Hypertensive LES

- Presentation: Chest pain/dysphagia/globus
  - May be an achalasia variant
- Dx: LES pressure > 35 mmHg AND failure to relax below IRP of 15 mmHg
  - Normal peristalsis
- More important than pressures: failure of full relaxation at LES
  - Incomplete bolus transfer
- Can overlap with other spastic esophageal conditions
  - May need additional provocation (bread swallow, multiple rapid swallows, solid swallows)
  - EUS recommended prior to therapy to exclude infiltrative or compressive disease (eg malignancy)
EGJ Outflow Obstruction

A: EGJOO: achalasia phenotype

- Locus of diverticulum above EGJ
- IRP = 22.3 mmHg

B: EGJOO: Mechanical

- Normal peristalsis
- Compartmentalized pressurization
- IRP = 27.2 mmHg

- Large diverticulum 4 cm above EGJ
- Barium tablet localized 12 mm restriction

EGJ Outflow Obstruction
Major Disorders of Peristalsis

The Chicago Classification

IRP is normal **AND**
short DL or high DCI or 100%
failed peristalsis

Yes

DES
- ≥ 20% of swallows with reduced DL(<4.5s)

Jackhammer esophagus
- DCI > 8,000 mmHg-s cm and normal DL

Absent contractility

Neurogastroenterology and Motility, 2015;27;160-74.
Non-achalasia motor abnormalities

Hypercontractile Esophagus  The diagnostic criteria for hypercontractile peristalsis have been redefined as at least one propagated swallow-induced contraction with a DCI of >8,000 mmHg-s-cm, as that value is extremely rare in asymptomatic subjects. The repetitive contractions sometimes seen in this presentation have led to the reference of “Jackhammer” esophagus.

Distal Esophageal Spasm  Distal Latency (DL) has been proposed as an improved measure to represent simultaneous contractions. The DL is currently used to define DES, although further evaluation of clinical outcomes is needed to support this metric.¹

Borderline Esophageal Motor Function  This category represents a range of abnormalities including borderline-normal to those conditions similar to ineffective esophageal motility (IEM) or absent contractility.
Nutcracker Esophagus

• Prolonged hypertensive contractions with normal swallow waveform propagation
• Etiology: possible hypercholinergic state resulting in incoordination of longitudinal and smooth muscle
• Presentation: episodic chest pain (may be mistaken with GERD or cardiac CP) – Dysphagia (solids or liquids) – Exacerbation with hot or cold 
• ENT: High dysphagia (globus)
• May exist with other abnormalities (LES HTN)
• Dx: DCI > 5000 (cont ampl x duration x length)
Hypercontractile or Jackhammer Esophagus (aka Spastic Nutcracker)

- Offshoot of Nutcracker
  - 4% manometry referrals; r/o mechanical obst
- Presentation: Chest pain/dysphagia
- Dx:
  - At least one DCI > 8000
  - Repeated high amplitude contractions
  - Normal DL (≥ 4.5 sec)
Distal Esophageal Spasm (DES)

- Unknown etiology; likely related to defects in inhibitory neural pathways of esophagus
  - Rarest manometric diagnosis (3%)
  - Classic corkscrew esophagus very rare
- Presentation: Chest pain/dysphagia
  - Symptom correlation poor
- Dx:
  - Normal median IRP (LES relaxation), ≥ 20% premature contractions with DCI > 450 mmHg x s x cm
  - Some normal peristalsis may be present
A: Latency described with conventional manometry

B: Latency measured with EPT

Normal swallow

C: DES: Rapid Premature Contraction

D: DES: Premature Contraction

E: Rapid Contraction with Normal latency
A) Normal: No breaks/ NL DCI

B) Jackhammer: No breaks/ Abnormal DCI

C) Absent Peristalsis

D) Absent/Failed Peristalsis

E) Weak Peristalsis- IEM

F) Weak Peristalsis- TZ Defect
Treatment of Achalasia

Untreated achalasia

- Candidate for surgery
  - Male <40 yo?
    - Graded Pneumatic Dilation
    - Myotomy
  - Failure
    - Repeat Treatment
      - Endstage
        - Pneumatic Dilation
        - Myotomy
        - Esophagectomy

- Surgery precluded
  - Botox
    - Surgery precluded
# Response Rates of Achalasia Treatments

*Patients categorized by pressure topography subtype*

<table>
<thead>
<tr>
<th>Author</th>
<th>Subtype</th>
<th>No. patients (%)</th>
<th>Success rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pandolfino</td>
<td>I</td>
<td>21 (21.2)</td>
<td>56*</td>
</tr>
<tr>
<td></td>
<td>II</td>
<td>49 (49.5)</td>
<td>96*</td>
</tr>
<tr>
<td></td>
<td>III</td>
<td>29 (29.3)</td>
<td>29*</td>
</tr>
<tr>
<td>Salvador (LHM)</td>
<td>I</td>
<td>96 (39)</td>
<td>84.6</td>
</tr>
<tr>
<td></td>
<td>II</td>
<td>127 (51.6)</td>
<td>95.3</td>
</tr>
<tr>
<td></td>
<td>III</td>
<td>23 (9.4)</td>
<td>69.3</td>
</tr>
<tr>
<td>Pratap (PD)</td>
<td>I</td>
<td>24 (47.1)</td>
<td>63</td>
</tr>
<tr>
<td></td>
<td>II</td>
<td>24 (47.1)</td>
<td>90</td>
</tr>
<tr>
<td></td>
<td>III</td>
<td>3 (5.8)</td>
<td>33.3</td>
</tr>
<tr>
<td>Rohof (PD &amp; LHM)</td>
<td>I</td>
<td>44 (25)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>II</td>
<td>114 (64.7)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>III</td>
<td>18 (10.2)</td>
<td></td>
</tr>
</tbody>
</table>

LHM, laparoscopic heller myotomy; PD, pneumatic dilatation.

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POEM (Per-oral endoscopic myotomy)
- Short-term success 82-100%
- LES pressures reliably lowered
- Lack of long-term data/comparisons to laparoscopic Heller myotomy (current data suggests equivalence)
Treatment of other Esophageal Hypertensive Conditions

- **Nutcracker/Jackhammer**
  - Trial of nitrates (SL or oral) + PPI
  - Calcium channel blockers (diltiazem, sildenafil) PRN
  - Tricyclic antidepressants

- **Hypertensive LES/EGJ Outflow obstruction**
  - Balloon dilation or botox injection
  - SAA
  - POEM

- **DES**
  - PPI + Botox injection
  - Surgical myotomy (when all else fails)
POEM for Spastic Disorders

- 73 patients
  - 9 DES; 10 Jackhammer, 54 type III (spastic) achalasia
  - Dysphagia, regurg, chest pain
  - 11 medcens

- 100% completion
  - 118 mins (mean)
  - 19 cm tunnel (mean)
  - 16 cm myotomy (mean)
  - 3.4 hospital days (mean)

- 8 AEs (11%) none severe

POEM for Spastic Disorders

Hypertensive Contractility Caveats

• There is no clear discriminator of symptomatic hypercontractility
• Propagation can appear normal
• Therapy focused on reducing peristaltic amplitude in altering symptoms is extremely limited
  • Smooth muscle relaxants
  • BOTOX
  • Treat visceral sensitivity
Minor Disorders of Peristalsis
Non-hypertensive Esophageal Motility Disorders

The Chicago classification

- IRP is normal AND ≥ 50% ineffective swallows
- ≥ 50% ineffective swallows
- ≥ 50% fragmented swallows with large (>5 cm) breaks in the 20-mmHg IBC and not effective

Minor Disorders of Peristalsis: Impaired clearance

Neurogastroenterology and Motility, 2015;27;160-74.
Minor Disorders of Peristalsis

Neurogastroenterology and Motility, 2015;27;160-74.
Utilizing HRM/EPT in the Management of Esophageal Symptoms

**Symptoms of dysphagia ± chest pain and bland regurgitation**

→ **Upper Endoscopy**

- Obstructive process: ring, stricture, etc.
- Normal
- Esophageal dilatation
- EGJ resistance
- Retained food
- Diverticulum

→ **High Resolution Manometry**

*Esophagram may be helpful when manometry is technically difficult to perform*

- **EGJ Outflow Obstruction**
  - EGD ± EUS/CT to rule out obstructive process
  - Potentially achalasia phenotype with preserved peristalsis

- **Absent Contractility**
  - If clinical scenario c/w achalasia, a timed barium esophagram should be performed
  - Potentially advanced GERD or scleroderma
  - Potentially achalasia phenotype with hypotensive LES

- **Achalasia I**
  - Severe dilatation associated with poor treatment response
  - Consider myotomy as initial therapy

- **Achalasia II**
  - Best treatment response
  - Esophagram can be normal without barium retention or esophageal dilatation

- **Achalasia III**
  - Worst treatment response
  - May benefit from treatment directed at spasm

- **DES**
  - Extremely rare
  - Difficult to treat
  - Many cases misdiagnosed Type III achalasia

- **Frequently misdiagnosed with conventional manometry**
- **Often diagnosed as DES on esophagram**
Approach to Patients with Esophageal Symptoms

Key Take Home Points

- Esophageal symptoms can have a number of overlapping etiologies
- Interaction between organic and functional influence should not be ignored
- Most disorders can be managed by a careful systematic evaluation
  - Exclude most dangerous causes first and then focus on the most likely cause
- Diagnose and treat in parallel
- Embrace the evolving technology available to you