

# ACHALASIA CARDIA

## Basics

- 6/1 lakh population
- MC in young women
- Classic Triad- DYSPHAGIA+ REGURGITATION+ WEIGHT LOSS
- Heart burn, Choking and NOCTURNAL Coughing are common
- Eat slowly and drink plenty of water.
- Severe pain is experienced until the LES opens and food enters stomach.
- Slow Progressive dysphagia\*\*

## Complications

- Regurgitation of undigested, foul smelling food particles is common
- Aspiration can become life threatening
- Pneumonia, lung abscess and bronchiectasis
- In 20 years, 8% chance of developing SCC seen where the undigested food particles cause mucosal irritation in Body of esophagus
- Even in patients treated with Achalasia there is ongoing cancer incidence risk is there. (Long term surveillance is advised but as of now no such program is available)

### Question

- What is the most common site of Adenocarcinoma in Achalasia cardia? (NBE repeat)
  - a. Lower 1/3rd
  - b. Upper 1/3rd esophagus
  - c. Above air fluid level in middle of esophagus
  - d. Below air fluid level in middle 1/3rd esophagus

Ans. D (Ref Sabiston 21st edition page 1026)

### Esophagogram findings

- Classical Bird beak
- Hurst Phenomenon
- Abrupt cut off contrast
- Lack of peristaltic waves in Body and failure of LES to open seen.
- Lack of gastric air bubble is a common finding in Upright portion of Esophagogram.
- Megaesophagus (tortuous/ Sigmoid Esophagus)

### Hurst Phenomenon

- Hurst Phenomenon--temporary transit through cardia provoked by hydrostatic pressure of barium column reaching above a critical level

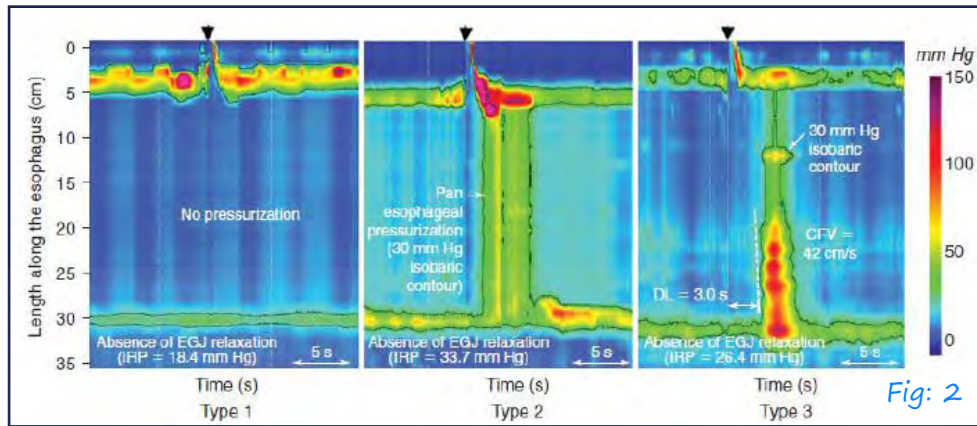


Fig: 1

### Manometry tracings of Achalasia Types

- In type I (classic) achalasia, the median IRP greater than 15 mm Hg and there is 100% failed peristalsis with minimal esophageal pressurization.
- In type II achalasia, the median IRP greater than 15 mm Hg, 100% failed peristalsis, and panesophageal pressurization with at least 20% of swallows.
- In type III (spastic or vigorous) achalasia, the median IRP greater than 15 mm Hg without normal peristalsis and there are spastic contractions (distal contractile integral  $>450$  mm Hg·s·cm) with at least 20% of swallows. Bailey says- Panesophageal pressurisation may be seen.

## Banding at 30 mmHg



## Endoscopy

- To look for Esophageal mucosal damage
- To look for secondary causes of Esophageal narrowing ( Pseudoachalasia)- GEJ tumors, Neuropathy, Strictures.

## Question

- False statement regarding the treatment of Achalasia cardia is
  - a. Sublingual CCB, Nitroglycerine and Nitrates provide hours of relief before and after meals.
  - b. Botox may offer symptomatic relief for years but symptoms recur within six months in more than 50%
  - c. Pneumatic Balloon dilatation provides 89% remission of symptom compared to Botox which provides 38% remission in 1 year.

*d. Balloon dilatation and Surgical treatments are curative for Achalasia cardia.*

**Ans. D**

- *There are surgical and nonsurgical treatment options for patients with achalasia; all are directed toward relieving the obstruction caused by the LES.*
- *Because none of them are able to address the issue of decreased motility in the esophageal body, they are all palliative treatments.*

### **Achalasia Management- Changed points**

- *In comparing balloon dilatation to Botox injections, remission of symptoms occurred in 89% versus 38% of patients at one year, respectively*
- *Studies done to compare balloon dilatation versus surgery have shown perforation rates of 4% and 1% and mortality rates of 0.5% and 0.2%, respectively.*
- *However, in a randomized controlled trial of the European Achalasia Trial Investigators, pneumatic dilatation was found to be equivalent to Laparoscopic Heller myotomy and Dor fundoplication with therapeutic success rates of 86% versus 90% at two years.*
- *Perforation occurred in 4% of the patients during pneumatic dilatations and mucosal tears occurred in*

12% during laparoscopic Heller myotomy, but all were repaired intraoperatively.

- Notably, patients in the pneumatic dilatation cohort had a 25% rate of re dilatation to achieve treatment success.

### **Sabiston 21st Edition Question**

- Regarding esophagectomy for Sigmoid mega esophagus – False statement is:
  - a. Indicated for cases where there is failure of more than 1 myotomy.
  - b. Indicated for cases with reflux stricture not amenable for dilatation.
  - c. Transhiatal esophagectomy with or without vagus preservation is done for end stage Achalasia.
  - d. In mega esophagus – transthoracic esophagectomy is advised.
  - e. POEM is not beneficial in End stage achalasia

**Ans. E** (Sabiston 21st Edition Page 1028)

### **Megaoesophagus**

- Esophagectomy is considered in any symptomatic patient with a tortuous dilated esophagus (megaesophagus), sigmoid esophagus, failure of more

than one myotomy, or reflux stricture that is not amenable to dilatation.

- A transhiatal esophagectomy with or without preservation of the vagus nerve offers a good long-term result in end stage Achalasia\*\*
- However, in the setting of megaesophagus, a total esophagectomy incorporating a transthoracic dissection may be safest, given the difficulty in palpating the borders of the esophagus through a transhiatal approach.
- In a recent study, two-year outcomes after POEM demonstrated treatment success in 96.8% of patients. Use of POEM may obviate the need for esophagectomy in this malnourished, often high-risk, patient population.

### *Sabiston 21st Edition update*

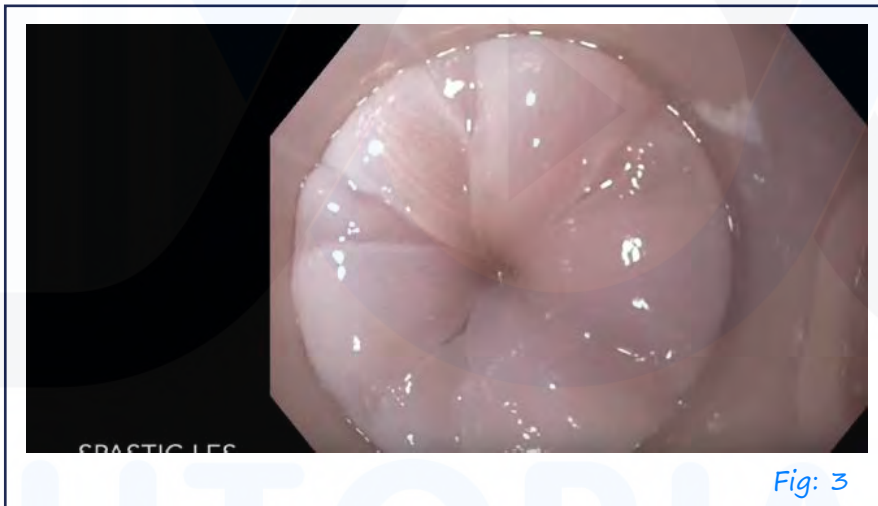
- The best treatment for Ineffective Esophageal Motility is
  - a. Heller Myotomy
  - b. Pneumatic Dilatation
  - c. Effective treatment of GERD
  - d. CCB

**Ans. C** (Ans. E Sabiston 21st Edition Page 1028)

## POEM for Achalasia

- Via a submucosal tunnel- Myotomy done.
- The major concerns are – Lack of anti-reflux procedure and increased incidence of Symptomatic reflux.
- As of now short-term outcomes are comparable to Heller myotomy, Long term results awaited.

## POEM



## Eckardt Scoring?

- Post Treatment Scoring for Achalasia Cardia
- Score more than 3 is failure of treatment
- Total Score = 12
- Mnemonic- “Regurgitate PWD”



Symptom				
Score	Weight loss	Dysphagia	Retrosternal pain	Regurgitation
0	None	None	None	None
1	<5kg	Occasional	Occasional	Occasional
2	5-10kg	Daily	Daily	Daily
3	>10kg	Each meal	Several times a day	Each meal

### *Ineffective Esophageal motility*

- It is defined as a contraction abnormality of the distal esophagus and is usually associated with GERD.
- It may be secondary to inflammatory injury of the esophageal body because of increased exposure to gastric contents
- Dampened Motility of Esophageal body results in poor acid clearance in distal esophagus
- Symptoms are usually mixed but patients presents with Reflux and Dysphagia.
- IEM is defined by greater than 50% of swallows being deemed ineffective (distal contractile integral <450 mm Hg·s·cm).
- The best treatment of IEM is prevention, which is associated with effective treatment of GERD.
- Scleroderma may manifest as IEM in HRM and best treated by addressing the underlying condition.