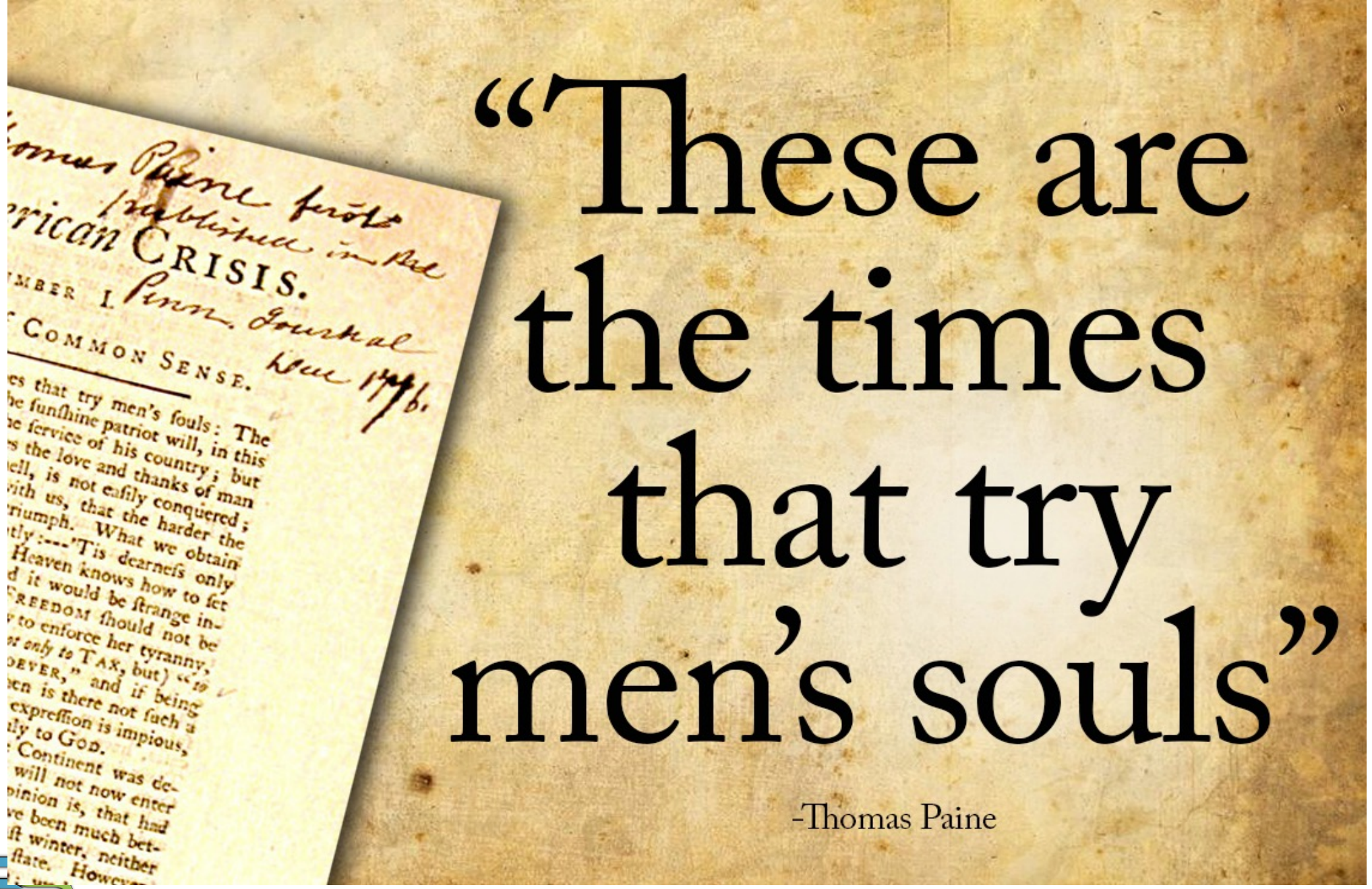
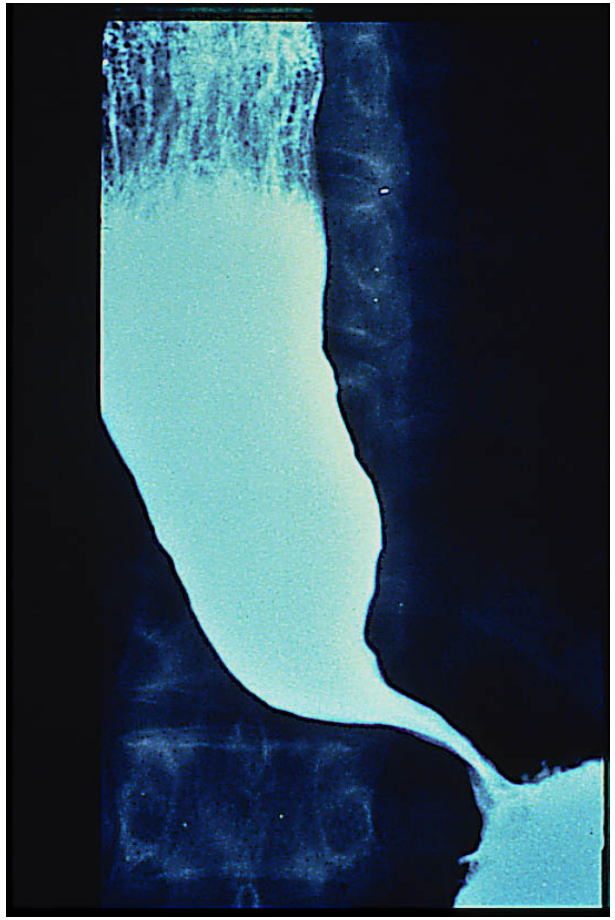


AMERICAN FOREGUT SOCIETY

Multidisciplinary Collaboration. Personalized Treatment Strategies. Patient Advocacy.

Clinical Spectrum of Achalasia



Philip Katz MD

Professor of Medicine

Weill Cornell Medicine

Achalasia: A disease with functional abnormalities of

- Esophagogastric Junction (incomplete relaxation and opening
- Esophageal contractions (loss of normal peristalsis and in many cases contractile vigor)
- Bolus emptying (transit thru esophageal body and EGJ delay)

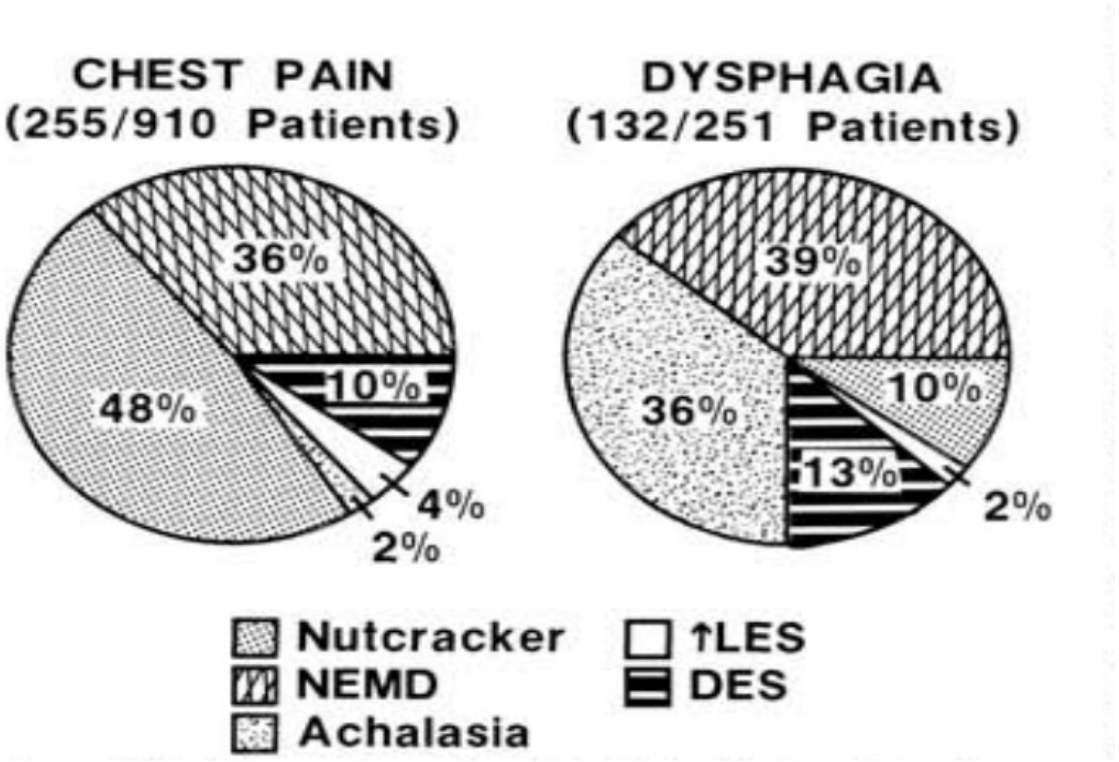
Is it True Achalasia? One disease with several presentations?

No doubt

Buyer beware

Courtesy Felice Schnoll

How Common is Achalasia (or any motility abnormality for that matter?)



Katz,P etal Annals 1987

When to Suspect Achalasia and make the diagnosis quickly

- Dysphagia solids>liquids, often both
- Heartburn not typical of GERD
- Regurgitation during (right after) a meal and supine
- Chest Pain (during a meal)
- Slow eating, standing, walking to move food into stomach, last person to finish a meal
- Waking up with oral debris
- Halitosis
- Globus

Other symptoms

- Recurrent pneumonia
- Aspiration
- Asthma
- Airway obstruction from megaesophagus
- Neck Mass
- Eating disorder symptoms
- Weight loss

Anyone Failing a PPI trial for an esophageal symptom

Should be considered as potentially having achalasia

Demographics

- Any age (30-60 most common)
- Any gender, sex
- No ethnic or racial predominance
- No geographic/regional predominance
- Twins reported

- Morbid Obesity does not preclude achalasia

Secondary Achalasia (Rare but not to be missed)

- Local malignancy
- Paraneoplastic
- Result of another intervention (lap band, wrap)
- Other mechanical issue

- Rapid weight loss (>10% body weight in 6 months)
- Rapid onset dysphagia
- Age >55

- Not a very sensitive triad

Tucker H, Annals Int Med 1978

Eckardt Score: Assess treatment not severity

Score	Weight loss	Retrosternal pain	Regurgitation	Dysphagia
0	None	None	None	None
1	<5 kg	Occasional	Occasional	Occasional
2	5–10 kg	Daily	Daily	Daily
3	>10 kg	Each meal	Each meal	Each meal

Eckardt V, Gastro 1992

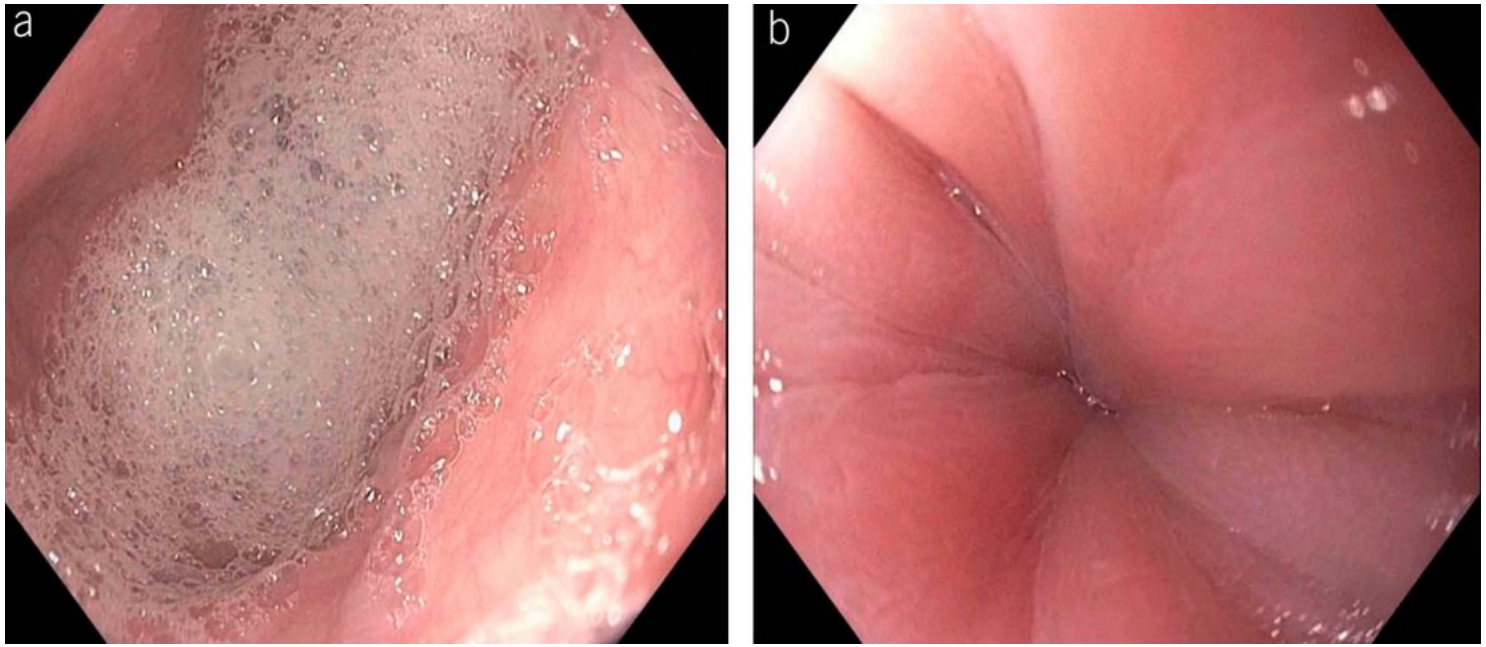
The Tools

- Endoscopy
- Barium Swallow (Timed)
- Manometry
- EndoFlip (Panometry)
- EUS/CT

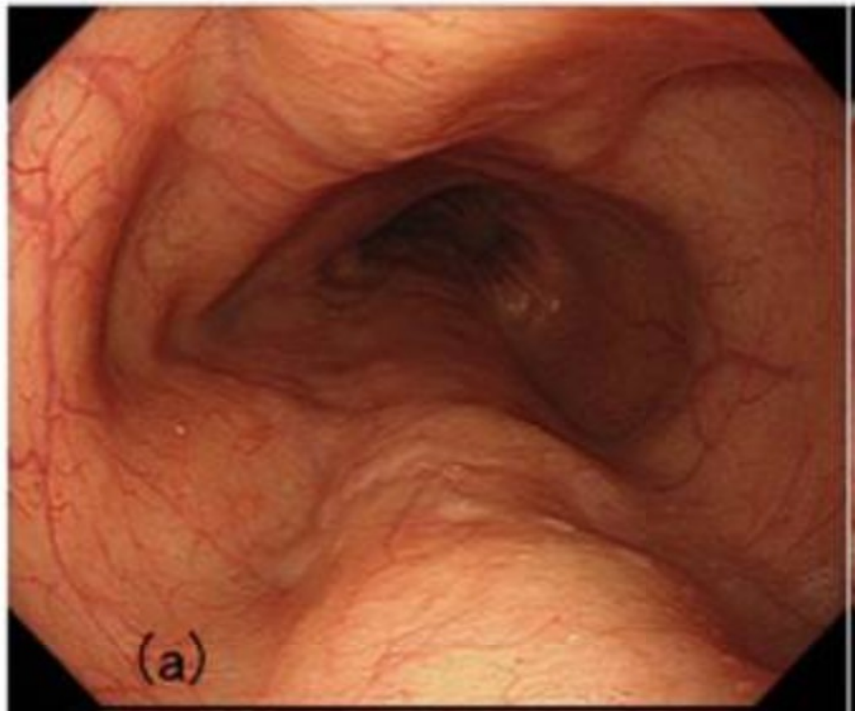
Endoscopy can tell me something about function: expect to see an abnormality

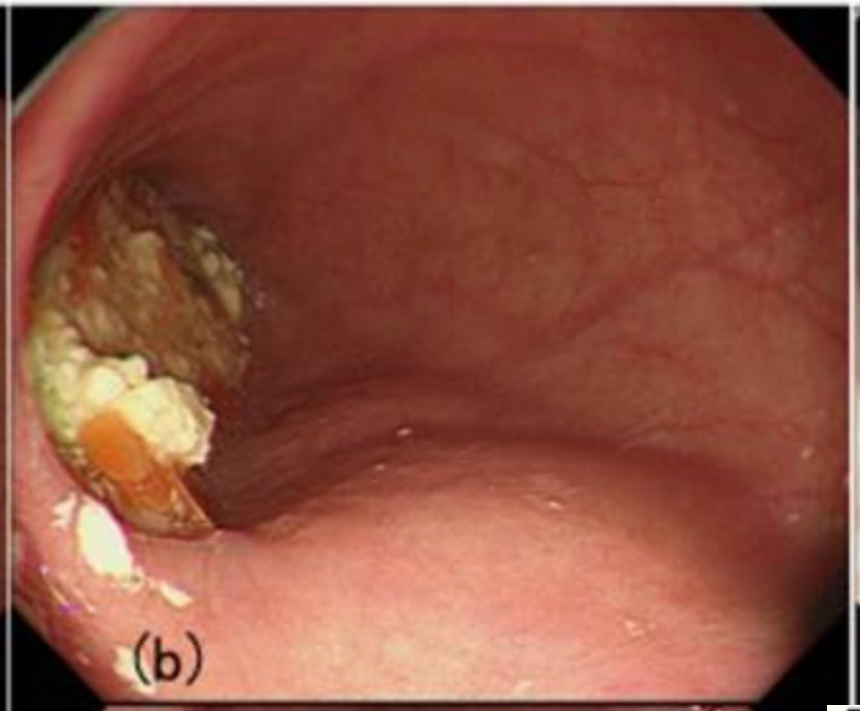
- Retained food/liquid after fasting reflects emptying delay (or mechanical obstruction)
- Excess saliva is a (soft) sign of poor emptying
- Esophageal Dilation
- Appearance of GEJ

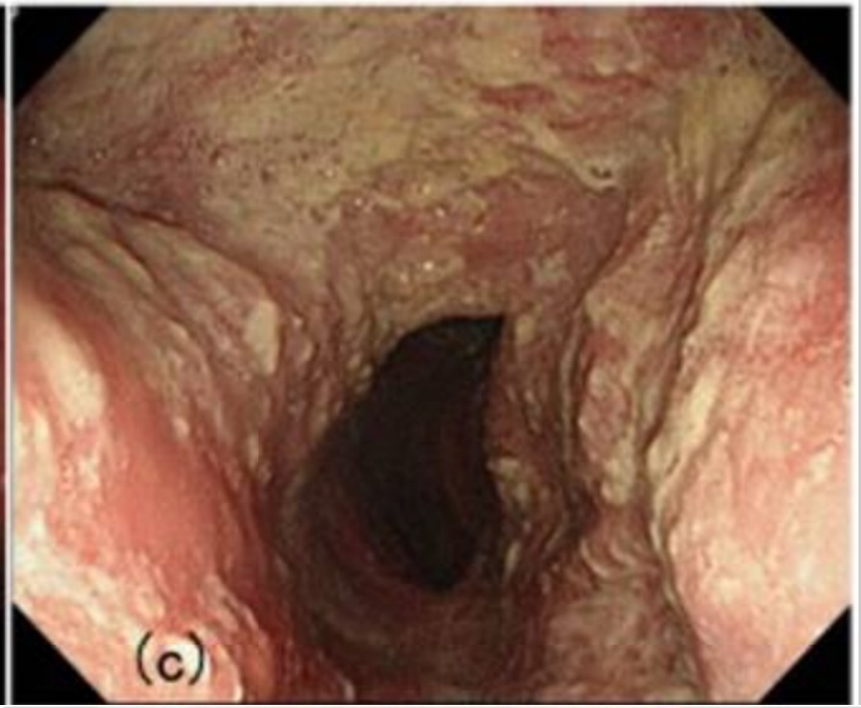
Endoscopic Findings

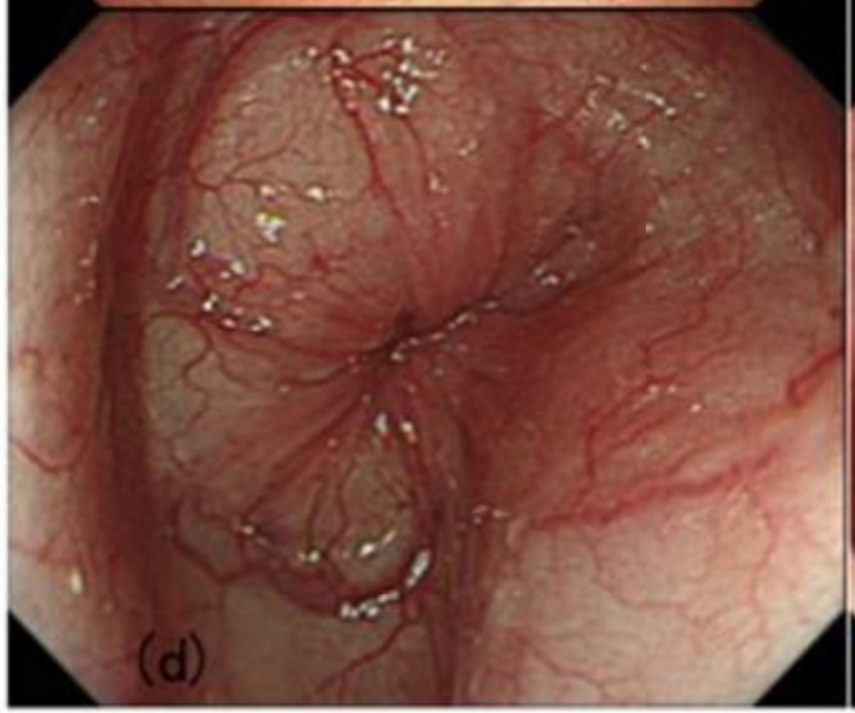


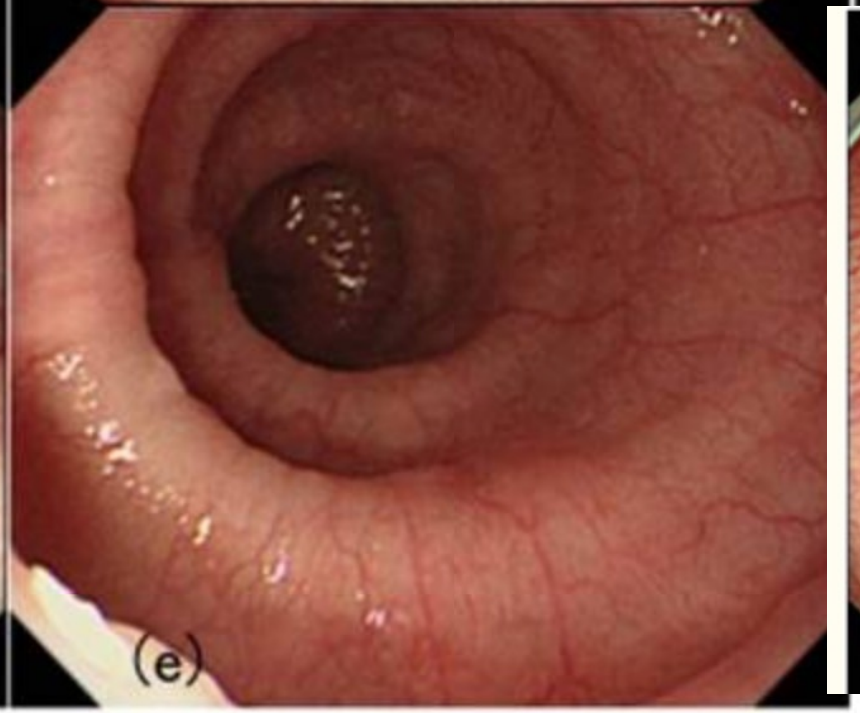
Vaezi, etal AJG 2020

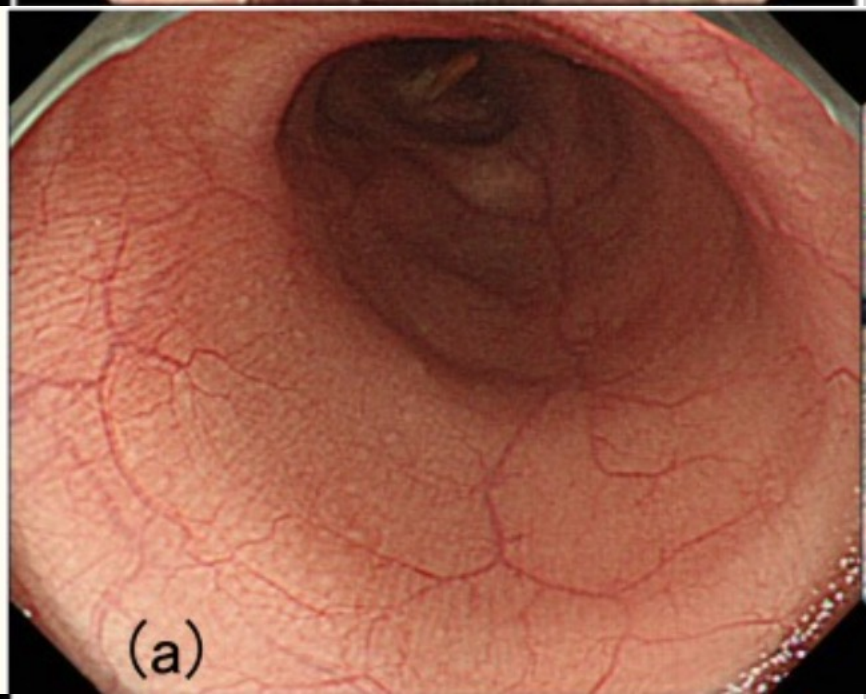

(a)


(b)


(c)


(d)

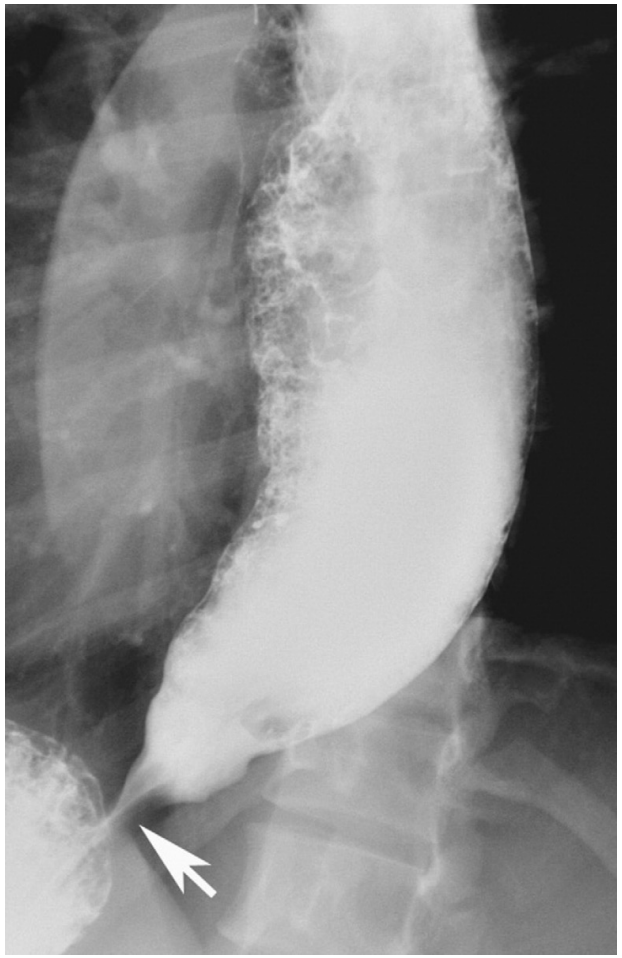

(e)


(a)

Barium is still relevant

- Sensitive for rings, subtle strictures, luminal diameter
- A good study will NOT miss achalasia
- A well performed normal study will almost always eliminate achalasia
- Likely best test for HH, para esophageal hernia

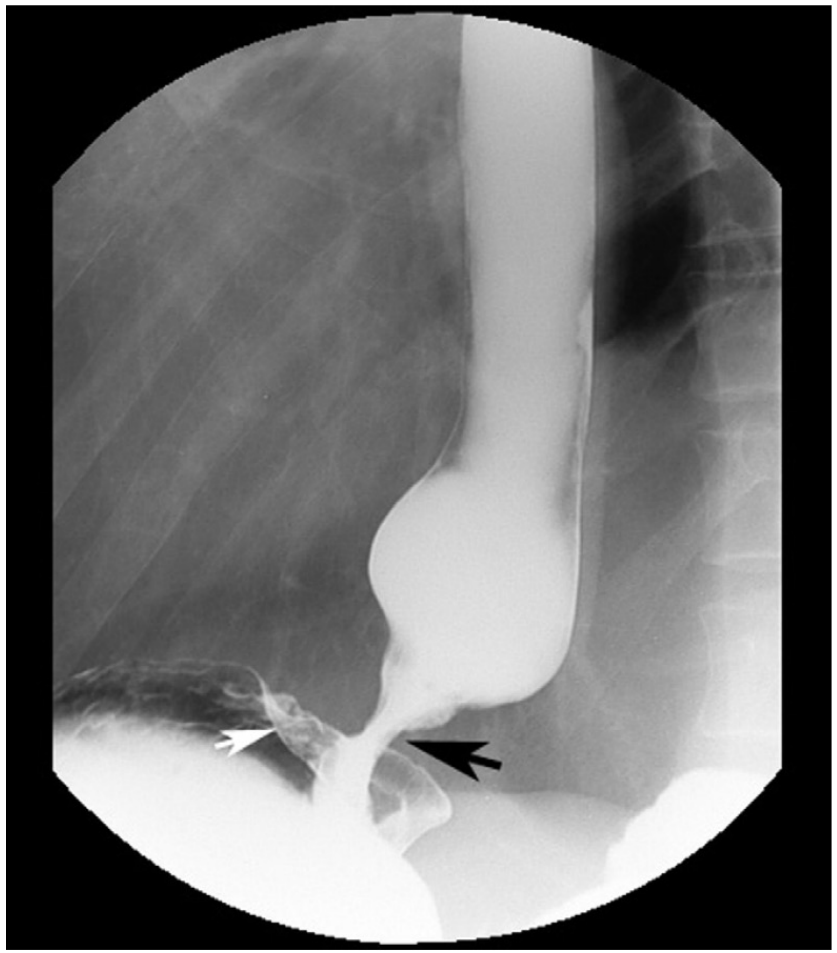
Achalasia



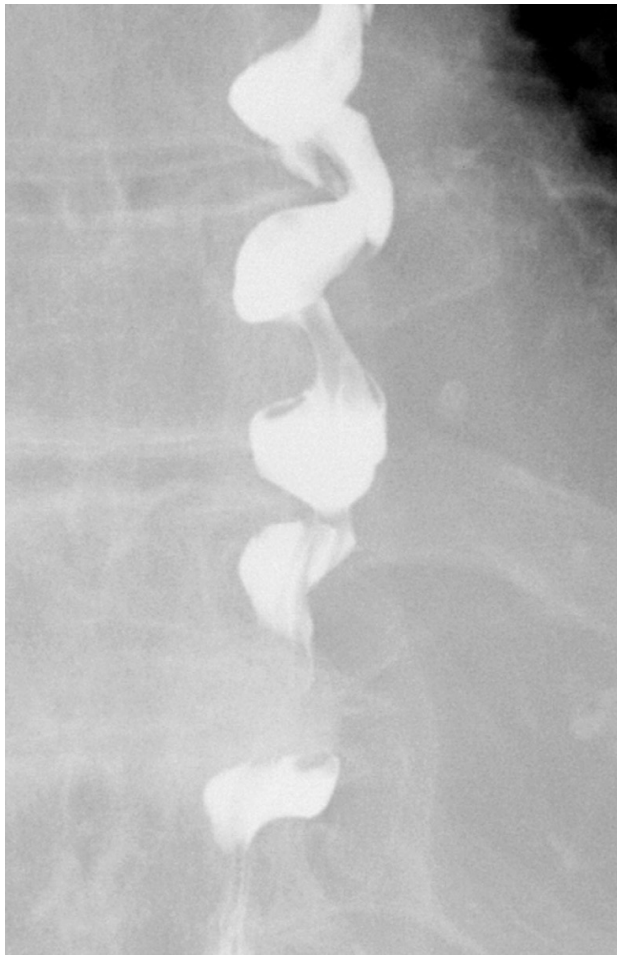
AMERICAN FOREGUT SOCIETY

Multidisciplinary Collaboration. Personalized Treatment Strategies. Patient Advocacy.

Secondary Achalasia



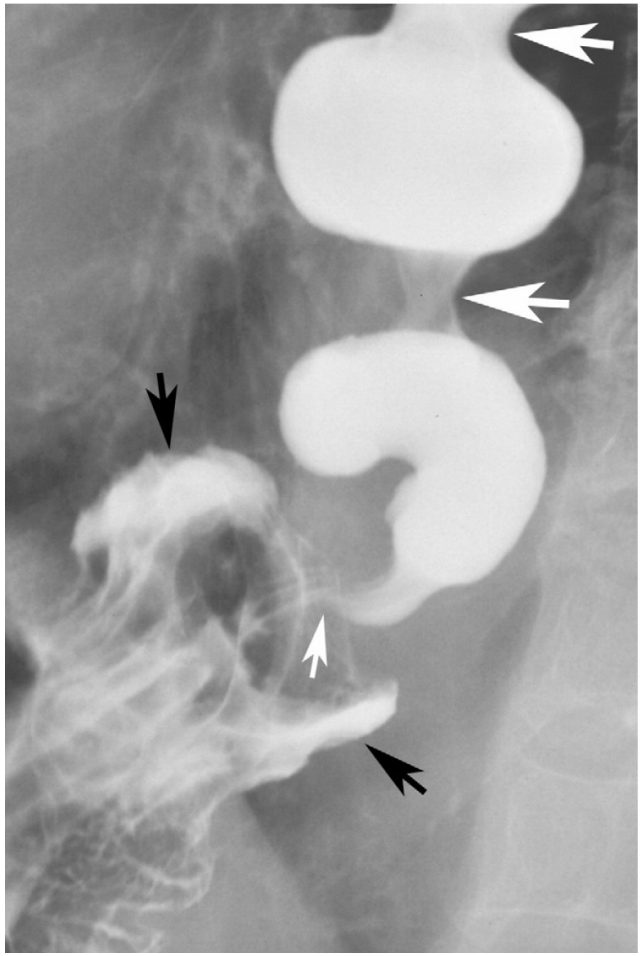
Achalasia



AMERICAN FOREGUT SOCIETY

Multidisciplinary Collaboration. Personalized Treatment Strategies. Patient Advocacy.

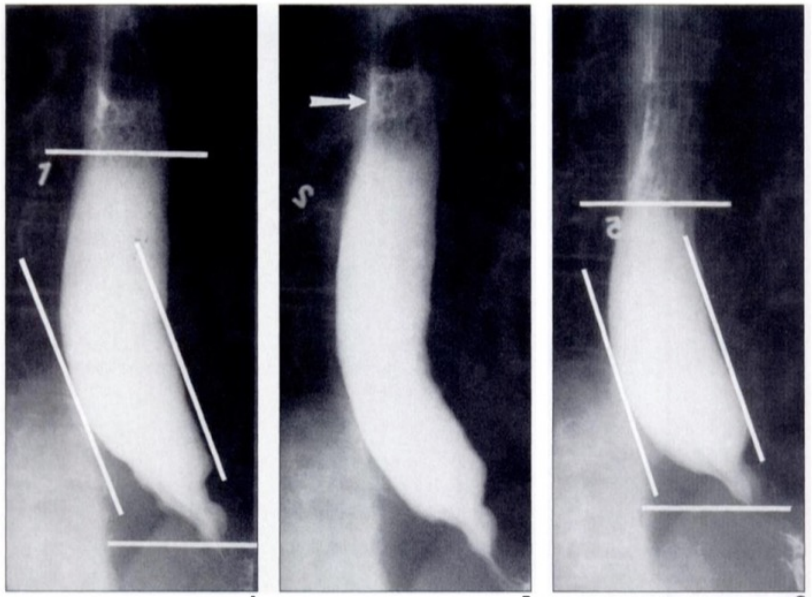
Achalasia



AMERICAN FOREGUT SOCIETY

Multidisciplinary Collaboration. Personalized Treatment Strategies. Patient Advocacy.

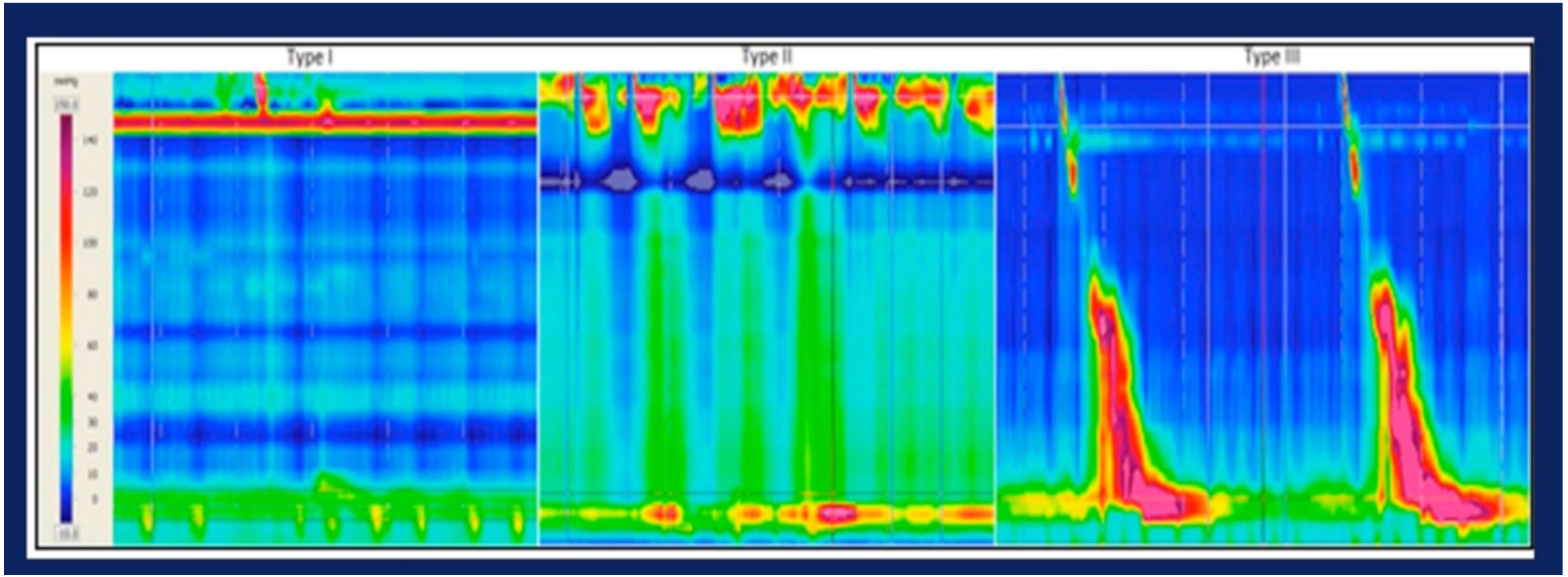
If you do nothing else get Timed Swallow



Vaezi MF. Gut 2002;50:765-770

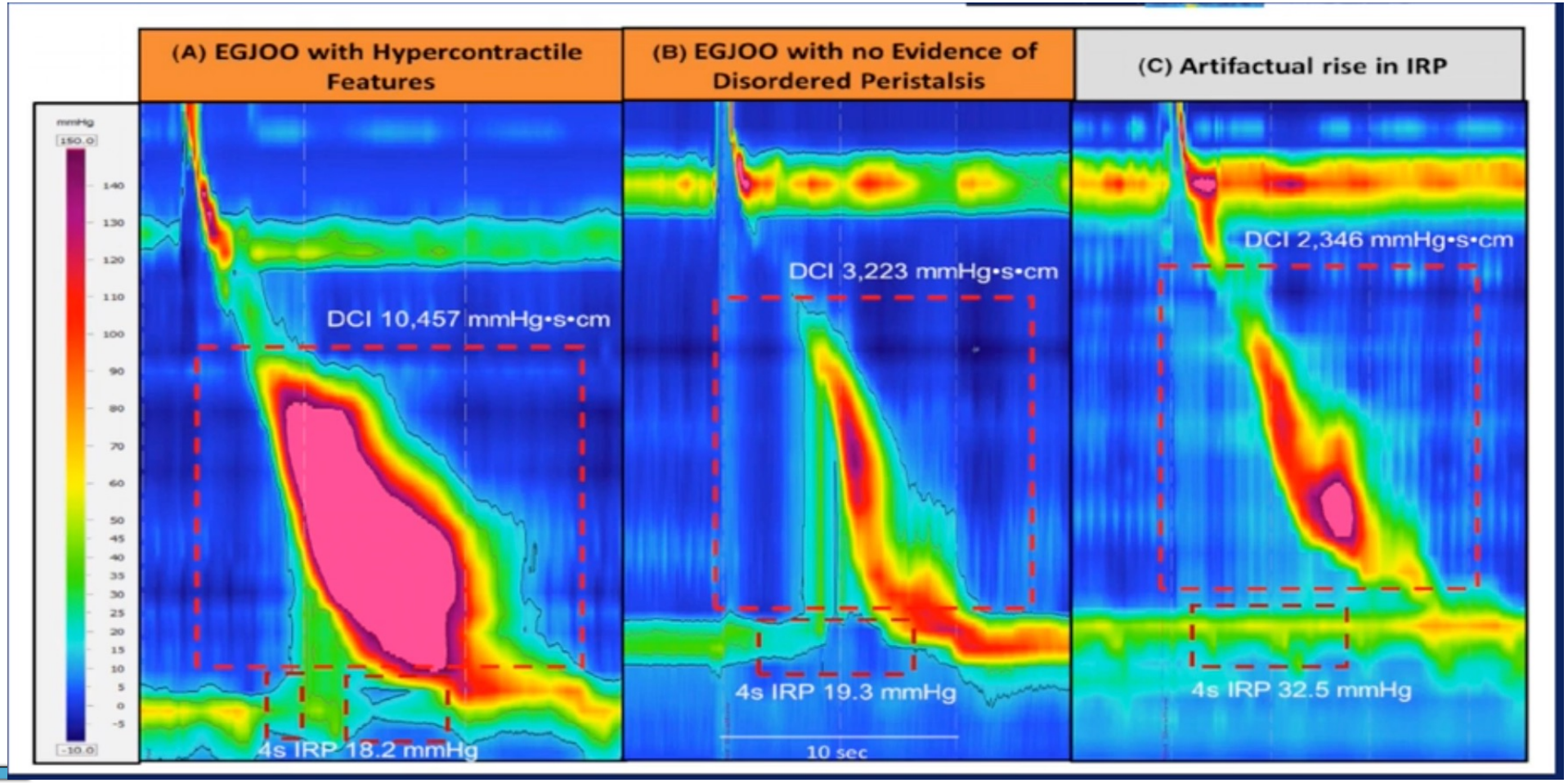
- Timed swallow measures esophageal emptying in the upright position, at 1,2 and 5 minutes after drinking 100-250 mL of low-density barium sulfate.
- Give a 13-mm tablet, repeating radiographs at 5 minutes
- Abnormal study defined as:
  - More than 1 cm of retained residual liquid barium in the esophagus at 1 and 5 minutes
  - Pill retention after 5 minutes

HRM: Three Phenotypes



From Yadlapati, etal

EGJOO which some treat like Achalasia



From Yadlapati etal

Some consider as “achalasia like” even if it does not definitely meet Chicago 4.0 definition

- Symptomatic EGJOO
- Absent peristalsis with normal IRP
- Elevated IRP with Jackhammer features
- Elevated IRP with spasm (DES variant not meeting Type 3 criteria)
- DES with normal IRP (Rapid drink challenge/Timed swallow pending)

- Manometry in patients on Opiates

Interpreting Manometry: Experts Do Not Always Agree

Inter-rater agreement between all raters by esophageal motility diagnoses.

Esophageal pressure topography		Conventional line tracings	
Motility diagnosis	**K (95% CI)**	**Motility diagnosis**	**K (95% CI)**
Type I achalasia	0.82 (0.78 – 0.86)	Classic achalasia	0.58 (0.54 – 0.62)
Type II achalasia	0.77 (0.73 – 0.81)		
Type III achalasia	0.39 (0.35 – 0.43)	ADLESR	0.10 (0.06 – 0.14)
EGJOO	0.45 (0.41 – 0.49)		
DES	0.32 (0.28 – 0.35)	DES	0.28 (0.25 – 0.32)
Jackhammer	0.62 (0.58 – 0.66)	Nutcracker	0.23 (0.19 – 0.27)
Absent peristalsis	0.87 (0.37 – 0.45)	Nutcracker w/HTN LES	0.15 (0.11 – 0.19)
HTN peristalsis	0.41 (0.37 – 0.45)		
Rapid contraction w/normal latency	0.18 (0.14 – 0.21)	Isolated HTN LES	0.09 (0.05 – 0.12)
Weak peristalsis	0.55 (0.51 – 0.59)	IEM	0.50 (0.47 – 0.54)
Normal	0.53 (0.49 – 0.57)	Normal	0.25 (0.21 – 029)

EGJOO – esophagogastric junction outflow obstruction. DES – distal esophageal spasm. HTN – hypertensive. LES – lower esophageal sphincter. ADLESR – Atypical disorder of LES relaxation. IEM – ineffective esophageal motility.

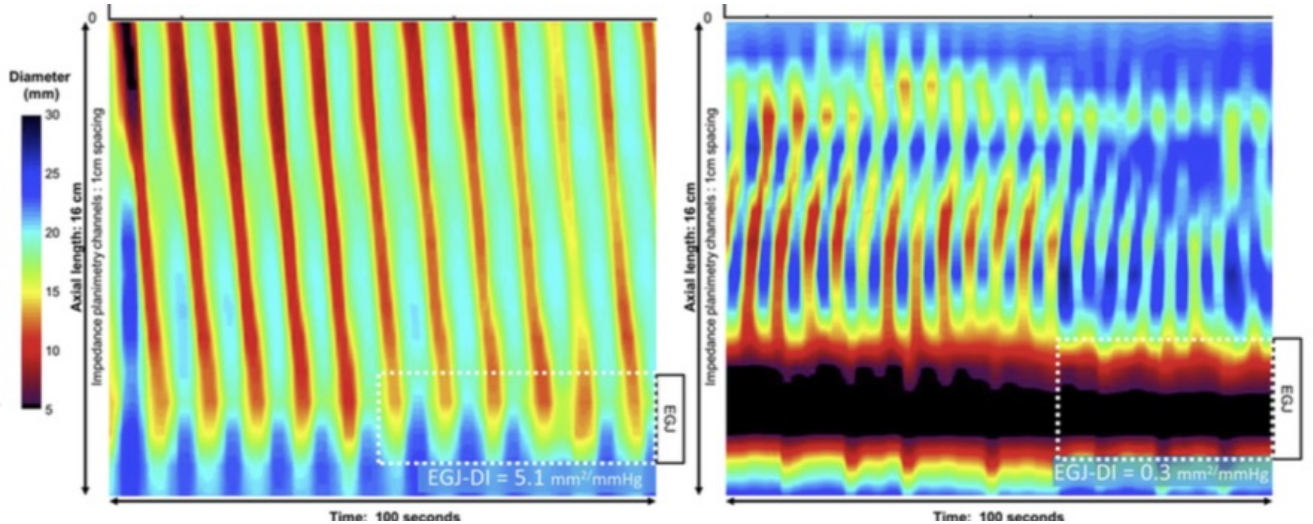
Carlson, D AJG 2016

Functional Lumen Imaging Probe (FLIP) Panometry

Two Assessments:

1) Distensibility of EGJ during volumetric distension (60ml balloon)
Normal DI >2.8mm^2/mmHg)

2) Panometry – esophageal contractions assesses motility



Normal:
Repetitive antegrade contractions
Normal Distensibility Index

Abnormal:
Repetitive retrograde contractions
Low Distensibility Index

Carlson DA, Am J Gastroenterol, 2016

Is Every EGJOO potentially Achalasia?

Test	EGJ outflow obstruction	Obstructive distal contractions
HRM	• IRP> upper limit of normal • Compartmentalized pressurization • Panesophageal pressurization	• Distal latency <4.5 s • DCI >8,000 mmHg•s•cm
HRM- rapid drink challenge	• Panesophageal pressurization • IRP> upper limit of normal	• Failed deglutitive inhibition
HRM- multiple repetitive swallows	• Compartmentalized pressurization	• Failed deglutitive inhibition
Timed barium esophagram	• Barium column >5 cm at 5 min • 12 mm tablet lodges at EGJ	• Tertiary contractions
FLIP	• EGJ distensibility index <2.8 mm^2/mmHg	
FLIP-panometry	• Low EGJ distensibility index at 60 ml distention	• Repetitive retrograde contractions
EUS or CT imaging	• Esophageal wall thickening at EGJ	• Distal esophageal wall thickening

From Kahrilas

THE END

AMERICAN FOREGUT SOCIETY

Multidisciplinary Collaboration. Personalized Treatment Strategies. Patient Advocacy.