Motility Disorders:
Choosing and Interpreting the Right Test for the Right Patient

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Did You Know?

1. GI Motility disorders account for nearly half of all outpatient gastroenterology clinic visits
2. Most gastroenterologists will see one new case of achalasia per year in their practice
3. Achalasia diagnosis is initially missed in 20-50% of cases

Two Main Questions

1. Who are the most appropriate patients that may benefit from esophageal motility testing?
2. What are the appropriate tests to offer these patients?

The Caveat

There are no perfect patients and there are no perfect tests!

**Test must be performed well and interpreted correctly**
What are the Right Tests?

- EGD +/- biopsy +/- EUS
- Barium esophagram
- High resolution manometry
- pH analysis +/- impedance
- Gastric emptying study
- EndoFlip

*Not necessarily all inclusive or in this order*

The Dying Art of the Esophagram
How to Evaluate Dysphagia on Esophagram

1. Titrate barium swallow protocol to 100–200 ml of low-density barium over 30 to 45 seconds at the upright position. Three 40° prone radiographs are obtained at 1, 2, and 3 seconds. A barium swallow McKee barium challenge is completed over 1–2 minutes, which assist individual patients to reach barium in the esophagus at the end of 3 minutes. Among swallowed patterns, the height of the swallow bolus volume changes with the amount of esophageal fluid and the depth of esophageal suctioning from 1 to 3 minutes with the degree of dysphagia.

2. Upright radiographic enema phase at least two normal details: esophageal, anatomic, mucosal, structural, and a transformed esophagus.

3. Assessment of residual in the supine position causes dysphagia to detect a residual amount (5–10 ml) of barium density of 1 hour, with each swallow repeated by at least 15–20 seconds. Radiopaque and xeroradiograms follow the course of cross-the upper esophageal sphincter. By radiopaque and xerographic imaging, measurement of residual, especially posterior, is at least 2 milliliter of barium, somewhat cloudy with esophageal mucosal irregularities.

4. Upright esophageal phase in the upright position causes dysphagia to detect a residual amount (5–10 ml) of barium density of 1 hour, with each swallow repeated by at least 15–20 seconds. Radiopaque and xeroradiograms follow the course of cross-the upper esophageal sphincter. By radiopaque and xerographic imaging, measurement of residual, especially posterior, is at least 2 milliliter of barium, somewhat cloudy with esophageal mucosal irregularities.

5. Solid food entering phase in the upright position. This is sampled down with a 15-mm barium swallow, measured, or finds which the patient belongs to the ingestion.

Richter JE. Clinical Gastro and Hepatology 2011;9:1010–1011

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Brief Primer on High Resolution Manometry
Chicago Classification 3.0v

1. Integrated Residual Pressure
2. Distal Latency
3. Distal Contractile Interval
4. Peristaltic Integrity
5. Pressurization Pattern
1. What is the IRP?

Deglutitive LES relaxation

Adapted from Kahrilas, PJ, Pandolfini JE

2. What is the Distal Latency?

Essential metric for spastic contractions

Adapted from Kahrilas, PJ, Pandolfini JE
3. What is the Distal Contractile Interval?

Measure of peristaltic vigor

4. What is the Peristaltic Integrity?

Measure of break size
5. What is the Pressurization Pattern?

What are we looking for by HRM?
Chicago Classification 3.0

Disorders of EGJ outflow obstructions
• Achalasia (Types I, II, III)
• EGJ outflow obstruction

Major disorders of peristalsis
• Hypercontractile esophagus (Jackhammer)
• Distal esophageal spasm
• Absent peristalsis

Minor disorders of peristalsis
• Ineffective esophageal motility
• Fragmented peristalsis
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Interpreting HRM with the Chicago Classification
The Hierarchial Approach

1. IFP ≥40N and 100% failed peristalsis or spasms → Yes
   - Achalasia
     - Type I: No contractility
     - Type II: ≥20% PEP
     - Type III: ≥20% systolic (PEP < 4.5 seconds)
   - Disorders with EGJ outflow obstruction

2. IFP ≥40N and not type I–III achalasia → No
   - EGG outflow obstruction
     - Incompletely expressed esophageal
     - Mechanical obstruction
   - Major disorders of peristalsis

3. IFP normal and short 11, or high DCE or 100% failed peristalsis → Yes
   - DES
     - ≥20% premature (≥4.5 seconds)
     - Jackhammer esophagus
     - ≥20% DCE >10000 contractions
     - Absent contractility
     - No visible contraction
     - Consider achalasia
   - Major disorders of peristalsis

4. IFP normal and ≥20% ineffective swallows → No
   - Ineffective motility (1EM)
     - ≥50% ineffective swallows
     - Fragmented peristalsis
     - ≥20% fragmented swallows and not ineffective
   - Minor disorders of peristalsis
     - Impaired clearance

5. IFP normal and >50% effective swallows → Yes
   - Normal

Adapted from Kahrilas, PJ, Pandolfino JE
What is the Utility of Esophagram versus HRM in the Detection of Esophageal Dysmotility?

Rourke AK. Otolaryngology-Head and Neck Surgery 2016;154(5) 888-891

Esophagram useful in assessment of structural and anatomic abnormalities but poor as screening exam for dysmotility

Dysphagia should be referred for HRM regardless of esophagram

Inter-rater Agreement Is Imperfect Too

Carlson DA. Am J Gastroenterol. 2015 July;110(7):967-978
So Who is The Right Patient?

- Nonobstructive dysphagia
- Noncardiac chest pain
- Regurgitation
- Refractory heartburn


Case #1

- 72 year old male former smoker who presents with history of GERD, HTN and a 8 month history of progressive dysphagia.
- EGD performed 14 months ago for worsening reflux revealed LA grade B esophagitis.
- Accommodated his diet and is currently eating only mechanical soft consistency of food.
- Lost 20 pounds over the past 4 months.
Case #2

What are you thinking is going on?

Is this.....
- Erosive esophagitis... refractory reflux...noncompliance with tx?
- Refractory reflux... esophageal stricture... subsequent dysphagia?
- Progressive dysphagia..... Functional obstruction?
- Progressive dysphagia..... Motility disorder?

Case #2

What are the right tests?
Barium esophagram revealed tapering of the distal esophagus consistent with "achalasia"
Hierarchial Approach to reading HRM

1. Assess IRP
   - Is it elevated? Yes.
2. Assess Peristalsis (on each swallow)
   - Is it failed? No.
   - Is there PEP? No.
   - Is the distal latency short? No.
3. Is there some evidence of peristalsis? Yes

EGJ Outflow Obstruction

Case #1

What does that mean? What is wrong with my patient?

Increased Area of IBP – Intra-Bolus Pressure >15 mmHg
Case #1

EGD repeated
EGJ distorted
Pressure to pass scope
Oozing following scope passage

Case #1

EUS
Hypoechoic lesion
Expanding submucosa and MP
No lymphadenopathy
What is my patient’s diagnosis?

Pseudoachalasia

- Older man, rapidly progressive dysphagia and profound weight loss.
- Pseudoachalasia is a term used to describe the clinical picture of gastroesophageal junction obstruction, most classically by tumor.
- The condition is present in as many as 5% of patients with the radiologic and manometric diagnosis of achalasia.
- A thorough workup includes an upper endoscopy with biopsies of any suspicious malignant process and imaging studies including CT scan, MRI and endoscopic ultrasound, as indicated to rule out a submucosal or extraluminal process.
Case #2

• 32 year old female with a 3 year history of Raynaud’s and reflux. In past her GERD had been controlled with daily PPI. Over the past 6 months her heartburn has increased significantly and she presents now with progressive dysphagia.
What are you thinking is going on?

Is this.....

• Erosive esophagitis... refractory reflux... noncompliance with tx?
• Refractory reflux... esophageal stricture... subsequent dysphagia?
• Progressive dysphagia..... Functional obstruction?
• Progressive dysphagia..... Motility disorder?

Case #2

• Initial improvement with BID PPI and strict life style GERD management
• In spite of this, dysphagia progresses and she incurs a 15 pound weight loss over 3 months
• Over past month she has had an episode of pneumonia
Case #2

What are the right tests?
Case #2

Esophagram Comparison

Case #2

High Resolution Manometry
Hierarchial Approach to reading HRM

1. Assess IRP
   • Is it elevated? No.

2. Assess Peristalsis (each swallow)
   • Is it absent? Yes. 100% failed swallows

   Major Disorder of Peristalsis
   Absent Contractility

HRM comparison
Case #2

Absent Peristalsis

Raynaud's
Erosive esophagitis
Pneumonia
Normal IRP
Absent peristalsis
Hypotensive LES

Absent peristalsis? What is that? What is wrong with my patient?

Case #2

What is my patient’s diagnosis? Scleroderma

• Esophageal dysmotility develops as the smooth muscles of the esophagus are replaced by scar tissue; smooth muscles atrophy and fibrosis occurs.

• Symptoms: Severe heartburn, regurgitation and dysphagia. Pulmonary involvement either as a primary manifestation or secondary to acid reflux and aspiration is well documented

• Classical manometric abnormalities include low LES pressure and low amplitude or absent peristaltic contractions in the distal esophagus

• Treatment is aimed at controlling the primary disease and GERD

Performed 24 hour pH analysis to assess for adequate control
More aggressive control of reflux with 2 years of symptom control

Case #3

- 23 year old woman referred for evaluation of regurgitation; rare heartburn
- Symptoms have progressed significantly over past year and have become noticeable to her spouse. He thinks she has an eating disorder because he sees her effortlessly bring up her food. When he ‘calls her out on it’ she spits it out.
- PPI treatment has made no difference in symptoms
- EGD normal
- 24-hour pH-impedance normal distal acid exposure

What is the right test for my patient?

Case #3

- Gastric strain overcomes the EGJ barrier
- Retrograde pressure gradient
- Pressure drives gastric contents through esophagus and UES
- Patient reports regurgitation
- Swallows mouth contents with effective clearance by primary peristalsis

Tucker et al, APT, 2013
The rumination syndrome in adults: A review of the pathophysiology, diagnosis and treatment

Papadopoulos V, Mimidis K*

J Postgrad Med 2007;53:203-6

Table 2: Diagnostic criteria of rumination syndrome according to Rome III classification[1]

A. Obligatory criteria
   (Present for the last three months with onset at least six months before diagnosis)
   1. Persistent or recurrent regurgitation of recently ingested food into the mouth with subsequent spitting or remastication and swallowing
   2. Regurgitation is not preceded by retching
B. Supportive criteria
   1. Regurgitation events are usually not preceded by nausea
   2. Cessation of the process when the regurgitated material becomes acidic
   3. Regurgitant contains recognizable food with a pleasant taste

Diaphragmatic Breathing

Take inspiratory and expiratory breaths with the abdominal muscles
Keep chest motionless

Behavior is incompatible with abdominal wall contraction
Case #4

• 75 year old female with a more than 20 year history of recurrent episodes of intermittent solid food dysphagia and rare chest pain. **Diagnosed with esophageal spasms.**
• Currently requires power swallows often. Self induces vomiting 3 times a week.
• Past treatments:
  • Esophageal dilations (18, 19, 20 mm at GEJ) no pneumatic
  • Botox – distal and mid esophagus. Last treatment in spiral fashion of lower 5 cm of esophagus
  • Levsin – lead to anxiety
  • NTG – offers some relief – takes SL every other week
Barium Swallow

- Diffuse esophageal spasm
- Mild corkscrew appearance with severe muscular contraction rings
- Loss of peristaltic wave in distal 1/3 of esophagus
- Distal esophagus tapers smoothly almost beak-like
- Stasis of barium bagel in distal esophagus (eventually cleared with multiple swallows of water)
 Hierarchial Approach to reading HRM

1. Assess IRP
   - Is it elevated? Yes.
   - Is there > 20% PEP? Yes.

Achalasia Type 2
Irony of life
There are 3 distinct achalasia subtypes which may respond differently to treatment

<table>
<thead>
<tr>
<th>Type I</th>
<th>Type II</th>
<th>Type III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absent Peristalsis</td>
<td>Panesophageal Pressurization</td>
<td>Spastic</td>
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</tbody>
</table>


Achalasia

- **Symptoms:** Dysphagia, regurgitation, heartburn, chest pain, coughing, choking, aspiration pneumonia, weight loss
- **All patients have at least 2 manometric abnormalities:** abnormal LES residual pressure and no normal peristalsis
- **Achalasia is never cured.** Treatment is directed to reducing the pressure gradient across the LES
  - Pneumatic dilation
  - Heller myotomy
  - POEM
  - Drug therapy: Botulinum toxin, calcium channel blockers, nitrates
Take Home Points

1. Do not use one technology as a sole arbitrar of a diagnosis
2. Must keep a high level of suspicion to diagnose motility disorders
3. Sometimes you have to repeat a test to make the right diagnosis
4. Listen to your patient (they are usually right)