ACHALASIA

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Esophageal Motility Disorders

Primary:

- Achalasia
- Distal Esophageal Spasm (DES)
- GERD

Secondary

- Pseudoachalasia (malignancy…paraneoplastic vs structural)
- Chagas
- Scleroderma
- Postoperative (fundoplication, gastric band)
- Parkinsons…and more

Achalasia- Introduction

- Deglutition initiates a peristaltic wave down the esophagus and also triggers the relaxation of the LES allowing food to enter the stomach
- Achalasia is greek for “does not relax”
- Patients primarily present with regurgitation and dysphagia

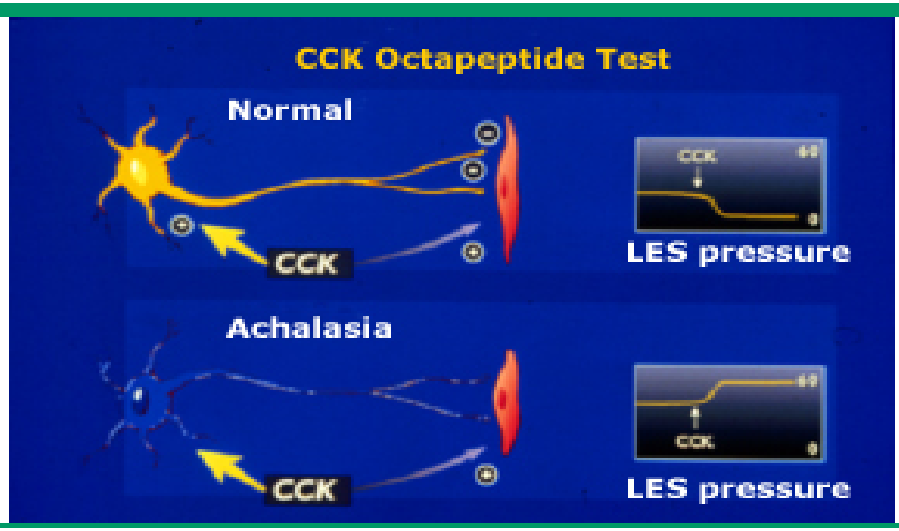
Achalasia

- Definition: Impaired relaxation of LES and aperistalsis
- 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)
  - CCK octapeptide test
  - Possible autoimmune disease(viral insult) involving latent HSV-1
  - Likely genetic component
  - Allgrove Syndrome(AAA): rare autosomal resessive disorder associated with Achalasia-Addisonianism-Alacrimia

The CCK octapeptide test for achalasia



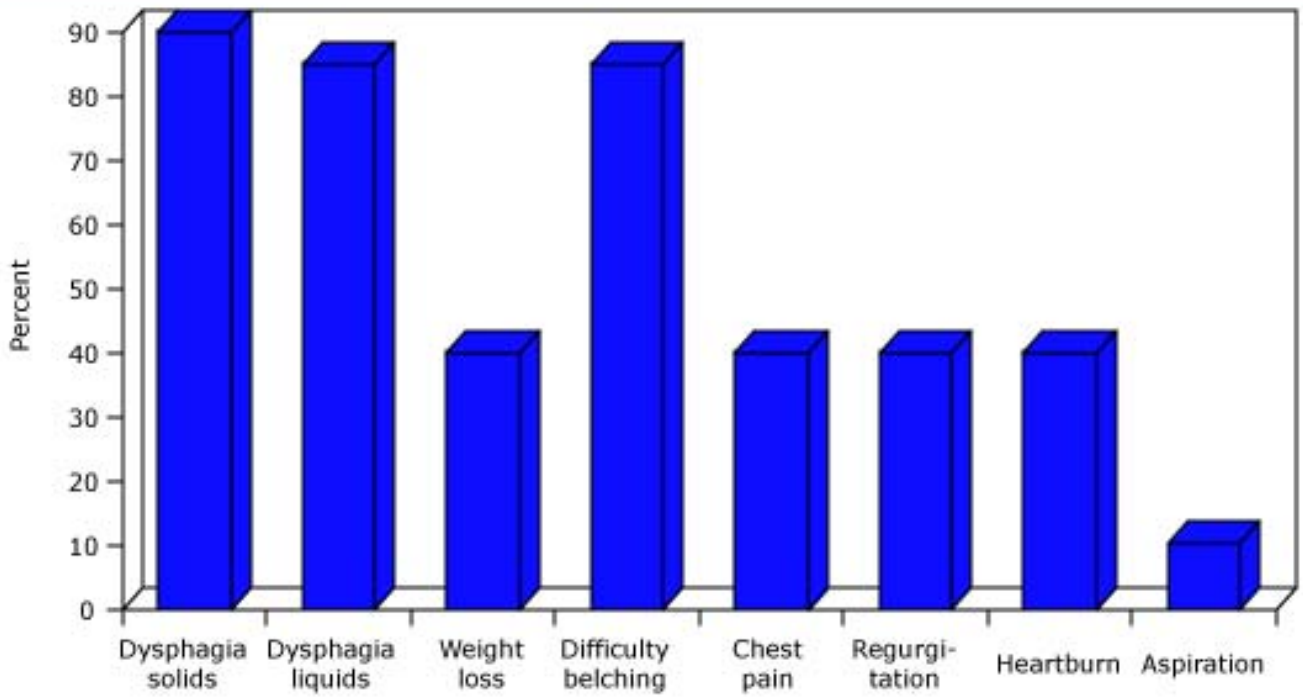
CCK has a dual effect on the lower esophageal sphincter (LES): it stimulates smooth muscle contraction and the release of inhibitory neurotransmitters. Administration of CCK normally causes the LES pressure to fall because the effect on inhibitory neurotransmitters is greater. In patients with achalasia, CCK causes the lower esophageal sphincter pressure to rise.

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

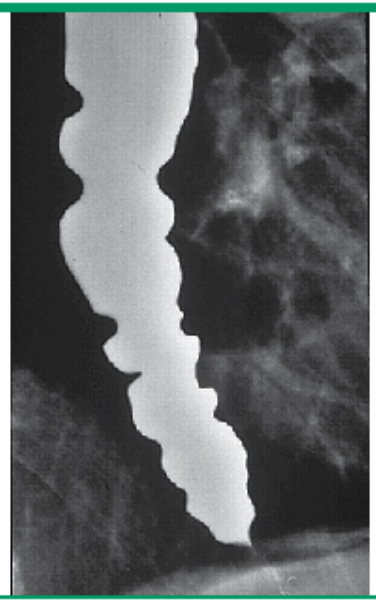
- CXR- widened mediastinum and absence of gastric bubble
- Manometry-confirmatory
- Barium swallow-Primary screening
- EGD- rule out pseudoachalasia

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

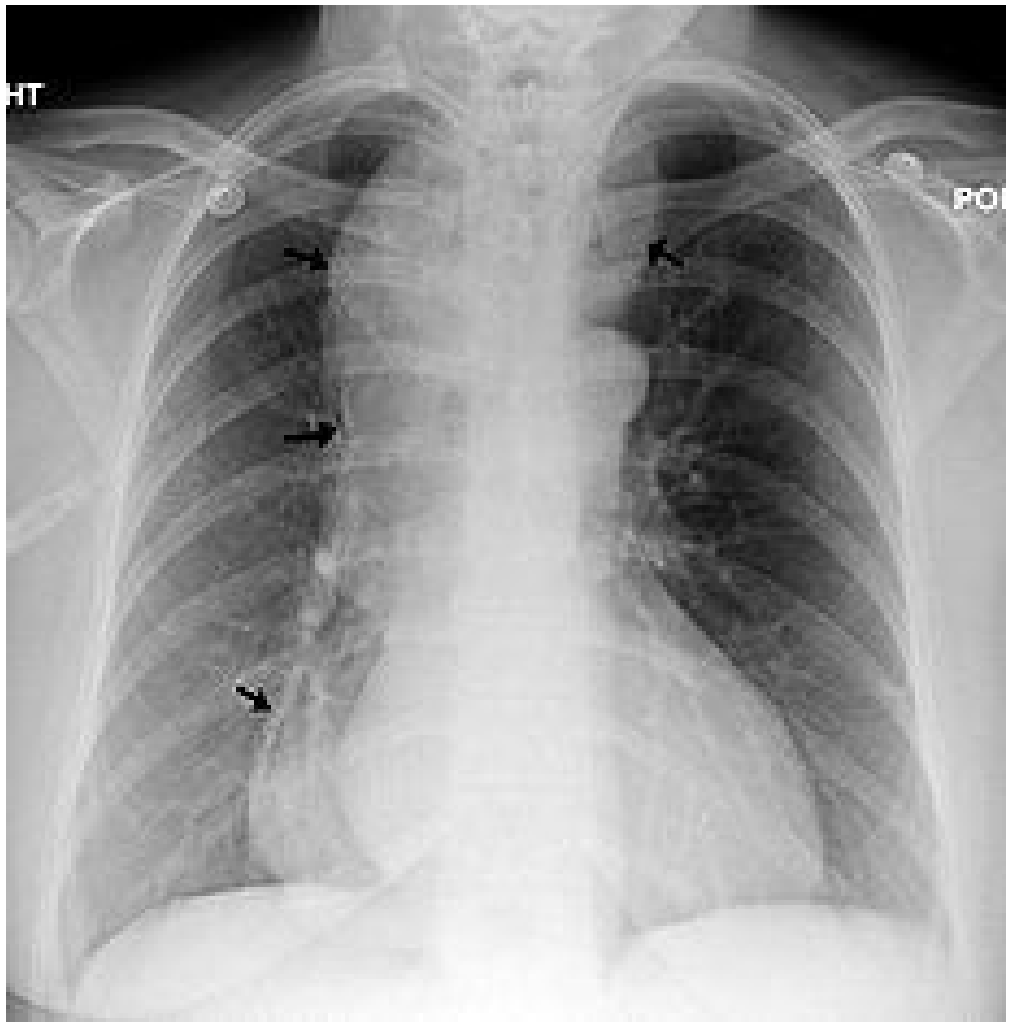
Barium swallow

Vigorous achalasia

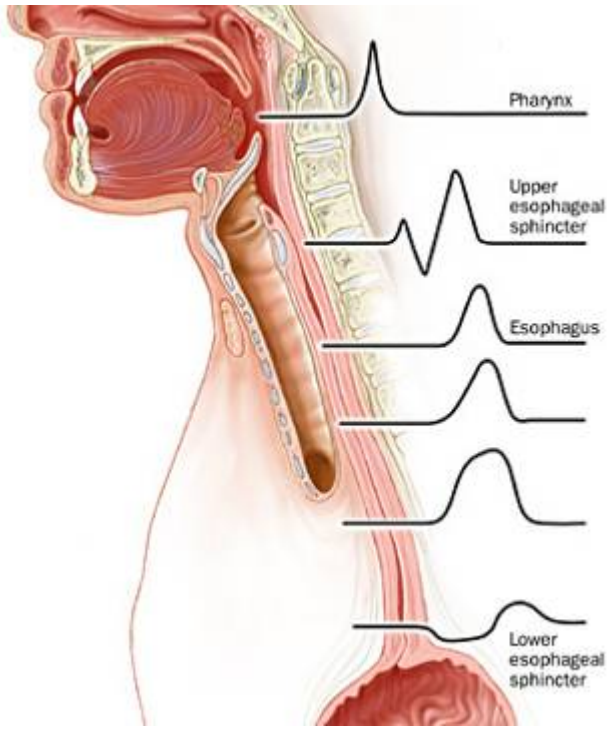


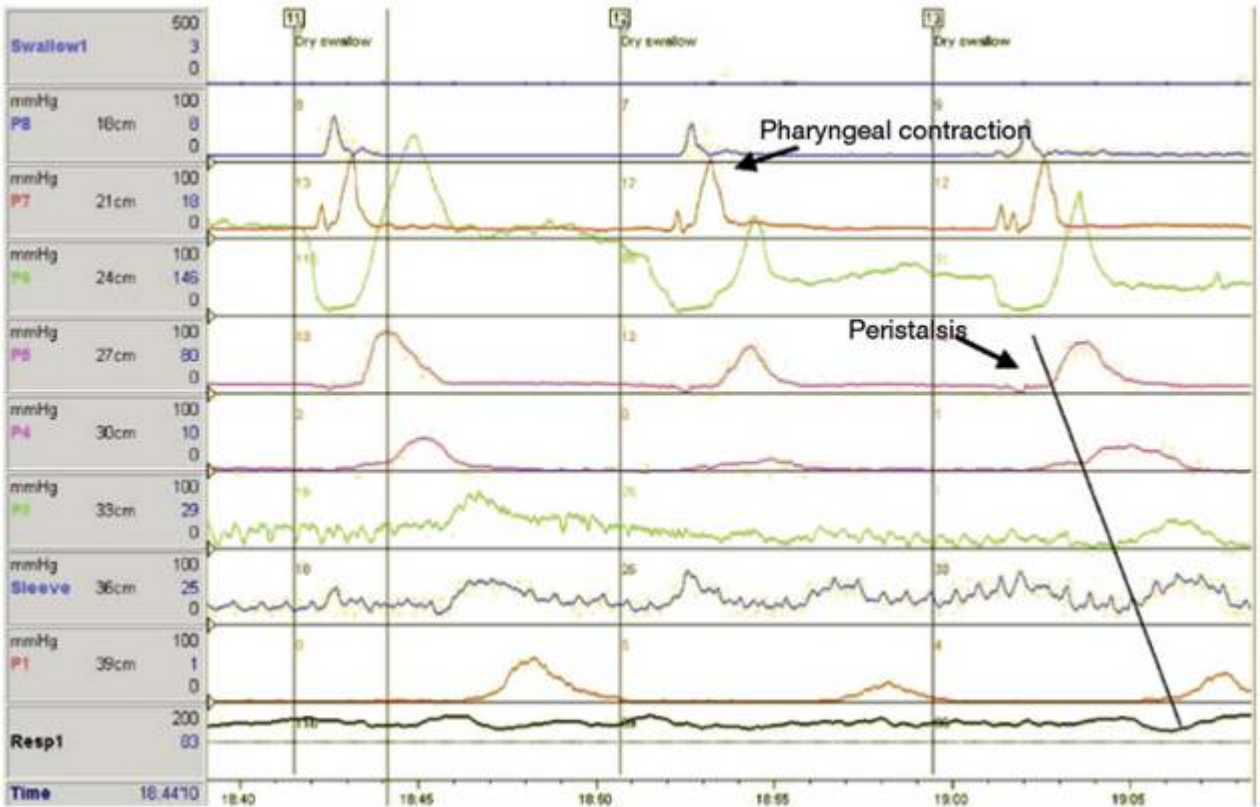
Barium swallow in a patient with "vigorous" achalasia. In addition to the narrowing at the esophagogastric junction, there are multiple, nonperistaltic muscular contractions in the dilated esophagus.

X-ray

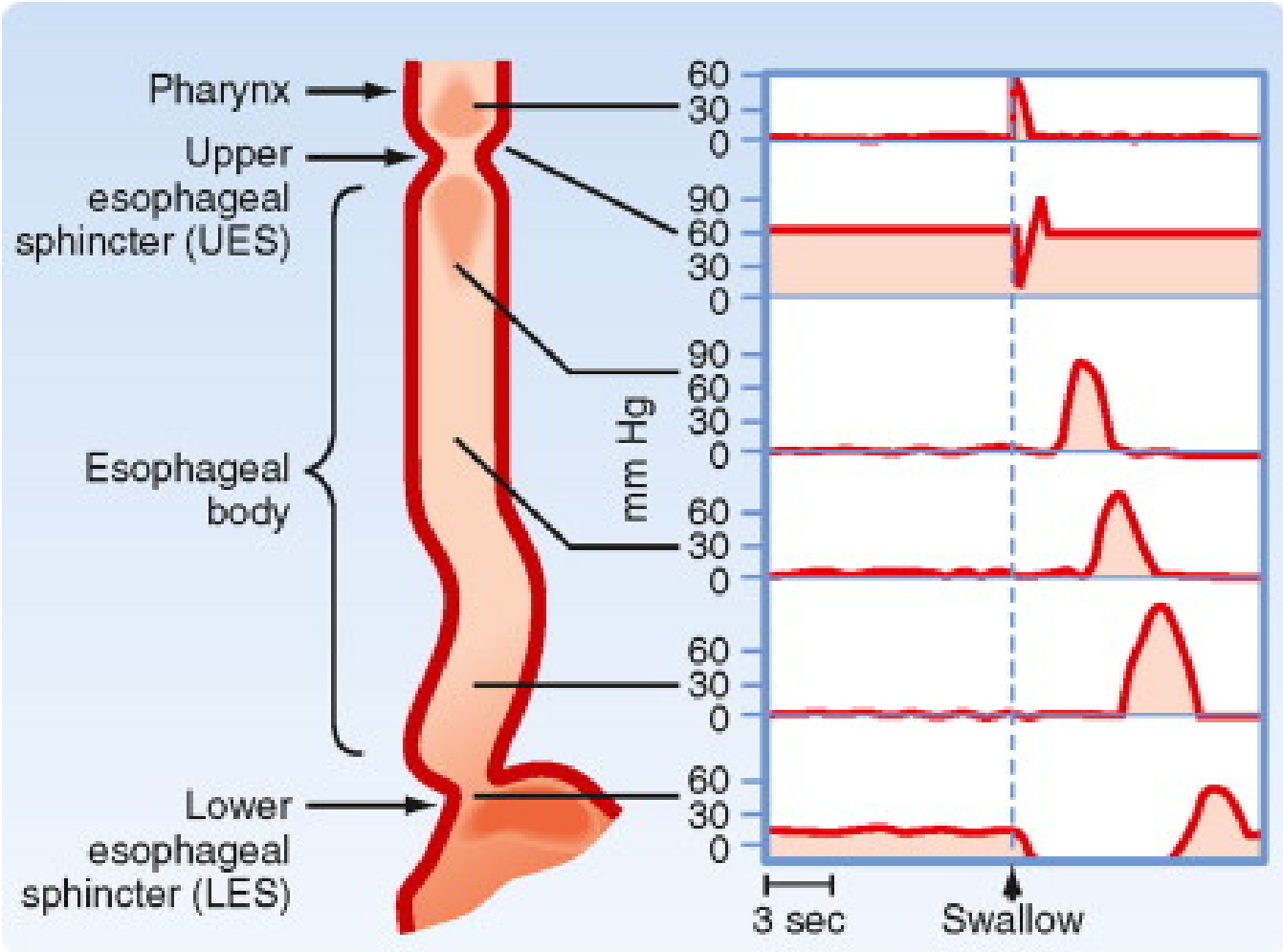


Normal Manometry





Normal Manometry



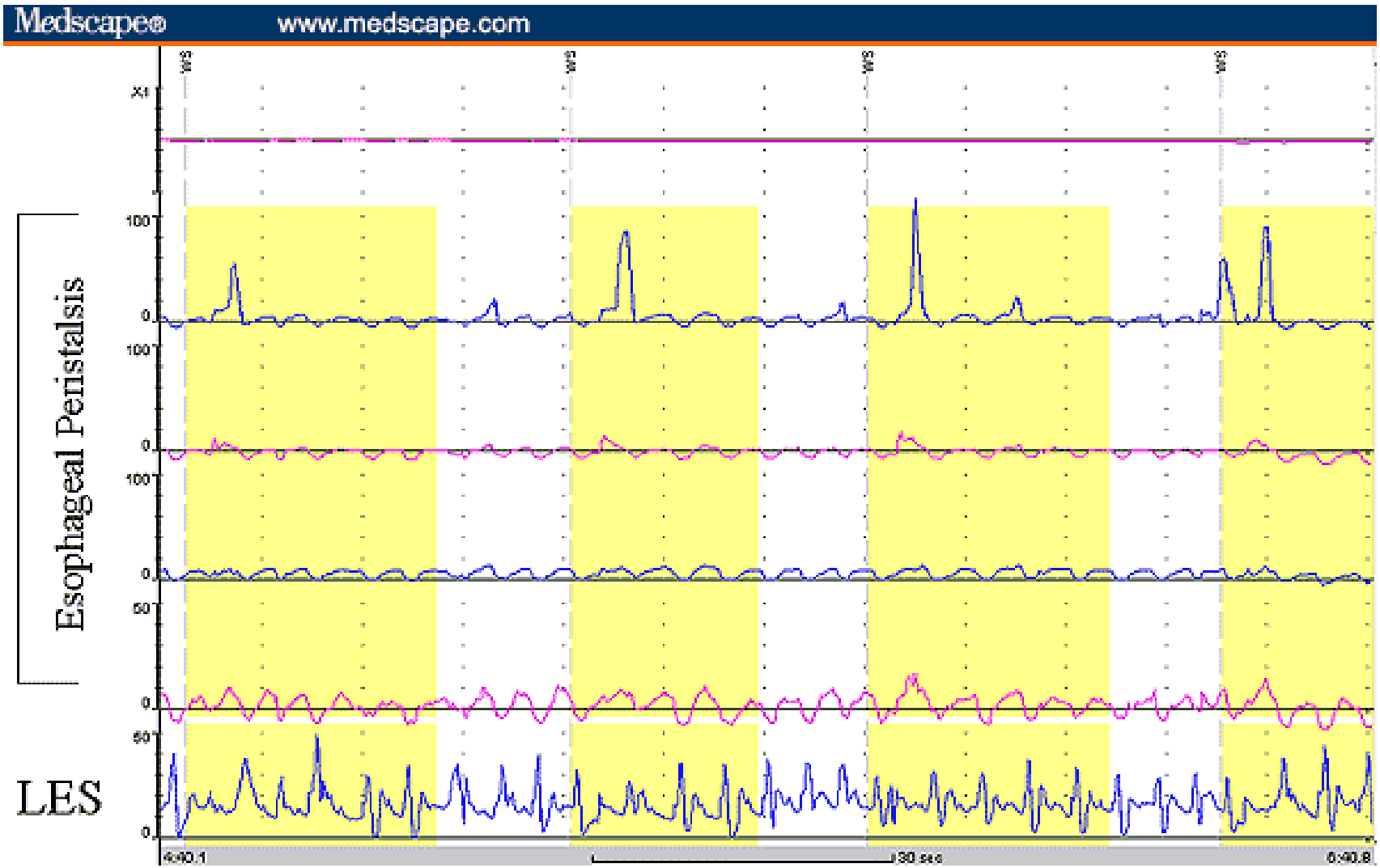

ZIM

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)

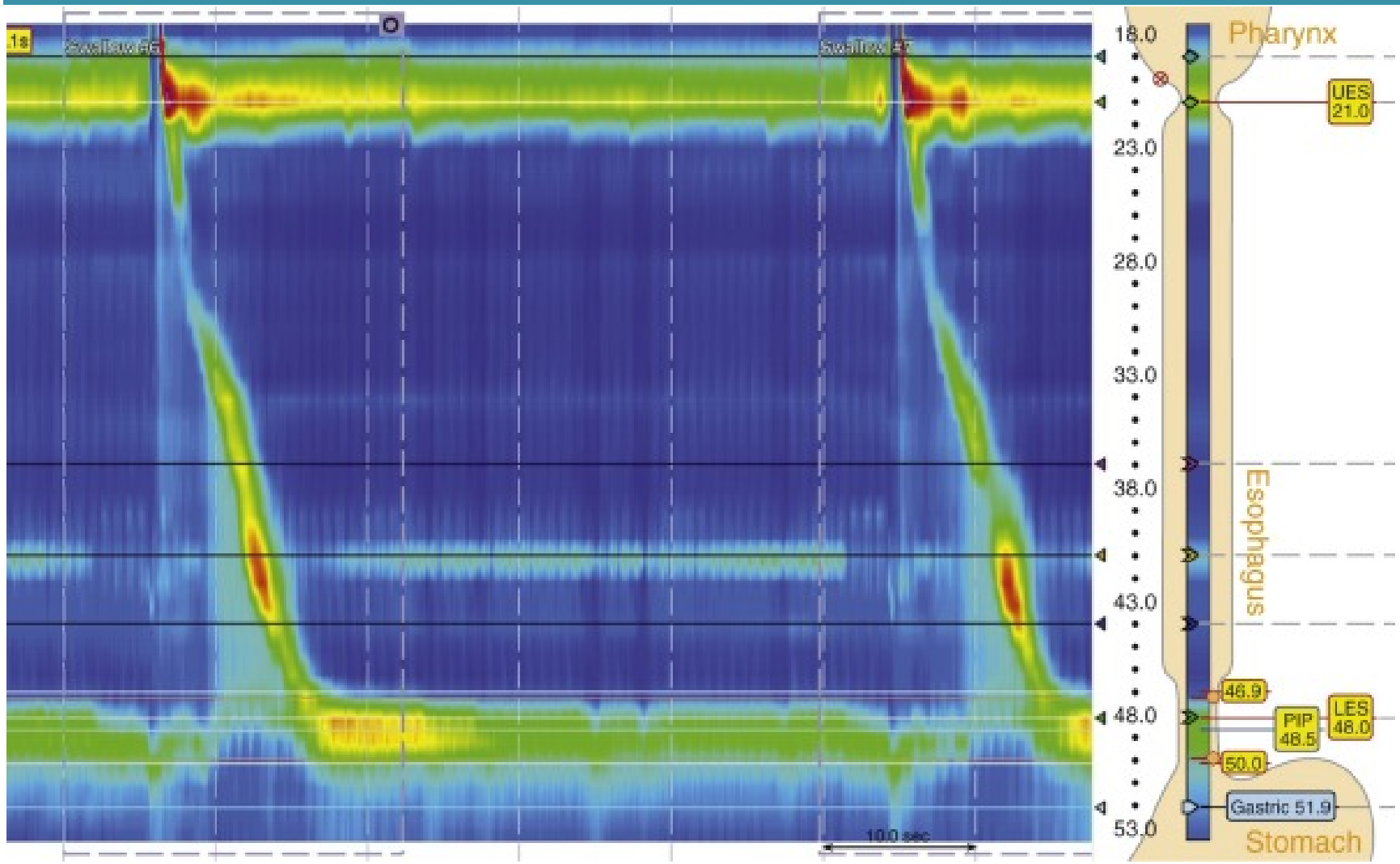
Manometry


Medscape®
www.medscape.com
Esophageal Peristalsis
LES
30 sec

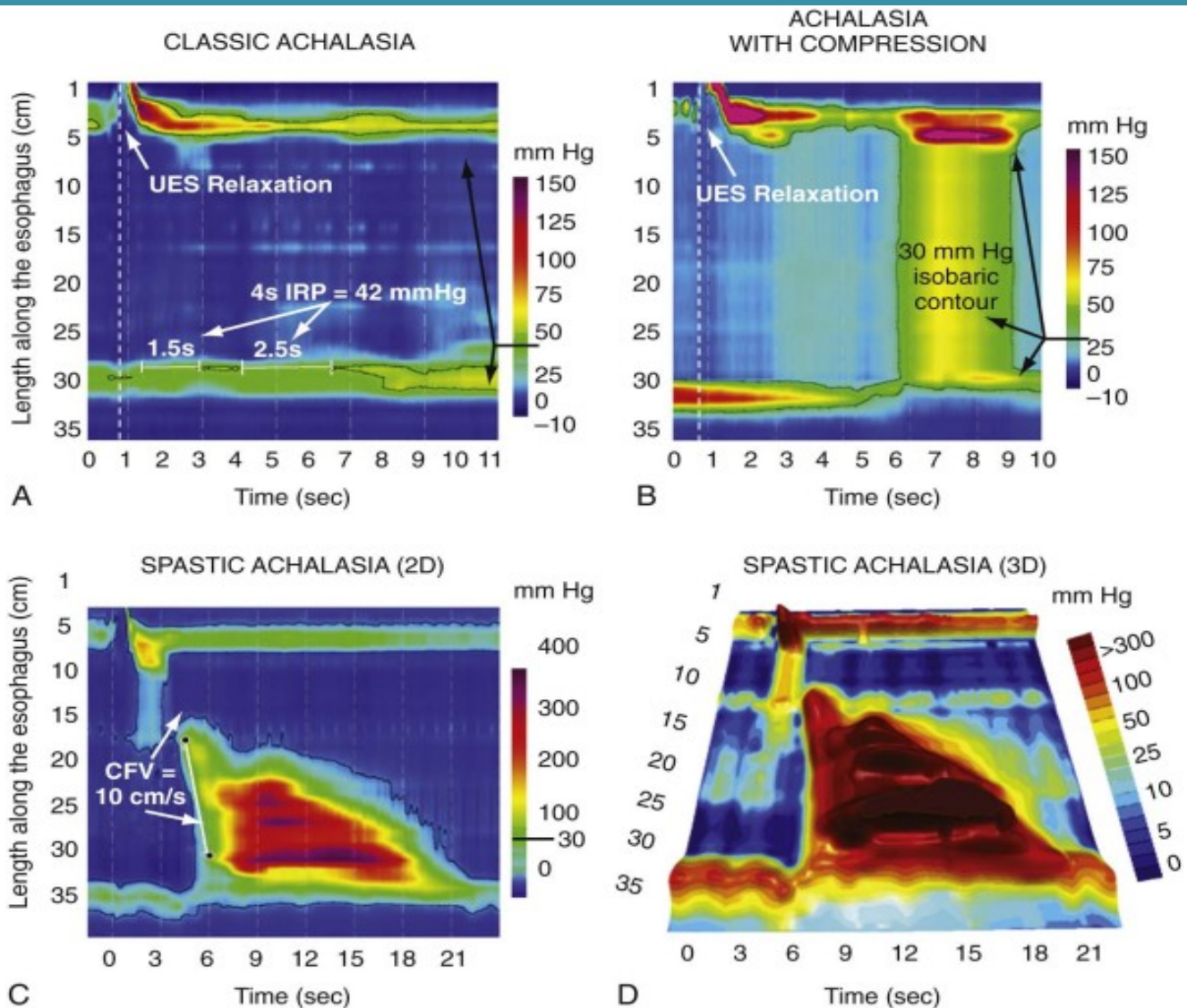
HRM

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

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
10.0 sec

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

Table 4. Response to Therapeutic Interventions Among Achalasia Subtypes

Achalasia subtype	Type I (n = 16)	Type II (n = 46)	Type III (n = 21)	All (n = 83)
Number of interventions, mean (*SD*)	1.6 (SD, 1.5)	1.2[a] (SD, 0.4)	2.4[a,b] (SD, 1.0)	1.8 (SD, 0.7)
Success with BoTox (first intervention) (%)	0 (0/2)	86 (6/7)	22 (2/9)	39 (7/18)
Success with dilation (first intervention with 30-mm balloon) (%)	38 (3/8)	73 (19/26)	0 (0/11)	53 (24/45)
Success with myotomy (first intervention) (%)	67 (4/6)	100 (13/13)	0 (0/1)	85 (17/20)
Success with first intervention (total) (%)	44 (7/16)	83 (38/46)	9 (2/21)	56 (47/83)
Success with last intervention (%) (last intervention type)	56 (B-0, P-10, M-6)	96[a] (B-6, P-25, M-15)	29[a,b] (B-8, P-8, M-5)	71 (B-14, P-43, M-26)

Gastroenterology. 2008 Nov;135(5):1526-33. Epub 2008 Jul 22. Pandolfino, et al

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

Achalasia Subgroups

- Felt that Type I represents progression from Type II

  -esophageal decompensation after prolonged outlet obstruction and continued destruction of myenteric plexus

  -treat early to try to prevent progression to Type I

Complications

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

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

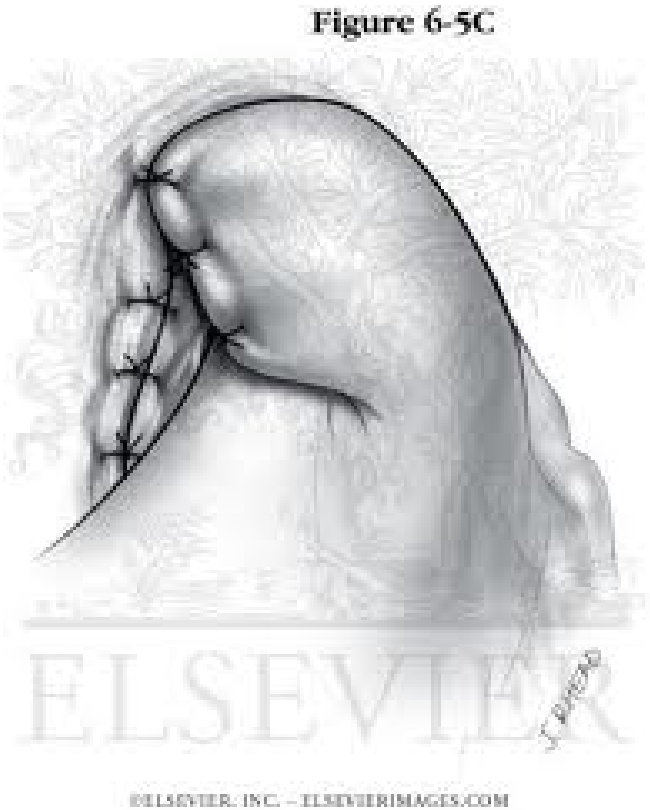
Treatment

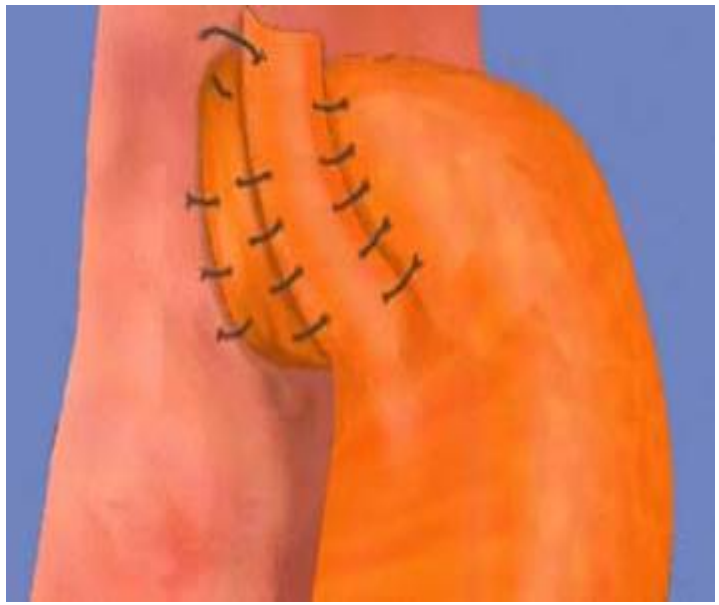
- Dilation
  - Bougie
    - Temporary relief but lower risk of perforation
  - Pneumatic
    - Forceful dilation, tears muscle fibers
    - Stepwise approach: 3.0cm ->3.5cm ->4.0cm
    - High success rate (85%)
    - 1.6% perforation rate
    - Gradual waning of success rate with repeat dilations
  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

Treatment

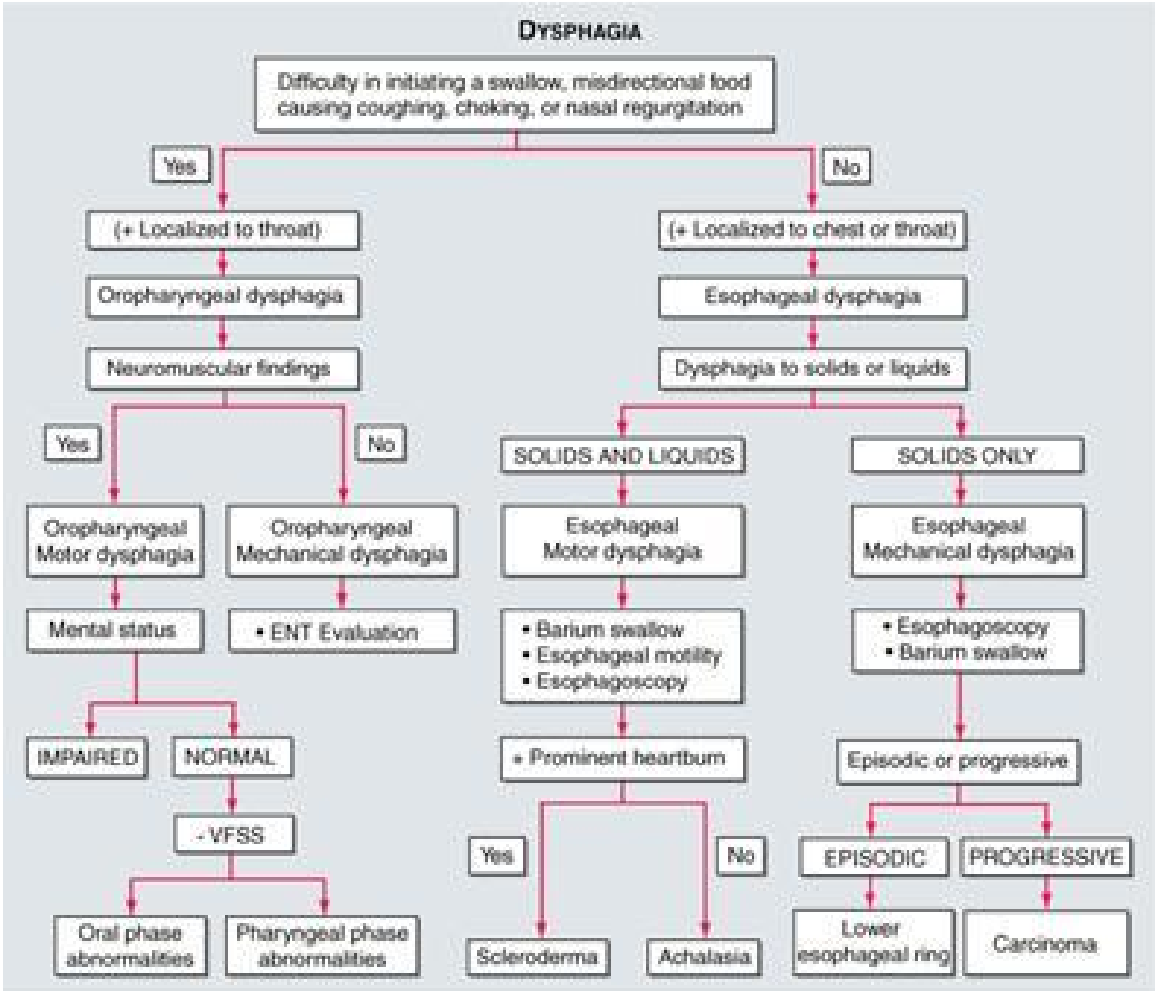
- Surgical Myotomy (Heller and Open myotomy)
  - LES is cut
  - Often partial wrap (Dor or Toupet fundoplication) to prevent reflux (No nissen, worse dysphagia)
  - High success rates (Open, 70%-85% at 10 yrs and 65-73% at 20-30 yrs) and low recurrence
  - Presence of low LES pressure or dilated esophagus 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

Dor and Toupet Fundoplication

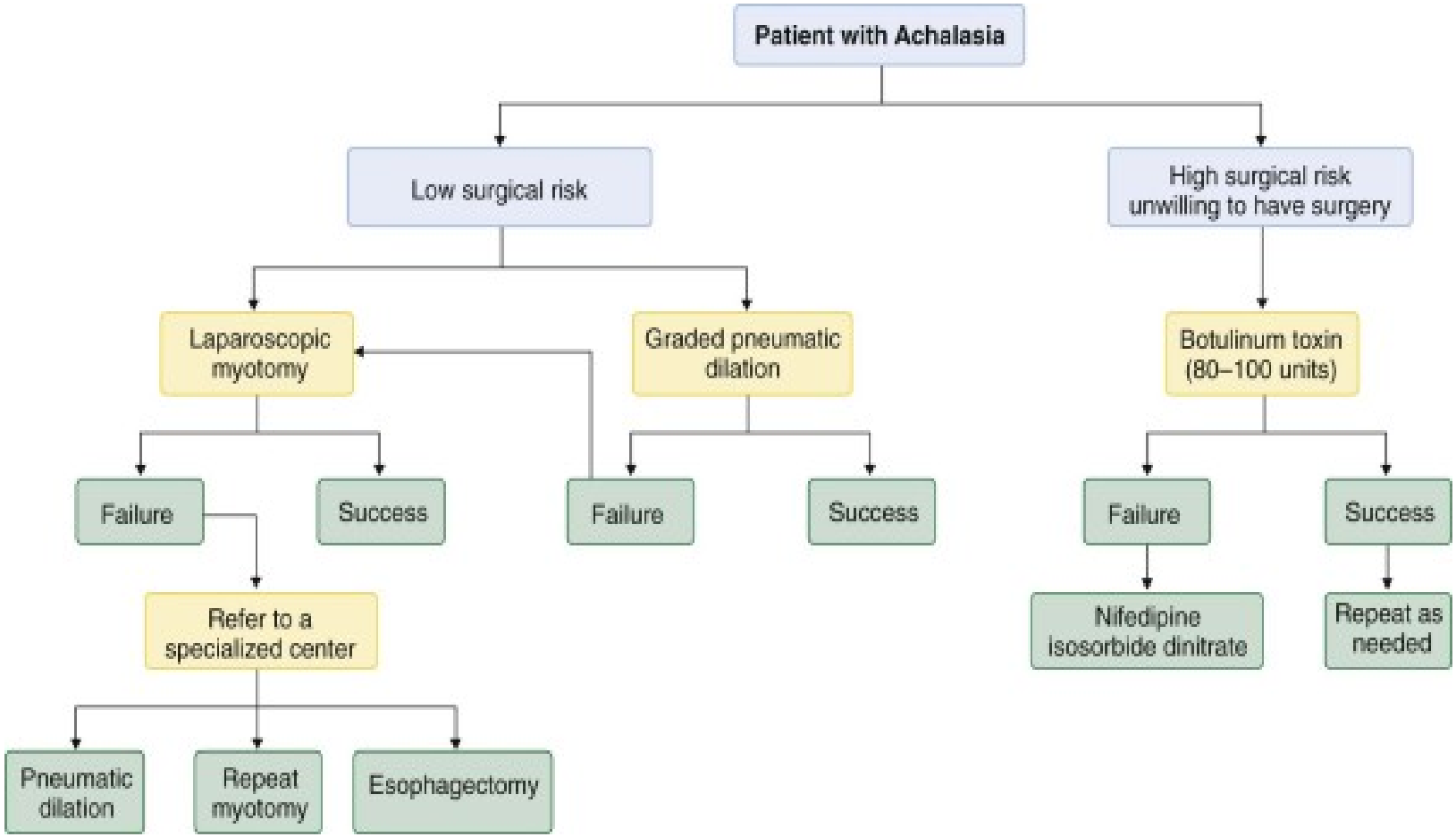




Treatment Algorithm



Treatment Algorithm (ACG)



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


... and finally
DENMARK: A patient broke wind while having surgery and set fire to his genitals. The 30-year-old man was having a mole removed from his bottom with an electric knife when his attack of flatulence was ignited by a spark. His genitals, which were soaked in surgical spirits, caught fire. The man, who is suing the hospital, said: 'When I woke up, my peniś and scrotum were burning like hell. Besides the pain; I can't have sex with my wife.' Surgeons at the hospital in Kjellerups
STOP LAUGHING