</tr>
<tr>
<td>2 yr Dysphagia improved</td>
<td>92.7%</td>
<td>90.0%</td>
<td>0.01</td>
</tr>
<tr>
<td>GERD symptoms</td>
<td>OR 1.69 (95% CI 1.33–2.14)</td>
<td></td>
<td>0.0001</td>
</tr>
<tr>
<td>Abnormal acid exposure</td>
<td>OR 4.30 (95% CI 2.96–6.27)</td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Erosive Esophagitis</td>
<td>OR 9.31 (95% CI 4.71–18.85)</td>
<td></td>
<td>&lt;0.0001</td>
</tr>
</tbody>
</table>

POEM results in lower rates of dysphagia than laparoscopic cardiomyotomy, but significantly higher rates of reflux

Schlottmann et al. Ann Surg 2018
RECOMMENDATIONS

1. Either graded pneumatic dilation (PD) or laparoscopic surgical myotomy with a partial fundoplication are recommended as initial therapy for the treatment of achalasia in those fit and willing to undergo surgery (strong recommendation, moderate-quality evidence).

2. PD and surgical myotomy should be performed in high-volume centers of excellence (strong recommendation, low-quality evidence).

3. The choice of initial therapy should be guided by patients’ age, gender, preference, and local institutional expertise (weak recommendation, low-quality evidence).

4. Botulinum toxin therapy is recommended in patients who are not good candidates for more definitive therapy with PD or surgical myotomy (strong recommendation, moderate quality evidence).

5. Pharmacologic therapy for achalasia is recommended for patients who are unwilling or cannot undergo definitive treatment with either PD or surgical myotomy and have failed botulinum toxin therapy (strong recommendation, low-quality evidence).
<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Preferred treatment, comments, and rationale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I and II achalasia</td>
<td>PD and LHM are both highly efficacious in RCT; PD has less morbidity and cost with PD, anticipate repeat dilations over the years. Insufficient data on efficacy of POEM for advanced esophageal dilation, sigmoidization, epiphrenic diverticulum, and hiatal hernia. POEM highly efficacious in RCT vs PD; only short-term data available. Expect more reflux after POEM, especially with hiatal hernia.</td>
</tr>
<tr>
<td>Type III achalasia</td>
<td>POEM, calibrate the myotomy length to the spastic segment imaged on HRM or thickened segment on EUS. Many cases resolve spontaneously. Image the EGJ (EUS, CT) to rule out obstruction. If achalasia therapies are applied, consider it type II achalasia.</td>
</tr>
<tr>
<td>EGJ outflow obstruction</td>
<td>Use FLIP, timed barium esophagram, or multiple repetitive swallows on HRM to establish need for treatment. If achalasia therapies are applied, consider it type I achalasia.</td>
</tr>
<tr>
<td>Absent contractility deemed to be achalasia</td>
<td>If achalasia therapies are applied, consider it type II achalasia. POEM, calibrate the length of myotomy to the spastic segment as imaged on HRM or thickened segment on EUS.</td>
</tr>
<tr>
<td>DES deemed to be achalasia</td>
<td>First choice, discontinue opioid; second choice: botox; third choice: POEM. Time course of reversal with opioid cessation is not known.</td>
</tr>
</tbody>
</table>
| Opioid effect                  | Many entities mimic achalasia, sometimes termed *pseudoachalasia*: eosinophilic esophagitis, cancer, reflux stricture, post-myotomy stricture. Conventional dilation. Operative reversal if relevant; directed medical therapy if relevant.