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Jointly provided by the Annenberg Center for Health Sciences at Eisenhower and Gastroenterology and Hepatology Advanced Practice Providers.
Esophageal Dysphagia

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Disclosures

Mary Clarke, NP-BC
• No financial relationships to disclose

Monica Nandwani, DNP, FNP-BC
• No financial relationships to disclose
RA is a 70 year old woman

• PMH:
  – Asthma
  – Anemia
  – Angina

• HPI:
  – Dysphagia to solids x 5 years, progressively worsening
  – Regurgitation
  – Retrosternal chest pain
  – Chronic cough, aspiration of food particles
  – Shortness of breath
Case Study #1: RA

What else do you want to know?
History Questions

Do you cough or choke when trying to eat?
- Oropharyngeal dysphagia

What do you have trouble swallowing?
- Solids only
  - Ring, web, stricture, malignancy
  - Hiatal hernia
- Solids and liquids
  - Motility disorder
  - Consider achalasia

Any associated heartburn?
- GERD, esophagitis, stricture, hypersensitivity, poor motility
- Hiatal hernia

Any associated regurgitation?
- Acidic or sour regurgitation
  - GERD
- Bland regurgitation
  - Achalasia, rumination

Where does food get stuck?
- Subxiphoid
  - Distal esophageal process
- Mid chest
  - Diffuse process like EoE or lichen planus
- Cervical
  - Cricopharyngeal bar or distal process referred proximally

Case Study #1: RA

What diagnostic testing do you recommend?
Non-GI Diagnostic Testing

Cardiac work up
- Normal EKG
- Normal echo

CBC, iron studies
- H/H: 9.9/32
- Ferritin: 12

Pulmonary testing
- Decreased FVC
Diagnostic Testing Considerations

- Barium swallow
- High-Resolution Esophageal Manometry
- Upper GI endoscopy
- pH study?
  - Wireless pH probe
  - 24 hour pH impedance
Barium Swallow Comparison

Normal Study

Patient RA

Stomach in Chest

esophagus in normal position
Case Study #1: Endoscopy

Diagnostic Testing: Upper Endoscopy

- Twisted gastric folds
- Difficulty advancing scope to distal gastric body
- Cameron’s ulcers near GEJ
Case Study #1: High Resolution Manometry
Diagnostic Evaluation: Esophageal pH Testing

Wireless pH probe

- Records esophageal pH for 48-96 hours
- Acid Exposure Time (AET)
  - > 6% is conclusive for pathologic reflux
  - 4 – 6% is borderline
  - < 4% is normal
Diagnostic Evaluation: Esophageal pH Testing

24 hour pH Impedance Testing
Case Study #1: RA

What is the diagnosis?
Hiatal/Paraesophageal Hernia

Prevalence
- Estimated 50% over age 60
- 15-20% pregnant women

10% experience symptoms

95% of hernias are type 1

SAGES (Society of Gastrointestinal and Endoscopic Surgeons) Recommendations:
- Repair of type 1 in the absence of reflux is not necessary (+++, strong)
- All symptomatic paraesophageal hernias should be repaired (+++, strong)
- Routine elective repair of completely asymptomatic paraesophageal hernias may not always be indicated (+++, weak)
- Acute gastric volvulus requires reduction of the stomach with limited resection if needed (+++, strong)
- Iron deficiency anemia can be seen in up to 50% of patients with a paraesophageal hiatal hernia
Hiatal/Paraesophageal Hernia Symptoms

- Mild nausea
- Bloating
- Dysphagia
- Retching
- Pain
- Dyspnea
- Postprandial fullness
- Gastric outlet obstruction
- Severe gastroesophageal reflux
- Anemia

Risk of progression from asymptomatic to symptomatic paraesophageal hernia:
- ~14%/year

Mortality rates:
- Emergency repair: 0 – 5.4%
- ~less than 2%/year

Risk of developing acute symptoms requiring emergency surgery:
- 17%
  - Primarily related to pulmonary complications, thromboembolic events and hemorrhage

Average mortality rates for emergency hiatal hernia surgery:
Hiatal/Paraesophageal Hernia: Comparison
SAGES Recommendations: When to Operate

Surgical Repair is reserved for patients with:

- Symptoms of gastric outlet obstruction
- Severe gastroesophageal reflux
- Anemia
- Possible gastric strangulation

Surgical repair of hernias for respiratory symptoms and symptoms of post-prandial fullness is less well studied.

Elective laparoscopic hiatal hernia repair in asymptomatic patients might actually decrease the quality-adjusted life expectancy for patients aged 65 years and older.
Considerations

• Postoperative nausea and vomiting should be treated aggressively to minimize poor outcomes (++, strong)

• Morbidity
  – Higher among elderly patients and those with co-morbidities
  – No increase in the recurrence rate based on age

• Mortality
  – High following emergency procedures among elderly patients undergoing PEH repair

• The larger the size of the hiatal hernia, as measured by the hiatal surface area, the more likely the recurrence
Surgical Treatment

Paraesophageal Hernia Repair
Success With Hiatal/Paraesophageal Hernia Repairs

28 – 43% recurrence

Re-operation only if symptomatic
Non-Surgical Management: Hiatal/Paraesophageal Hernia

- Weight loss/maintaining normal BMI (21-25)
- Avoid tight fitting clothing
- PPI (treating GERD)
- H₂ Blockers
- Antacids
Case Study #2

YZ is a 44 year old gentleman

• PMH
  – HTN

• HPI
  – Dysphagia to solids & intermittently liquids x 5 years
  – Nocturnal cough
  – Weight loss of 10 pounds
  – Episodic chest pain
  – Unable to belch
  – Travel to Mexico 1 month before symptom onset
What else do you want to know?
Good history is
**History Questions**

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History Questions

When did it start?

- Recent onset, rapidly progressive, weight loss
  - **Malignancy**
- Chronic, slowly progressive, no associated weight loss
  - **Non-malignant disease process**

Do you have a history of allergies?

- Eosinophilic Esophagitis (EoE)

Medications?

- Tetracyclines, bisphosphonates, potassium, NSAIDs
  - **Pill or caustic esophagitis**
- Opiates
  - **Opioid-induced esophageal dysfunction (OIED)**

# Eckardt Score

<table>
<thead>
<tr>
<th>Score</th>
<th>Weight loss (kg)</th>
<th>Dysphagia</th>
<th>Retrosternal pain</th>
<th>Regurgitation</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>1</td>
<td>&lt; 5</td>
<td>Occasional</td>
<td>Occasional</td>
<td>Occasional</td>
</tr>
<tr>
<td>2</td>
<td>5 – 10</td>
<td>Daily</td>
<td>Daily</td>
<td>Daily</td>
</tr>
<tr>
<td>3</td>
<td>➢ 10</td>
<td>Each meal</td>
<td>Each meal</td>
<td>Each meal</td>
</tr>
</tbody>
</table>

**Patient Response:**
- Weight loss (kg): 1
- Dysphagia: 3
- Retrosternal pain: 2
- Regurgitation: 2

**Total Eckardt Score:** 8

Dysphagia

Oropharyngeal
- ‘Transfer’ dysphagia
- Difficulty in transferring food from mouth to esophagus

Esophageal
- Difficulty of food or liquid passage from the UES to stomach

Esophageal Dysphagia

Solids and/or liquids

Motor Disorder

Intermittent
- Primary esophageal motility disorder
- Secondary esophageal motility disorder

Progressive
- Chronic heartburn
- Regurgitation, respiratory symptoms, weight loss

Solids

Mechanical obstruction

Intermittent/Non-progressive
- Esophageal ring
- Eosinophilic esophagitis

Progressive
- Chronic heartburn
- Peptic stricture, Hiatal hernia
- Esophageal/cardia cancer

Chronic heartburn
Scleroderma
Achalasia

Weight loss
Eosinophilic esophagitis
Peptic stricture, Hiatal hernia

Older adult, significant weight loss, anemia
Esophageal/cardia cancer

What diagnostic testing do you recommend?
Case Study #2: YZ

Diagnostic Testing: Upper Endoscopy

- Mildly dilated esophagus
- Some retained saliva
- Tight gastroesophageal junction with no obvious stricture
- Biopsies show 5-10 eos/hpf

(a)
What next?
Case Study #2: YZ

Diagnostic Testing:

- Barium Esophagram (timed)
- High Resolution Esophageal Manometry
High Resolution Esophageal Manometry (HRM)
High Resolution Esophageal Manometry (HRM)

- Evaluates esophageal motility
- Establishes landmarks for pH probe placement
- Recommended for preoperative evaluation prior to antireflux surgery
24 Hour pH Impedance Testing

Below line = pH <4
Impedance Planimetry

Provides real-time information about distensibility of the esophageal segments in the form of a distensibility index (DI)

Information about esophageal motility through FLIP topography

Tool that can help guide treatment and predict treatment response

Spechler SJ. *UpToDate*. 2020.
What is the diagnosis?
Achalasia

Rare esophageal disorder

- Incidence: 1.6 cases per 100,000
- Prevalence: 10 cases per 100,000

Thought to result from inflammation and degeneration of neurons in the esophageal wall resulting in:

- Absence of relaxation of the LES
- Absence of peristalsis along esophageal body

Achalasia

Progressive dilation of esophagus can develop without treatment

- Late- or end-stage achalasia is characterized by esophageal tortuosity, angulation, and megaesophagus (diameter >6 cm)

Increased risk for developing esophageal cancer

Spechler SJ. *UpToDate*. 2020.
Chicago Classification, v3.0

Hierarchical Analysis

IRP ≥ ULN and 100% failed peristalsis or spasm

- Yes: Achalasia
  - Type I: No contractility
  - Type II: ≥ 20% PEP
  - Type III: ≥ 20% spasm (DL < 4.5s)

- No: IRP ≥ ULN and not Type I-III achalasia

IRP ≥ ULN and not Type I-III achalasia

- Yes: EGJ outflow obstruction
  - Incompletely expressed achalasia
  - Mechanical obstruction

- No: IRP ≥ normal and Short DL or high DCI or 100% failed peristalsis

IRP ≥ normal and Short DL or high DCI or 100% failed peristalsis

- Yes: DES
  - ≥ 20% premature (DL < 4.5s)
  - Jackhammer esophagus: ≥ 20% DCI > 8000 mmHg*s*cm
  - Absent contractility
  - No scorable contraction
  - Consider achalasia

- No: IRP normal and ≥ 50% ineffective swallows

IRP normal and ≥ 50% ineffective swallows

- Yes: Ineffective motility (IEM)
  - ≥ 50% ineffective swallow
  - Fragmented peristalsis
  - ≥ 50% fragmented swallows and not ineffective

- No: IRP normal and > 50% effective swallows

IRP normal and > 50% effective swallows

- Yes: Normal

Disorders with EGJ outflow obstruction

Major disorders of peristalsis
- Entities not seen in normal subjects

Minor disorders of peristalsis
- Impaired clearance

The Chicago Classification v3.0
## Chicago Classification, v3.0

### Table 4. The Chicago Classification of esophageal motility v3.0

<table>
<thead>
<tr>
<th>Achalasia and EGJ outflow obstruction</th>
<th>Criteria</th>
</tr>
</thead>
</table>
| **Type I achalasia (classic achalasia)** | Elevated median IRP (> 15 mmHg*), 100% failed peristalsis (DCI < 100 mmHg•s•cm)  
Premature contractions with DCI values less than 450 mmHg•s•cm satisfy criteria for failed peristalsis |
| **Type II achalasia (with esophageal compression)** | Elevated median IRP (> 15 mmHg*), 100% failed peristalsis, panesophageal pressurization with ≥ 20% of swallows  
Contractions may be masked by esophageal pressurization and DCI should not be calculated |
| **Type III achalasia (spastic achalasia)** | Elevated median IRP (> 15 mmHg*), no normal peristalsis, premature (spastic) contractions with DCI > 450 mmHg•s•cm with ≥ 20% of swallows  
May be mixed with panesophageal pressurization |

<table>
<thead>
<tr>
<th>EGJ outflow obstruction</th>
<th>Elevated median IRP (&gt; 15 mmHg*), sufficient evidence of peristalsis such that criteria for types I-III achalasia are not met†</th>
</tr>
</thead>
</table>

### Major disorders of peristalsis

- **Absent contractility**
  - Normal median IRP, 100% failed peristalsis  
  - *Achalasia should be considered when IRP values are borderline and there is evidence of esophageal pressurization*  
  - Premature contractions with DCI values less than 450 mmHg•s•cm meet criteria for failed peristalsis

- **Distal esophageal spasm**
  - Normal median IRP, ≥ 20% premature contractions with DCI > 450 mmHg•s•cm*. Some normal peristalsis may be present.

- **Hypercontractile esophagus (jackhammer)**
  - At least two swallows with DCI > 8000 mmHg•s•cm*.†  
  - *Hypercontractility may involve, or even be localized to, the LES*

- **Ineffective esophageal motility (IEM)**
  - ≥ 50% ineffective swallows  
  - *Ineffective swallows can be failed or weak (DCI < 450 mmHg•s•cm)*  
  - Multiple repetitive swallow assessment may be helpful in determining peristaltic reserve

- **Fragmented peristalsis**
  - ≥ 50% fragmented contractions with DCI > 450 mmHg•s•cm

- **Normal esophageal motility**
  - Not fulfilling any of the above classifications

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*Cutoff value dependent on the manometric hardware; this is the cutoff for the Sierra device. †Potential etiologies: early achalasia, mechanical obstruction, esophageal wall stiffness, or manifestation of hiatal hernia. ‡Hypercontractile esophagus can be a manifestation of outflow obstruction as evident by instances in which it occurs in association with an IRP greater than the upper limit of normal.
What is the best treatment option for YZ?
Achalasia: Treatment Options

Medical
- Nitrates
- CCB
- Phosphodiesterase

Pharmacologic

Botulinum Toxin Injection (BTI)

Endoscopic

Pneumatic dilatation (PD)

Peroral endoscopic myotomy (POEM)

Alternative treatments
- Retrievable or absorbable stents
- Intrasphincteric injection with ethanolamine oleate or polidocanol

Surgical

Laparoscopic Heller Myotomy (LHM)
- With partial fundoplication

Esophagectomy
- End stage achalasia

Zaninotto et al. Dis Esophagus. 2018;31(9).
Achalasia: Treatment Options

Medical treatment with nitrates, CCB, phosphodiesterase inhibitors

- International Society for Diseases of the Esophagus (ISDE) Achalasia Guidelines recommend against use for treatment of achalasia

- No convincing evidence that these are effective for symptomatic relief

- 2020 ACG Clinical Guidelines for achalasia state that pharmacologic therapy should be reserved for those who have failed botulinum toxin injection and cannot undergo definitive therapy

Achalasia: Treatment Options

Botulinum Toxin Injection (BTI)

• Limited application in young patients (<50)

• Should be reserved for patient unfit for surgery

• Can be a bridge to more effective therapies

• Repeated treatments safe, but less effective

• 2020 ACG Clinical Guidelines for achalasia state that they recommend that treatment with BTI does not significantly affect performance and outcomes of myotomy

Achalasia: Treatment Options

Pneumatic Dilatation (PD)

• Serial PD is the most effective non-surgical treatment option

• Retreatment may be required (¼ to 1/3 of dilated patients will require redilation)

• Patients wishing long-term remission (without further dilation) may opt for surgical treatment

• Perforation is a potential serious complication

Achalasia: Treatment Options

Peroral Endoscopic Myotomy (POEM)

- Medium term follow-up (2-4 years) has shown similar outcomes to LHM or PD
- ISDE recommends that patients are informed of GERD risk post-POEM and that acid suppressive therapy is discussed
- No evidence that prior BTI or PD reduces feasibility
- Appropriate for symptom persistent/recurrence after LHM
- 2020 ACG Clinical Guidelines for achalasia suggest that POEM would be a better treatment option for Type III achalasia

Achalasia: Treatment Options

Alternative Treatments

- Little evidence to support stent placement or use of sclerotherapy as effective treatment
- ISDE & ACG guidelines recommend against these treatment options for achalasia

Achalasia: Treatment Options

Laparoscopic Heller Myotomy (LHM)

- Best outcomes are achieved in Chicago type I & II
- Partial fundoplication (Dor or Toupet) should be added to LHM to reduce GERD risk
- Should be considered as a first-line treatment option in achalasia patients with sigmoid esophagus

Achalasia: Treatment Options

Esophagectomy

• Last resort when conservative strategies failed

• Consider in those with megaesophagus (larger than 8 cm)

• Indicated in patients with persistent or recurrent achalasia after failure of PD, POEM, LHM and radiologic progression of disease

• Associated with high rate of complications and surgical mortality

Case Study #2: YZ

- Patient preferences
- Local expertise
- Prior therapies
2020 ACG Clinical Guidelines for Diagnosis and Management of Achalasia

Symptoms of dysphagia ± chest pain and bland regurgitation
GERD symptoms not responsive to PPI therapy

Upper Endoscopy

Mechanical Obstruction/Eosophagitis:
Treat appropriately

Normal

Esophageal dilatation, EGJ resistance,
Retained food, Diverticulum

High Resolution Manometry,
Timed Barium Esophagram
(FLIP may be helpful in patients unable
to tolerate HRM)

Type I or II Achalasia

Definitive Therapy:
- Pneumatic dilation (30/35/40 mm)
  - May start with 35 mm in young males
  - Routine gastrografin is not needed
  - Repeat in 2-4 weeks if no response
- Laparoscopic Heller Myotomy
- POEM
- Standard myotomy length
- All patients discharged with PPI therapy

Type III Achalasia

Definitive Therapy
- Tailored myotomy via POEM or tailored Heller myotomy

Patients unfit for definitive therapy:
- Botulinum toxin
- Smooth muscle relaxants
Thank You!


