Case Presentation

Joel A. Ricci MD
SUNY Downstate Medical Center
Lutheran Medical Center
Department of Surgery
June 26, 2009
Xx year old female with worsening dysphagia and solid food regurgitation for 2 days

Other symptoms included: poor appetite and occasional epigastric tenderness

Denied weight loss, fever, dysuria or changes in bowel habits

Known hx of Achalasia (2006): refused Tx at the time

PMHx: NIDDM, HTN, hyperlipidemia, GERD?

No Tobacco or EtOH use

Meds: Lipitor, Glipizide, Zolpidem, Toprol XL, Prilosec
Physical Exam

- Vital Signs: T: 97.8°F, BP: 169/81 mmHg, HR: 100 b/m
- Gen: AAO x 3, NAD
- HEENT: PERRL, moist mucous membranes, no icterus
- CV: RRR, S₁S₂, no murmurs or gallops
- Lungs: CTA b/l
- Abd: Soft, NT/ND, +BS
- Ext: 2+ pulses throughout, no edema

* Labs were unremarkable
Imaging

- Chest x-ray: Large bullae in Right upper lobe extending into lower neck

- Chest CT Scan: Diffusely dilated esophagus with food residues and mildly compressed airway
Chest X-Ray
CT Scan
CT Scan
CT Scan
Procedure

- Thoracoscopic Heller myotomy with intra-operative Esophago-gastroduodenoscopy (EGD)
  - EGD: Dilated esophagus with spastic LES
  - Left side decubitus
  - 4 VATS incisions
  - Adhesiolysis; Esophagus encircled w/ penrose drain
  - Narrowed tapering of LES visualized
  - Muscular layer divided w/ scissors 5 cm onto proximal esophagus and 2 cm beyond the GEJ
  - EGD confirmed adequate passage
  - Gas insufflation confirmed no mucosal perforation
Post-op Course

- **POD #1:**
  - UGI Series: No obstruction or leakage
  - Tolerated clear liquids

- **POD #2:**
  - Tolerated full liquids
  - Discharged home

- Currently (4 months post-op):
  - Adequate relief of dysphagia, no complaints
Upper GI Series
Upper GI Series
Upper GI Series
Esophageal Motility Disorders

Joel A. Ricci M.D.
SUNY Downstate Medical Center
Department of Surgery
June 26, 2009
Esophageal Disease

- Motility Disorders
- GERD
- Barrett’s Esophagus
- Esophageal Cancer

See previous presentations by yours truly

www.downstatesurgery.org
Motility Disorders

*Classification based on manometry*

- Achalasia
  - *Inadequate LES relaxation*
- Diffuse Esophageal Spasm
  - *Uncoordinated contraction*
- Nutcracker Esophagus
  - *Hypercontraction*
- Ineffective Esophageal Motility
  - *Hypocontraction*

*Spechler et al. Gut 49:145-151, 2001*
Achalasia

Symptomatology

- Progressive dysphagia
  - Liquids to solids
- Chest pain
- Aspiration
- Regurgitation
- Weight loss
Achalasia

Etiology

- Complex motor abnormality of the LES
- 6 in 100,000 individuals
- 2nd most common functional disorder of the esophagus requiring surgery (GERD = 1st)
- Cause is unknown: data suggests different theories
  - Hereditary
  - Degenerative
  - Autoimmune
  - Infectious
Achalasia

Pathophysiology

- T-lymphocyte, eosinophil, and mast cell infiltration in the myenteric (Auerbach) plexus
  - Myenteric neural fibrosis
  - Hypertrophy of the two muscle layers and nerve fibers
  - Degeneration of NO and producing inhibitory neurons

Affects relaxation of LES  Basal LES pressure rises
Achalasia

**Manometry**

- Manometric features:
  - Incomplete LES relaxation
  - Elevated resting pressure (>45 mmHg)
  - Aperistalsis of esophageal body
  - Elevated lower esophageal pressure
Achalasia

Work-up

- Chest X-ray
  - Absence of gastric bubble
  - Dilated fluid filled esophagus
  - Right side posterior mediastinal shadow
Achalasia

Work-up

- Barium Swallow
  - Air fluid level
  - “Bird’s beak”

- Fluoroscopic imaging
  - Flaccid non-peristaltic esophagus
  - Absence of “stripping” waves
Achalasia

Work-up

- EGD
  - Narrowed distal lumen
  - “Stuck” solid food particles
  - Rule out “pseudo-achalasia” caused by obstructing tumor in distal esophagus
Achalasia

Treatment

- **Pharmacologic Treatment**
  - **Isosorbide dinitrate**
    - Reduces LES 66% for 90 minutes
  - **Nifedipine**
    - Reduces LES pressure 30-40% for > 60 minutes
- **Botulinum Toxin Injection**
  - Inhibits acetylcholine release
  - 60 – 80% relief
  - 50% recurrence within 6 months
  - Obliterates plane btwn mucosa & submucosa
  - Increased rate of perforation during surgery
Achalasia

Treatment

- **Pneumatic Dilatation**
  - Success increases with repeat dilatations
  - 60-80% success rate; 5yr recurrence rate: 50%

Achalasia

Management Algorithm

Clinical symptoms (dysphagia, regurgitation, weight loss)

Barium swallow

Manometry

Low-surgical-risk patient

Laparoscopic myotomy

Failure

Repeat myotomy or esophagectomy

Success

Pneumatic dilation

Failure

Repeat as needed

Success

Botox injection

Success

Nifedipine or isosorbide

High-surgical-risk patient (or refuses surgery)
Achalasia

*Surgical Treatment*

- **Heller Myotomy**
  - Thoracoscopic
  - Laparoscopic
- **Fundoplication**
  - GERD prevention
  - Nissen
  - Dor
  - Toupet
Achalasia

Which treatment is better?

- **Zaninotto et al**: Randomized controlled trial
  - Botox (n = 40) vs Myotomy w/ fundoplication (n = 40)
  - Both groups initial improvement of symptoms
  - 6 months: 45% recurrence in Botox group
  - 2 years (symptom free): 87% myotomy; 34% Botox

- **Csendes et al**: Prospective randomized trial
  - Pneumatic dilatation (n = 20) vs Myotomy (n = 18)
  - 3.5 years (no dysphagia): 100% myotomy
  - 60% pneumatic dilatation

Achalasia

Heller Myotomy

- Ernest Heller; 1913
  - Both ant & post LES muscle fibers disrupted
- Modified version
  - Single, anterior, longitudinal myotomy
- Standard operative technique
Achalasia

Thoracoscopic Esophagomyotomy

Port placement

Division of muscle fibers
Achalasia

*Laparoscopic Heller Myotomy*

**Patient positioning**

**Port placement**
Achalasia

*Laparoscopic Heller Myotomy*

- Identification of LES
- Division of muscle fibers
Achalasia

*Fundoplication*

Dor (Anterior fundoplication)  Toupet (Posterior fundoplication)
Achalasia

Extent of Myotomy

- From GEJ: Proximal (5 – 6 cm); Distal (1.5 – 3 cm)

- Chen et al: 7 to 16 years post myotomy and fundoplication
  - 67% incidence of epiphrenic pseudo-diverticulum
  - Likely caused by absence of coverage over proximal extent of myotomy

- Oelschlager et al: Standard (1.5 cm) vs Extended (3 cm) distal myotomy (n = 110 pts)
  - Lower post-op LES pressures with extended (9.5 mmHg) vs standard (15.8 mmHg)
  - Improved dysphagia
  - 24-hr pH monitoring: no increase in GERD

Achalasia

Addition of Fundoplication

- Myotomy:
  - Lowers esophageal outflow resistance
  - Improves esophageal emptying
  - Increases propensity for GERD

Is Fundoplication always needed?
Achalasia

Addition of Fundoplication

- Richards et al: Randomized trial
  - Heller myotomy w/ Dor vs w/o Dor (n = 43)
  - Post-op GERD (by 24-hr pH monitoring)
    - 47.5% in pts w/ Heller myotomy alone
    - 9.1% in pts w/ added Dor fundoplication
    - No difference in LES pressure or dysphagia scores

- Rice et al: Retrospective study
  - Heller with and w/o Dor (n = 149)
  - Decreased incidence of GERD (by 24-hr pH monitoring) following fundoplication
  - Fundoplication did not decrease esophageal emptying time (assessed by barium esophagography)

Achalasia

Which Fundoplication?

- Complete fundoplication (Nissen) should be avoided due to aperistaltic esophagus
- Partial: Dor (anterior) versus Toupet (posterior)
  - Minimal difference
  - Arain et al: No difference in relief of
    - Dysphagia, Heartburn, Chest pain
    - Need of proton pump inhibitors
- Advantage Dor
  - Technically easier
  - Preservation of natural posterior attachments

Achalasia

Thoracoscopic vs Laparoscopic

- Patti et al: Thoracoscopic Heller myotomy versus Laparoscopic Heller w/ Dor fundoplication
  - 60 pts (30 in each group)
  - Avg hospital stay: 84 hrs (thorac.); 42 hrs (lap)
  - Relieved dysphagia: 87% vs 90%
  - Abnormal reflux (by pH monitoring)
    - 60% thoracoscopic pts
    - 10% laparoscopic/fundoplication pts

# Achalasia

**Outcomes following Laparoscopic Heller Myotomy**

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Patients</th>
<th>Follow-up months</th>
<th>Relief of Dysphagia (%)</th>
<th>Length of Stay (median)</th>
<th>Perforation (%)</th>
<th>Reflux (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Portale</td>
<td>2005</td>
<td>248</td>
<td>43 (1–131)</td>
<td>88</td>
<td>5 (3–11)</td>
<td>4.0</td>
<td>7*</td>
</tr>
<tr>
<td>Bonatti</td>
<td>2005</td>
<td>75</td>
<td>64 (10–131)</td>
<td>84</td>
<td>2 (1–6)</td>
<td>4.0</td>
<td>11</td>
</tr>
<tr>
<td>Khajanchee</td>
<td>2005</td>
<td>121</td>
<td>9 (6–48)</td>
<td>91</td>
<td>1.7 (na)</td>
<td>6.6</td>
<td>13*</td>
</tr>
<tr>
<td>Arain</td>
<td>2004</td>
<td>78</td>
<td>24 (6–100)</td>
<td>77</td>
<td>na</td>
<td>0</td>
<td>17</td>
</tr>
<tr>
<td>Perrone</td>
<td>2004</td>
<td>100</td>
<td>26 (6–72)</td>
<td>96</td>
<td>1.2 (1–4)</td>
<td>3.0%</td>
<td>na</td>
</tr>
<tr>
<td>Oelschlager</td>
<td>2003</td>
<td>110</td>
<td>26 (1–85)</td>
<td>90</td>
<td>na</td>
<td>na</td>
<td>23</td>
</tr>
<tr>
<td>Donahue</td>
<td>2002</td>
<td>81</td>
<td>45 (1–70)</td>
<td>84</td>
<td>1 (na)</td>
<td>14.0%</td>
<td>4</td>
</tr>
<tr>
<td>Sharp</td>
<td>2002</td>
<td>100</td>
<td>11 (na)</td>
<td>93</td>
<td>1.5 (na)</td>
<td>8.0%</td>
<td>4</td>
</tr>
<tr>
<td>Patti</td>
<td>2001</td>
<td>102</td>
<td>25 (na)</td>
<td>89</td>
<td>1.5 (na)</td>
<td>5.0%</td>
<td>na</td>
</tr>
<tr>
<td>Zaninotto</td>
<td>2001</td>
<td>113</td>
<td>24 (1–83)</td>
<td>91</td>
<td>na</td>
<td>na</td>
<td>6</td>
</tr>
<tr>
<td>Patti</td>
<td>1999</td>
<td>133</td>
<td>23 (na)</td>
<td>89</td>
<td>2</td>
<td>5.0%</td>
<td>17</td>
</tr>
</tbody>
</table>
## Heller Myotomy

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Before Laparoscopic Heller Myotomy</th>
<th>After Laparoscopic Heller Myotomy†</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heartburn while sleeping</td>
<td>5.0, 4.9 ± 3.86</td>
<td>1.0, 2.2 ± 2.76</td>
</tr>
<tr>
<td>Postprandial heartburn</td>
<td>4.0, 4.8 ± 3.87</td>
<td>1.0, 1.6 ± 1.48</td>
</tr>
<tr>
<td>Nausea after meals</td>
<td>5.0, 4.7 ± 4.24</td>
<td>0.0, 1.0 ± 2.12</td>
</tr>
<tr>
<td>Vomiting after meals</td>
<td>6.0, 5.2 ± 4.26</td>
<td>0.0, 0.8 ± 1.91</td>
</tr>
<tr>
<td>Regurgitation after meals</td>
<td>9.0, 7.5 ± 3.04</td>
<td>0.0, 1.5 ± 2.80</td>
</tr>
<tr>
<td>Food stuck in throat</td>
<td>10.0, 8.0 ± 3.17</td>
<td>1.0, 2.2 ± 2.65</td>
</tr>
<tr>
<td>Food stuck in chest</td>
<td>10.0, 8.1 ± 2.88</td>
<td>1.0, 1.9 ± 2.62</td>
</tr>
<tr>
<td>Chest pain with swallowing</td>
<td>5.0, 5.0 ± 4.01</td>
<td>0.0, 1.2 ± 2.12</td>
</tr>
<tr>
<td>Postprandial bitter taste</td>
<td>2.0, 4.0 ± 4.18</td>
<td>0.0, 1.1 ± 2.28</td>
</tr>
<tr>
<td>Bitter taste while sleeping</td>
<td>5.0, 4.6 ± 4.09</td>
<td>0.0, 1.4 ± 2.75</td>
</tr>
<tr>
<td>Asthma/coughing</td>
<td>3.0, 3.9 ± 3.95</td>
<td>0.0, 1.1 ± 2.17</td>
</tr>
<tr>
<td>Gas/bloating</td>
<td>5.0, 4.9 ± 5.40</td>
<td>2.0, 2.7 ± 2.99</td>
</tr>
</tbody>
</table>

*Data are presented as median, mean ± SD when appropriate.
†Less than before laparoscopic Heller myotomy, $P < 0.01$, paired Student $t$ test.

Source: Ann Surg © 2005 Lippincott Williams & Wilkins
Achalasia

Outcome Predictors

- Degree of improvement in resting LES
  - If reduced to < 10 mmHg = long-lasting relief of dysphagia (5 years or more)
  - > 20 mmHg = recurrent dysphagia within 12 to 24 months following surgery

Laparoscopic Myotomy

Outcome Predictors

- High pre-operative resting LES pressure increases relief of dysphagia

  - $> 35 \text{ mmHg} = 21.3$ times more likely to have relief than those with $< 35 \text{ mmHg}$

- The greater the decrease in LES pressure following surgery, better improvement in dysphagia
Diffuse Esophageal Spasm

- Rapid progression of:
  - Abnormally high amplitude waves
  - Longer duration contractions
  - Aperistalsis during more than 20 swallows

- Nutcracker esophagus:
  - Variant of diffuse esophageal spasm
  - Rapid progression of esophageal pump with high amplitude waves (> 180 mmHg) of the distal esophagus
Diffuse Esophageal Spasm

- LES may show:
  - Normal resting pressure w/ relaxation after deglutition
  - Intermittent episodes of incomplete relaxation
  - Higher risk of epiphrenic diverticulum

- Treatment:
  - Medical Tx and lifestyle adjustments
  - If persistent symptoms: surgical intervention
    - Long esophageal myotomy via thoracoscopic approach
    - Myotomy throughout entire distance of manometric abnormality (from aortic arch to LES)
Esophageal Hypercontraction

- Resting pressure in LES exceeds upper limit of normal
- Esophageal body peristalsis remains normal
- Symptoms:
  - Dysphagia
  - Chest pain
  - GERD
- Occasionally 2ry to GERD and/or type III hiatal hernia
  - Alteration of configuration of the cardia
- Treatment:
  - Laparoscopic myotomy w/ partial fundoplication
  - Nissen fundoplication (if 2ry to GERD/hiatal hernia)
Esophageal Hypocontraction

- Typically secondary to systemic illness
  - Scleroderma
  - Rheumatoid Arthritis
  - SLE
  - Alcoholism
- Abnormally low amplitude (< 30 mmHg) contractions
- Discoordination leads to ineffective peristalsis
- Normal LES
- GERD due to lack of peristalsis
- Heartburn and Reflux more common than dysphagia
Esophageal Hypocontraction

- Need to rule out:
  - Mechanical obstruction
  - Malignant disorders
- Contrast esophagograms
- Manometry
- 24-hr pH monitoring

- Treatment: directed towards GERD
  - Surgical Anti-reflux procedures
Conclusion

- Lap Heller myotomy has become the standard surgical approach for patients w/ achalasia.
- Surgical myotomy provides superior long-term symptom relief compared to non-surgical interventions.
- Extended distal myotomy with partial fundoplication has been found to provide greater dysphagia relief with minimal development of GERD.
- A high pre-operative LES pressure portends a better symptomatic outcome following surgery.
- Persistent or recurrent symptoms following myotomy can be treated effectively with pneumatic dilatation.
A 34 y.o man has a progressive hx of dysphagia to solids and liquids. Over the last several weeks he has regurgitated food that is several days old. Barium swallow reveals narrowed tapering of distal esophagus and EGD reveals retained food particles. The most appropriate management for this pt is:

a) Botulinum toxin injection of the LES
b) Calcium channel blocker therapy
c) Esophageal myotomy
d) Esophageal myotomy with fundoplication
e) Pneumatic dilation of the LES
A 70 y.o man presents with dysphagia and intermittent regurgitation of mucoid material. He has lost 35 lbs since the onset of symptoms 3 months ago. Barium swallow reveals a “bird’s beak” appearance. All of the following are pertinent to his work-up EXCEPT:

a) Esophagoscopy
b) 24-hr pH monitoring
c) Esophageal manometry
d) Serum albumin
e) Chest x-ray
Question 3

A 52 y.o woman presents c/o several year hx of progressive dysphagia to liquid and solid food. She describes a sensation of food “sticking” and not passing into her stomach. Barium swallow reveals narrow tapering of distal esophagus. Manometry reveals LES pressure that remains constant thru a swallow test. You diagnose achalasia. Which of the following is TRUE regarding LES in healthy pts?

a) LES is a specific anatomic sphincter  
b) Gastric distention causes decreased LES tone  
c) LES pressure decreases during the initiation of a swallow  
d) The LES serves to prevent air from entering the stomach during a swallow  
e) The LES can be visualized by upper endoscopy