Esophageal Motility Disorders

DARIUS A. JAHANN, M.D.
DIRECTOR OF ADVANCED ENDOSCOPY AND PANCREATIC DISEASES
DIVISION OF GASTROENTEROLOGY
CREIGHTON UNIVERSITY SCHOOL OF MEDICINE
CATHOLIC HEALTH INITIATIVES

Objectives

• Review the symptoms of esophageal motility disorders
• Identify the indications for High Resolution Manometry (HRM)
• Define esophageal motility disorders based on HRM data by using the Chicago Classification
• Discuss the role of Pneumatic Dilation (PD) and Botulinum Toxin (BT) in the treatment of achalasia
• Define and delineate the treatment of Hypercontractile Esophageal Motility Disorders, specifically Distal Esophageal Spasm and Jackhammer Esophagus.
Common Signs of Esophageal Motility Disorders

- Dysphagia – generally to solids and liquids
- Chest pain – retrosternal, parasternal, etc.
- Heartburn

**UPPER ENDOSCOPY**

- **POSITIVE:** Stricture, Malignancy, EoE
- **NEGATIVE**

**HRM**
CHEST PAIN  Cardiac Etiology?  Proton-Pump Inhibitor Trial  Symptoms resolve?
Yes  No  No

OTHER ENDOSCOPY

Negative

HRM

Other Indications to Order HRM

• Exclude motility disorder before esophageal surgery
• Evaluate for esophageal involvement of systemic diseases affecting smooth muscle (ie. scleroderma)
Basic Anatomy and Physiology

- Motor function of GI tract depends on contraction of smooth muscle, controlled by enteric and extrinsic nerves
- Upper Esophageal Sphincter
  - Striated muscle
  - Prevents swallowing of air
  - Secondary barrier to prevent reflux
- Primary Peristalsis
  - Initiated by a swallow
- Secondary Peristalsis
  - Elicited in response to esophageal distention
- Lower Esophageal Sphincter
  - Circular muscle band
  - Maintains a tonic contraction at rest to prevent reflux of gastric contents
  - Relaxes when a food bolus is propagating downward

High-Resolution Manometry (HRM)

- Best test for diagnosis of motility disorders
- Sensors every 1 cm
- 10 swallow protocol with 5 mL of water
- Calculates an esophageal pressure topography as well as various metrics
HRM Metrics

- **Integrated Relaxation Pressure (IRP):** adequacy of EGJ relaxation with swallowing
  - \( \rightarrow \) *Is the bottom of your esophagus relaxing to let food down?*

- **Distal Contractile Integral (DCI):** vigor of distal esophageal contraction
  - \( \rightarrow \) *How strong is your esophagus contracting to push a food bolus down?*

- **Distal Latency (DL):** measure of timing of peristalsis.
  - \( \rightarrow \) *Is the next contraction of your esophagus starting too early before the last one is finished?*

- Plug these into an algorithm known as the **Chicago Classification** to facilitate diagnosis of esophageal motility disorders

---

**Flowchart Diagram:**

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

- IRP ≥ ULN and not Type I-III achalasia
  - Yes: DES
    - ≥20% premature (DL<4.5s)
    - Jackhammer esophagus
    - ≥20% DCI >8,000 mmHg*s/cm
    - Absent contractility
    - No scorable contraction
    - Consider achalasia
  - No: Ineffective motility (IEM)
    - ≥50% ineffective swallows
    - Fragmented peristalsis
    - ≥50% fragmented swallows and not ineffective

- IRP normal and ≥50% ineffective swallows
  - No: IRP normal and ≥ 50% effective swallows
    - Yes: Normal
    - No: Major disorders of peristalsis
      - Entities not seen in normal subjects

- Minor disorders of peristalsis
  - Impaired clearance

---

Disorders with EGJ outflow obstruction

---

5/9/2019
Achalasia: Overview

• Failure of LES to relax completely
• Aperistalsis in smooth muscle of esophagus
• Degeneration of ganglion cells in myenteric plexus of LES -> Loss of inhibition to the muscle -> LES muscle does NOT relax
• Exact mechanism unknown: autoimmune, viral, neurodegenerative, Chagas
Pneumatic Dilation

- Tearing of muscle fibers of LES with a radial force delivered by a balloon catheter
- 71-90% respond. Graded dilation if there is no response (30->35->40 mm)
- 20-30% 5-year recurrence (50% in prospective studies)
- In patient symptom-free at 12 months after first PD, 60% remained symptom-free at 10 years.
- Complications:
  - Perforation: ~3-5%
  - Pain: 15%
  - Bleeding
Botulinum Toxin (BT)

- Botulinum neurotoxin type A blocks acetylcholine release from nerve endings -> Relaxation of LES muscle (blocks the unopposed excitation of the LES muscle)
- Injected endoscopically into 4 quadrants at the LES
- Response rate: 66%. Usually requiring multiple sessions for the best response.
- Discouraging duration of response: 6 – 9 months
- Low risk procedure, but can cause fibrosis with repeated injections, making surgery difficult
- Reserved for:
  - Elderly
  - Those who are not candidates for definitive therapy
- Complications:
  - Chest pain 25%
  - Heartburn 5%
  - Other more rare complications (wall injury, mediastinitis, ulceration)

4-quadrant BT injection for achalasia
Medications for Achalasia

- Calcium channel blockers
- Nitrates
- 5’-phosphodiesterase inhibitors

- Limiting adverse effects
- Variable absorption
- Short lasting effects without any long term efficacy
- Reserved for poor candidates for definitive therapy

Hypercontractile Esophageal Motility Disorders: Distal Esophageal Spasm (DES) and Jackhammer Esophagus (JE)

- Underlying pathophysiology not well understood
- **DES:** Impaired inhibitory innervation -> premature and rapidly propagated contractions in the distal esophagus
- **JE:** excessive excitation and/or smooth muscle hypertrophy
- Symptoms: **Dysphagia** (solids and liquids) and/or **Chest pain** and/or **Heartburn**
- Differentiated by HRM metrics. Imaging varies as well.
DES and JE: Treatment

- Goal of therapy is to provide relief of symptoms
- Control GERD symptoms: proton pump inhibitor twice daily. Why?
  - Esophageal hyperperistalsis may coexist with GERD
  - Dysmotility may be induced by acid reflux
  - Some treatments for esophageal hyperperistalsis may worsen GERD symptoms
- Other therapies aimed more at Chest Pain and Dysphagia:
  - Peppermint oil as needed for chest pain
  - Calcium channel blockers
  - Phosphodiesterase inhibitors
  - Nitroglycerin
  - Low dose Tricyclic antidepressants
  - BT injection as with achalasia (for DES)
Empiric Dilation for Dysphagia

- First study in 1996 (retrospective in 40 pts): there was a benefit in patients with solid food dysphagia
- 3 subsequent randomized clinical trials (2000, 2001, 2004): No benefit associated with empiric dilation compared to control
- American Society of Gastrointestinal Endoscopy no longer recommends it in their most recent guideline in 2006
- Despite the lack of evidence, demographic/endoscopic data suggests that empiric dilation is still widely practiced.

Conclusions

- If a patient has dysphagia or noncardiac chest pain and undergoes an upper endoscopy that is normal, the next step in their workup should involve HRM.
- Pneumatic Dilation is one of the options for definitive treatment of achalasia.
- Botulinum Toxin is reserved for high risk candidates or elderly patients in whom a definitive therapy is not an option.
- The management of Distal Esophageal Spasm and Jackhammer Esophagus is targeted at the symptoms the patient is experiencing, often a combination of heartburn, dysphagia, and chest pain.
Other Resources

Guidelines for Diagnosis and Management of GERD. Am J Gastro 2013

