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

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## Achalasia

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

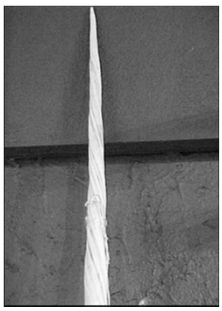
## Epidemiology

- **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**

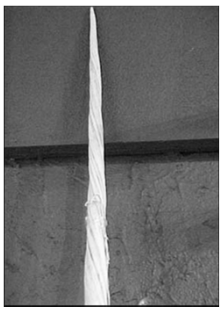
## Cancer Risk

- **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



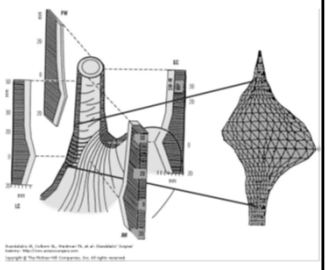
## Historical Perspective



**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, valsalva**

  **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**

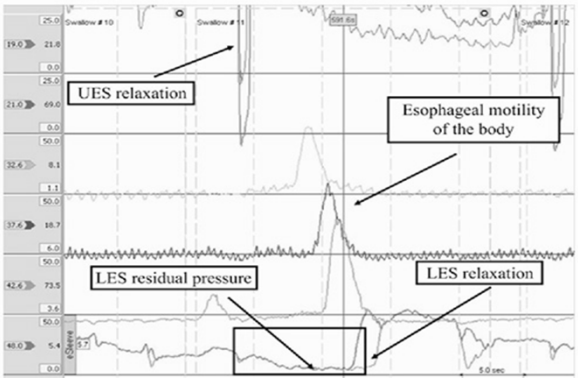
## Evaluation

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

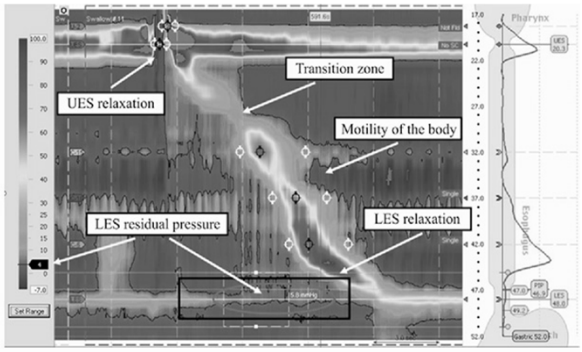
# Esophageal Manometry

- **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



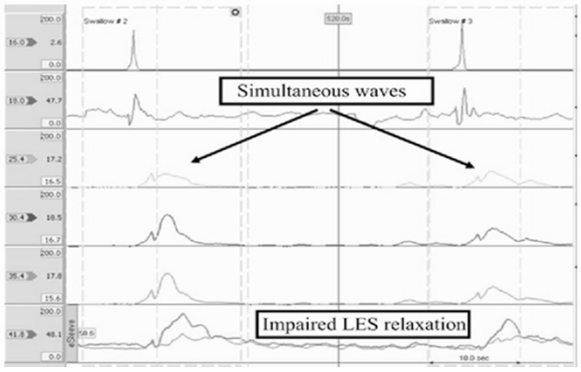

# Normal High-Res Manometry



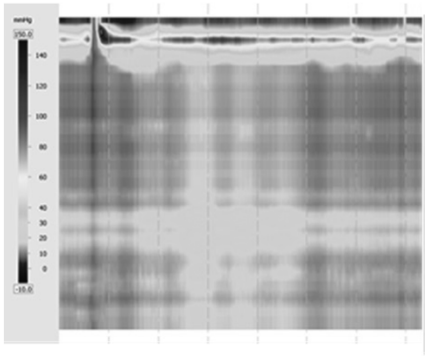

# Manometry in Achalasia

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

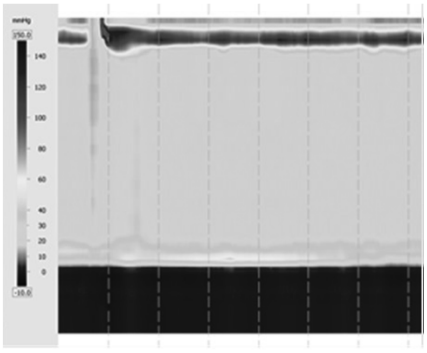
## Manometry



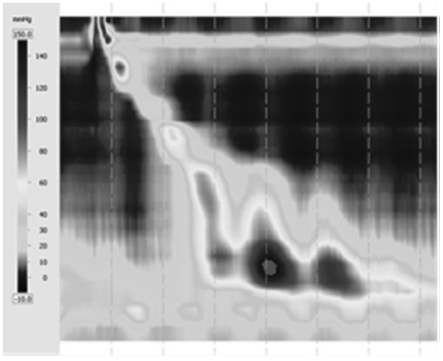

## High-Res Manometry: Type I Achalasia



## High-Res Manometry: Type II Achalasia



## High-Res Manometry: Type III Achalasia



## Barium Swallow

- Dilated, possibly tortuous esophagus (late finding)
- "Bird's beak" tapered appearance of distal esophagus
- Air fluid level with retained food

## Endoscopy

- Necessary to rule out malignancy
- Requires retroflexed view of cardia/GEJ
- Scope should pass through easily – if not, consider malignancy
- EUS may help to evaluate esophageal wall when CA suspected

## pH Study

- May help when diagnosis is in question, especially when heartburn is major sx
- Interpretation may be difficult due to fermentation
- Achalasia shows gradual decline in pH *vs.* rapid drop seen with GERD

## Management

- Pharmocologic
- Botox injection
- Pneumatic dilation
- 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

Campos, GM et al. Ann Surg 2009; 249:45-57

## Pneumatic Esophageal Dilation

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

## Pneumatic Esophageal Dilation

- 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.

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

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## 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: Thoracosopic 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**

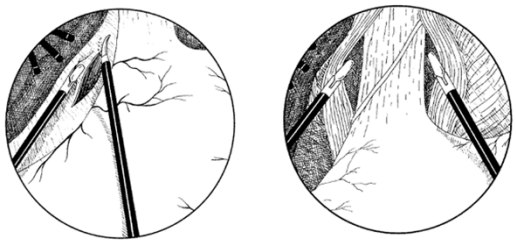
## Surgical Principles

- **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

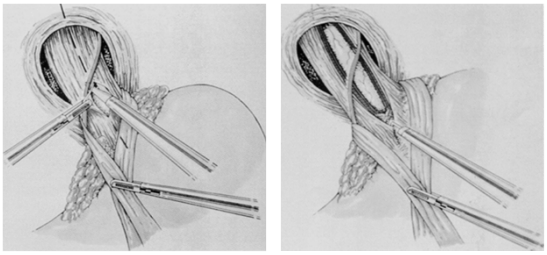
- **Prompted by reports of up to 40% reflux rate after thorascopic approach**
- **Partial fundoplication**
  **Anterior wrap (Dor)**
  **Posterior wrap (Toupet)**
- **No difference in rates of dysphagia or reflux b/w two procedures**

## Surgical Technique



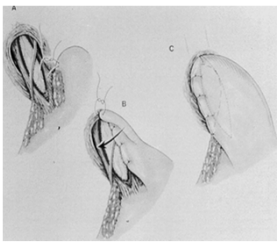
**Dissection of Gastrohepatic Ligament and Crura**

# Surgical Technique

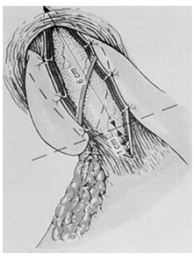


Performing the Myotomy

# Fundoplication



Dor



Toupet

# 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**

# LHM: Long-Term Results

| | RHM (n=33) | LHM (n=11) | p-value |
|---|---|---|---|
| **Follow-up Interval, median (range), years** | 9.1 (3.9-12.8) | 9.9 (4.4-14.8) | 0.49 |
| **Dysphagia, No. (%)** | | | |
| **Absent or mild** | **26 (79)** | **8 (80)** | 1.00 |
| **Moderate or severe** | 7 (21) | 2 (20) | |
| **Heartburn, No. (%)** | | | |
| **Absent or mild** | 26 (79) | 9 (90) | 0.66 |
| **Moderate or severe** | 7 (21) | 1 (10) | |
| **PPI use, No (%)** | **18 (56.3)** | **8 (80%)** | 0.27 |
| **GERD-HRQL score, median (range)** | 11 (0-36) | 12 (6-20) | 0.55 |
| **Satisfied, No. (%)** | **32 (95.5)** | **10 (90.9)** | 0.44 |
| Heller again, No. (%) | 30 (90.9) | 10 (90.9) | 1.00 |

Perry et. al. Surg Endosc 2014; DOI: 10.1007/s00464-014-3576-9.

## LHM: Long-Term Results

| Primary Author | Patients (No.) | Follow-up (years) | Absent or Mild Dysphagia (%) | Retreatment (%) | Mild to Mod Heartburn (%) | PPI Use (%) | Satisfaction (%) |
|---|---|---|---|---|---|---|---|
| **Cowgill** | 47 | 10.6 | **92** | 12.8 | NR | NR | **92** |
| **Jeansonne** | 17 | 11.2 | **94** | 17.7 | 23.5 | NR | **94** |
| **Kilic** | 46 | 6.4 | **80** | 20 | NR | NR | NR |
| **Sasaki** | 34 | 7.8 | **100** | 5.9 | 0 | NR | NR |
| **Zaninotto** | 177 | 10.0 | **82** | 9.6 | NR | NR | NR |

## 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***

## Per Oral Endoscopic Myotomy

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

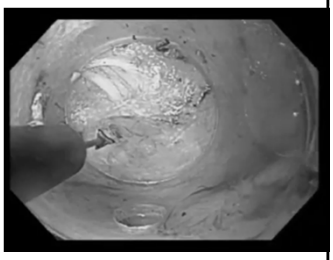
**Inoue H et al. Endoscopy. 2010;42:265-71.**

## 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**

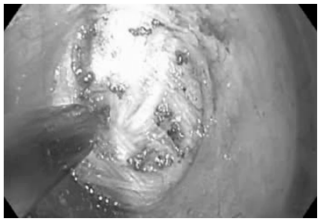
# POEM Technique

- 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



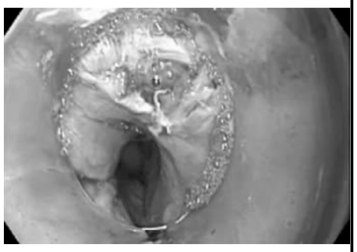
# POEM Technique

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



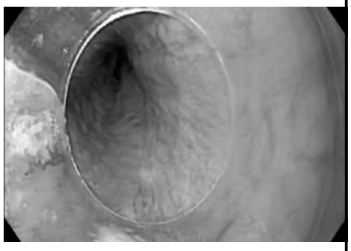
# POEM Technique

- 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

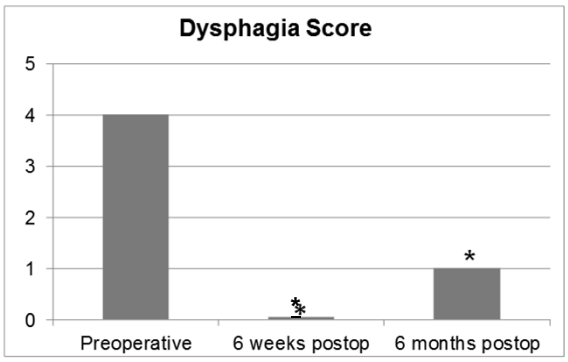
- Between August 2012 and October 2013, 26 patients underwent POEM, 25 for achalasia and 1 long myotomy for diffuse esophageal spasm
  - Median age: 54 years
  - Median BMI: 28.4 kg/m²
  - 14/26 (54%) male



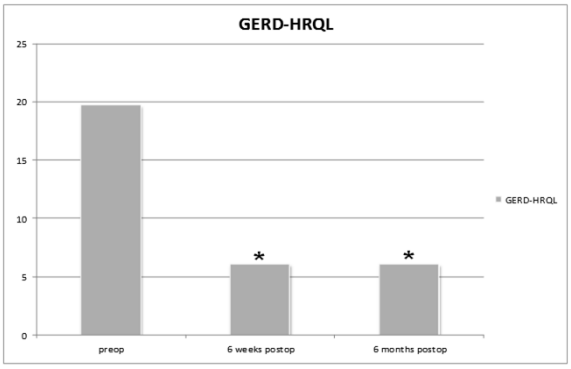
## Operative Experience

- **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**

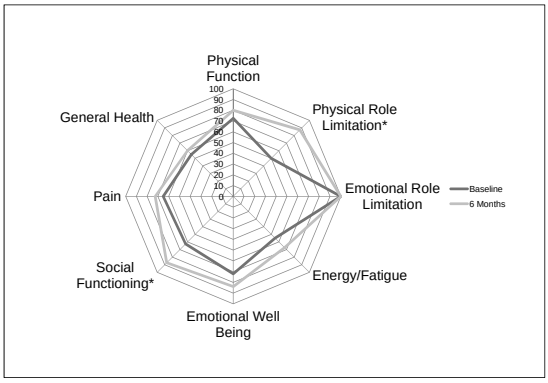
## Dysphagia




## Disease Specific QoL




## Global QoL: 6 Months



# POEM vs. LHM

**Population: Preoperative Descriptors**

| | Heller n = 64 | POEM n = 37 | *P* |
|---|---|---|---|
| Age, mean (SD), yr | 57 (20) | 56 (16) | 0.7 |
| Female, % | 52 | 48 | 0.8 |
| Eckardt Score, mean (SD) | 5.9 (2.4) | 5.4 (2.2) | 0.5 |
| Manometry, median (mm Hg) | | | |
| Resting pressure | 37 | 41 | 0.2 |
| Relaxation pressure | 20 | 19 | 0.4 |
| Distal esophageal contraction amplitude | 32 | 29 | 0.9 |
| Symbol score ≥2, % | | | |
| Heartburn | 19 | 10 | 0.5 |
| Dysphagia to solid | 62 | 32 | 0.06 |
| Dysphagia to liquid | 49 | 30 | 0.4 |
| Reflux | 45 | 17 | 0.01 |
| Chest pain | 23 | 13 | 0.6 |

Bhayani NH et al. Ann Surgery 2014;259:1098-1103

# POEM vs. LHM: Operative Data

**Operative Details**

| | Heller n = 64 | POEM n = 37 | *P* |
|---|---|---|---|
| Operative time, min | | | |
| Median | 160 | 120 | 0.003 |
| Range | 100-280 | 60-215 | |
| Full-thicknesss injury, n | | | |
| Esophagus | 8 | 4 | 0.1 |
| Stomach | 3 | 0 | 0.8 |
| Return to the OR, n | | | |
| Bleeding | 1 | 1 | |
| Length of stay, mean days (SD) | 2.5 (1.9) | 1.1 (0.6) | <0.0001 |
| | | | |
| OR indicates operating room. | | | |

Bhayani NH et al. Ann Surgery 2014;259:1098-1103

# POEM vs. LHM: Acid Reflux

**Long-Term pH Testing**

| | Heller n = 31 (48%) | POEM n = 23 (76%) | *P* |
|---|---|---|---|
| DeMeester score, median | 2 | 4 | 0.2 |
| DeMeester score ≥14.7,% | 10 | 9 | 0.4 |
| Number of reflux episodes, median | 4 | 12 | 0.4 |
| | | | |
| Testing >6 mo after surgery | | | |
| | | | |
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Bhayani NH et al. Ann Surgery 2014;259:1098-1103

# 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

Swanstrom, et al. Ann Surg 2012

## POEM: What's next?

- Procedure has proven safe and effective in the short-term
- → Comparison of outcomes with pneumatic dilation and laparoscopic myotomy (dysphagia relief and reflux)
- → Long-term studies to assess durability and cost-effectiveness

## 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)

## 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**