

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**

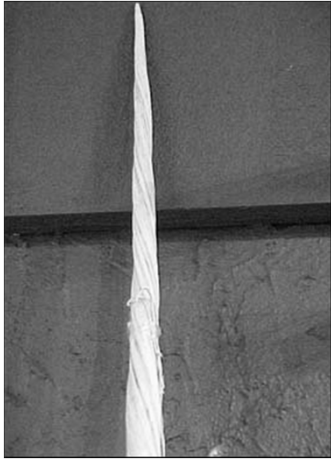
Epidemiology

- **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**

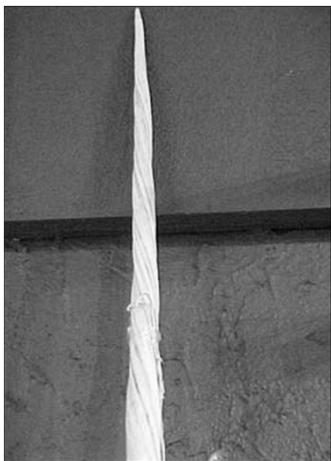
Cancer Risk

- **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

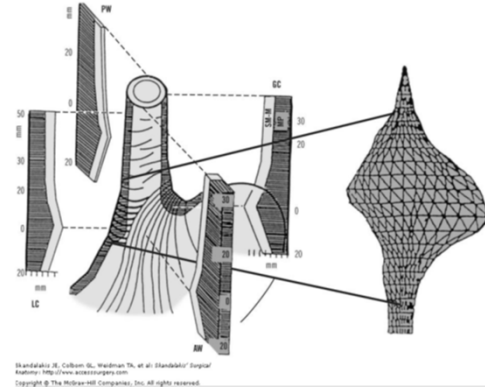


Historical Perspective



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

- 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



- **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% 2° 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**

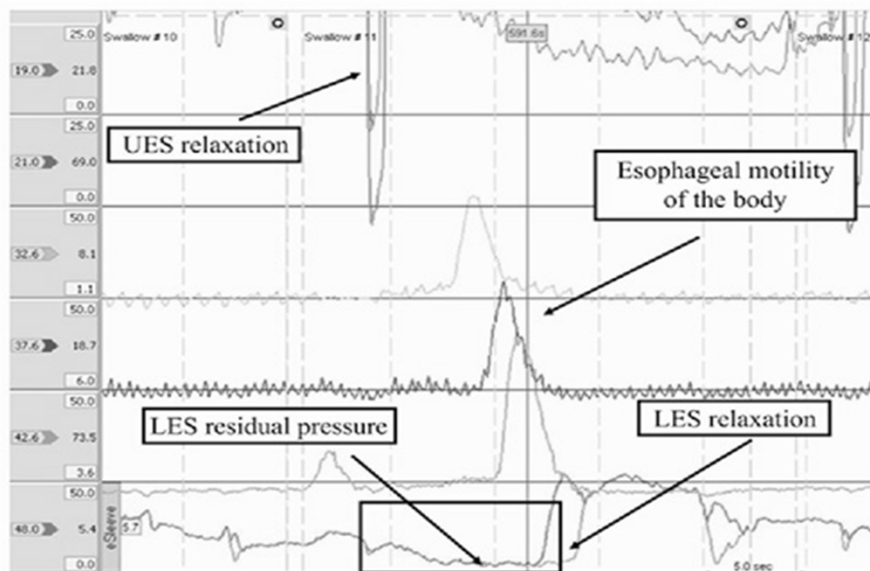
Evaluation

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

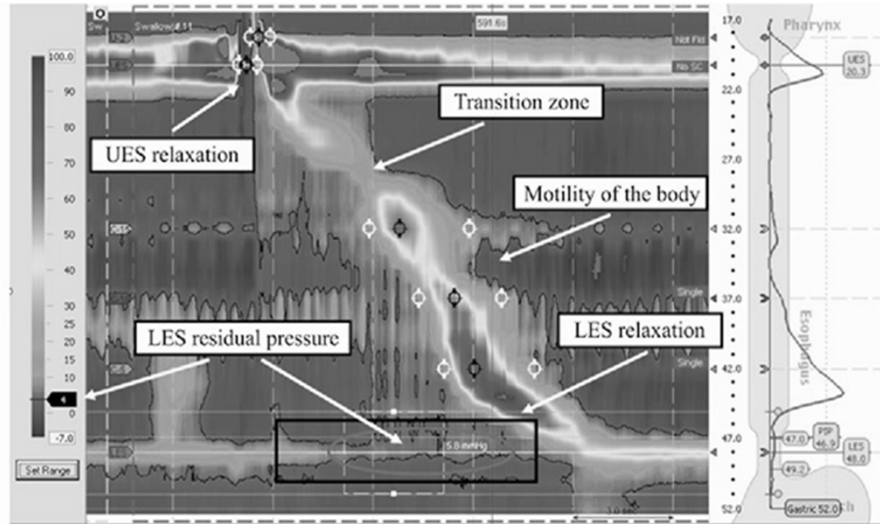
Esophageal Manometry

- 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



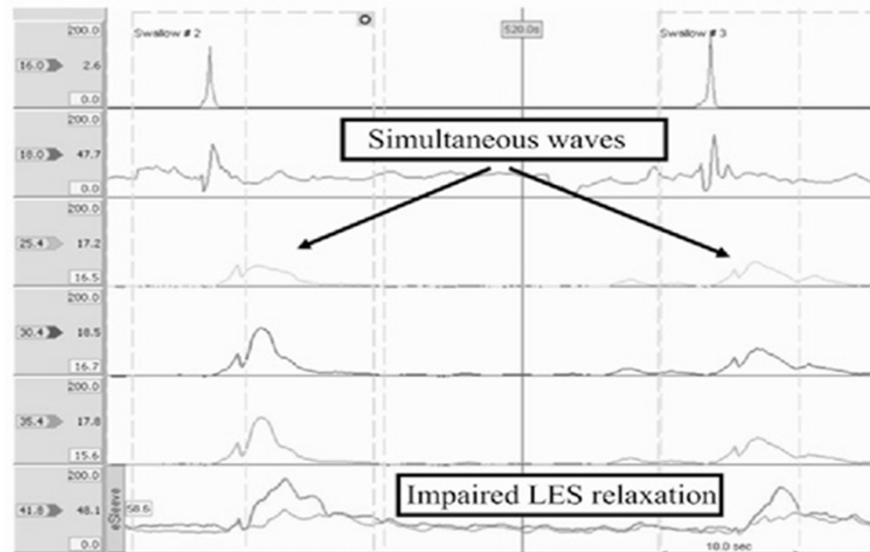
Normal High-Res Manometry



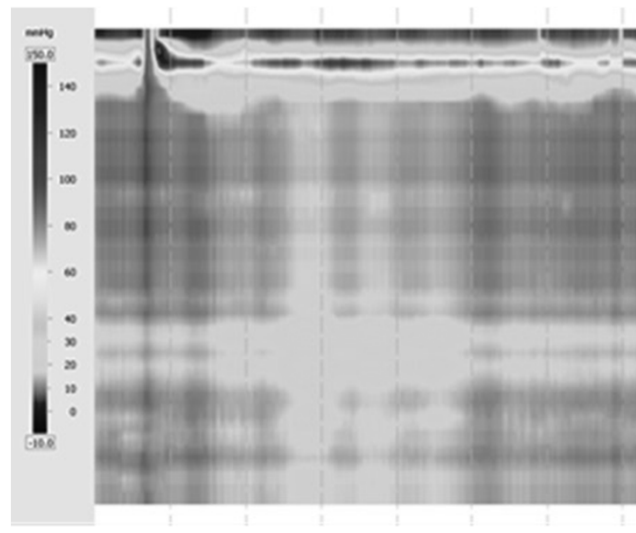
Manometry in Achalasia

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

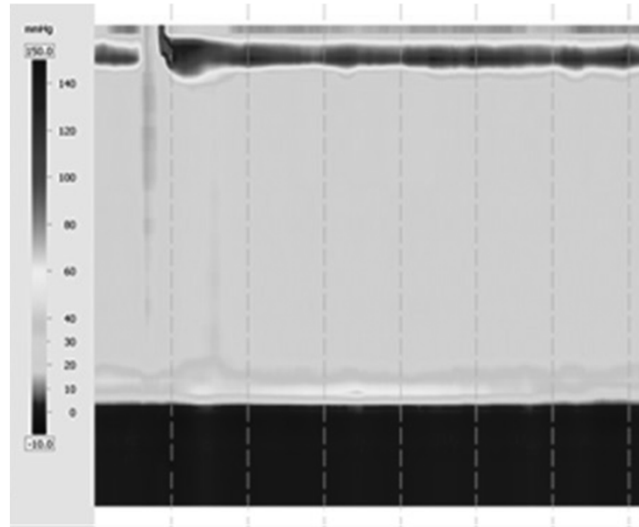
Manometry



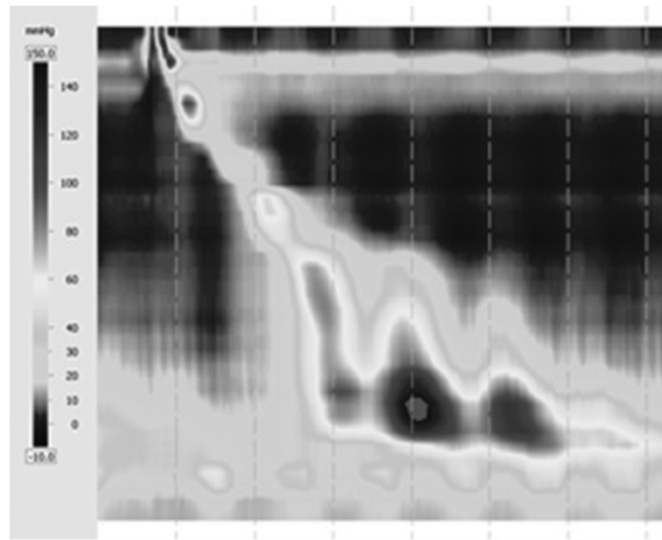
High-Res Manometry: Type I Achalasia



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

- **Pharmacologic**
- **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

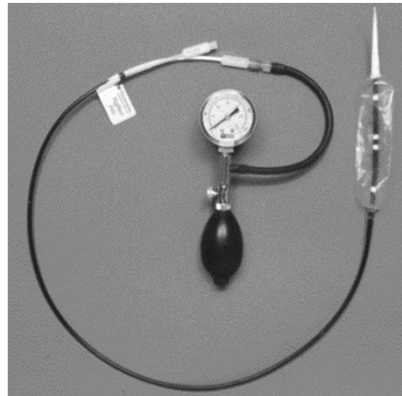
Treatment Strategies

- 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

- | | |
|---|---|
| <ul style="list-style-type: none">• Dilation<ul style="list-style-type: none">• outpatient procedure• minimal pain• Rapid return to work• May treat any patient population (frail, pregnant, ect...)• Less expensive• Does not preclude myotomy | <ul style="list-style-type: none">• Myotomy<ul style="list-style-type: none">• 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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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**

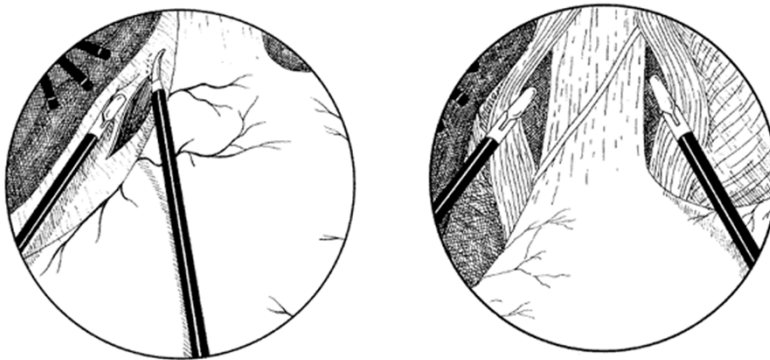
Surgical Principles

- **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

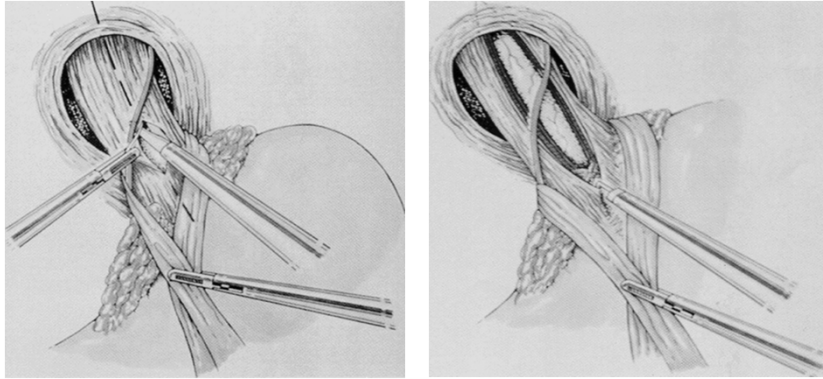
- Prompted by reports of up to 40% reflux rate after thorascopic approach
- Partial fundoplication
 - Anterior wrap (Dor)
 - Posterior wrap (Toupet)
- No difference in rates of dysphagia or reflux b/w two procedures

Surgical Technique



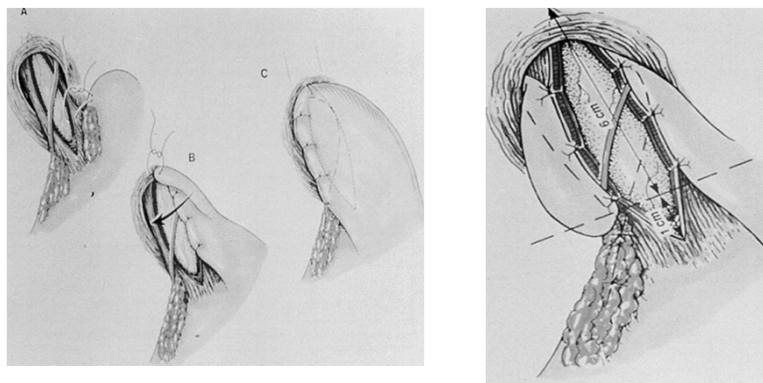
Dissection of Gastrohepatic Ligament and Crura

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

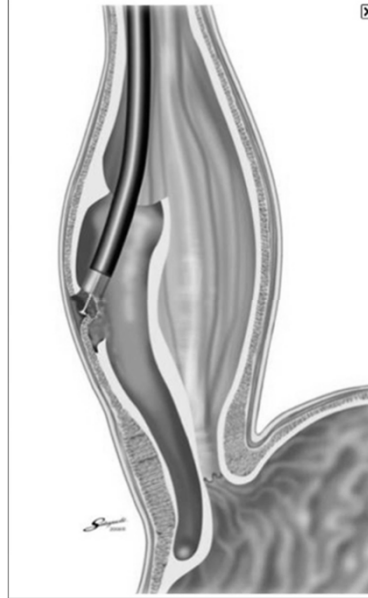
Primary Author	Patients (No.)	Follow-up (years)	Absent or Mild Dysphagia (%)	Retreatment (%)	Mild to Mod Heartburn (%)	PPI Use (%)	Satisfaction (%)
Cowgill	47	10.6	92	12.8	NR	NR	92
Jeansonne	17	11.2	94	17.7	23.5	NR	94
Kilic	46	6.4	80	20	NR	NR	NR
Sasaki	34	7.8	100	5.9	0	NR	NR
Zaninotto	177	10.0	82	9.6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

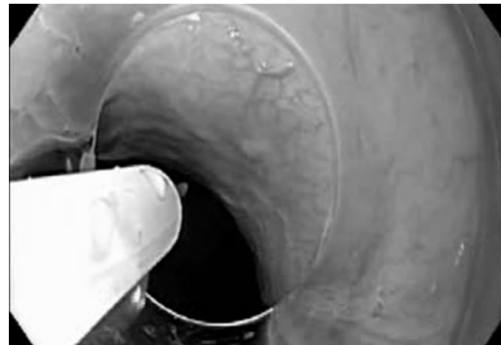
- Described by Inoue in 2009
- Surgical principles
 - Mucosal flap
 - Submucosal dissection
 - Adequate distal myotomy
 - Secure closure of mucosotmy
- Supine position
- General anesthesia
- CO2 insufflation



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

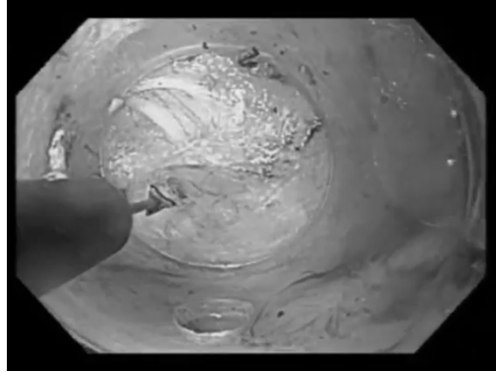
POEM Technique

- 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



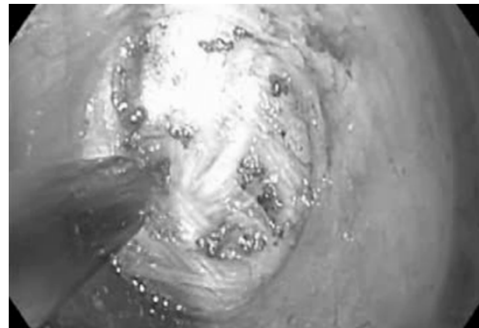
POEM Technique

- 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



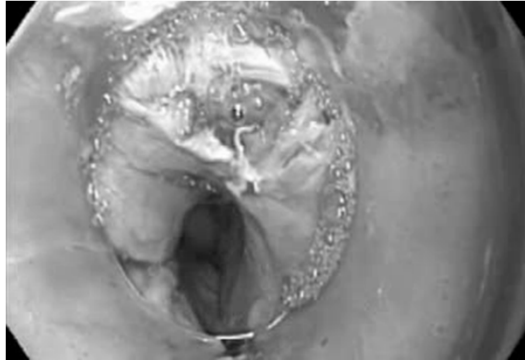
POEM Technique

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



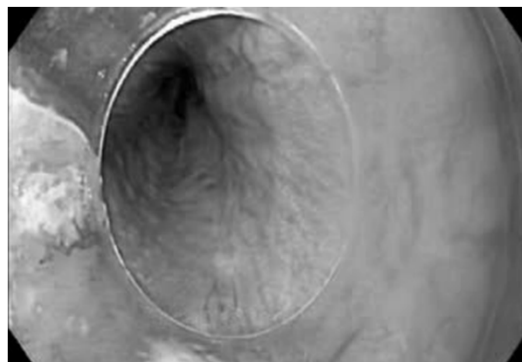
POEM Technique

- 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

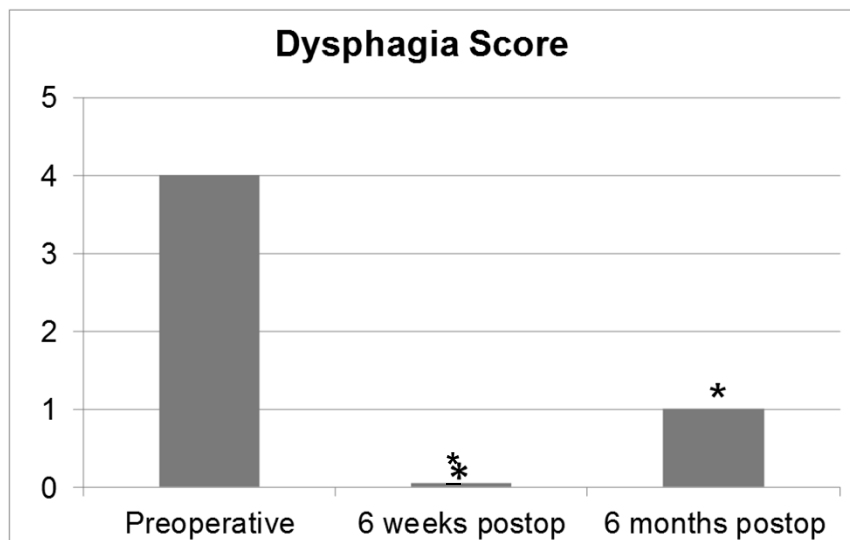
- Between August 2012 and October 2013, 26 patients underwent POEM, 25 for achalasia and 1 long myotomy for diffuse esophageal spasm
 - Median age: 54 years
 - Median BMI: 28.4 kg/m²
 - 14/26 (54%) male



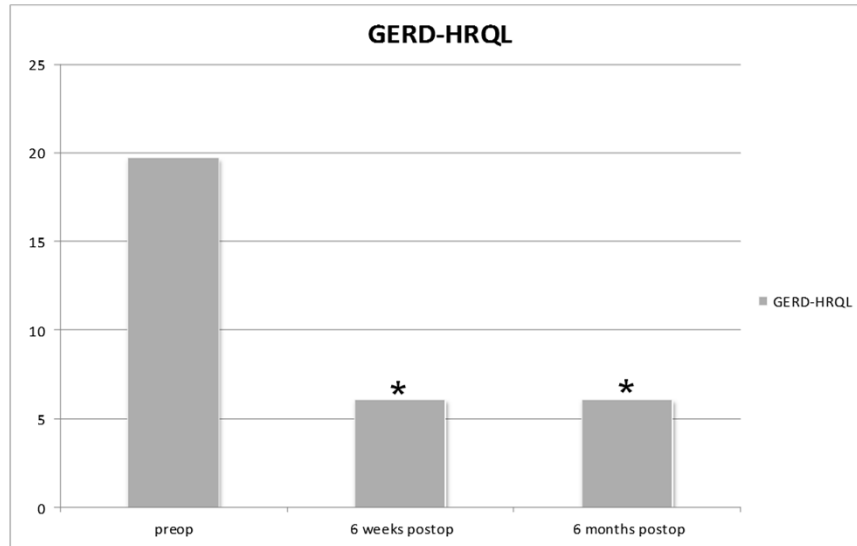
Operative Experience

- 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

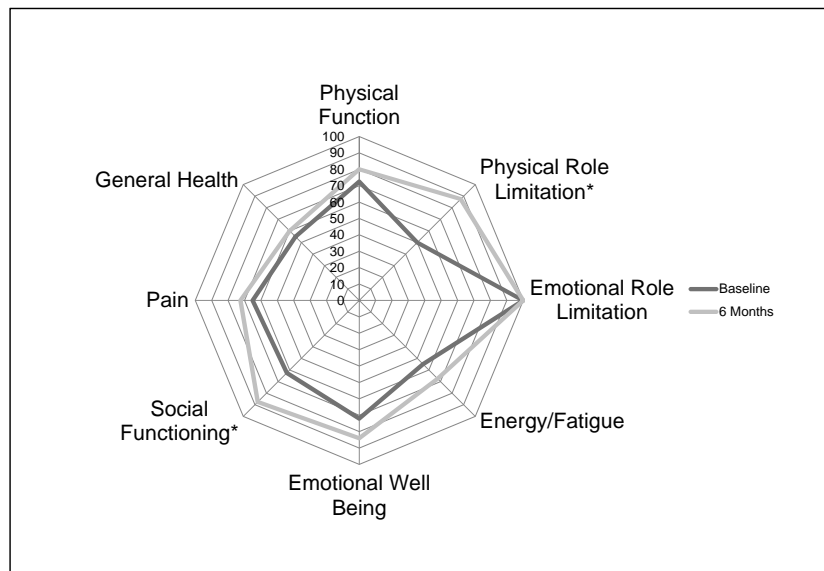
Dysphagia



Disease Specific QoL



Global QoL: 6 Months



POEM vs. LHM

Population: Preoperative Descriptors

	Heller n = 64	POEM n = 37	<i>P</i>
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	<i>P</i>
Operative time, min			
Median	160	120	0.003
Range	100-280	60-215	
Full-thickness 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			

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**