## Diagnosis and Management of Achalasia: Past, Present, & Future

Kyle A. Perry, MD, FACS  
Assistant Professor of Surgery  
Division of General & Gastrointestinal Surgery  
The Ohio State University Wexner Medical Center

### Achalasia

- Motor disorder of esophagus  
  - Aperistalsis  
  - Impaired LES relaxation  
- Causes dysphagia, pain, regurgitation  
- Leads to weight loss, pulmonary complications  
- Increased risk of esophageal CA

### Epidemiology

- Prevalence 7.9-12.6/100,000  
- Incidence 0.4-1.1/100,000  
- Mean age at diagnosis 30 to 60 years  
- Peak age in 40’s

### Cancer Risk

- Achalasia series report 0-33% increased risk of esophageal CA (mostly SCCA)  
- Swedish population-based study of 1062 achalasia patients with 9864 pt-years f/u  
  → 16-fold increased risk of esoph CA  
- Surveillance not recommended – would require >400 endoscopies to find one cancer
Historical Perspective

1674 Sir Thomas Willis (England) -- Successful treatment of “cardiospasm” with serial esophageal dilation using a sponge attached to a whale bone (baleen)

Lower Esophageal Sphincter

- High pressure zone 2 to 4 cm long at GEJ
- Parasympathetic and sympathetic innervation mostly in myenteric plexus
- Provides barrier to reflux of gastric juice

Pathophysiology of Achalasia

- Dysfunction of myenteric plexus
  - Early: lymphocytic inflammation
  - Late: loss of ganglion cells
- Selective loss of inhibitory neurons (VIP/NO)
- Sparing of stimulatory cholinergic innervation
- Failure of LES to relax with swallow
Clinical Presentation

- Progressive solid food dysphagia (variable for liquids)
- Chest pain – more frequent early decreases with progressive dilation
  Regurgitation of undigested, nonbilious food (esp. at night)
- Heartburn – not relieved with acid suppression

Clinical Features

- Eating maneuvers augment food passage
  Head back, upright posture, valsala
  Warm, carbonated/alcoholic beverages
- Food fermentation → acidification with esophageal ulceration/ heartburn
- Pulmonary complications -- aspiration
- Weight loss – up to 84% of patients

Differential Diagnosis

- Pseudoachalasia
  Over 50% 2º to GEJ/cardia tumor
  Other causes: pseudocysts, GEJ obstruction after hiatal surgery, paraneoplastic syndromes
- Scleroderma
  Aperistalsis with low LES pressure
- Neurologic disorders (Parkinson’s)
- Chagas’ Disease

Evaluation

- Manometry
- Barium esophagogram
- EGD
- +/- pH study
**Esophageal Manometry**

- Used to assess esophageal motility and LES function (pressure, length, relaxation)
- Multichannel water-perfused or solid state catheter connected to pressure transducers
- Records pressure at various points along the esophagus during bolus swallow

**Normal Manometry**

- UES relaxation
- Esophageal motility of the body
- LES residual pressure
- LES relaxation

**Normal High-Res Manometry**

**Manometry in Achalasia**

- Aperistalsis of body
- Incomplete relaxation of LES
- Normal to elevated LESP
- Simultaneous low amplitude contractions
Manometry

High-Res Manometry: Type I Achalasia

High-Res Manometry: Type II Achalasia

High-Res Manometry: Type III Achalasia
<table>
<thead>
<tr>
<th><strong>Barium Swallow</strong></th>
<th><strong>Endoscopy</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>- Dilated, possibly tortuous esophagus (late finding)</td>
<td></td>
</tr>
<tr>
<td>- “Bird's beak” tapered appearance of distal esophagus</td>
<td></td>
</tr>
<tr>
<td>- Air fluid level with retained food</td>
<td></td>
</tr>
<tr>
<td>- Necessary to rule out malignancy</td>
<td></td>
</tr>
<tr>
<td>- Requires retroflexed view of cardia/GEJ</td>
<td></td>
</tr>
<tr>
<td>- Scope should pass through easily – if not, consider malignancy</td>
<td></td>
</tr>
<tr>
<td>- EUS may help to evaluate esophageal wall when CA suspected</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>pH Study</strong></th>
<th><strong>Management</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>- May help when diagnosis is in question, especially when heartburn is major sx</td>
<td></td>
</tr>
<tr>
<td>- Interpretation may be difficult due to fermentation</td>
<td></td>
</tr>
<tr>
<td>- Achalasia shows gradual decline in pH vs. rapid drop seen with GERD</td>
<td></td>
</tr>
<tr>
<td>- Pharmocologic</td>
<td></td>
</tr>
<tr>
<td>- Botox injection</td>
<td></td>
</tr>
<tr>
<td>- Pneumatic dilation</td>
<td></td>
</tr>
</tbody>
</table>
| - Surgical myotomy  
  - Laparoscopic  
  - Endoscopic |
| - Esophagectomy |
### Pharmacologic Therapy
- **Goal** – decrease LES pressure to allow esophagus to empty
- **Nitrates** – use limited by side effects
- **Nifedipine** – reduces LESP but minimal improvement in symptoms vs. placebo in 2 out of 3 RCT’s
- **Reserved** for mild disease or patients unable to tolerate dilation/surgery

### Botulinum Toxin
- **Intrasphincteric injection of LES**
- Blocks release of acetylcholine at neuromuscular junction
- 70-100% effective at 1 month
- Ease of administration and low rate of adverse effects contribute to popularity
- Most benefit seen in elderly, debilitated patients

### Botulinum Toxin – The Downside
- Usually requires repeat injection at 6 to 9 months – 30% remission at 1 year
- Response to repeat injections limited by antibody formation
- Reported to increase scarring of distal esophagus, increasing difficulty of surgery
- Long term results inferior to dilation in several RCT’s

### Treatment Strategies
- **Medical Therapy** (Nitrates, nifedipine)
- **Botulinum Toxin**
- **Endoscopic pneumatic dilation**
  - **Advantages**: Effective symptom relief, outpatient procedure
  - **Disadvantages**: Repeat dilations often needed, increased risk of esophageal perforation
- **Laparoscopic Heller Myotomy**
  - **Advantages**: durable symptom relief
  - **Disadvantages**: invasive surgical procedure

**Pneumatic Esophageal Dilation**

- 3-4 cm balloon rapidly inflated in distal esophagus under fluoro guidance
- Relies on rupture of LES fibers

**Trials with f/u > 2 years report good to excellent results in 65-80%**

- Repeat dilation required in > 50%
- West *et. al.* reported on 125 patients
  - 12 year f/u → 50% remission with median 4 tx
  - 15 year f/u → 40% remission *Am J Gastro 2002*
- Improved results in older patients (over 40) and those with post-dilation LESP < 10

**Complications of Dilation**

- Perforation
  - Most series report 0 to 4%

- Gastroesophageal reflux
  - Symptomatic in 7-17% of cases

**Surgery vs. Dilation**

- **Dilation**
  - outpatient procedure
  - minimal pain
  - Rapid return to work
  - May treat any patient population (frail, pregnant, ect…)
  - Less expensive
  - Does not preclude myotomy

- **Myotomy**
  - single procedure
  - dysphagia relief is longer at the cost of more heartburn
  - may be more effective treatment in younger patients
Surgery vs. Dilation

• Over a two year horizon, the clinical success of pneumatic dilation and laparoscopic myotomy are comparable in a recent large European randomized trial.

• However, at 5 year follow-up, surgery is favored due to higher rate of recurrent dysphagia requiring retreatment with increased complications in patients undergoing endoscopic dilation.

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Jeffrey W. Hazey, MD, FACS
Associate Professor of Surgery
Center for Minimally Invasive Surgery
Division of General & Gastrointestinal Surgery
The Ohio State University Wexner Medical Center

Surgical Therapy – Historical Perspective

• 1913 Heller: Anterior and posterior myotomy via abdominal approach
• 1918 Groeneveldt: Single anterior myotomy
• 1958 Ellis: Thoracic approach
• 1991 Cuschieri: Laparoscopic myotomy
• 1992 Pellegrini: Thoracoscopic approach

Surgery – Laparoscopic vs. Open

• Laparoscopic approach associated with significant reduction in
  Length of stay
  Post-operative narcotic use
  Time to return to work
• Long term efficacy and LES pressure/relaxation equivalent in retrospective comparison studies
Laparoscopic vs. Thoracoscopic

- Lap approach → better symptom relief and less reflux in 3 large retrospective series
- Thoracoscopic limited by need for single lung ventilation and postop chest tube
- Easier to extend myotomy onto cardia with laparoscopic approach
- Thoracic approach used for hostile abdomen or when need to extend myotomy higher onto esophagus

Surgical Principles

- Goal – decrease LES pressure and improve esophageal emptying
- Transect longitudinal and circular fibers of esophagus, and sling fibers of cardia
- Myotomy should extend 1 to 2 cm on stomach and up to normal esoph muscle
- Cut edges of muscle should be widely separated to prevent reapproximation

Addition of Antireflux Procedure

- Prompted by reports of up to 40% reflux rate after thorascopic approach
- Partial fundoplication
  - Anterior wrap (Dor)
  - Posterior wrap (Toupet)
- No difference in rates of dysphagia or reflux b/w two procedures

Surgical Technique

- Dissection of Gastrohepatic Ligament and Crura
**Surgical Technique**

Performing the Myotomy

**Fundoplication**

Dor

Toupet

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**Complications of Surgery**

- Overall low morbidity
- Esophageal/ gastric mucosal perf (4-5%)
  Usually inconsequential when recognized at time of operation
- Other complications (3%)
  Pneumothorax, bleeding, abscess

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**LHM: Long-Term Results**

<table>
<thead>
<tr>
<th></th>
<th>RHM (n=33)</th>
<th>LHM (n=11)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Follow-up Interval, median (range), years</td>
<td>9.1 (3.9-12.8)</td>
<td>9.9 (4.4-14.8)</td>
<td>0.49</td>
</tr>
<tr>
<td>Dysphagia, No. (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent or mild</td>
<td>26 (79)</td>
<td>8 (80)</td>
<td>1.00</td>
</tr>
<tr>
<td>Moderate or severe</td>
<td>7 (21)</td>
<td>2 (20)</td>
<td></td>
</tr>
<tr>
<td>Heartburn, No. (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent or mild</td>
<td>26 (79)</td>
<td>9 (90)</td>
<td>0.66</td>
</tr>
<tr>
<td>Moderate or severe</td>
<td>7 (21)</td>
<td>1 (10)</td>
<td></td>
</tr>
<tr>
<td>PPI use, No (%)</td>
<td>18 (56.3)</td>
<td>8 (80%)</td>
<td>0.27</td>
</tr>
<tr>
<td>GERD-HRQL score, median (range)</td>
<td>11 (0-36)</td>
<td>12 (0-20)</td>
<td>0.55</td>
</tr>
<tr>
<td>Satisfied, No. (%)</td>
<td>32 (95.5)</td>
<td>10 (90.9)</td>
<td>0.44</td>
</tr>
<tr>
<td>Heller again, No. (%)</td>
<td>30 (90.9)</td>
<td>10 (90.9)</td>
<td>1.00</td>
</tr>
</tbody>
</table>

# LHM: Long-Term Results

<table>
<thead>
<tr>
<th>Primary Author</th>
<th>Patients (No.)</th>
<th>Follow-up (years)</th>
<th>Absent or Mild Dysphagia (%)</th>
<th>Retreatment (%)</th>
<th>Mild to Mod Heartburn (%)</th>
<th>PPI Use (%)</th>
<th>Satisfaction (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cowgill</td>
<td>47</td>
<td>10.6</td>
<td>92</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>92</td>
</tr>
<tr>
<td>Jeansonne</td>
<td>17</td>
<td>11.2</td>
<td>94</td>
<td>17.7</td>
<td>23.5</td>
<td>NR</td>
<td>94</td>
</tr>
<tr>
<td>Kilic</td>
<td>46</td>
<td>6.4</td>
<td>80</td>
<td>20</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Sasaki</td>
<td>34</td>
<td>7.8</td>
<td>100</td>
<td>5.9</td>
<td>0</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Zaninotto</td>
<td>177</td>
<td>10.0</td>
<td>82</td>
<td>9.6</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
</tbody>
</table>

# Per Oral Endoscopic Myotomy

- **Per Oral Endoscopic Myotomy (POEM)**
  - Submucosal plane accessed via esophageal mucosotomy
  - Totally endoscopic procedure
  - Allows complete surgical myotomy under direct visualization

- Has the potential to offer the advantages of both surgical myotomy and endoscopic balloon dilation

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## Per Oral Endoscopic Myotomy

- **Described by Inoue in 2009**
- **Surgical principles**
  - Mucosal flap
  - Submucosal dissection
  - Adequate distal myotomy
  - Secure closure of mucosotomy
- **Supine position**
- **General anesthesia**
- **CO2 insufflation**

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## POEM Technique

- **Inspect and washout esophagus/stomach**
- **Identify GE junction**
- **Inject mucosa at 12 o’clock position 13cm proximal to GE junction**
- **Mucosotomy should be at least 3cm proximal to start of myotomy**
- **Incise mucosa**
- **Use TT knife**
- **Enter submucosal space**
POEM Technique

- Submucosal dissection past 2cm on cardia
- GE junction usually narrows and then widens on gastric side
- The scope length can also be checked to approximate the GE junction
- Important to go back into the stomach lumen and look for the length of dissection
- Transillumination

POEM Technique

- Complete circular myotomy created using electrocautery
- Begin ~6cm above GEJ
  - At least 2cm distal to mucosotomy
  - Extend 2cm onto the gastric cardia

POEM Technique

- Close mucosal incision with clips or suturing device
- Upper GI series POD #0 or 1
- Follow-up: 2 weeks, 6 weeks, 6 months, 1 year

POEM Outcomes

- Between August 2012 and October 2013, 26 patients underwent POEM, 25 for achalasia and 1 long myotomy for diffuse esophageal spasm
- Median age: 54 years
- Median BMI: 28.4 kg/m²
- 14/26 (54%) male
**Operative Experience**

- Successfully completed in all cases
- Mean operative time: 105 ± 30 minutes
- Median of 7 (5-16) endoscopic clips were required for mucosotomy closure
- Pneumoperitoneum requiring Veress needle decompression occurred in 9 (35%) cases
- No intraoperative complications or esophageal leaks
- Median hospital stay: 1 (1-2) day

**Dysphagia**

**Dysphagia Score**

<table>
<thead>
<tr>
<th>Preoperative</th>
<th>6 weeks postop</th>
<th>6 months postop</th>
</tr>
</thead>
<tbody>
<tr>
<td>4</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

**Disease Specific QoL**

**GERD-HRQL**

<table>
<thead>
<tr>
<th>preop</th>
<th>6 weeks postop</th>
<th>6 months postop</th>
</tr>
</thead>
<tbody>
<tr>
<td>20</td>
<td>15</td>
<td>10</td>
</tr>
</tbody>
</table>

**Global QoL: 6 Months**

- Physical Function
- Physical Role Limitation
- Emotional Role Limitation
- Energy/Fatigue
- General Health
- Social Functioning
- Emotional Well Being
POEM vs. LHM

### Population: Preoperative Descriptors

<table>
<thead>
<tr>
<th></th>
<th>Heller n = 64</th>
<th>POEM n = 37</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, mean (SD), yr</td>
<td>57 (20)</td>
<td>56 (16)</td>
<td>0.7</td>
</tr>
<tr>
<td>Female, %</td>
<td>52</td>
<td>48</td>
<td>0.8</td>
</tr>
<tr>
<td>Eckardt Score, mean (SD)</td>
<td>5.9 (2.4)</td>
<td>5.4 (2.2)</td>
<td>0.5</td>
</tr>
<tr>
<td>Manometry, median (mm Hg)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Voltage pressure</td>
<td>37</td>
<td>41</td>
<td>0.2</td>
</tr>
<tr>
<td>Distal esophageal contraction amplitude</td>
<td>32</td>
<td>29</td>
<td>0.9</td>
</tr>
<tr>
<td>Heartburn</td>
<td>19</td>
<td>10</td>
<td>0.3</td>
</tr>
<tr>
<td>Dysphagia to solid</td>
<td>62</td>
<td>34</td>
<td>0.05</td>
</tr>
<tr>
<td>Dysphagia to liquid</td>
<td>49</td>
<td>30</td>
<td>0.4</td>
</tr>
<tr>
<td>Reflux</td>
<td>45</td>
<td>17</td>
<td>0.01</td>
</tr>
<tr>
<td>Chest pain</td>
<td>23</td>
<td>13</td>
<td>0.6</td>
</tr>
</tbody>
</table>


### POEM vs. LHM: Operative Data

<table>
<thead>
<tr>
<th></th>
<th>Heller n = 64</th>
<th>POEM n = 37</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Operative time, min</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median</td>
<td>180</td>
<td>120</td>
<td>0.003</td>
</tr>
<tr>
<td>Range</td>
<td>100-280</td>
<td>60-215</td>
<td></td>
</tr>
<tr>
<td>Full-thickness injury, n</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Esophagus</td>
<td>11</td>
<td>4</td>
<td>0.1</td>
</tr>
<tr>
<td>Stomach</td>
<td>0</td>
<td>0</td>
<td>0.8</td>
</tr>
<tr>
<td>Return to the OR, n</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bleeding</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Length of stay, mean days (SD)</td>
<td>2.5 (1.5)</td>
<td>1.1 (0.6)</td>
<td>&lt;0.0001</td>
</tr>
</tbody>
</table>

OR indicates operating room.


### POEM vs. LHM: Acid Reflux

<table>
<thead>
<tr>
<th></th>
<th>Heller n = 31 (48%)</th>
<th>POEM n = 23 (76%)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>DeMeester score, median</td>
<td>4</td>
<td>4</td>
<td>0.2</td>
</tr>
<tr>
<td>DeMeester score ≥14.7 %</td>
<td>10</td>
<td>9</td>
<td>0.4</td>
</tr>
<tr>
<td>Number of reflux episodes, median</td>
<td>4</td>
<td>12</td>
<td>0.4</td>
</tr>
<tr>
<td>Testing &gt;6 mo after surgery</td>
<td></td>
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</tr>
</tbody>
</table>


### POEM vs. LHM: Conclusions

- POEM is less invasive than LHM and enhances recovery with decreased pain, shorter length of hospital stay, and faster return to work
- Similar efficacy and post-operative reflux compared to LHM at 1 year
### POEM Outcomes
- 18 patients with 1 year follow-up
  - 3 intraoperative perforations managed with clips
  - Durable dysphagia relief at 11 months in all patients
  - 46% had positive pH study at 6 months

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### POEM: What’s next?
- Procedure has proven safe and effective in the short-term
  - \(\rightarrow\) Comparison of outcomes with pneumatic dilation and laparoscopic myotomy (dysphagia relief and reflux)
  - \(\rightarrow\) Long-term studies to assess durability and cost-effectiveness

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### Persistent or Recurrent Dysphagia
- Seen in 8 to 13%
- DDX includes incomplete myotomy, CA, stricture from reflux, severe dysmotility
- Evaluation similar to achalasia workup
- Treatment may include dilation or repeat myotomy (70-80% successful)

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### Esophagectomy
- Required in 1 to 2% of cases
- Indications: tortuous megaesophagus, failure of myotomy, stricture from reflux
- Orringer et. al. reported on 93 patients
  - 10% leak rate/ 2% mortality
  - 50% required dilation of anastomosis
  - 95% eating well
## Summary

- Rare disorder of esophageal motility
- Progressive and debilitating
- Treatment aimed at reducing LES pressure and improving esophageal clearance
- Myotomy and dilation most effective trx
- Laparoscopic approach favored, endoscopic emerging
- Addition of antireflux procedure controversial
- Esophagectomy rarely required