Dysmotility disoders of the eosophagus

- Dr Anas Bani Hani
- Associate Professor, The Hashemite University
- MBBS, MRCP, ESEGH

Introduction

- Symptoms related to the esophagus are among the most common in general medical as well as gastroenterologic practice. For example, dysphagia becomes more common with aging and affects up to 15% of persons age 65 or older.
- Heartburn, regurgitation, and other symptoms of GERD are also common.
- Mild symptoms of GERD rarely indicate severe underlying disease but must be addressed, especially if they have occurred for many years. Frequent or persistent dysphagia or odynophagia suggests an esophageal problem that necessitates investigation and treatment.
- Other less specific symptoms of possible esophageal origin include globus sensation, chest pain, belching, hiccups, rumination, and extraesophageal complaints like wheezing, coughing, sore throat, and hoarseness, especially if other causes have been excluded.
- A major challenge in evaluating esophageal symptoms is that the degree of esophageal damage often does not correlate well with the patient's or physician's impression of symptom severity. This is a particular problem in older patients, in whom the severity of gastroesophageal reflux–induced injury to the esophageal mucosa is increased despite an overall decrease in the severity of symptoms.

Dysphagia

- Definition:

Refers to the sensation that food is hindered in its passage from the mouth to the stomach. Most patients complain that food sticks, hangs up, or stops, or they feel that the food “just won’t go down right.”

Patients with a dilated esophagus, particularly due to achalasia, may incorrectly interpret dysphagia as regurgitation or even vomiting.

Pathophysiology

- Problem with the strength or coordination of the muscles required to move material from the mouth to the stomach or by a fixed obstruction somewhere between the mouth and stomach.
- The oropharyngeal swallowing mechanism and the primary and secondary peristaltic contractions of the esophageal body that follow usually transport solid and liquid boluses from the mouth to the stomach within 10 seconds.
- Mechanical narrowing of the esophageal lumen may interrupt the orderly passage of a food bolus despite adequate peristaltic contractions.

Differential Diagnosis and Approach

- Most patients can localize dysphagia to the upper or lower portion of the esophagus, although occasional patients with a distal esophageal cause of dysphagia will present with symptoms referred only to the suprasternal notch or higher.
- The approach to dysphagia can be divided into oropharyngeal and esophageal dysphagia,

Oropharyngeal Dysphagia

- With processes that affect the mouth, hypopharynx, and upper esophagus, the patient is often unable to initiate a swallow and repeatedly has to attempt to swallow.
- Patients frequently describe coughing or choking when they attempt to eat.
- Dysphagia that occurs immediately or within 1 second of swallowing suggests an oropharyngeal abnormality.
- Abnormalities of speech like dysarthria or nasal speech may be associated with oropharyngeal dysphagia

Box 13-1
Causes of Oropharyngeal Dysphagia

Neuromuscular Causes*

- Amyotrophic lateral sclerosis (ALS, Lou Gehrig's disease)
- CNS tumors (benign or malignant)
- Idiopathic UES dysfunction
- Manometric dysfunction of the UES or pharynx†
- Multiple sclerosis
- Muscular dystrophy
- Myasthenia gravis
- Parkinson's disease
- Polymyositis or dermatomyositis
- Postpolio syndrome
- Stroke
- Thyroid dysfunction

Structural Causes

- Carcinoma
- Infections of pharynx or neck
- Osteophytes and other spinal disorders
- Prior surgery or radiation therapy
- Proximal esophageal web
- Thyromegaly
- Zenker's diverticulum

CNS, central nervous system; UES, upper esophageal sphincter.

Esophageal Dysphagia

- Most patients with esophageal dysphagia localize their symptoms to the lower sternum or, at times, the epigastric region.
- To clarify the origin of symptoms of esophageal dysphagia, the answers to 3 questions are crucial:
- What type of food or liquid causes symptoms?
- Is the dysphagia intermittent or progressive?
- Does the patient have heartburn?

Common Causes of Esophageal Dysphagia

Motility (Neuromuscular) Disorders

Primary

- Achalasia
- Distal esophageal spasm
- Hypercontractile (jackhammer) esophagus
- Hypertensive LES
- Nutcracker (high-pressure) esophagus
- Other peristaltic abnormalities*

Secondary

- Chagas' disease
- Reflux-related dysmotility
- Scleroderma and other rheumatologic disorders

Structural (Mechanical) Disorders

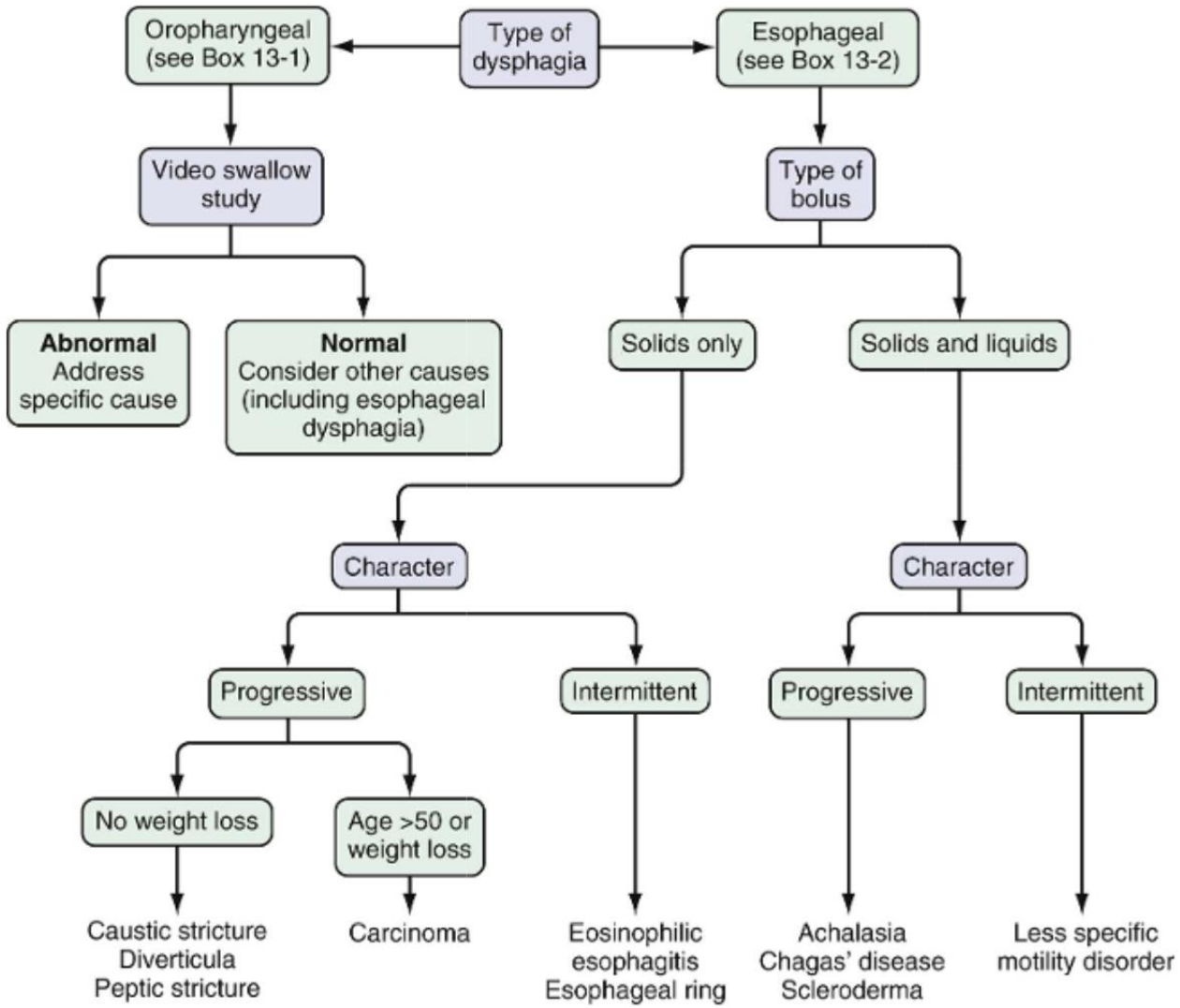
Intrinsic

- Carcinoma and benign tumors
- Diverticula
- Eosinophilic esophagitis
- Esophageal rings and webs (other than Schatzki ring)
- Foreign body
- Lower esophageal (Schatzki) ring
- Medication-induced stricture
- Peptic stricture

Extrinsic

- Mediastinal mass
- Spinal osteophytes
- Vascular compression

LES, lower esophageal sphincter.


Type of dysphagia
Oropharyngeal (see Box 13-1)
Esophageal (see Box 13-2)
Video swallow study
Type of bolus
Abnormal
Address specific cause
Normal
Consider other causes (including esophageal dysphagia)
Solids only
Solids and liquids
Character
Character
Progressive
Intermittent
Progressive
Intermittent
No weight loss
Age >50 or weight loss
Caustic stricture
Diverticula
Peptic stricture
Carcinoma
Eosinophilic esophagitis
Esophageal ring
Achalasia
Chagas' disease
Scleroderma
Less specific motility disorder

Eosinophilic Esophagitis

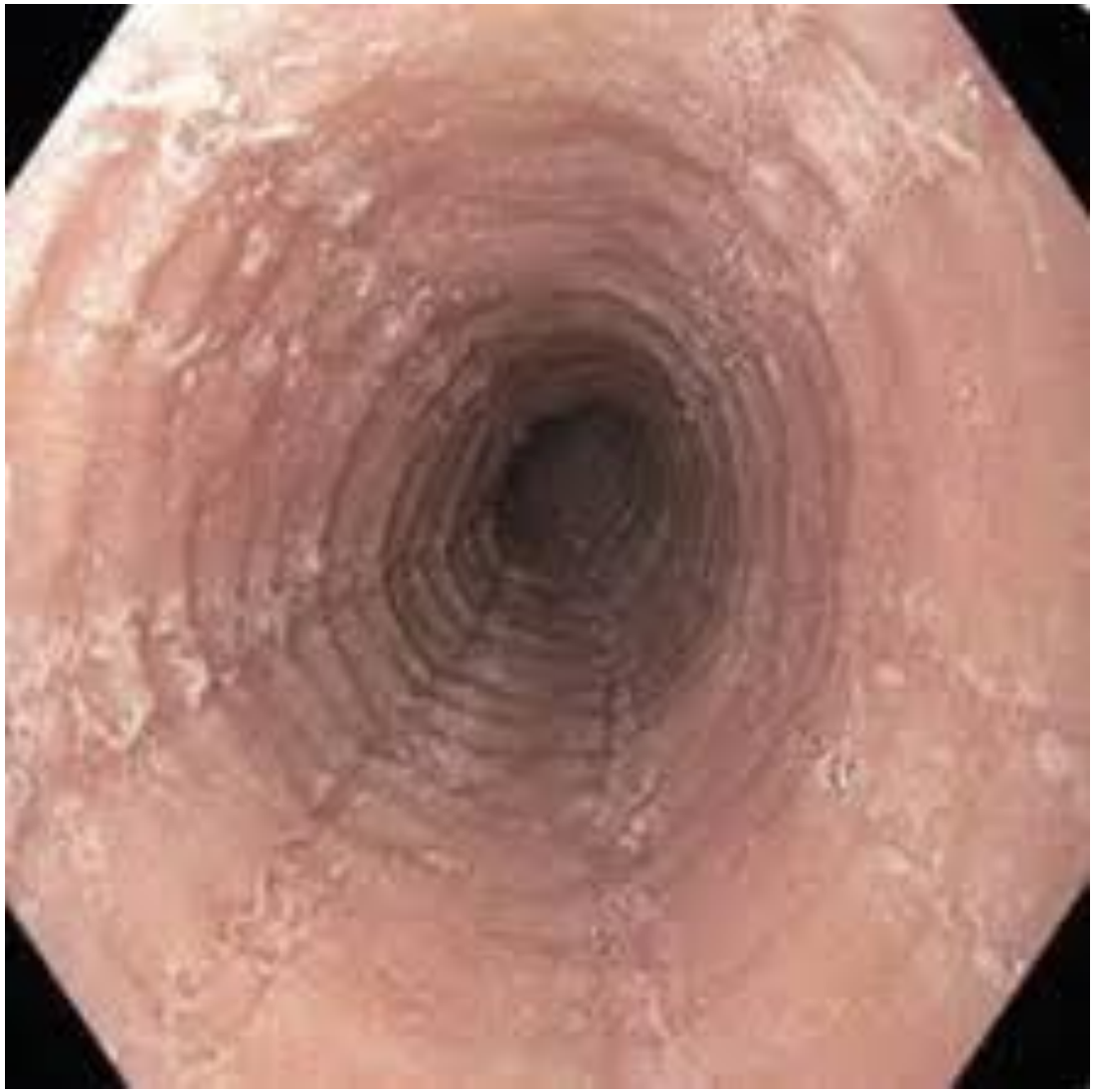
- Eosinophilic esophagitis (EoE) is a clinicopathologic disease characterized by symptoms resulting from esophageal dysfunction accompanied by pathologic evidence of a predominantly eosinophilic inflammatory response confined to the esophagus.
- Many disorders are accompanied by eosinophil infiltration in the esophagus: EoE, eosinophilic gastroenteritis, GERD, parasitic and fungal infections, IBD, HES, esophageal leiomyomatosis, myeloproliferative disorders, carcinomatosis, polyarteritis, allergic vasculitis, collagen vascular diseases (e.g., scleroderma), pemphigus vegetans, and drug injury.

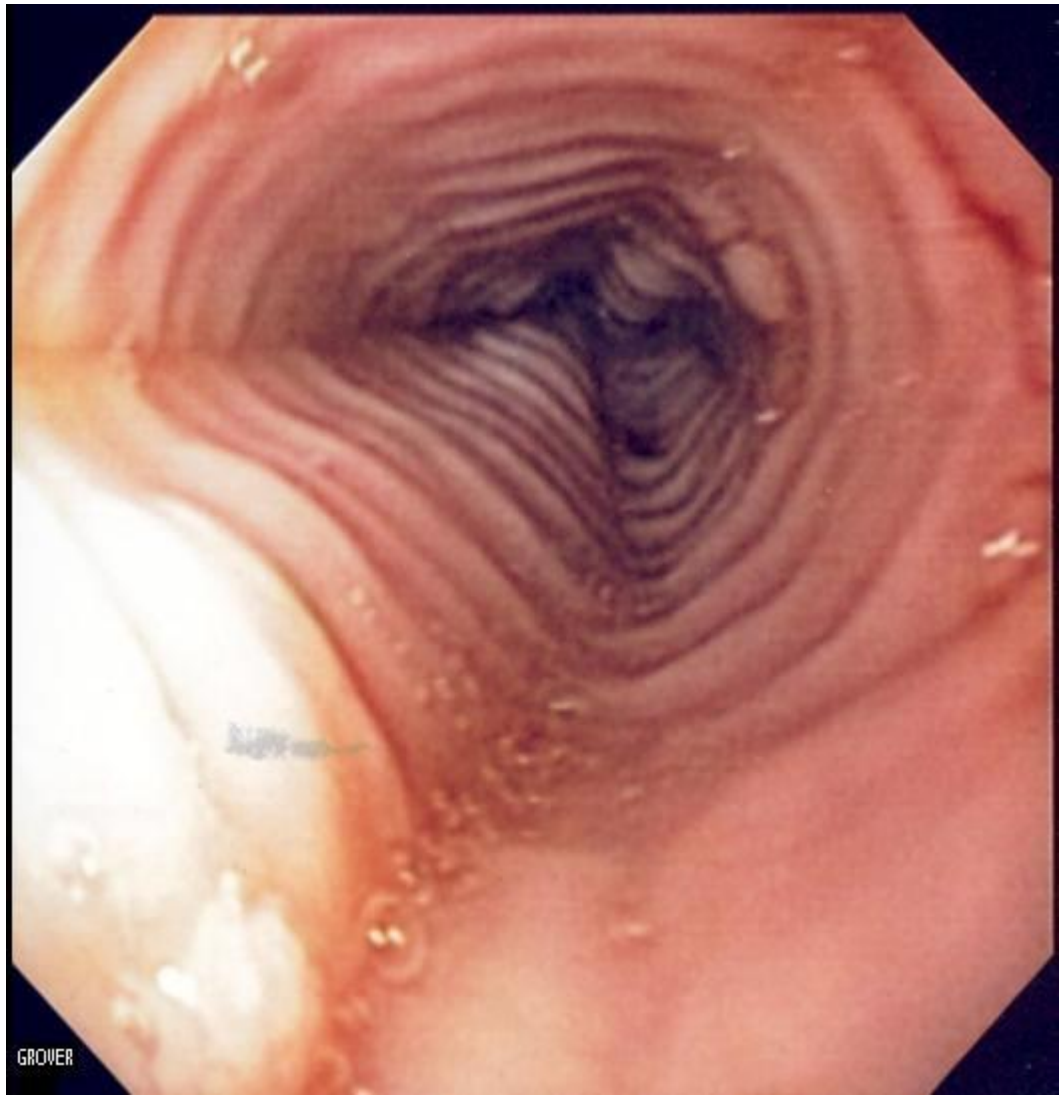
Etiology

- The etiology of EoE is poorly understood, but food allergy has been implicated as a primary contributor.
- EoE representing the second most common cause of chronic esophagitis.

Clinical Features and Diagnosis

- The new consensus criteria published in 2011 emphasize that EoE is an antigen-driven, immune-triggered disease that requires adequate treatment with PPI therapy prior to finding more than 15 eosinophils/high-power field (HPF; peak value) in the esophagus.
- Symptoms include difficulties with eating, failure to thrive, chest and/or abdominal pain, dysphagia, and food impaction.
- Solid-food dysphagia continues to be the most common presenting symptom.
- Food impaction necessitating endoscopic bolus removal occurs in 33% to 54% of adults with EoE.
- Endoscopy with esophageal biopsy remains the only reliable diagnostic test for EoE.




GROVER

Treatment

- No drug therapy is currently approved for the treatment of EoE by the US Food and Drug Administration.
- It has been shown that dietary therapy frequently improves symptoms and reduces the number of eosinophils in esophageal biopsies of patients with primary EoE (allergic or non-allergic subtypes).
- A diet consisting of an amino acid–based formula, termed an *elemental diet*, or avoidance of the most common allergenic foods (cow's milk, soy, wheat, egg, peanut/tree nuts, and seafood/shellfish), termed the *six-food elimination diet* (SFED).
- Systemic or topical glucocorticoids have also been used to treat EoE with satisfactory results
- Systemic glucocorticoids are used for acute exacerbations, whereas topical glucocorticoids are used to provide long-term control.

Prognosis

- EoE requires prolonged treatment, similar to allergic asthma.
- chronic EoE, if left untreated, can develop into progressive esophageal scarring and dysfunction.
- EoE complications include food impaction, esophageal stricture, narrow-caliber esophagus, and esophageal perforation.

ACHALASIA

Esophageal Motility Disorders

- Retained material within the esophagus or excessive reflux from the stomach is indicative of a disorder of peristalsis or of sphincter competence.
- Processes of esophageal dysmotility may be either dysfunction of deglutitive relaxation (including failure of sphincter relaxation) and/or of propagated excitation.
- Esophageal motor disorders are not diagnosed on histopathological grounds; diagnosis depends upon identification of functional aberrations. A possible exception is achalasia in which tissue may be available and the condition is characterized by a quantifiable reduction in the number of ganglion cells in the myenteric plexus. Hence, esophageal manometry, in conjunction with radiologic findings, is typically used to evaluate peristaltic function.

Achalasia

- Deglutition initiates a peristaltic wave down the esophagus and also triggers the relaxation of the LES allowing food to enter the stomac
- Achalasia is greek for “does not relax”
- Definition: Failure of the LES to relax appropriately with swallowing and absent peristalsis in the smooth muscle esophagus.
- Achalasia occurs as a result of degeneration of ganglion cells in the myenteric plexus of the distal esophageal body and LES.
- Epidemiology:
  - Incidence 1/100,000
  - Affects males and females equally
  - Presents ages 25-60

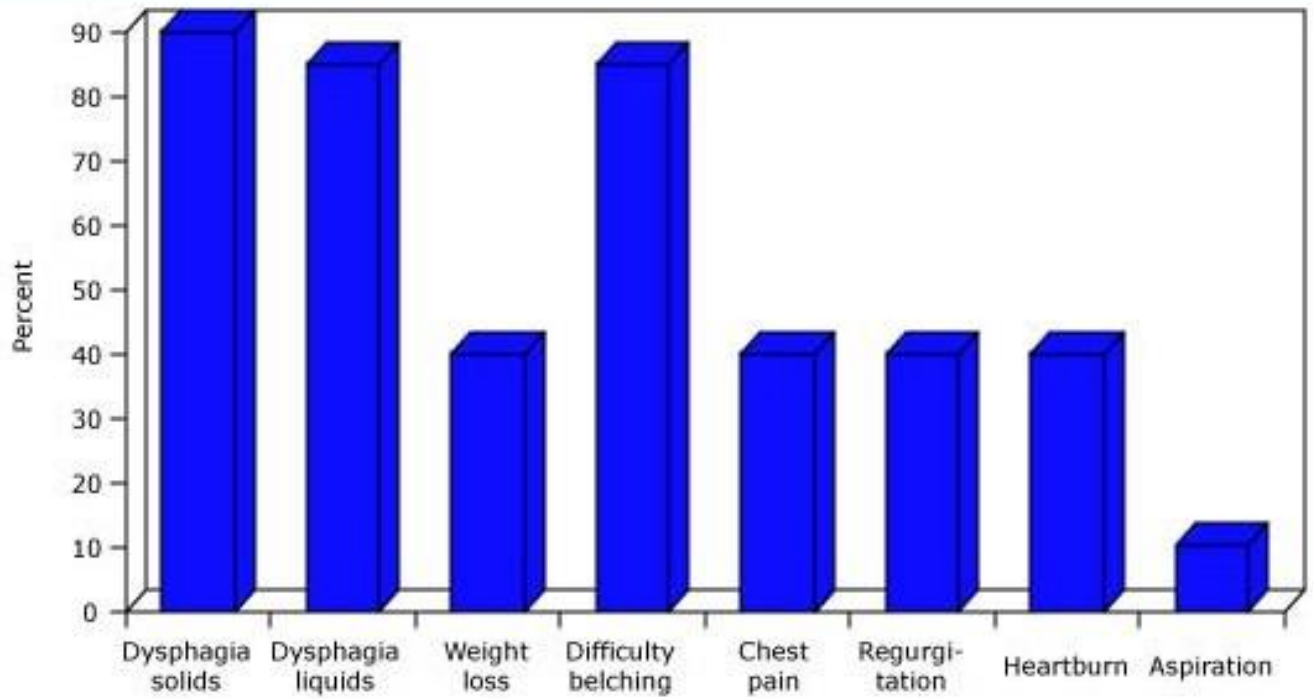
Achalasia

- Pathogenesis
  - Loss of ganglion cells within the myenteric (Auerbach's) plexus (longer the disease the fewer ganglion cells present)
  - Loss of inhibitory (NO) ganglion function resulting in impaired relaxation. Intact cholinergic (excitatory)
  - Possible autoimmune disease(viral insult) involving latent HSV-1
  - Likely genetic component
  - Allgrove Syndrome(AAA): rare autosomal resessive disorder associated with Achalasia-Addisonianism-Alacrimia

Achalasia- Clinical Presentation

- Symptoms:
  - Dysphagia to solids and liquids (91% and 85% respectively)- Most common
  - Regurgitation- 2nd most common
  - Difficult belching
  - Chest pain
  - Heartburn
- +/- weight loss (mild)

Frequency of the symptoms of achalasia



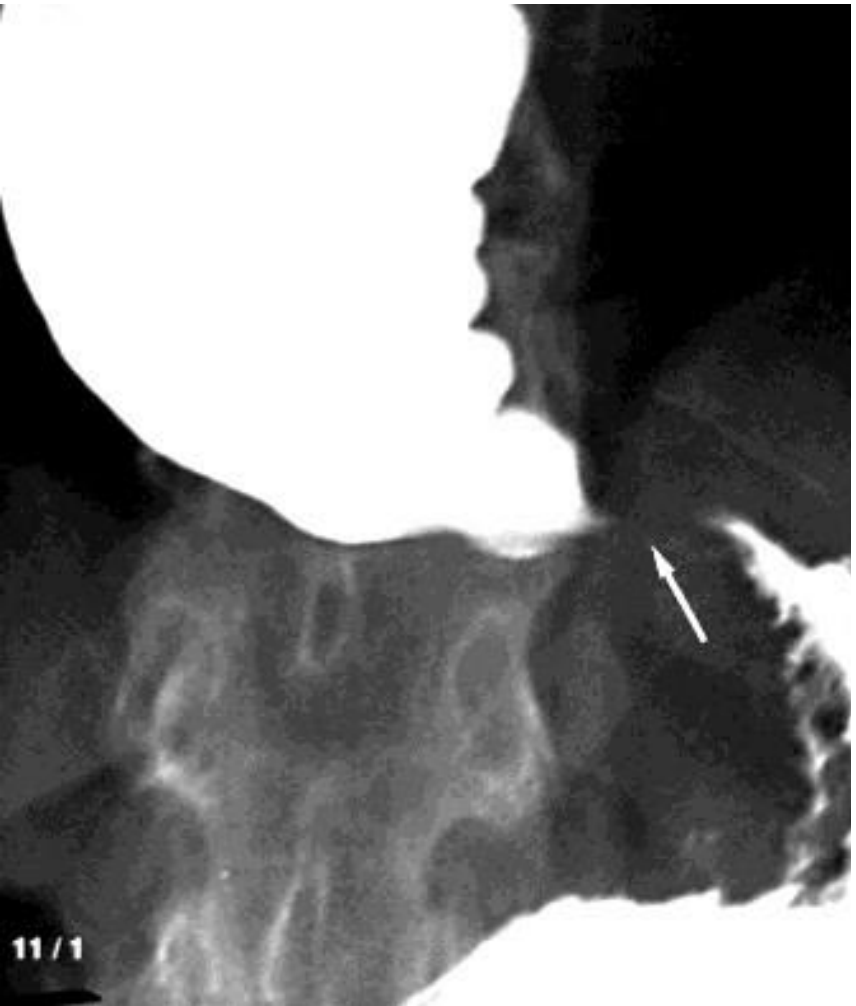
Achalasia- Diagnosis

- CXR- widened mediastinum and absence of gastric bubble
- Manometry-confirmatory
- Barium swallow-Primary screening
- EGD- rule out pseudoachalasia

EGD

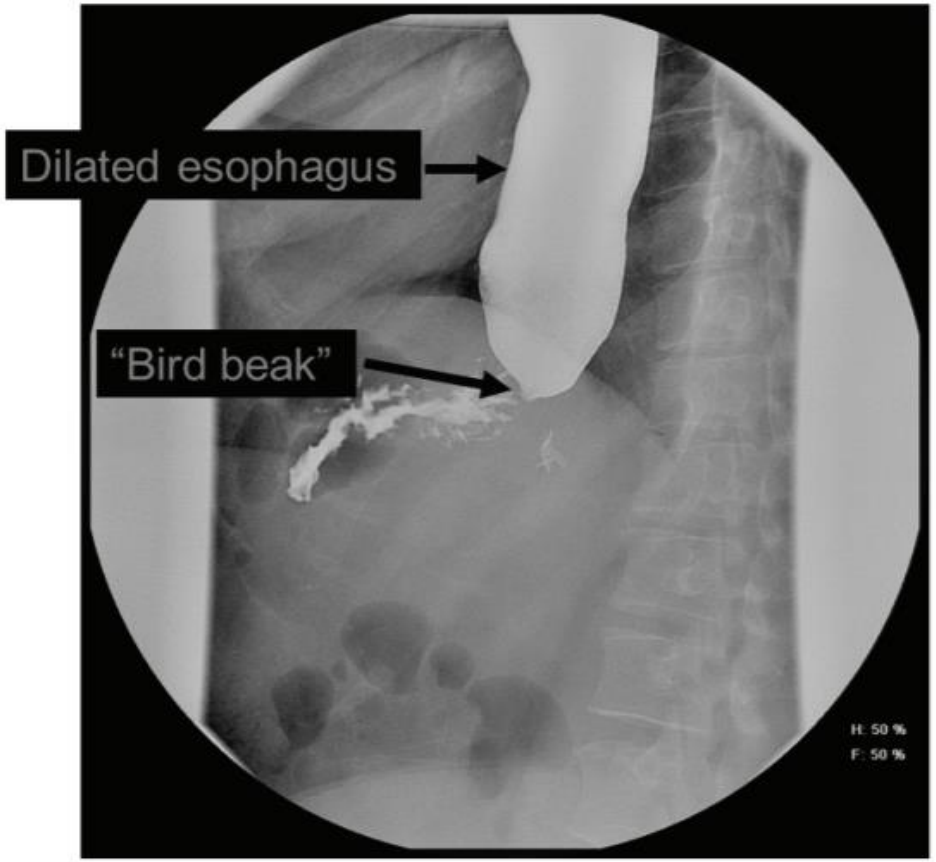
- Recommended in all achalasia patients to rule out malignancy or “pseudoachalasia” (excessive wt loss, symptoms <6mos, >60 yo)
- Examine cardia of stomach well for malignancy
- Dilated esophagus with retained food
- Esophageal stasis predisposes to candida esophagitis

Barium swallow

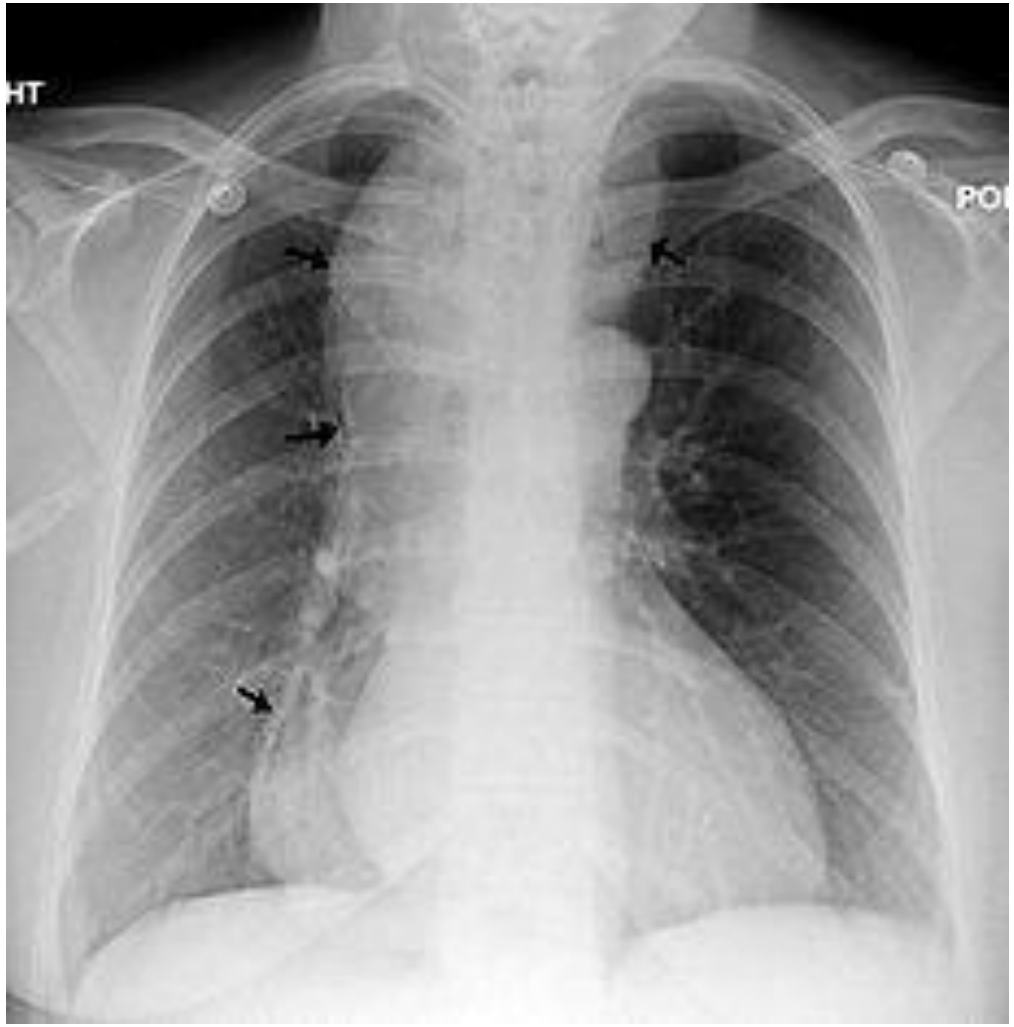


Vigorous achalasia

Barium swallow in a patient with "vigorous" achalasia. In addition to the narrowing at the esophagogastric junction, there are multiple, nonperistaltic muscular contractions in the dilated esophagus.



X-ray

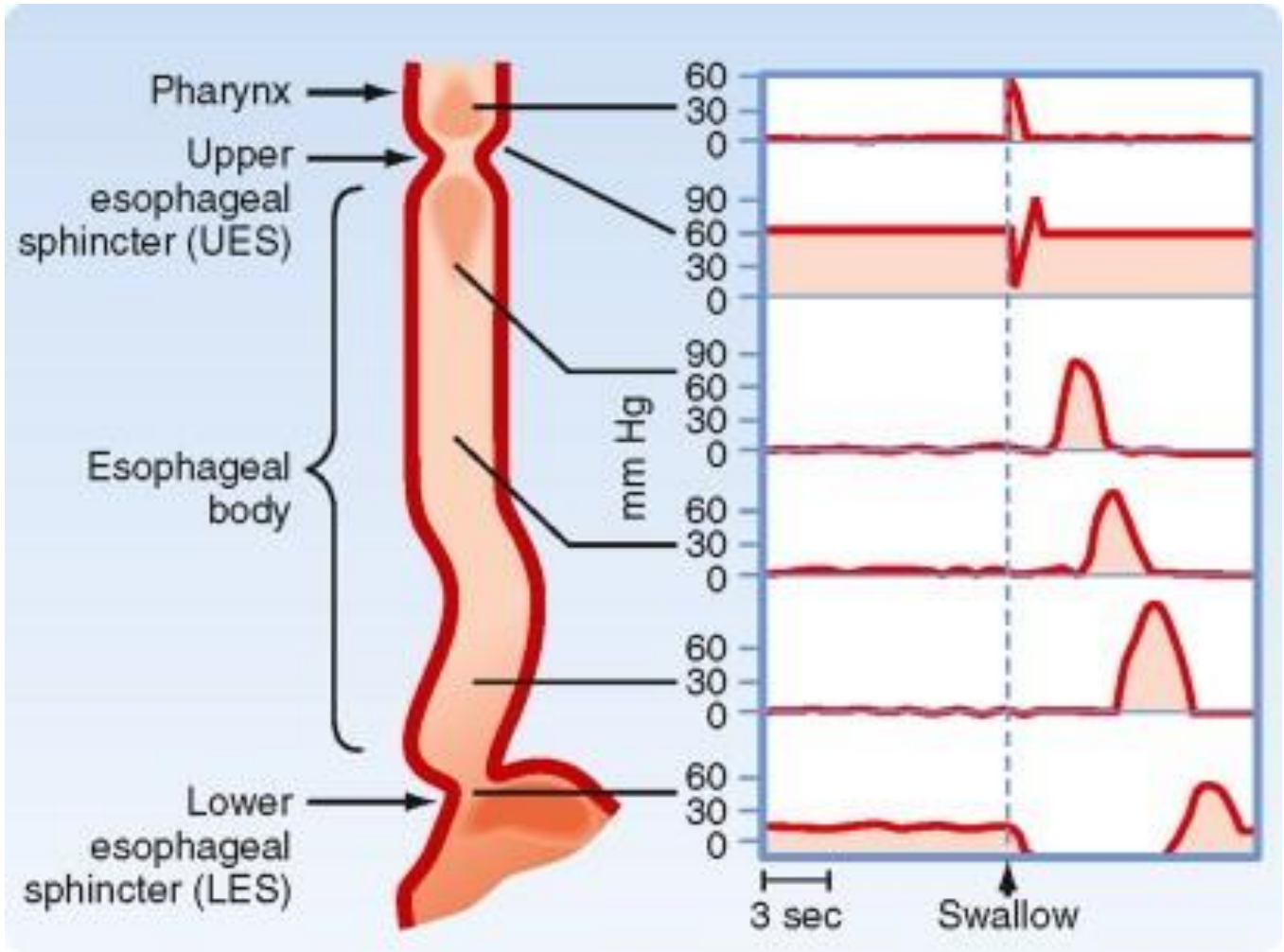


Standard Manometry

- Elevated LES pressure (usually >45mmHg)
- Incomplete LES relaxation (normal <8mmHg)
- Aperistalsis (there can still be contractions)

- "Vigorous" Achalasia- most contractions in Achalasia are low amplitude but some patient's have high amplitude contractions (37-60mmHg)

Normal Manometry

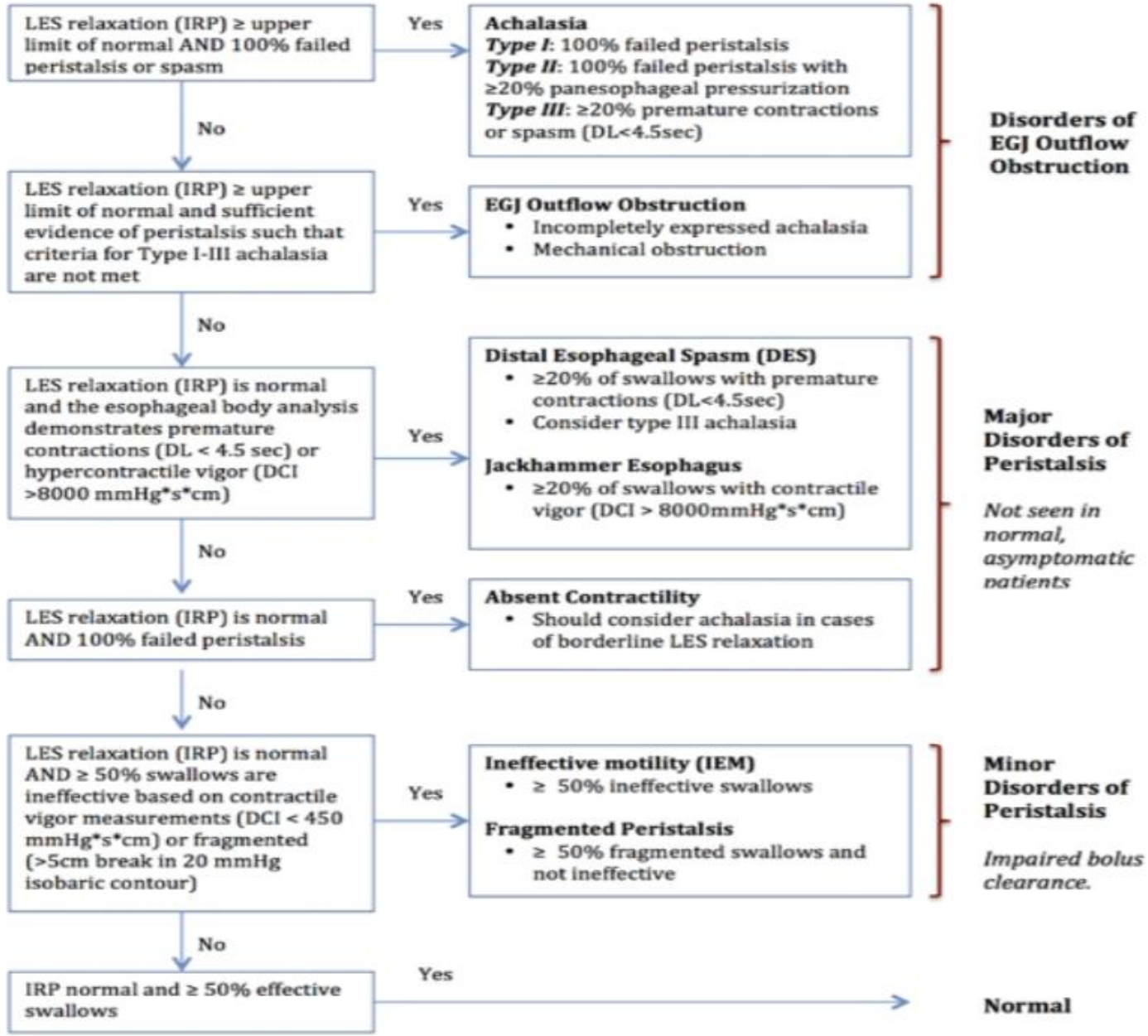


HRM

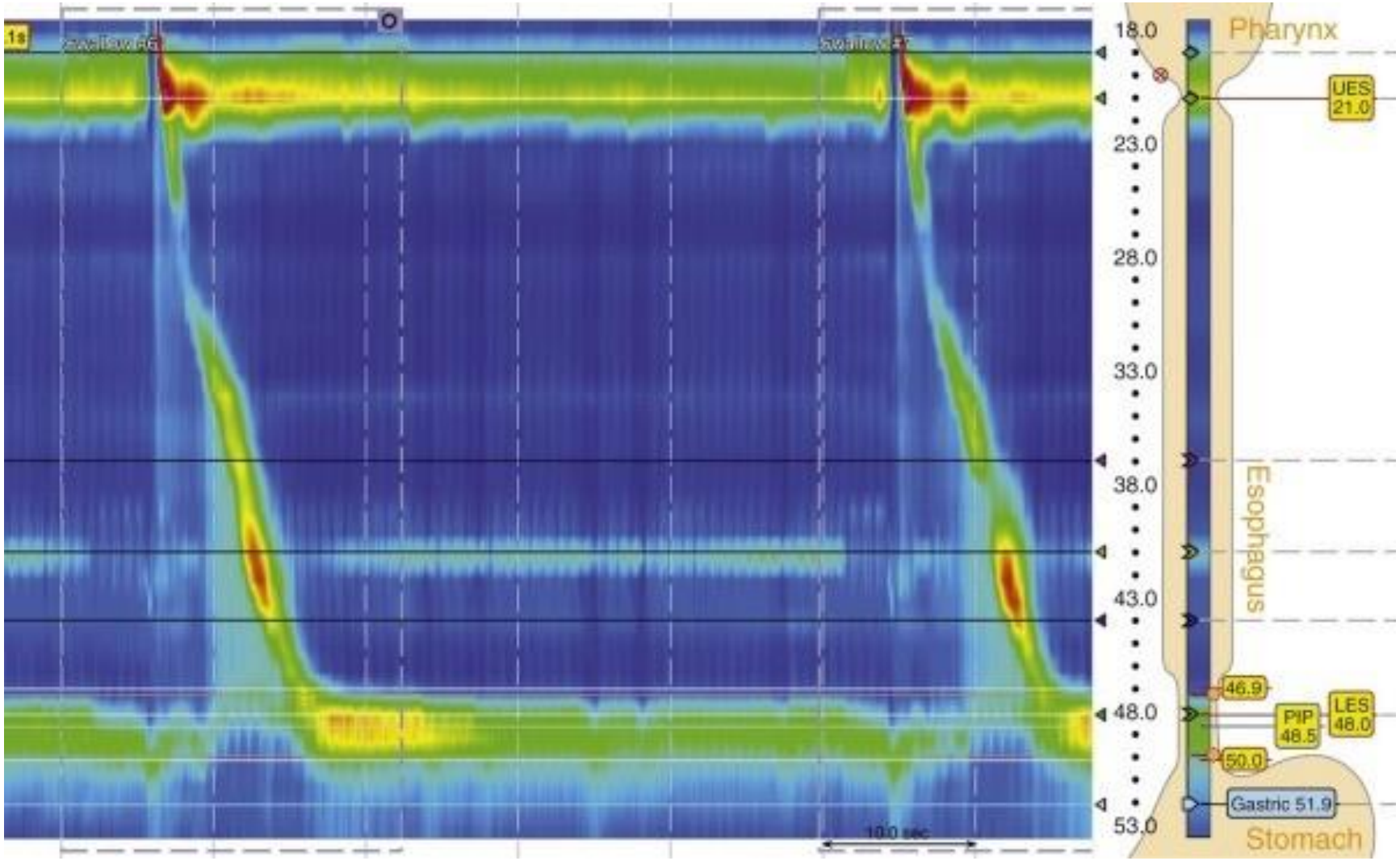
- Standard Manometry has 3-8 sensors at 3-5cm apart
- HRM-36 sensors 1cm apart
- Chicago Classification is used to categorize esophageal motility disorders

Figure 10.3

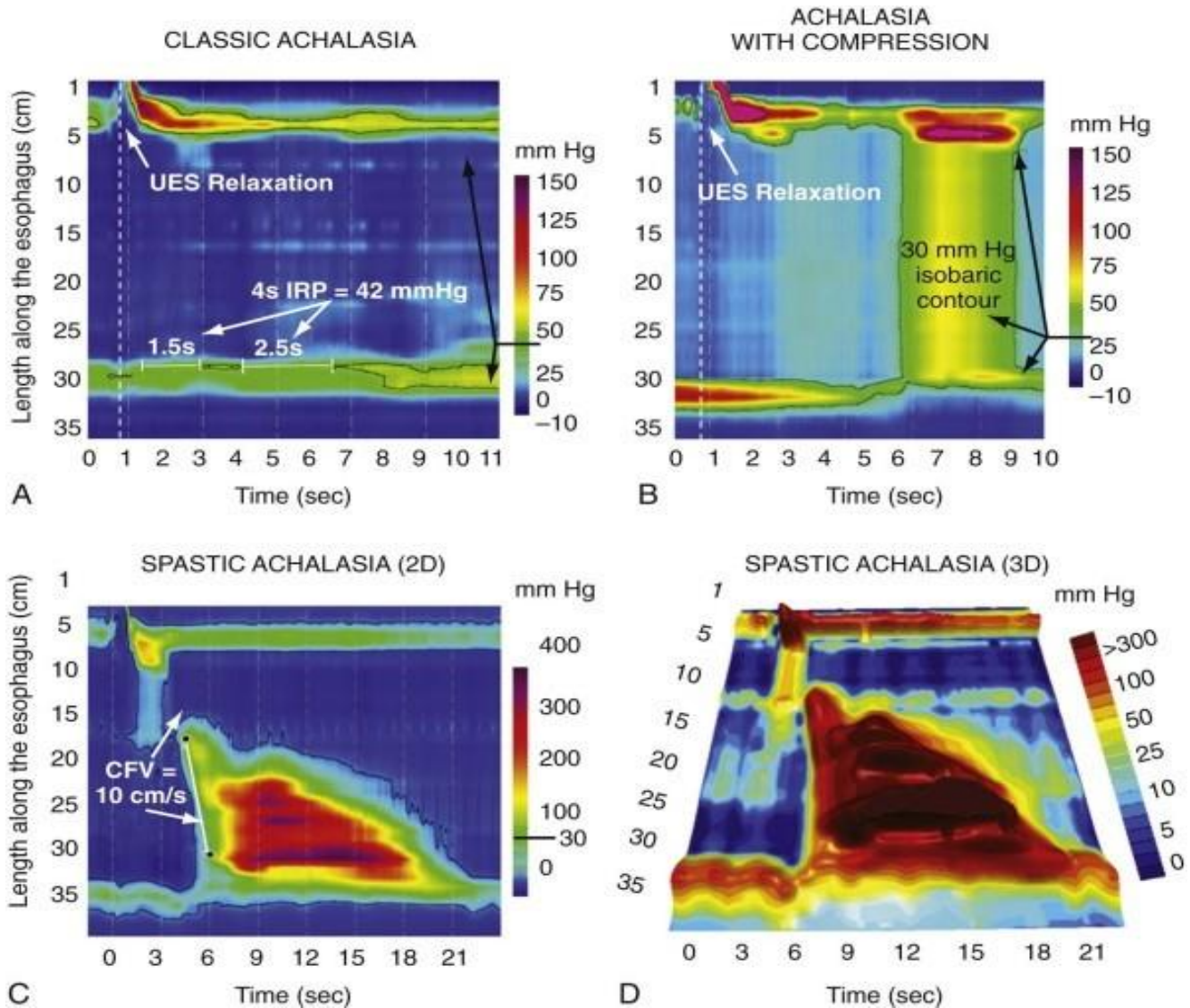
Chicago classification (v3) algorithm



Normal HRM


18.0
23.0
28.0
33.0
38.0
43.0
48.0
53.0
Pharynx
UES
21.0
Esophagus
46.9
PIP
48.5
LES
48.0
50.0
Gastric 51.9
Stomach

Achalasia- HRM



Achalasia Subgroups

- Type I: Classic achalasia
- Type II: Achalasia with compartmentalized panesophageal pressurization (>30mmHg)
- Type III: Achalasia with spastic contractions (with or without compartmentalized pressurization)
- Type II and III="vigorous achalasia" Historical correlation:
- Chest pain more prevelant with Type II and III

Treatment

- Medical Therapy
  - Nitrates and CCB (nifedipine), relax smooth muscle
  - Used for pt's unwilling or unable to undergo more invasive therapy
  - Variable success
- Botulinum Toxin
  - Injected into LES (25units in 4 quadrants)
  - Poisons the excitatory (acetylcholine) neurons
  - Success rates of 80% however relief wanes gradually to 41% at 12 mos. Requires repeat injections
  - ? Increases intraoperative perforation and myotomy failure

Treatment

- Dilation
  - Bougie
    - Temporary relief but lower risk of perforation
  - Pneumatic
    - Forceful dilation, tears muscle fibers
    - Stepwise approach: 3.0cm ->3.5cm ->4.0cm
    - High success rate (85%)
    - 1.6% perforation rate
    - Gradual waning of success rate with repeat dilations
    - Consider additional therapy for persistent dysphagia after 2-3 dilations
    - Consider PPI therapy

Treatment

- Surgical Myotomy (Heller and Open myotomy)
  - LES is cut
  - Often partial wrap (Dor or Toupet fundoplication) to prevent reflux (No nissen, worse dysphagia)
  - High success rates (Open, 70%-85% at 10 yrs and 65-73% at 20-30 yrs) and low recurrence
  - Presence of low LES pressure or dilated esophagus predicted higher rate of failure with Heller
  - High cost, longer recovery, GERD
  - Complication rate is higher if surgical myotomy performed after endoscopic therapy
  - Pneumatic dilation and Heller are comparable

Treatment

- Type I- Heller Myotomy
- Type II- good response to all therapy
- Type III- poor response to all therapy

Complications

- Aspiration PNA
- Epiphrenic diverticulum
- SCC>Adenocarcinoma
  - 16 fold increase
  - Presents ~15 years after diagnosis of achalasia

Conclusion

- Suspect achalasia in patients with dysphagia to solids and liquids along with regurgitation
- If patient can tolerate, preferred treatment is surgical myotomy or pneumatic dilation…discuss risks and benefits with the patient and consider institutional expertise.
- Post PD LES pressure <10mmHg is goal

Thank you

Questions???