# EGJ Outflow obstruction and spastic disorders

Sravanya Gavini, MD, MPH
Associate Professor
Division of Digestive and Liver Diseases
University of Texas Southwestern Medical Center
Dallas, Texas

# Conflicts of Interest - None

# Outline

- Esophagogastric junction outflow obstruction
  - Definition by high resolution manometry (HRM) and associated conditions
  - Presentation and diagnostic evaluation
  - Management
- Non-achalasia spastic disorders
  - Definitions
  - Presentation and diagnostic work up
  - Management

# EGJOO – heterogenous disorder

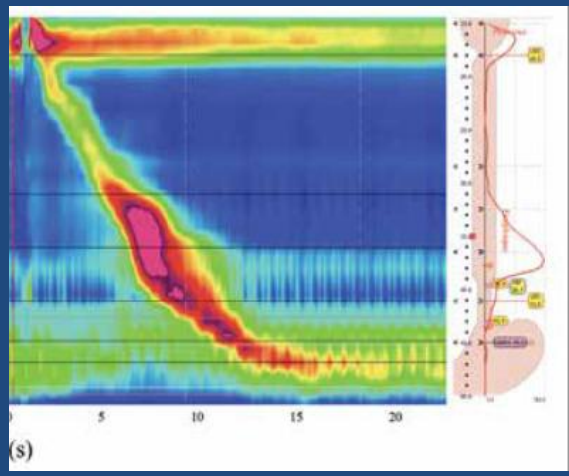
- Anywhere from 15%-73% of patients have spontaneous recovery without intervention
- Only a minority of patients progress to Achalasia
- Can overlap with other disorders

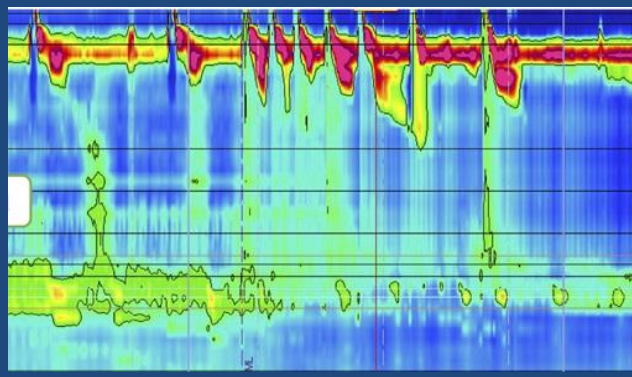
Beveridge C.,et.al. Gastroenterol and Hep 2020
Clayton S., et.al Clin Gastroenterol and Hep 2016

# HRM definition and considerations

- Elevated median IRP in both supine and upright positions, >20% elevated intrabolus pressure (IBP), peristalsis is present
- Provocative maneuvers:
  - Rapid drink challenge
  - Pharmacologic provocation

    e.g. Amyl nitrite inhalation, Cholecystokinin IV



Yadlapati et.al Neurogastro Motil. 2021
Sanagapalli et.al. Am J Gastroenterol 2021

# Establish clinical relevance of EGJOO

- EGJOO can coexist with other motility disorders such as ineffective esophageal motility, distal esophageal spasm (DES), hypercontractile esophagus
- Manometric diagnosis of EGJOO is always considered clinically inconclusive
  - > 1/3$^{rd}$ of the cases may be clinically irrelevant and related to benign etiologies
  - EGJOO can be a result of structural, post surgical, infiltrative etiologies

Jodorkovsky et.al J Clin Gastroenterol 2021
Yadlapati et.al Neurogastro Motil. 2021
Beveridge et.al. Gastroenterol Hepatol. 2020

# Etiologies

- Idiopathic
- Secondary causes:

| | |
|---|---|
| **Structural causes** | Hiatal hernia, esophageal stricture, esophageal ring, esophageal web, esophageal diverticula, gastric volvulus, esophageal varices, vascular compression |
| **Post-surgical** | Nissen fundoplication, laparoscopic gastric band, other bariatric surgery |
| **Infiltrative/ inflammatory** | Systemic sclerosis, eosinophilic esophagitis, amyloidosis |
| **Medications** | Chronic opiates, first- and second-generation antipsychotics |
| **Malignant** | Esophageal cancer, gastric cancer, metastatic disease |

Beveridge et.al. Gastroenterol Hepatol. 2020

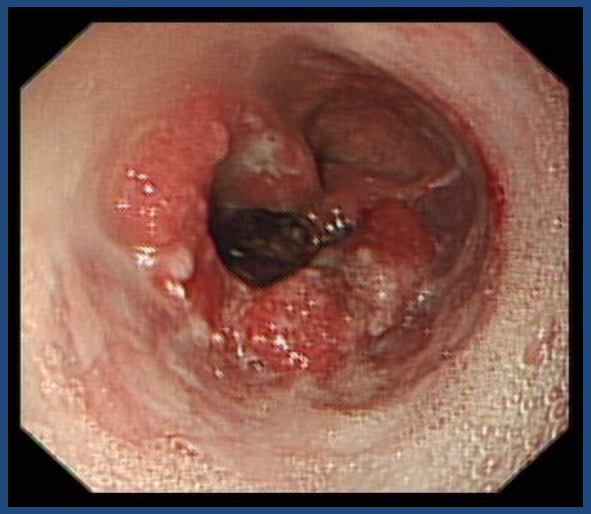
# Clinical presentation

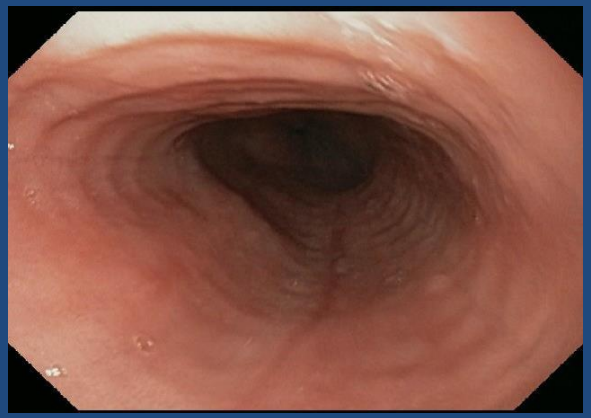
- Esophageal symptoms
  - Dysphagia (40-75%)
  - Chest pain (7%-55%)
  - Regurgitation
  - Heartburn

Beveridge et.al. Gastroenterol Hepatol. 2020

# Diagostic workup - Upper endoscopy

- Rule out secondary causes:
  - Mechanical obstruction
    - Malignancy
    - Stricture, rings
  - Assess and biopsy abnormal mucosa
    - Eosinophilic esophagitis
- No significant data to support need for EUS unless there is a clinical suspicion for submucosal/infiltrative process

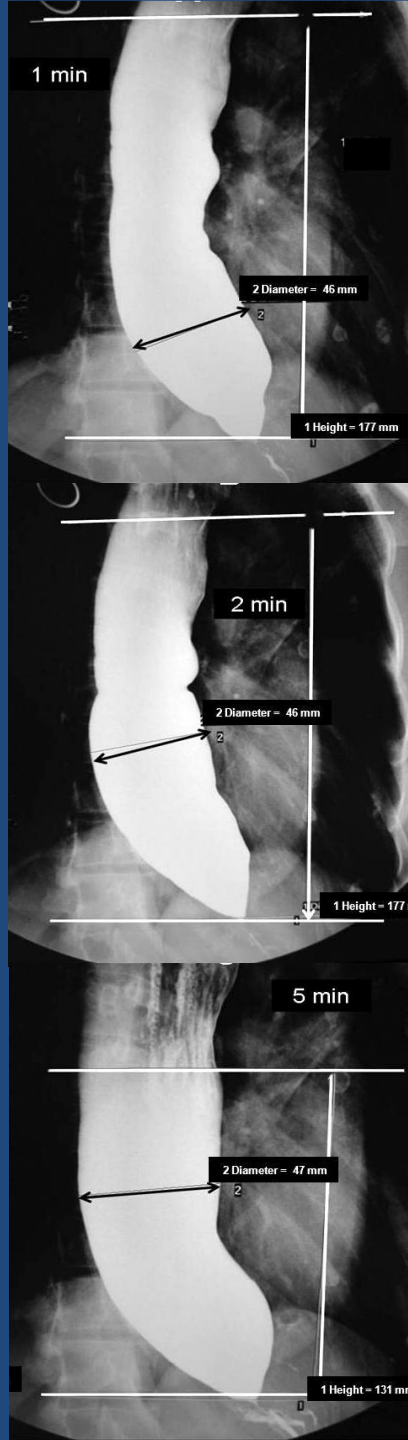




Beveridge et.al. Gastroenterol Hepatol 2020
Liu et.al. Neurogastroenterol Motil 2019

# Timed Barium Esophagram

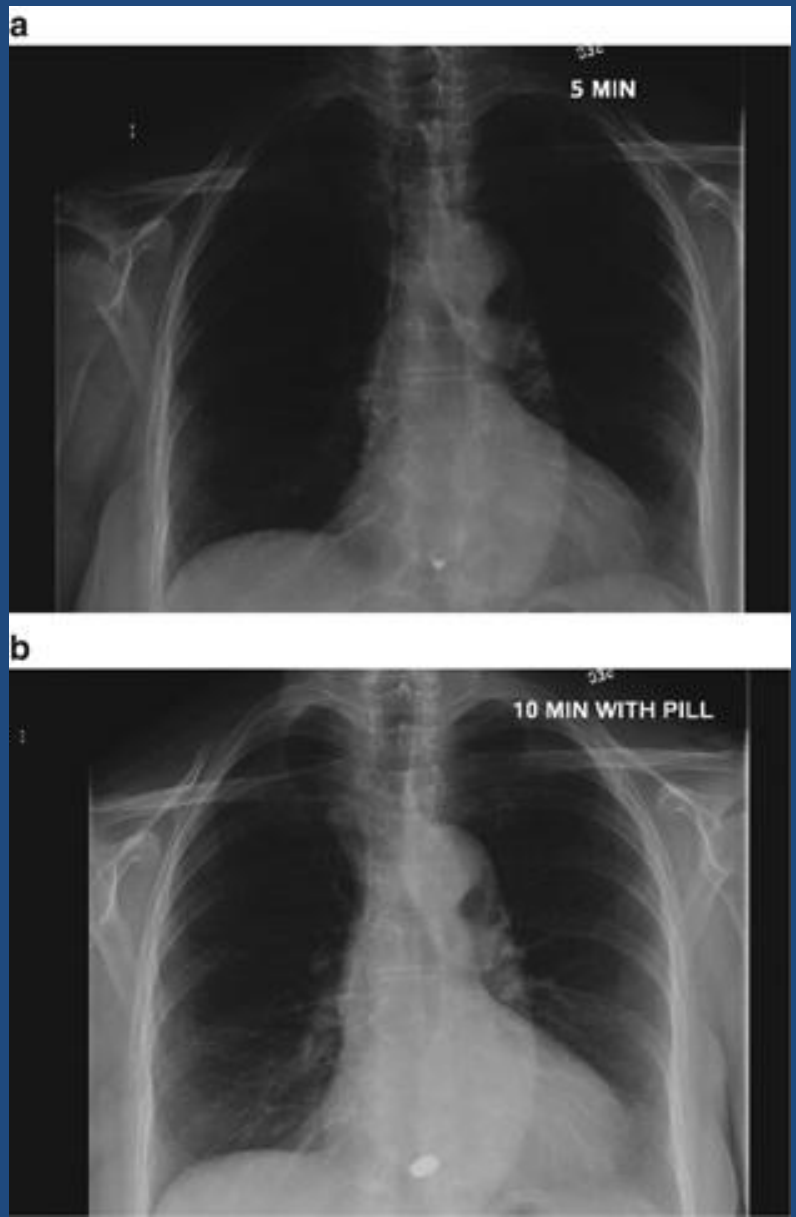
- Evaluate for secondary causes of EGJOO
- Confirm delayed esophageal transit or functional obstruction at EGJ
- Barium completely empties from esophagus in 1 min in most and in 5 min in all healthy individuals




Stefan et.al J Thoracic and Cardiovasc Surg 2018
Blonski et.al. A Journal of Gastroenterol 2018

# Timed Barium Esophagram

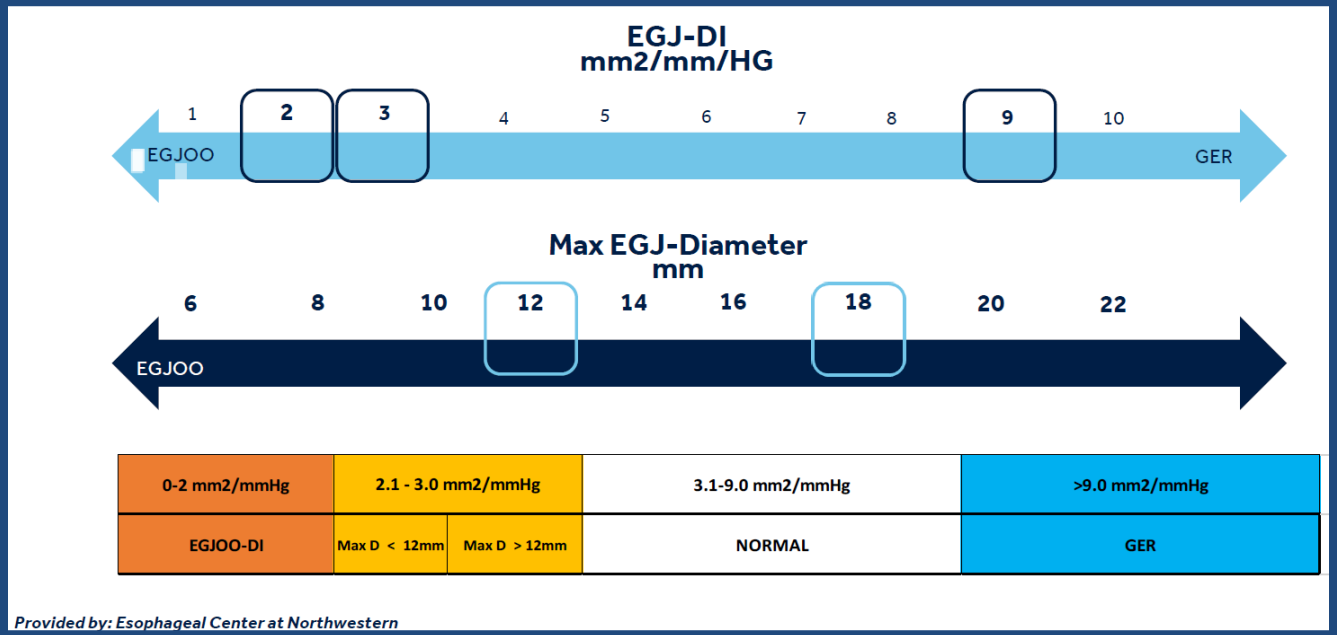
- Combining liquid Barium with barium tablet increases diagnostic yield from 48.9%→ 60% in EGJOO




Blonski et.al. A Journal of Gastroenterology 2018

# EndoFLIP

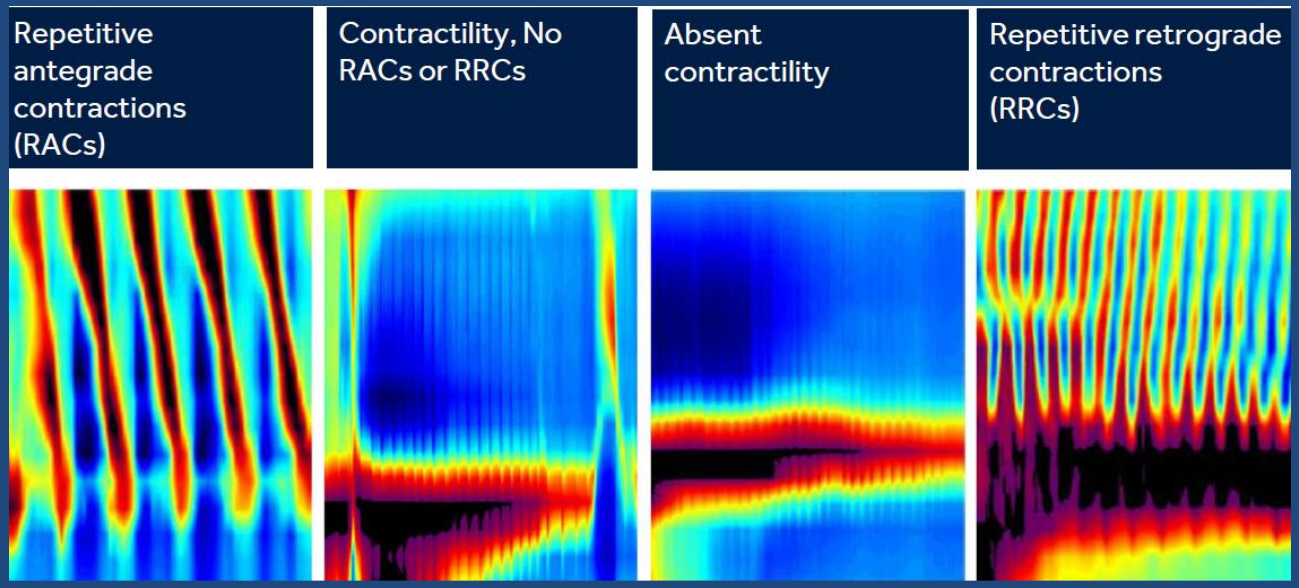
- Popular supportive investigation to assess for EGJOO that might respond to EGJ disruption therapies
- Uses high-resolution impedance planimetry to evaluate calculate the distensibility and cross-sectional area of the EGJ.




Carlson DA, Kahrilas PJ, Lin Z, et al. Am J Gastroenterol. 2016
Savarino E, Am J Gastroenterol 2020

# EndoFLIP/EsoFLIP

- Can evaluate response to volume distention (RRC can be seen in EGJOO)
- Pneumatic dilation with EsoFLIP catheter up to 30 mm




Carlson DA, Kahrilas PJ, Lin Z, et al. Am J Gastroenterol. 2016
Savarino E, Am J Gastroenterol 2020

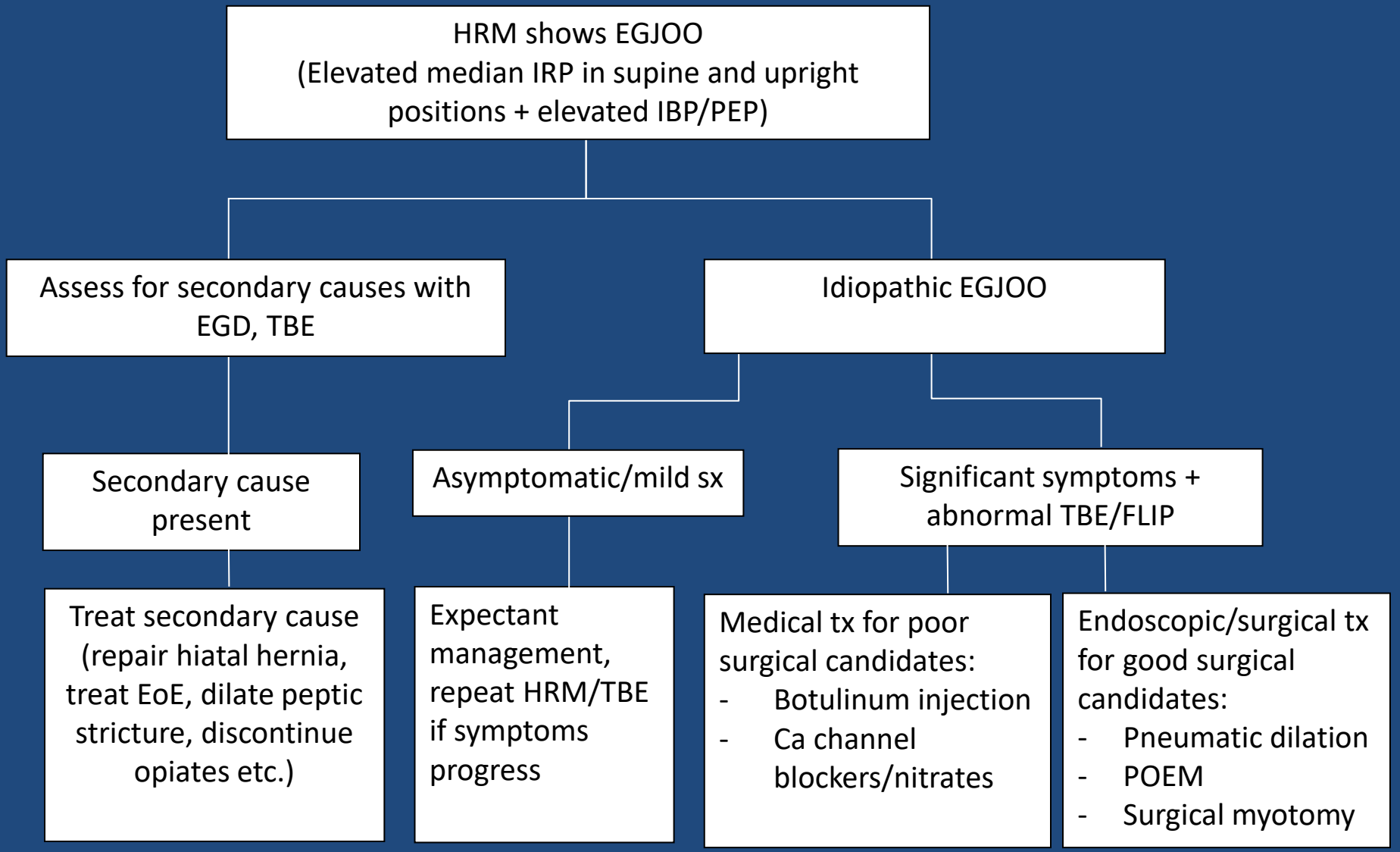
# Management considerations

- Spontaneous resolution occurs in ~43% patients
- Only a minority of patients progress to achalasia
- Obstructive symptoms (dysphagia) vs. perceptive symptoms (atypical chest pain)

# Treatment options

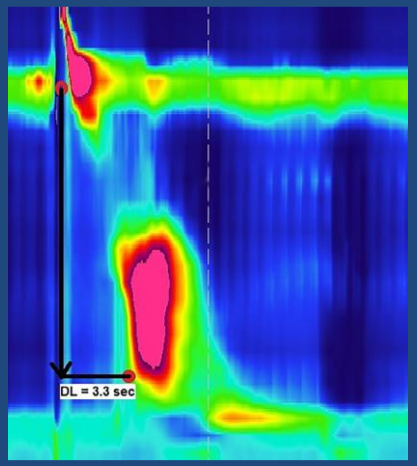
| | |
|---|---|
| **Response** | |
| **Complications** | |
| **Advantages** | |
| **Disadvantages** | |

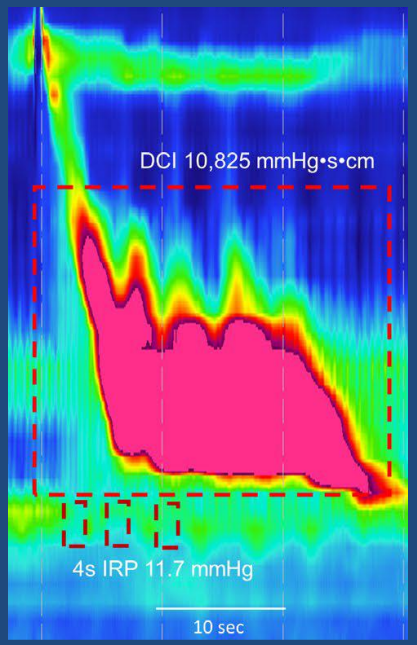
Beveridge et.al. Gastroenterol Hepatol. 2020
Samo et. Al. World J Gastroenterol 2019


HRM shows EGJOO
(Elevated median IRP in supine and upright positions + elevated IBP/PEP)
Assess for secondary causes with EGD, TBE
Idiopathic EGJOO
Secondary cause present
Asymptomatic/mild sx
Significant symptoms + abnormal TBE/FLIP
Treat secondary cause (repair hiatal hernia, treat EoE, dilate peptic stricture, discontinue opiates etc.)
Expectant management, repeat HRM/TBE if symptoms progress
Medical tx for poor surgical candidates:
- Botulinum injection
- Ca channel blockers/nitrates
Endoscopic/surgical tx for good surgical candidates:
- Pneumatic dilation
- POEM
- Surgical myotomy

# Spastic disorders (non achalasia)

- Distal esophageal spasm
  - ≥20% premature contractions
    (Distal latency <4.5 s)
- Hypercontractile esophagus
  - ≥ 20% DCI >8000 mmHg.s.cm
- Normal IRP ( in most cases)




Khalaf et.al Am J Medicine. 2018
Patel et al. (eds.), Evaluation and Management of Dysphagia. 2020
Yadlapati et.al Neurogastro Motil. 2021

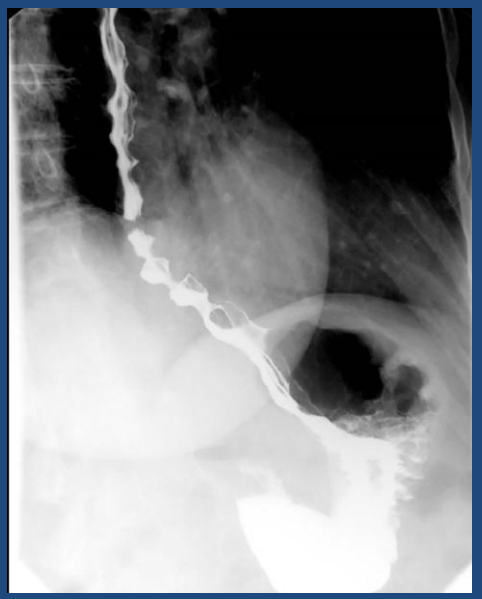
# Clinical presentation

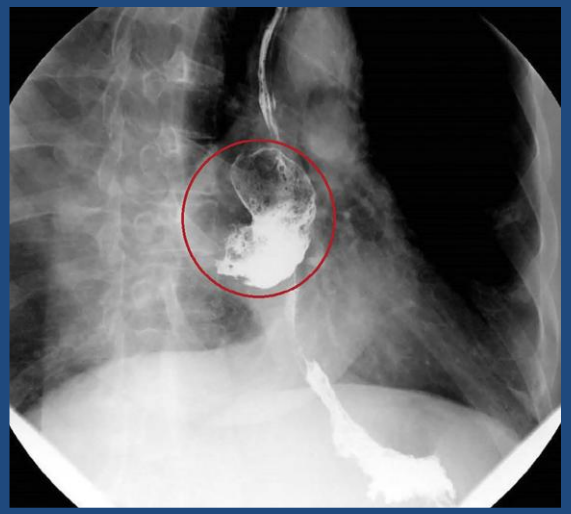
- Dysphagia
- Non-cardiac chest pain (can mimic angina)
- **Gastroesophageal reflux disease (GERD) can coexist with DES and hypercontractile esophagus**
- Epiphrenic diverticula can result from spastic disorders especially in patients with connective tissue disorders
- In rare cases DES can progress to develop achalasia

Patel et al. (eds.), Evaluation and Management of Dysphagia. 2020

# Diagnostic workup

- Upper endoscopy to rule out mechanical obstruction, biopsy to rule out eosinophilic esophagitis
- Timed barium swallow
  - DES can have “corkscrew” appearance
  - Epiphrenic diverticula





Patel et. al. (eds.) Evaluation and Management of Dysphagia 2020
Khalaf et.al Am J Medicine. 2018

# Treatment considerations

- Evaluate and treat GERD
  - GERD can be associated with spastic disorders
  - Smooth muscle relaxants may worsen untreated reflux
- Treat potential secondary causes of hypercontractile esophagus (mechanical obstruction/EGJOO)
- Opioids can be associated with shortened DL (DES), and increased DCI (hypercontractile esophagus)

Patel et. al. (eds.), Evaluation and Management of Dysphagia. 2020
Khalaf et.al Am J Medicine. 2018

# Treatment

- Medical therapy is first line > surgical therapy
- Tailor treatment based on symptoms
  - Obstructive symptoms (abnormal motor function) respond better to endoscopic therapy/surgery
  - Perceptive symptoms (consider hypersensitivity)

# Medical therapy

## Smooth muscle relaxants

- Concentrated peppermint oil
  - 5 drops in 10 ml water
  - 2 Altoid mints sublingual
- Nitrates, phosphodiestase-5 inhibitor
  - Sildenafil 25-50mg bid
- Calcium channel blockers
  - Diltiazem 60-90mg QID
  - Nifedipine 10mg 30 min qAC

## Neuromodulators

- Tricyclic antidepressants
  - Imipramine 50mg QHS
  - Amitriptyline/ Nortriptyline 10-25 mg upto 50-75mg QHS
- Anxiolytic
  - Trazodone 100-150mg QHS
- Selective Serotonin reuptake inhibitors
  - Fluoxetine 10-20 mg
  - Paroxetine 10-20 mg
  - Sertraline 25-50 mg daily
- Theophylline 200mg bid

Patel et. al. (eds.), Evaluation and Management of Dysphagia. 2020
Khalaf et.al Am J Medicine. 2018

# Treatment- Endoscopic/surgical

- Botulinum toxin injection to LES and 7 cm above (50% response)
- Extended surgical myotomy (14 cm in esophagus, 2 cm below EGJ)
  - 90-100% response in DES; 80% in hypercontractile esophagus
- POEM is emerging as a preferred therapy
  - 88% response in DES; 72% response in hypercontractile esophagus)
  - Potential complications:
    - Hyperactive spastic contractions complicate creation of the submucosal tunnel
    - Extended duration, increased postoperative pain
    - GERD

Patel et. al. (eds.), Evaluation and Management of Dysphagia. 2020
Khalaf et.al Am J Medicine. 2018
Khan et.al Dig Dis Sci 2017

# Summary

- Diagnosis of spastic disorders of the esophagus such as DES and hypercontractile esophagus requires a manometric diagnosis as well as relevant clinical symptoms
- Exclude secondary causes, treat associated GERD, wean off opiates
- Tailor therapy based on quality of symptoms
- Endoscopic and surgical therapy yields better results for obstructive symptoms