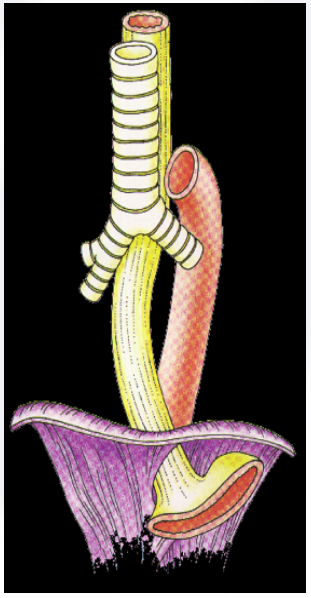

بسم الله الرحمن الرحیم

Esophageal Motility Disorders



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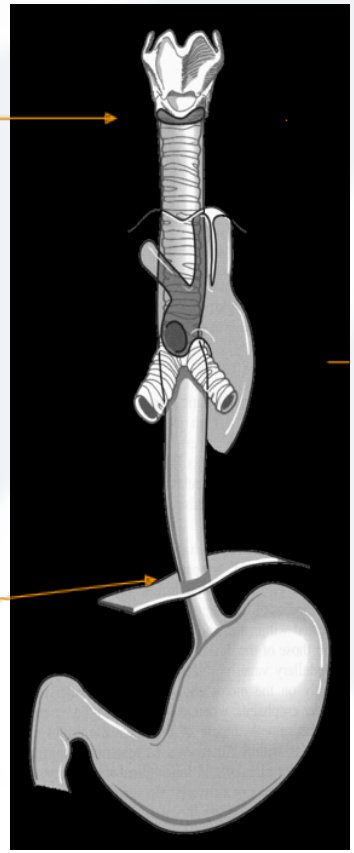
Esophageal Anatomy

Upper Esophageal Sphincter (UES)

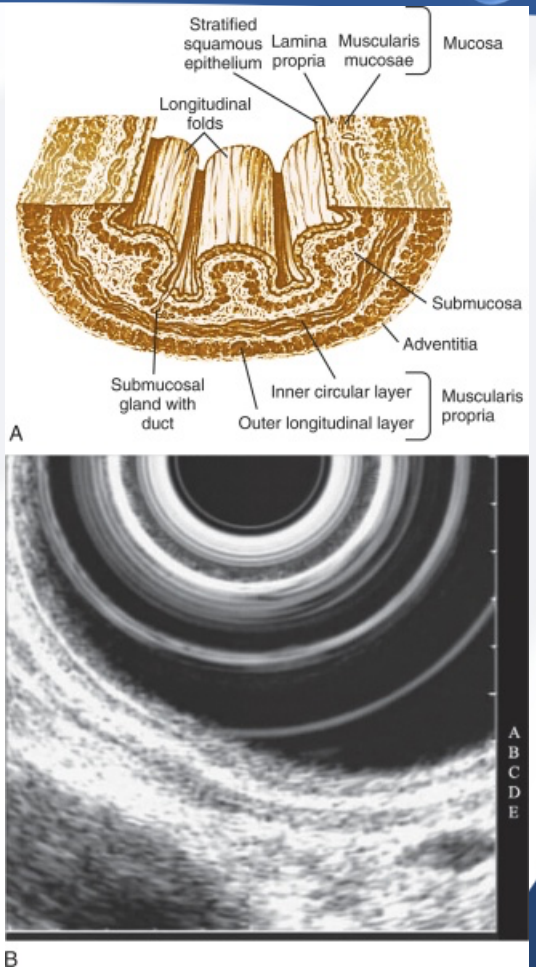
Esophageal Body (cervical & thoracic)

Lower Esophageal Sphincter (LES)



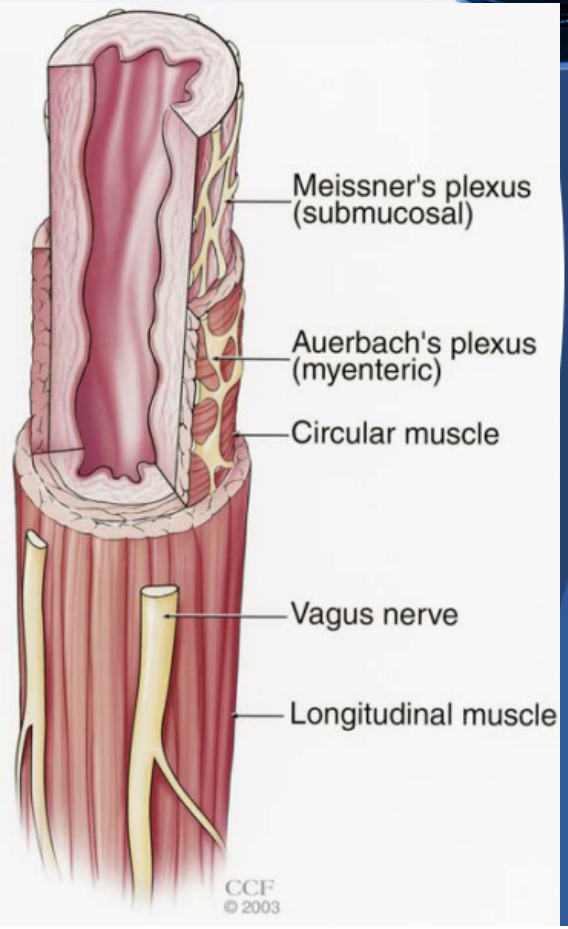
18 to 24 cm

- Four layers:
  - Mucosa
  - Submucosa
  - Muscularis propria
  - Adventitia; no serosa.



Muscularis propria

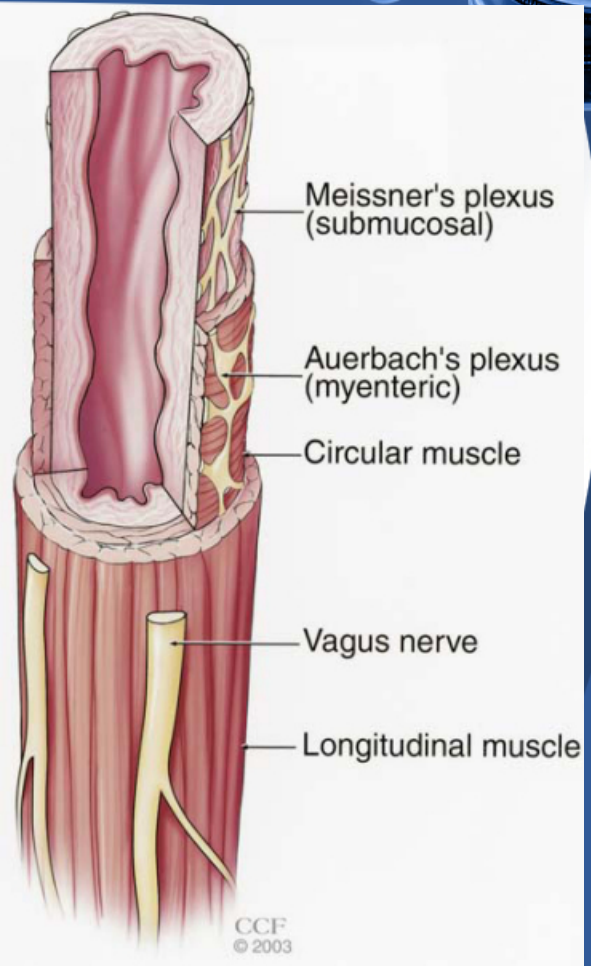
- Skeletal and smooth muscle
  - Skeletal muscle (Proximal 1/3)
  - Mixed (Middle 1/3)
  - Smooth muscle (Distal 1/3)
- Inner circular
- Outer longitudinal layers.



Innervation mainly by Vagus n.

- Auerbach’s (myenteric) plexus
  - Between the two muscle layers
  - Controls esophageal peristalsis
  - Acetylcholine mediates contraction
  - Nitric oxide relaxation

- Meissner's plexus
  - Submucosal layer
  - Sensory input
  - Pain sensation overlap with the heart and respiratory system



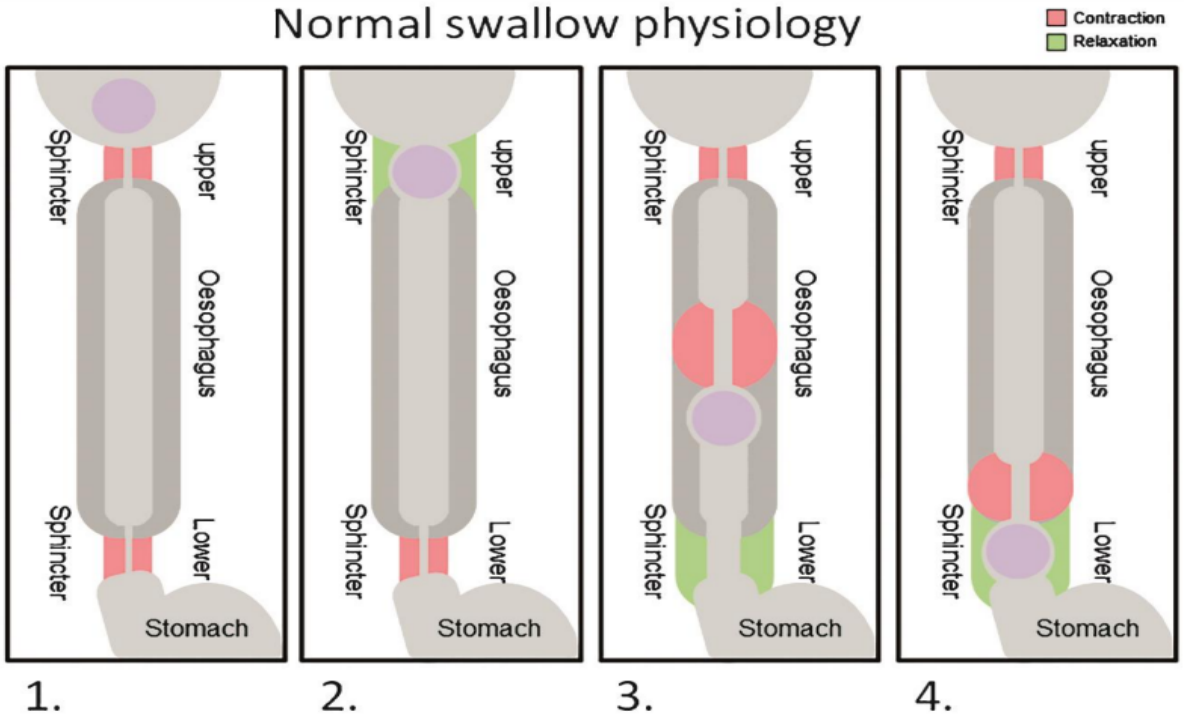
Normal Phases of Swallowing

- **Voluntary**
  - oropharyngeal phase – bolus is voluntarily moved into the pharynx
- **Involuntary**
  - UES relaxation
  - peristalsis (aboral movement)
  - LES relaxation
- **Between swallows**
  - UES prevents air entering the esophagus during inspiration and prevents esophagopharyngeal reflux
  - LES prevents gastroesophageal reflux

Peristalsis

Sequential, coordinated contraction wave that travels along the whole length of the esophagus, propelling intraluminal contents downstream.

- **Primary wave** strips from proximal to distal, triggered by swallowing center, 2cm/sec.
- **Secondary wave** induced by distension of bolus, acts to clear esophagus of retained food.
- **Tertiary contractions** are dysfunctional and have no role.



شکل ۱. فیزیولوژی بلع طبیعی.

۱. اسفنکترهای فوقانی (UES) و تحتانی مری (LES) در حالت استراحت منقبض می‌مانند.

۲. بلع با شل شدن اسفنکتر فوقانی مری (UES) شروع می‌شود، تا لقمه‌غذا عبور کند.

۳. یک انقباض قوی در اسفنکتر فوقانی مری (UES)، محتوای آن را به سمت پایین مری حرکت می‌دهد. در همین حال، اسفنکتر تحتانی مری (LES) زود شل می‌شود تا اجازه عبور هوای گیر افتاده در مری را بدهد.

۴. لقمه‌ی غذا قبل از انقباض اسفنکتر تحتانی مری (LES) برای جلوگیری از بازگشت محتویات معده به مری، از اسفنکتر تحتانی مری (LES) شل شده عبور کرده، وارد معده می‌شود.

- UES: upper esophageal sphincter; LES: lower esophageal sphincter

Motor (neuromuscular) dysphagia

➢ **Disorders of pharyngeal and esophageal striated muscle**

- Muscle weakness((bulbar paralysis) CVA ,Motor neuron disease,Poliomyelitis, postpolio syndrome ,Polyneuritis Amyotrophic lateral sclerosis ,Myasthenia gravis
- Muscle disorders(Polymyositis ,Dermatomyositis)

➢ **Disorders of esophageal smooth muscle**

- sclerodermy
- Achalasia

Motility Disorders

upper esophageal
- UES disorders
- neuromuscular disorders

esophageal body
- achalasia
- diffuse esophageal spasm
- nutcracker esophagus
- nonspecific esophageal dysmotility

LES
- achalasia
- hypertensive LES

primary disorders
- achalasia
- diffuse esophageal spasm
- nutcracker esophagus
- nonspecific esophageal dysmotility

secondary disorders
- scleroderma
- diabetes
- Parkinson's
- stroke

Upper Esophageal Motility Disorders

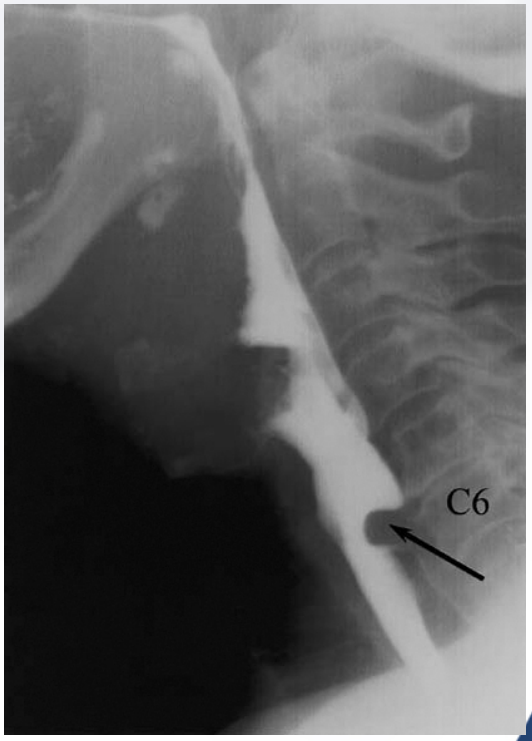
- Cause **oropharyngeal** dysphagia (transfer dysphagia)
  - patients complain of *difficulty swallowing*
  - tracheal aspiration may cause symptoms
- **pharyngoesophageal** neuromuscular disorders
  - stroke
  - Parkinson's
  - poliomyelitis
  - ALS
  - multiple sclerosis
  - diabetes
  - myasthenia gravis
  - dermatomyositis and polymyositis
- **upper esophageal sphincter** (cricopharyngeal) dysfunction

upper esophageal sphincter (cricopharyngeal) dysfunction

- **cricopharyngeal hypertension**
  - elevated UES resting tone
  - poorly understood
- **cricopharyngeal achalasia**
  - incomplete UES relaxation during swallow
  - may be related to Zenker's diverticula in some patients
- clinical manifestations
  - localizes as upper (cervical) dysphagia
  - within seconds of swallowing
  - coughing, choking, immediate regurgitation, or nasal regurgitation
- diagnosis: swallow evaluation & modified barium swallow

Cricopharyngeal Dysfunction

- The cricopharyngeus remains contracted between swallows
- **Cricopharyngeal achalasia**
  - Muscle fails to completely relax
  - smooth posterior impression on the hypopharynx



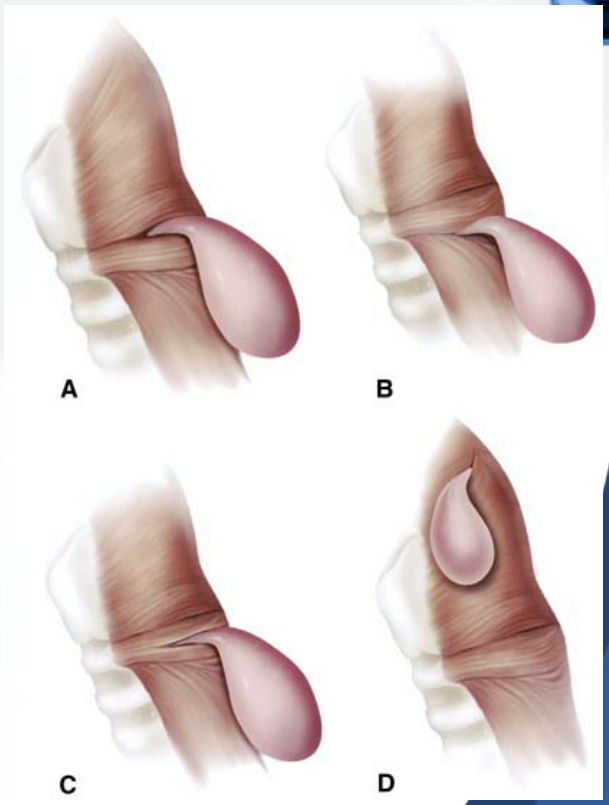
- Esophageal diverticula are categorized by location with the most common being epiphrenic, hypopharyngeal (Zenker's), and mid esophageal.
- Epiphrenic and Zenker's diverticula are false diverticula involving herniation of the mucosa and submucosa through the muscular layer of the esophagus. ***These lesions result from increased intraluminal pressure associated with distal obstruction.***
- In the case of Zenker's, the obstruction is a stenotic cricopharyngeus muscle (upper esophageal sphincter) and the hypopharyngeal herniation most commonly occurs in an area of natural weakness known as *Killian's triangle*
- Small Zenker's diverticula are usually asymptomatic but when they enlarge sufficiently to retain food and saliva they can be associated with dysphagia, halitosis, and aspiration.
- Treatment is by surgical diverticulectomy and cricopharyngeal myotomy or a marsupialization procedure in which an endoscopic stapling device is used to divide the cricopharyngeus.

Zenker’s Diverticulum

- Esophageal diverticula are classified based on:
  - Anatomic location
  - Mechanism of origin (pulsion or traction).
- Zenker's diverticulum (ZD)
  - Pulsion type diverticulum
    - Herniation of esophageal mucosa and submucosa through an area of weakened esophageal musculature
  - Annual incidence of 2 per 100,000 people per year
  - Males predominance (2 to 3 times)

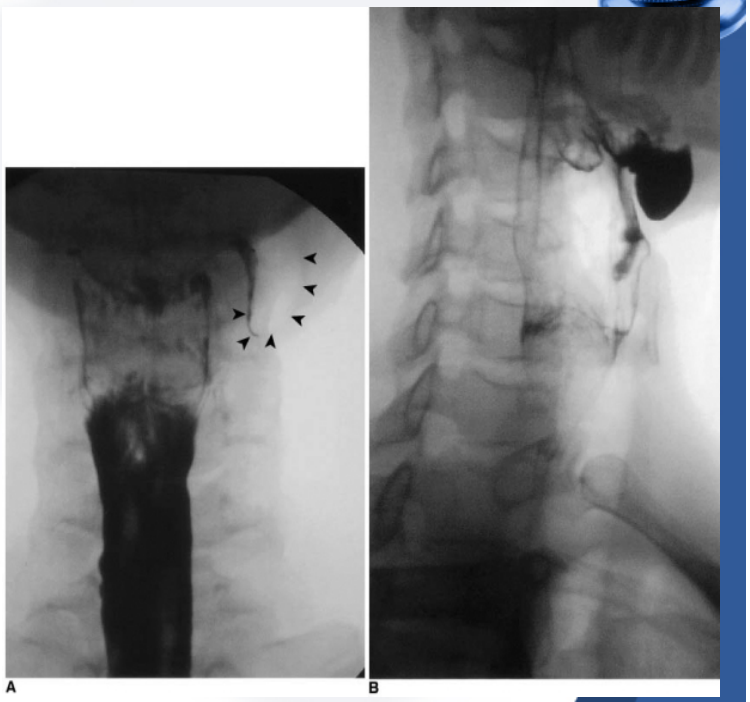
Zenker’s Diverticulum

- **Killian's dehiscence or triangle**
  - Between the cricopharyngeal muscle and inferior constrictor muscle
- Killian-Jamieson's area
  - between the oblique and transverse fibers of the cricopharyngeal muscle
- Laimer's triangle
  - Between the cricopharyngeal muscle and the most superior esophageal wall circular muscles



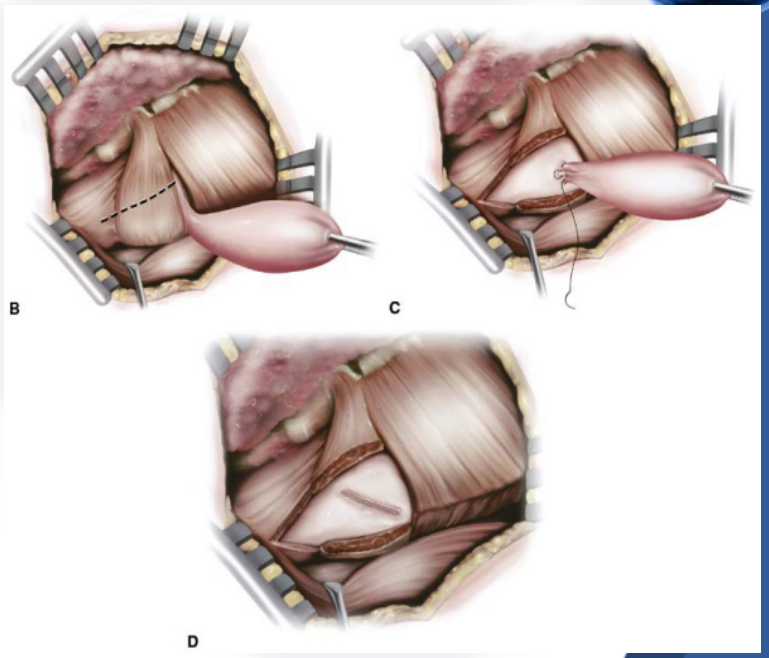
Diagnosis

- History
  - Progressive dysphagia
    - 90% of patients presenting with ZD
  - Regurgitation of food
  - Unprovoked aspiration
  - Noisy deglutition
- Barium Esophagogram

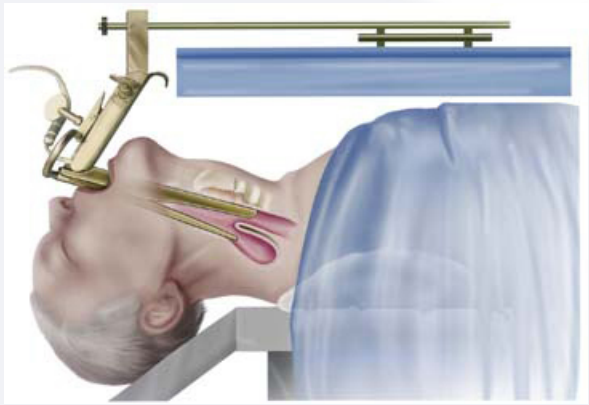


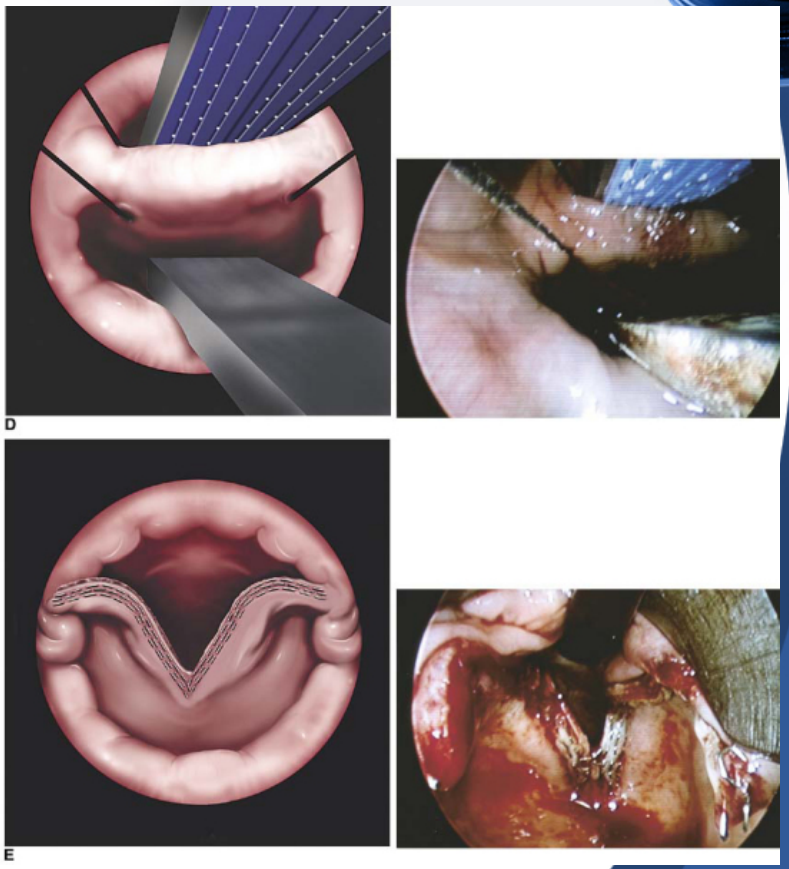
Treatment

- Surgery
  - Cricopharyngeal myotomy
- External
  - cricopharyngeal myotomy
  - Diverticulum is excised and the defect closed



Endoscopic





Motility Disorders of the Body & LES

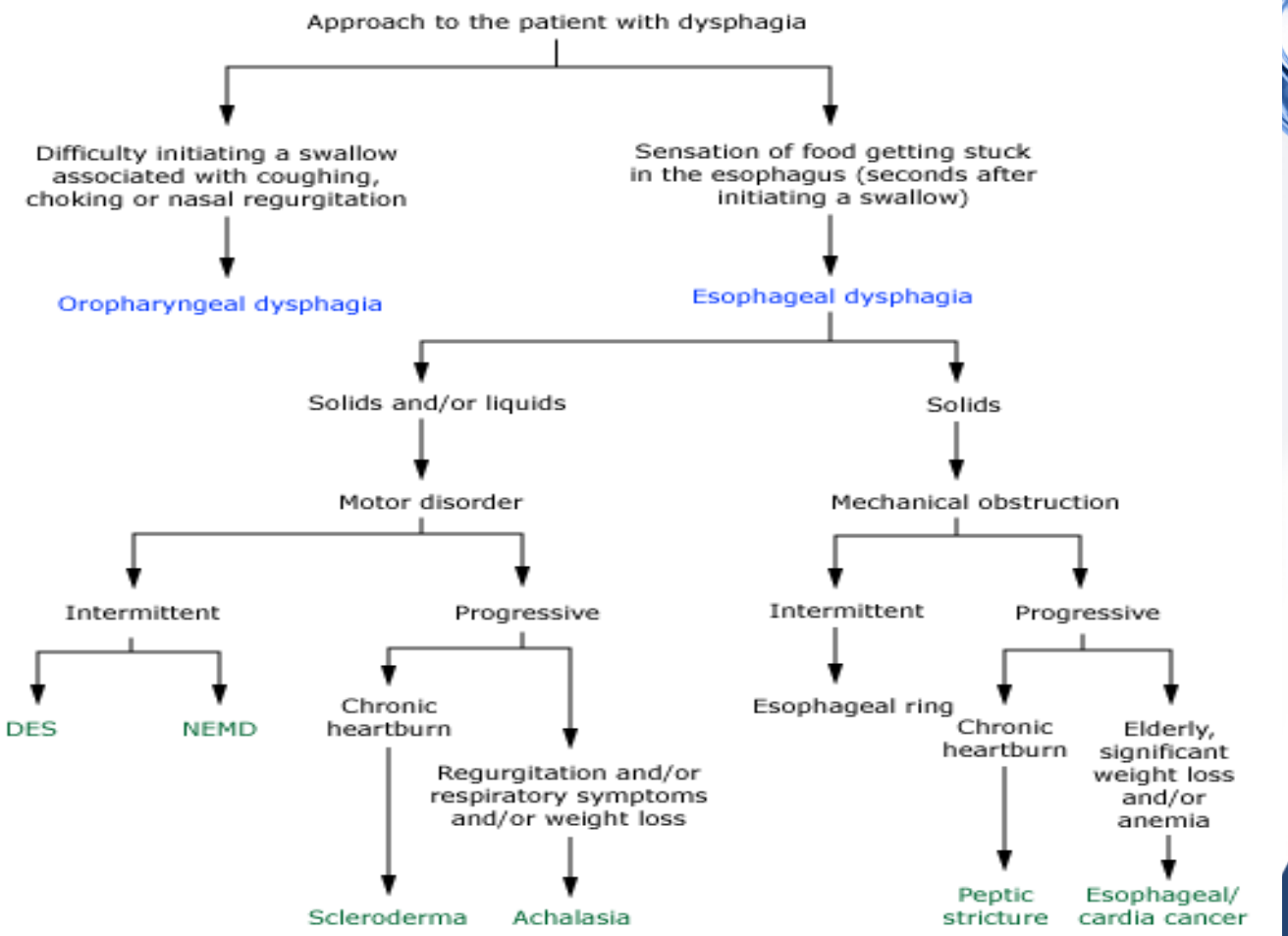
Motility Disorders of the Body & LES

- symptoms: usually dysphagia (intermittent and occurring with liquids & solids)
- diagnostic tests
  - barium esophagram
  - endoscopy
  - esophageal manometry
- disorders
  - achalasia
  - diffuse esophageal spasm (DES)
  - nutcracker esophagus
  - hypertensive LES
  - nonspecific esophageal dysmotility
    - hypomotility
    - hypermotlity

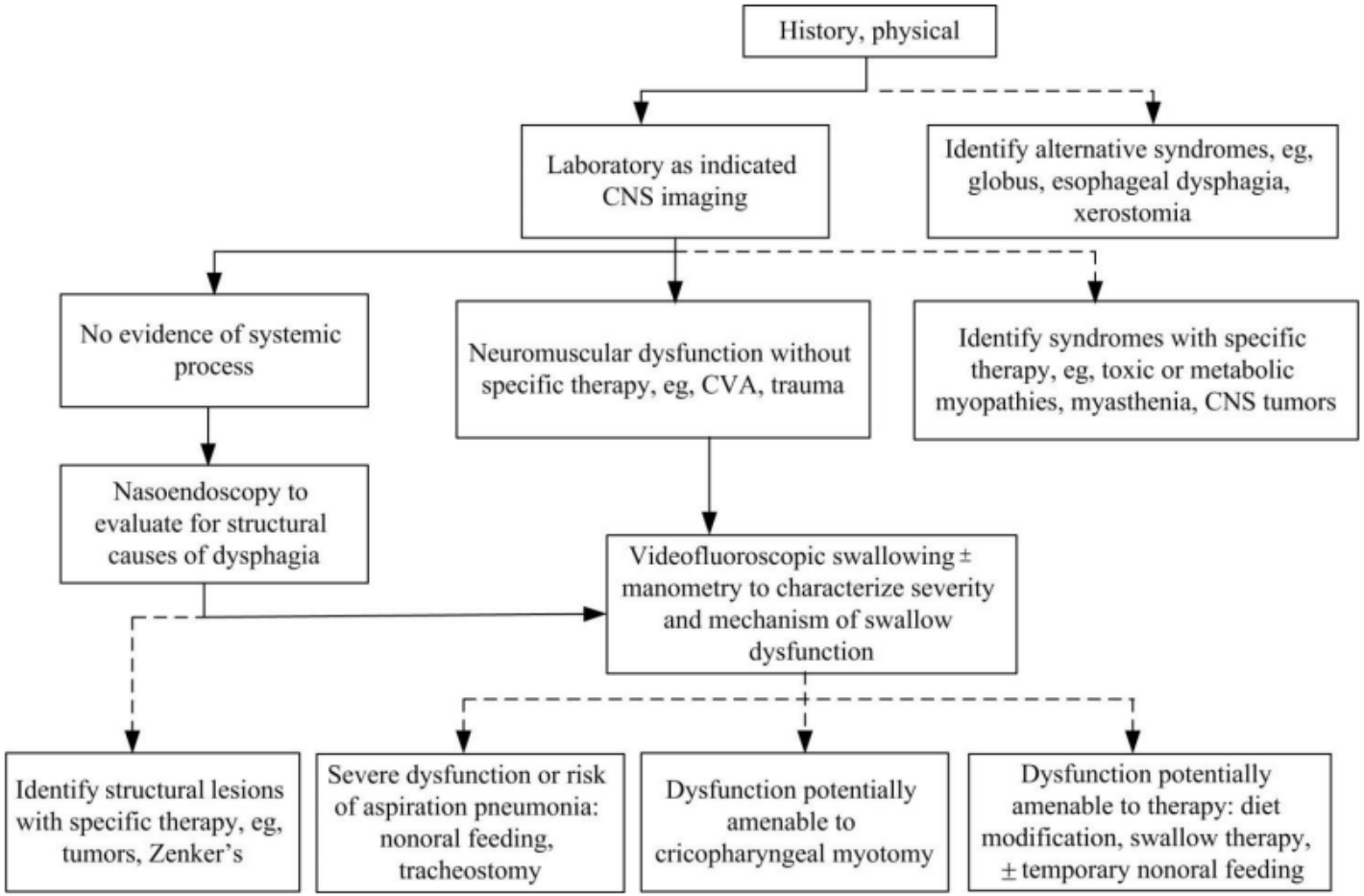
➢ Esophageal motility disorders are diseases attributable to *esophageal neuromuscular dysfunction* commonly associated with dysphagia*, chest pain*, or heartburn

➢ The major entities are ***achalasia, diffuse esophageal spasm (DES)***, and ***GERD***.

➢ Motility disorders can also be secondary to broader disease processes as is the case with pseudoachalasia, Chagas' disease, and scleroderma..

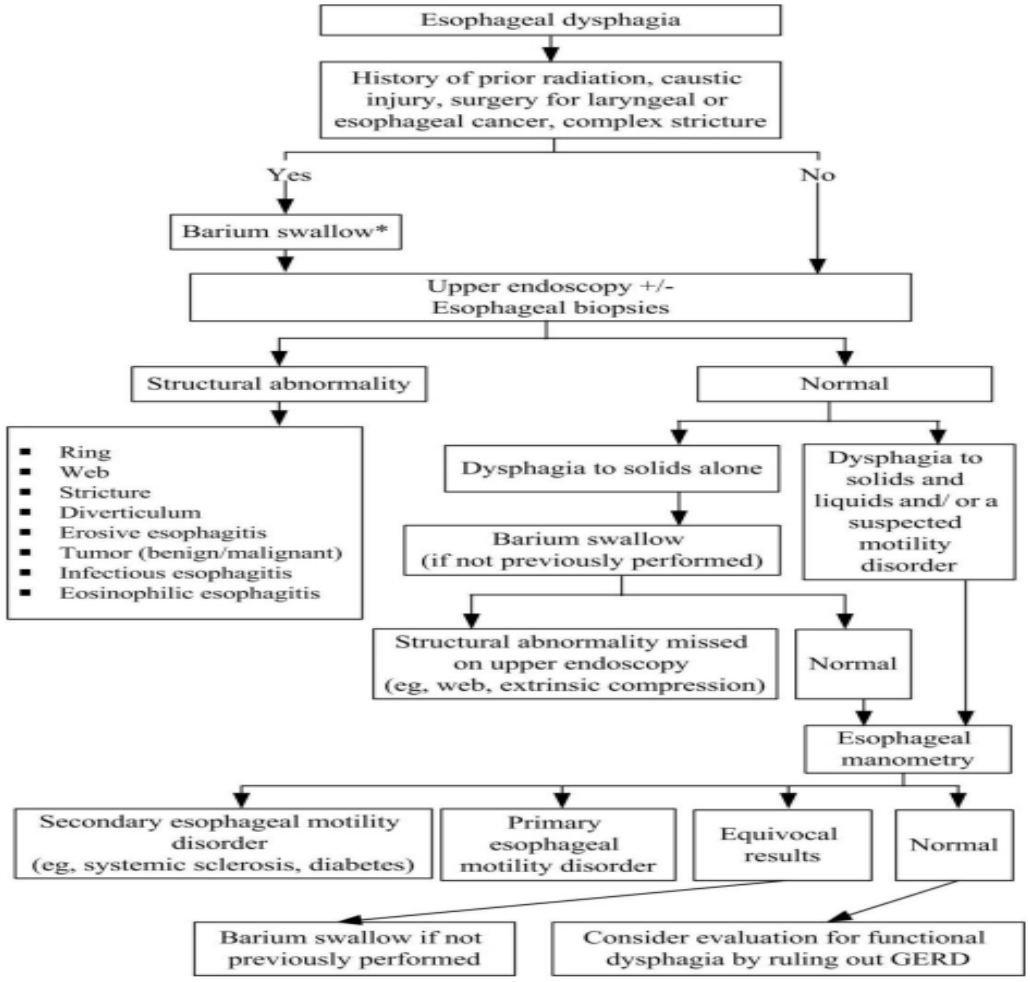


DES: diffuse esophageal spasm; NEMD: nonspecific esophageal motility disorder.



الگوریتم ۲. ارزیابی دیسفاژی Oropharyngeal.

- CNS: central nervous system; CVA: cerebrovascular accident.



الگوریتم ۳. ارزیابی بیمار با دیسفاژی Esophageal.

* انجام بلع باریم قبل از اِندوسکوپی فوقانی مورد بحث (contraversial) است.

- GERD: gastroesophageal reflux disease.

- Fass R, Feldman M, Robson KM. Approach to the evaluation of dysphagia in adults (Topic 2241). UpToDate (2022).

Achalasia

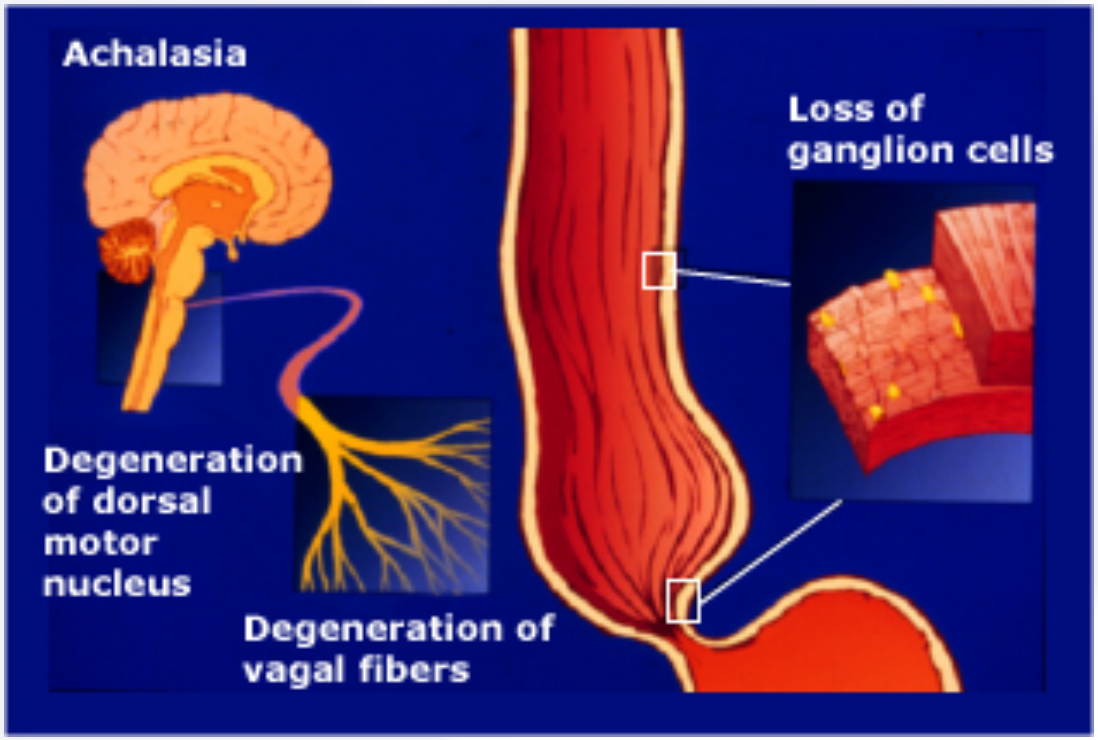
Achalasia

- **Epidemiology**
  - 1-2 per 200,000 population
  - usually presents between ages 25 to 60
  - male=female
  - average symptom duration at diagnosis: 2-5 years

Pathogenesis

Pathophysiology

- Achalasia results from the degeneration of neurons in the esophageal wall.
- Histologic examination reveals decreased numbers of neurons (ganglion cells) in the myenteric plexuses, and the ganglion cells that remain often are surrounded by lymphocytes
- This inflammatory degeneration preferentially involves the *nitric oxide-producing*, inhibitory neurons that effect the relaxation of esophageal smooth muscle.
- However the cholinergic neurons that contribute to LES tone by causing smooth muscle contraction are relatively spared.

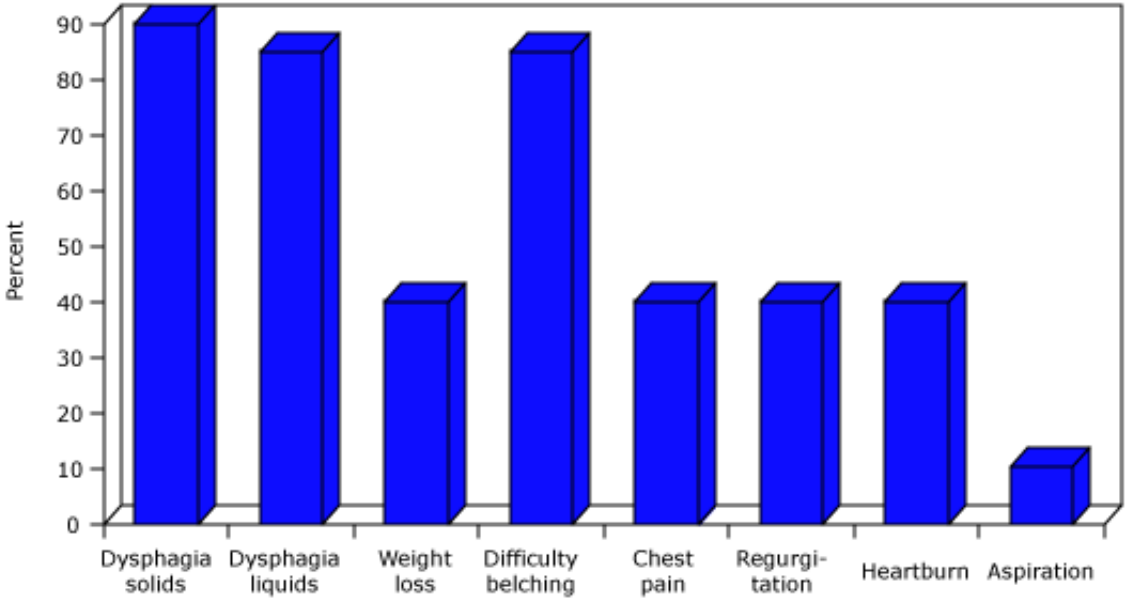

Achalasia
Loss of ganglion cells
Degeneration of dorsal motor nucleus
Degeneration of vagal fibers

Achalasia -aetiology

Achalasia

- ***Clinical presentation***
  - solid dysphagia 90-100% (75% also with dysphagia to liquids)
  - post-prandial regurgitation 60-90%
  - chest pain 33-50%
  - pyrosis 25-45%
  - weight loss
  - nocturnal cough and recurrent aspiration

Frequency of the symptoms of achalasia



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Diseases with manometric findings of achalasia

Diseases with manometric findings of achalasia

Malignancy, especially gastric carcinoma
Chagas disease
Amyloidosis
Sarcoidosis
Neurofibromatosis
Eosinophilic esophagitis
Multiple endocrine neoplasia, type 2B
Juvenile Sjögren's syndrome with achalasia and gastric hypersecretion
Chronic idiopathic intestinal pseudo-obstruction
Anderson-Fabry disease

Achalasia

- ***DDX***

- DES,
- Chagas' disease
- pseudoachalasia :

Tumor infiltration, most commonly seen with carcinoma in the gastric fundus or distal esophagus can mimic idiopathic achalasia.

Is more likely with advanced age>60, abrupt onset of symptoms (<1 year), and weight loss. Endoscopy should be part of the evaluation of achalasia.

➢ When the clinical suspicion for pseudoachalasia is high and endoscopy nondiagnostic, *CT scanning or EUS* may be of value

Diagnosis

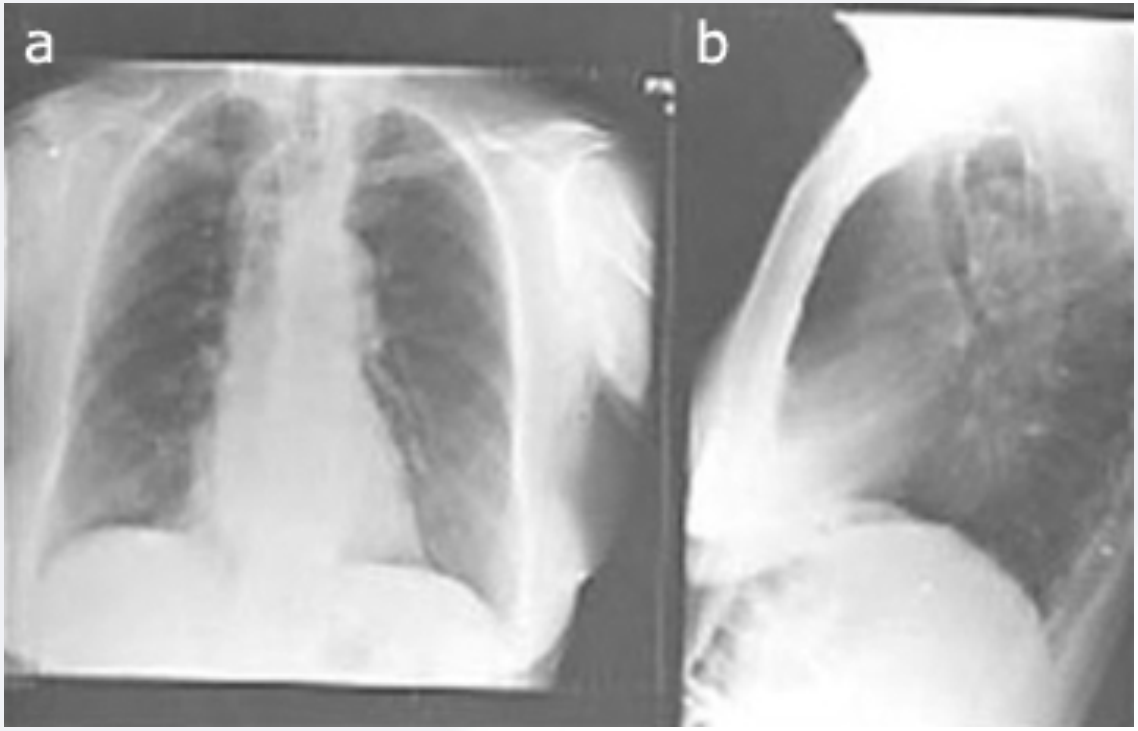
- The symptoms of achalasia often are insidious in onset and gradual in progression.
- As a result, patients typically experience symptoms for years before seeking medical attention.
  - Many patients are treated for other disorders such as gastroesophageal reflux disease before the diagnosis of achalasia is made.

Achalasia

- ***Diagnosis***
  - plain film (air-fluid level, wide mediastinum, absent gastric bubble, pulmonary infiltrates)
  - barium esophagram
  - endoscopy (rule out GE junction tumors, esp. age>60)
  - esophageal manometry (absent peristalsis, ↓ LES relaxation, & resting LES >45 mmHg)

Achalasia

Chest film of a patient with longstanding achalasia



Barium esophagram

- Dilation of the esophagus
- Narrow esophagogastric junction with "bird-beak" appearance caused by the persistently contracted LES
- Aperistalsis
- Poor emptying of barium

However barium esophagram may be falsely negative in one-third of patients

Achalasia

- esophagram

Timed barium esophagram (TBE)

- A timed barium esophagram (TBE) can be useful for documenting bolus retention for patients with untreated achalasia in addition to assessing response to treatment.

- To perform TBE, the patient drinks approximately 100 ml of low-density barium sulphate suspension, and oblique films are taken in the upright position at 1, 2, and 5 minutes after barium ingestion .
- Most patients with untreated achalasia exhibit barium retention (measured by the height of the barium column) at one or more of these time points.
- After treatment, complete esophageal emptying at one minute post-barium ingestion is consistent with treatment success

Timed barium Esophagogram(TBE)

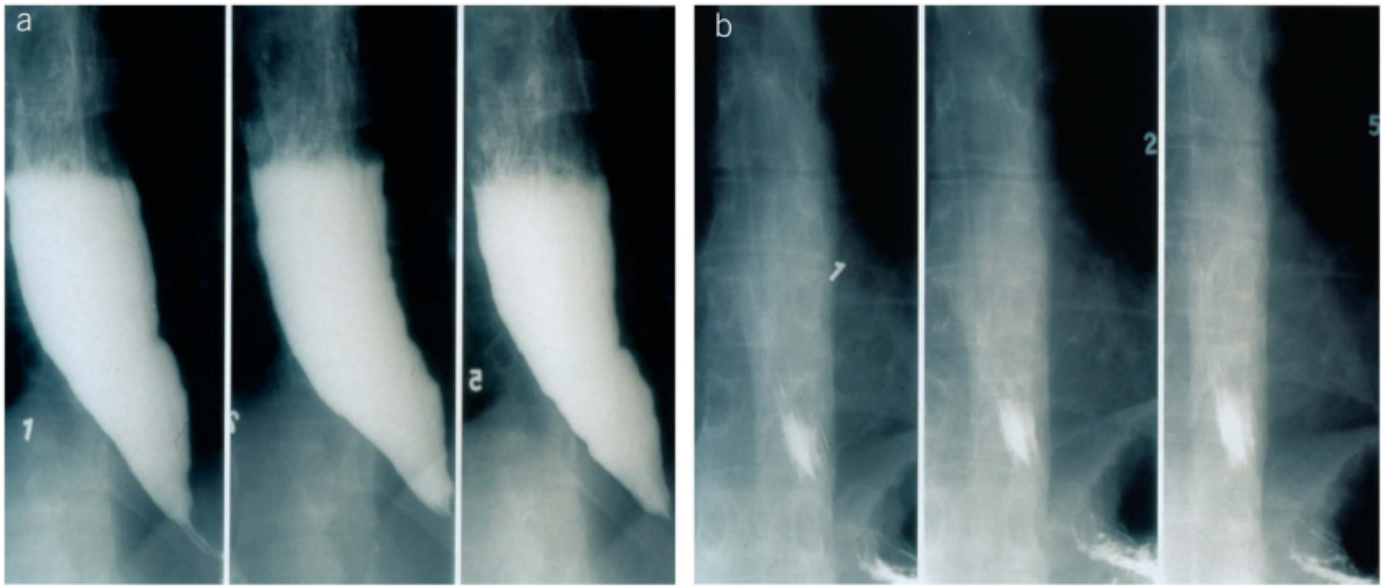
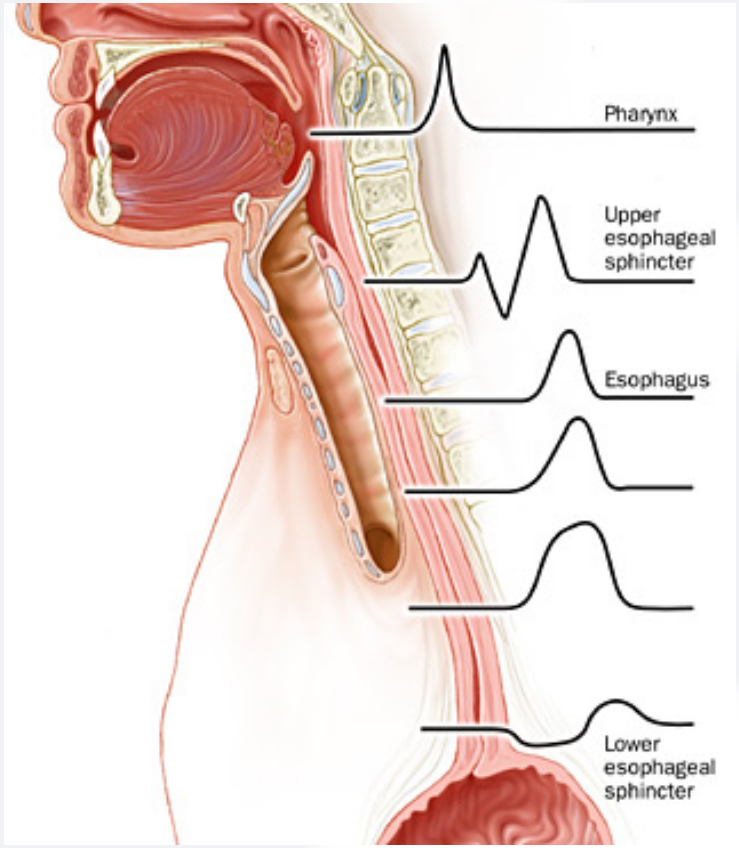


Figure 7. Timed barium swallow (**a**) before and (**b**) after pneumatic dilation showing retention of barium in the former and complete emptying posteffective therapy.

Oesophageal Manometry

Esophageal Manometry

- Absence of Esophageal peristalsis
- Partial or no relaxation of LES


Pharynx
Upper esophageal sphincter
Esophagus
Lower esophageal sphincter

Conventional manometry

Typical manometric findings

- **Aperistalsis** in the distal two-thirds of the esophagus

simultaneous contractions with amplitudes <40 mmHg

- **Incomplete LES relaxation**

Normally, there is complete relaxation of the LES after a swallow to a level <8 mmHg

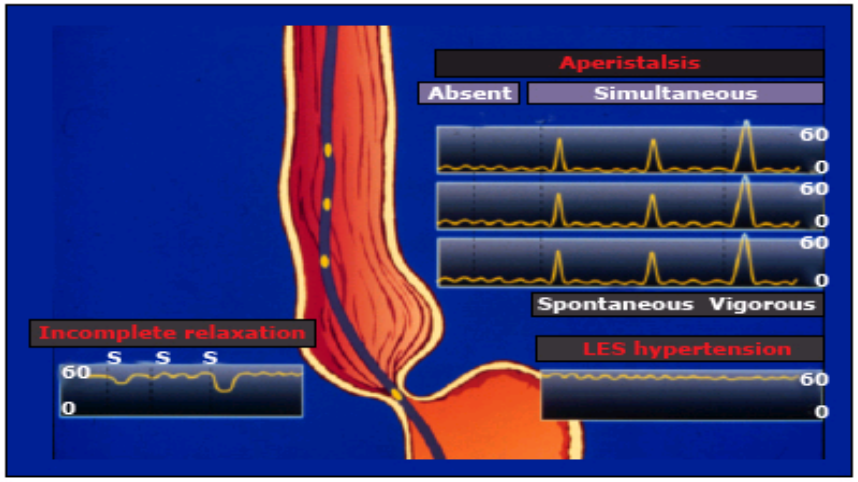
- **Elevated resting LES pressure** resting LES pressures to rise to hypertensive levels (>45 mmHg).

Conventional manometry

Atypical manometric findings

- ***Vigorous***
- "normal" to high-amplitude simultaneous esophageal body contractions >40 mmHg in the presence of a nonrelaxing LES

Manometric features of achalasia



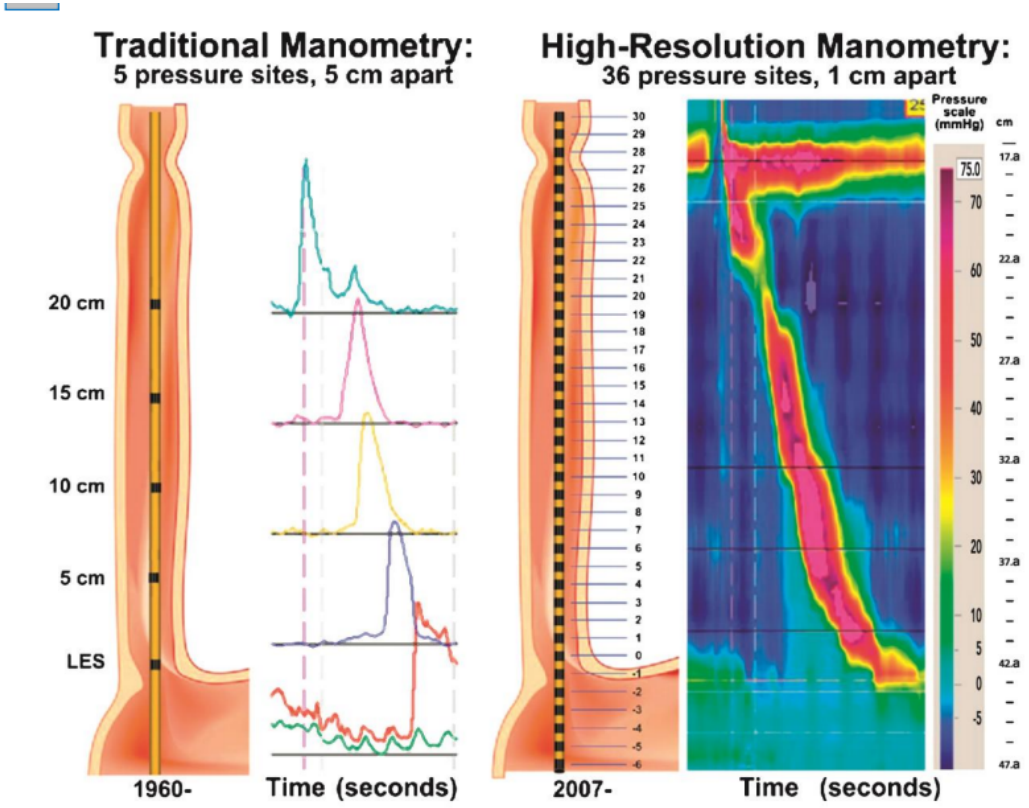
There are three characteristic manometric features of achalasia: elevated resting lower esophageal sphincter (LES) pressure (above 45 mmHg); incomplete LES relaxation after a swallow (S); aperistalsis in the smooth muscle portion of the body of the esophagus. Swallows may elicit no esophageal contraction or may be followed by simultaneous contractions. The esophagus may also contract spontaneously in a simultaneous fashion. In some cases, the simultaneous esophageal contractions have amplitudes >60 mmHg, a condition known as "vigorous" achalasia.

In contrast to conventional manometry where three to eight sensors are spaced at 3 to 5 cm intervals,

HRM sensors are typically spaced 1 cm apart along the length of the manometric assembly.

Catheters with up to 36 sensors allow for simultaneous pressure readings spanning both sphincters and the entire interposed esophagus.

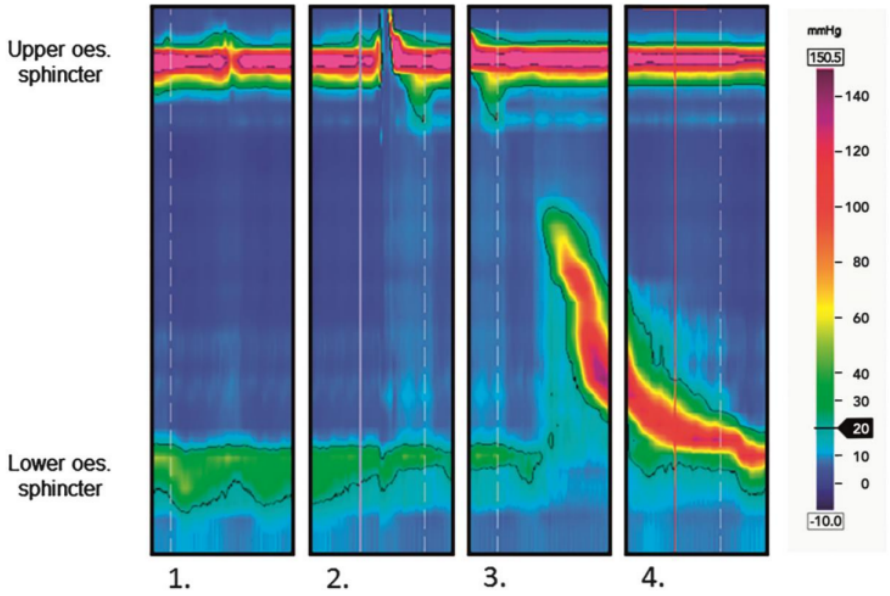
Esophageal pressure topography (EPT) is a three-dimensional plotting format devised for depiction of HRM studies. EPT interpolates pressure values between sensors to create a pressure continuum



شکل ۲. مقایسه‌ی مانومتری مری با وضوح بالا (HRM) با مانومتری معمولی (conventional).

برخلاف مانومتری معمولی (conventional) که در آن ۳ تا ۸ حس‌گر (sensor) در فواصل ۳ تا ۵ سانتی‌متری قرار می‌گیرند، HRM دارای ۳۶ حس‌گر (sensor) با فاصله ۱ سانتی‌متری از یکدیگر در طول مری است. EPT، اندازه فشار (pressure magnitude) گزارش شده توسط حس‌گرها (sensor) را با استفاده از رنگ‌های سرد برای نشان دادن فشار کم و رنگ‌های گرم برای نشان دادن فشارهای بالاتر، بر اساس زمان (محور X) و مکان (محور Y) نشان می‌دهد.

- HRM: high-resolution manometry; cm: centimeter; mmHg: millimeter of mercury.



شکل ۶. مانومتری مری با وضوح بالا (HRM) در بلع نرمال.

۱. اسفنکترهای فوقانی (UES) و تحتانی مری (LES) در حالت استراحت منقبض می‌مانند.

۲. بلع با شل شدن اسفنکتر فوقانی مری (UES) شروع می‌شود، تا لقمه‌غذا عبور کند.

۳. یک انقباض قوی در اسفنکتر فوقانی مری (UES)، محتوای آن را به سمت پایین مری حرکت می‌دهد. در همین حال، اسفنکتر تحتانی مری (LES) زود شل می‌شود تا اجازه عبور هوای گیر افتاده در مری را بدهد.

۴. لقمه‌ی غذا قبل از انقباض اسفنکتر تحتانی مری (LES) برای جلوگیری از بازگشت محتویات معده به مری، از اسفنکتر تحتانی مری (LES) شل شده عبور کرده، وارد معده می‌شود.

- HRM: high-resolution manometry; mmHg: millimeter of mercury; UES: upper esophageal sphincter; LES: lower esophageal sphincter.

High resolution manometry

- Achalasia is diagnosed on (HRM) by impaired EGJ relaxation defined as a mean four-second integrated relaxation pressure (IRP) ≥15 mmHg

- **Type I (classic)** swallowing results in no significant change in esophageal pressurization
- **Type II achalasia**, swallowing results in simultaneous pressurization that spans the entire length of the esophagus
- **Type III (spastic)** swallowing results in abnormal, lumen-obliterating contractions or spasms

- ●**Type I (classic achalasia)** – Swallowing results in no significant change in esophageal pressurization. By CC-4 criteria, type I achalasia has 100 percent failed peristalsis as indicated by a distal contractile integral (DCI, an index of the strength of distal esophageal contraction) <100 mmHg·s·cm.
- ●**Type II** – Swallowing results in simultaneous pressurization that spans the entire length of the esophagus. According to CC-4, type II achalasia has 100 percent failed peristalsis and pan-esophageal pressurization seen in ≥20 percent of swallows.
- ●**Type III (spastic achalasia)** – Swallowing results in premature and often lumen-obliterating contractions or spasms. By CC-4 criteria, type III achalasia has no normal peristalsis and premature (spastic) contractions with distal latency <4.5 seconds and DCI >450 mmHg·s·cm seen in ≥20 percent of swallows.
- These subtypes have important implications for management, and HRM is preferred over conventional manometry for the diagnosis of achalasia

Table 2. Comparison of manometric abnormalities in conventional and high-resolution manometry

Manometric features of achalasia	Conventional manometry Line tracing format	High-resolution manometry Esophageal pressure topography
LES		
	Impaired LES relaxation[a] • Mean swallow induced fall in resting LES pressure to a nadir value of >8 mm Hg above gastric pressure • Complete relaxation to gastric baseline with a short duration (<6 s)[b] *Basal pressure*[b] • >45 mm Hg	*Impaired EGJ relaxation* • Mean 4 s IRP ≥10 mm Hg over test swallows[a]
Esophageal peristalsis		
	Aperistalsis in distal 2/3 of the esophagus • No apparent contractions • Simultaneous contractions with amplitudes <40 mm Hg	*Aperistalsis* • Absent peristalsis (type I) • Pan-esophageal pressurization (type II)
Atypical/variants		
	Vigorous • Preserved peristalsis with esophageal contractions >40 mm Hg • Simultaneous contractions >40 mm Hg -Isobaric -Nonisobaric	• Spastic achalasia (type III)

EGJ, esophagogastric junction; IRP, integrated relaxation pressure; LES, lower esophageal sphincter.
[a]Required for diagnosis.
[b]Supportive for the diagnosis.

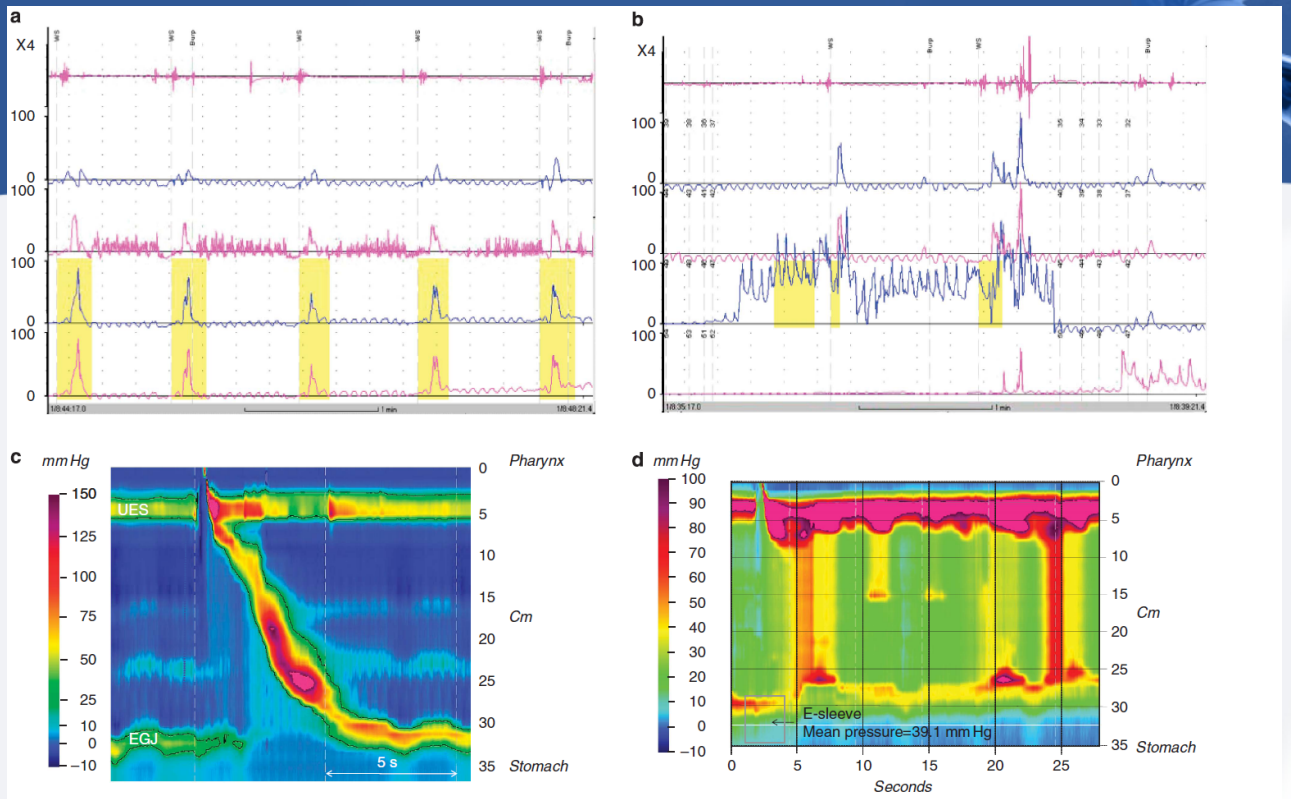


Figure 1 . Manometric tracings of normal and achalasia. (a) Simultaneous esophageal contractions associated with high lower esophageal sphincter (LES) pressure and (b) incomplete relaxation noted during conventional water-perfused manometry. High-resolution manometry (HRM) tracings of (c) normal esophageal peristalsis and (d) achalasia showing simultaneous contractions along the esophagus with high E-sleeve LES pressure and incomplete relaxation. EGJ, esophagogastric junction; UES, upper esophageal sphincter

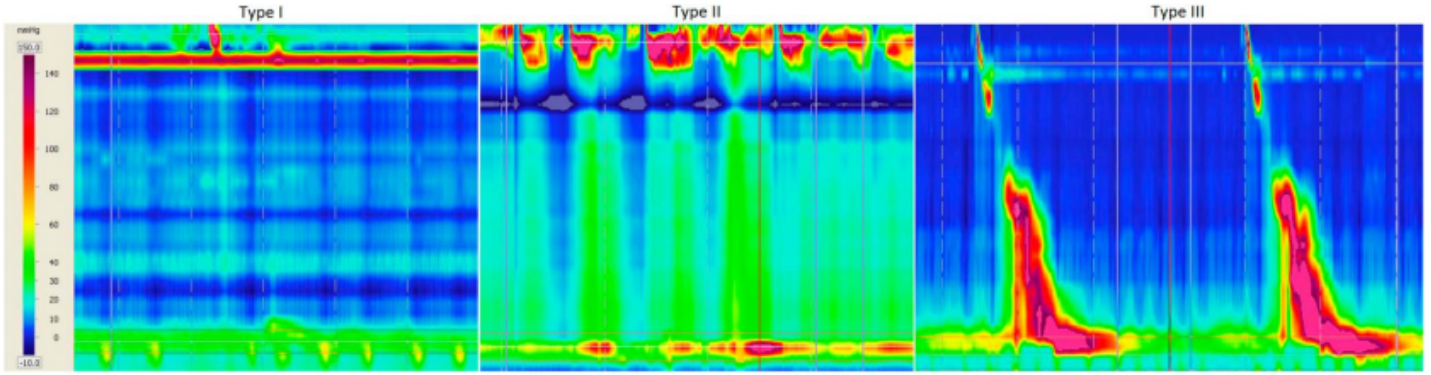


Figure 2. High resolution manometry of achalasia phenotypes: type I-absent pressurization (left), type II-pan pressurization (middle), and type III-spastic contractions (right). Lower esophageal sphincter relaxation is impaired for all subtypes.

EVALUATION

- Achalasia should be suspected in patients with dysphagia to solids and liquids and in those with regurgitation unresponsive to a trial of (PPI) therapy for 4 weeks

- Manometry is required to establish the diagnosis In patients with equivocal motility testing

- **Barium esophagram** should be performed to assess esophageal emptying and EGJ morphology.
- Endoscopic evaluation should be performed in patients with suspected achalasia to exclude a malignancy at EGJ that can mimic primary achalasia
- evaluation with endoscopic (EUS FNA) when pseudoachalasia due to malignancy is suspected

EGD

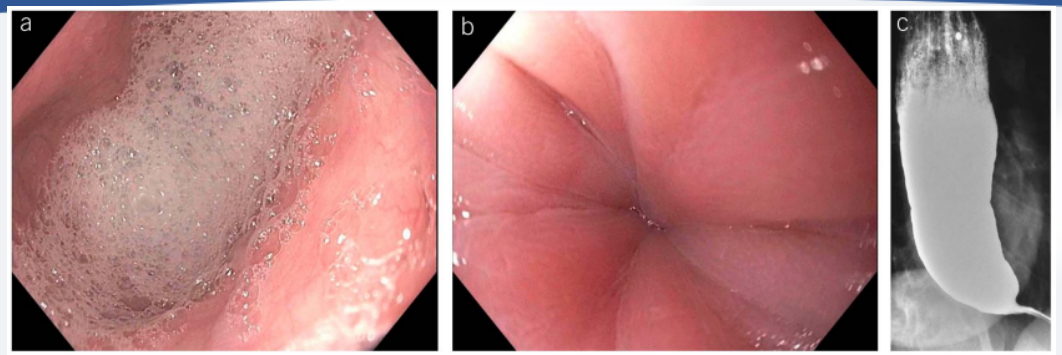
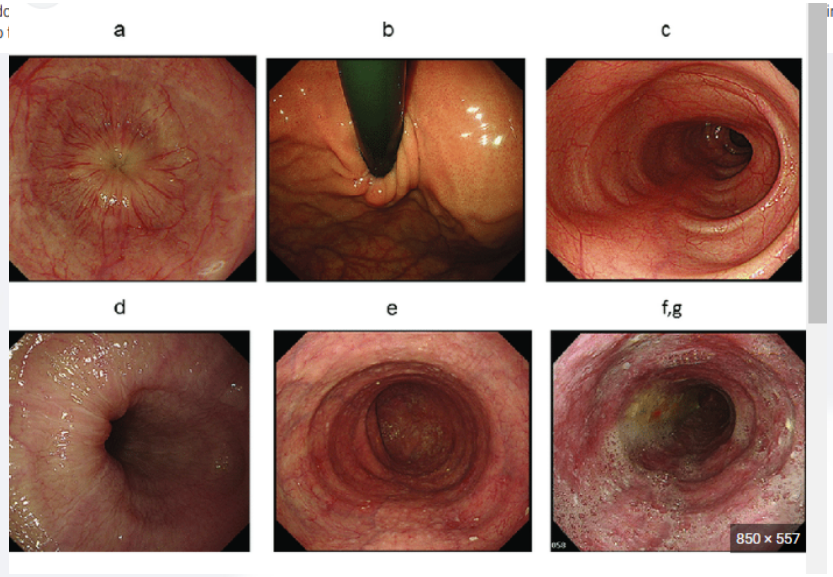


Figure 1. (**a**) Endo ing more than usual pressure to

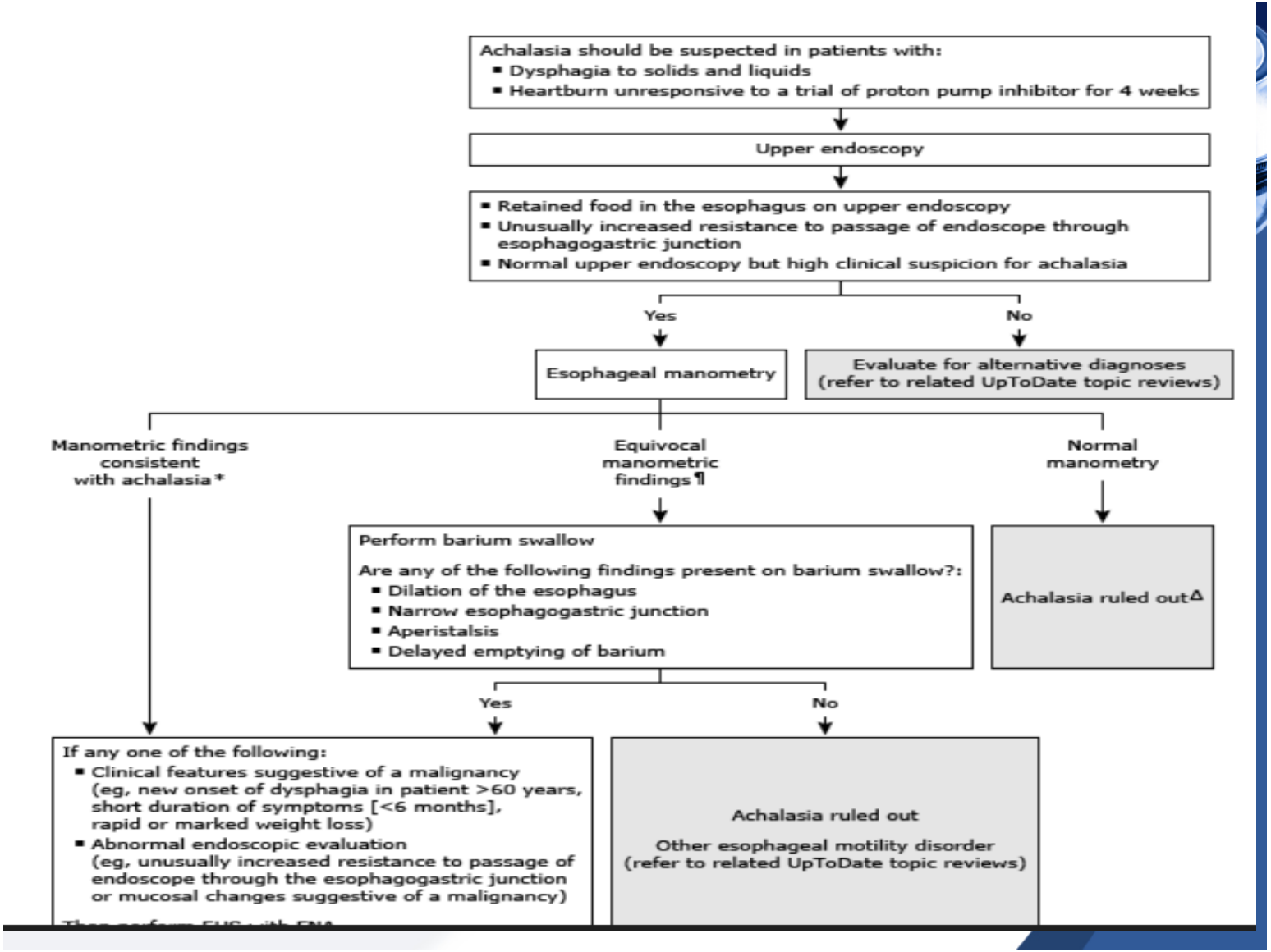


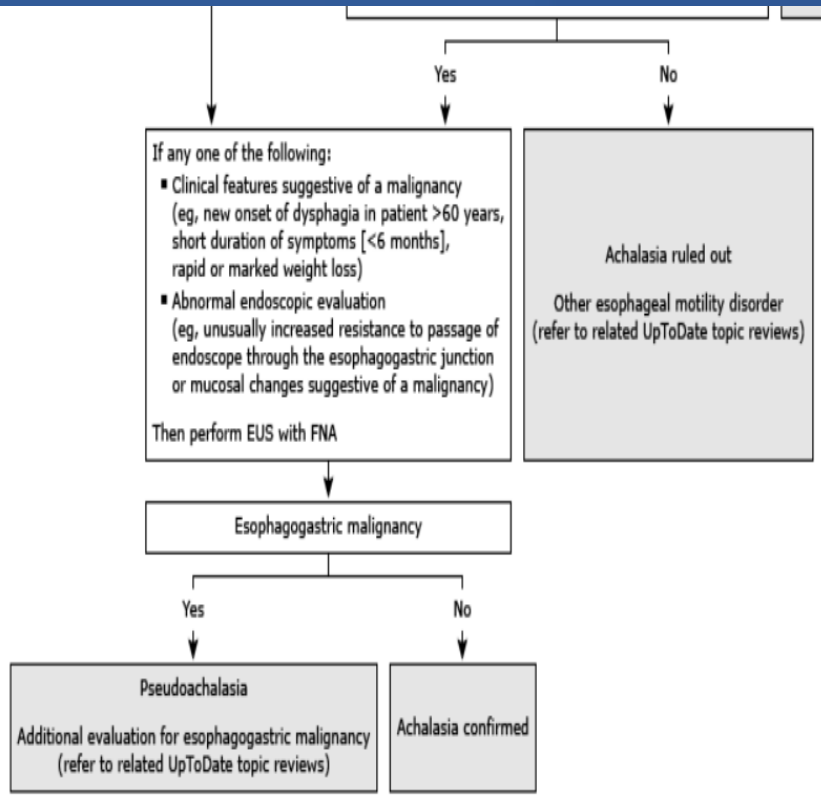
EUS

- **Endoscopic ultrasound**
- Findings of achalasia on (EUS) can include:
- A thickened circular muscle layer at the LES and through the smooth muscle esophagus.
- Although the accuracy of EUS in distinguishing achalasia from pseudoachalasia has not been established,
- EUS is useful for characterizing tumors of the distal esophagus and gastric cardia .
- *EUS findings of marked (>10 mm) and/or asymmetric esophageal wall thickening are suggestive of an underlying malignancy that may be confirmed by fine-needle aspiration*

Functional lumen imaging probe (FLIP)

- (FLIP) is a catheter-based instrument with an attached, fluid-filled balloon that surrounds a series of impedance planimetry electrodes.
- The FLIP catheter is passed into the esophagus, and it displays the diameter of various esophageal segments in real-time images. In addition, FLIP provides real-time information about the distensibility of the esophageal segments in the form of a distensibility index, and information about esophageal motility in the form of FLIP topography.
- *In achalasia, FLIP reveals a narrowed esophagogastric junction segment that has a low distensibility index*, and inflation of the FLIP balloon results in abnormal or no esophageal contractions.
- FLIP may provide useful data to establish the diagnosis of achalasia in equivocal cases


Achalasia should be suspected in patients with:
▪ Dysphagia to solids and liquids
▪ Heartburn unresponsive to a trial of proton pump inhibitor for 4 weeks
Upper endoscopy
▪ Retained food in the esophagus on upper endoscopy
▪ Unusually increased resistance to passage of endoscope through esophagogastric junction
▪ Normal upper endoscopy but high clinical suspicion for achalasia
Yes
No
Esophageal manometry
Evaluate for alternative diagnoses (refer to related UpToDate topic reviews)
Manometric findings consistent with achalasia*
Equivocal manometric findings¶
Normal manometry
Perform barium swallow
Are any of the following findings present on barium swallow?:
▪ Dilation of the esophagus
▪ Narrow esophagogastric junction
▪ Aperistalsis
▪ Delayed emptying of barium
Achalasia ruled out△
Yes
No
If any one of the following:
▪ Clinical features suggestive of a malignancy (eg, new onset of dysphagia in patient >60 years, short duration of symptoms [<6 months], rapid or marked weight loss)
▪ Abnormal endoscopic evaluation (eg, unusually increased resistance to passage of endoscope through the esophagogastric junction or mucosal changes suggestive of a malignancy)
Achalasia ruled out
Other esophageal motility disorder (refer to related UpToDate topic reviews)



LES: lower esophageal sphincter; EUS: endoscopic ultrasound; FNA: fine needle aspiration; IRP: integrated relaxation pressure.

* Diagnostic manometric findings of achalasia are incomplete relaxation of the LES (IRP above the upper limit of normal) and aperistalsis in the distal two-thirds of the esophagus

¶ Equivocal manometric findings include incomplete LES relaxation but some preserved peristalsis; some complete LES relaxation with aperistalsis.

Disease course

- Without treatment, patients with achalasia develop progressive dilation of the esophagus

- Late- or end-stage achalasia is characterized by esophageal tortuosity, angulation, and severe dilation or megaesophagus (diameter >6 cm)
- Approximately 10 to 15 % of patients who have undergone treatment for achalasia will develop late- or end-stage achalasia and 5 % require esophagectomy

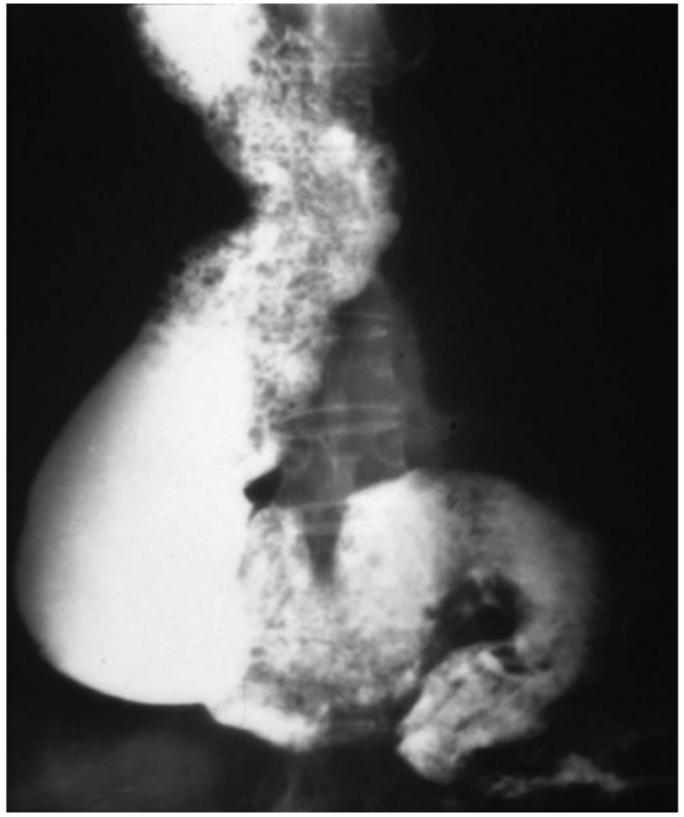


Figure 6. Dilated sigmoid esophagus representing end-stage achalasia with retained saliva and barium.

Cancer risk

- The risk of esophageal SCC is significantly increased in achalasia, and the estimated incidence rate is approximately 1 cancer per 300 patient years. This represents a hazard ratio of 28 for developing esophageal SCC.

- *We recommend against routine endoscopic surveillance for esophageal carcinoma in patients with achalasia.*

Achalasia

Treatment

- There is no known way of preventing or reversing achalasia.

- LES pressure can be reduced by :
  - pharmacologicals therapy,
  - pneumatic dilation
  - surgical myotomy
  - POEM

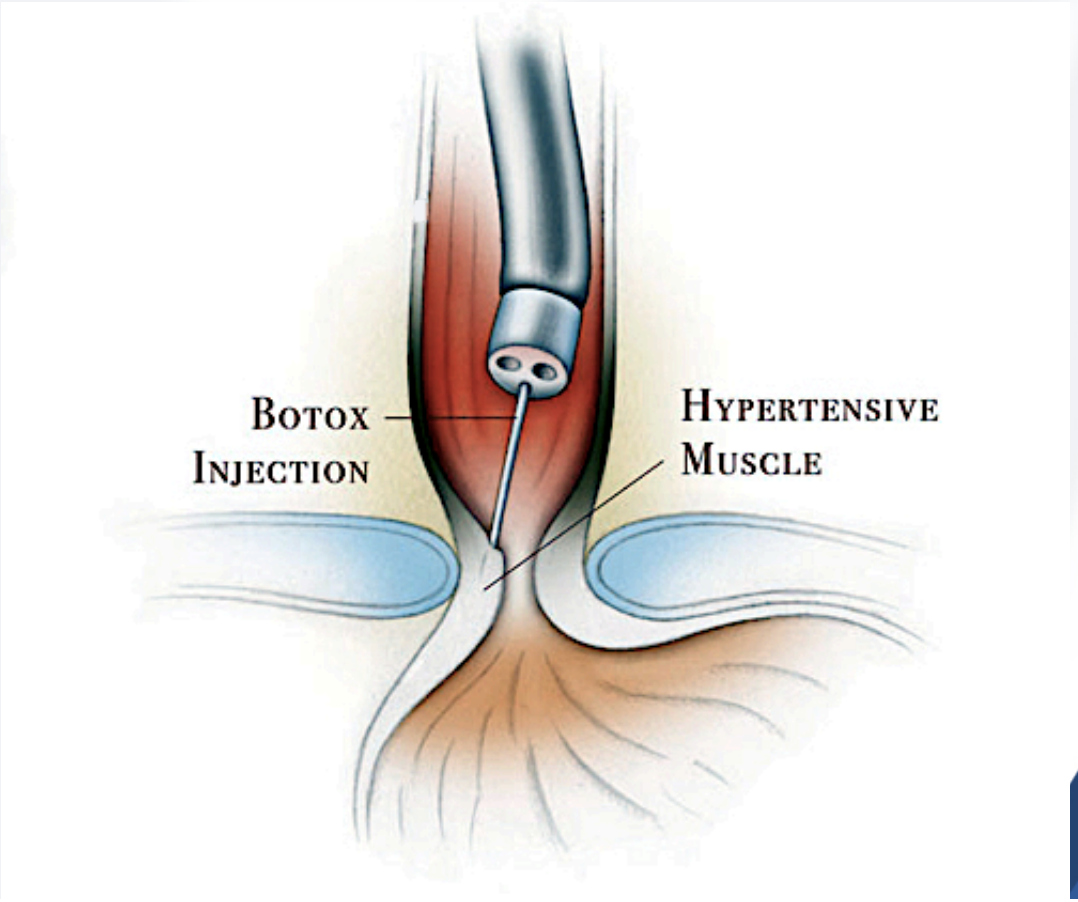
Achalasia

- **pharmacological therapy**

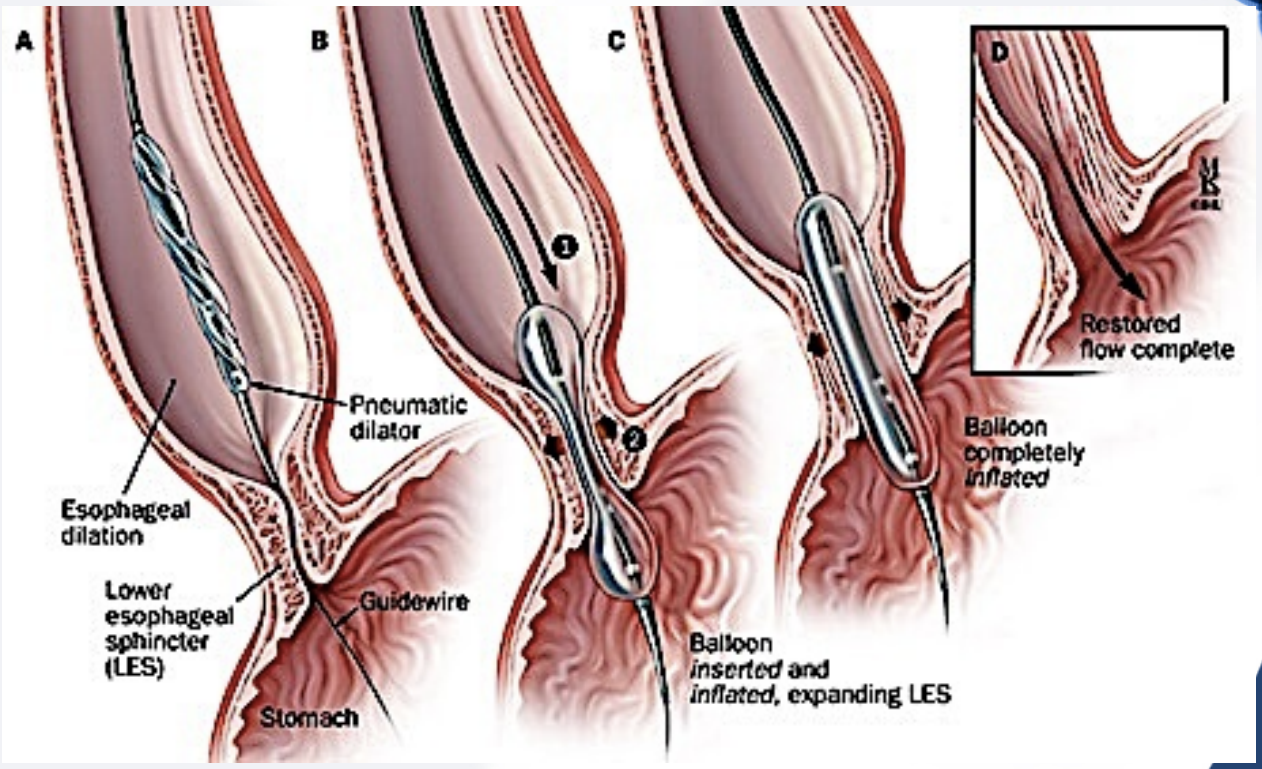
- ***Oral pharmacologic*** *therapies are the least effective treatment* options in achalasia
- Calcium channel blockers and long-acting nitrates are the two most common medications used to treat achalasia

Botulinum toxin (prevents ACH release at NM junction)

Intersphincteric Injection of Botulinum Toxin



Pneumatic Dilatation



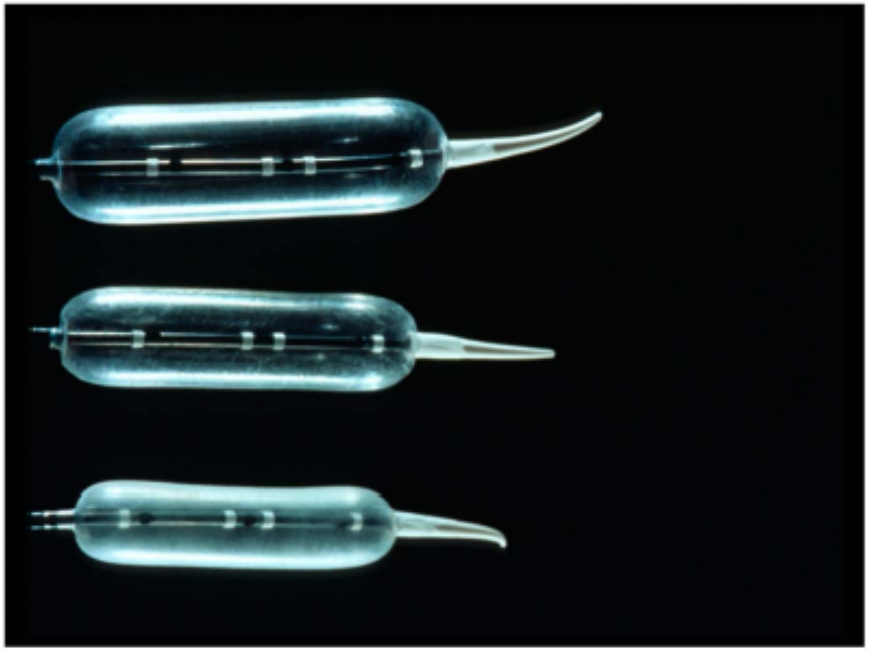
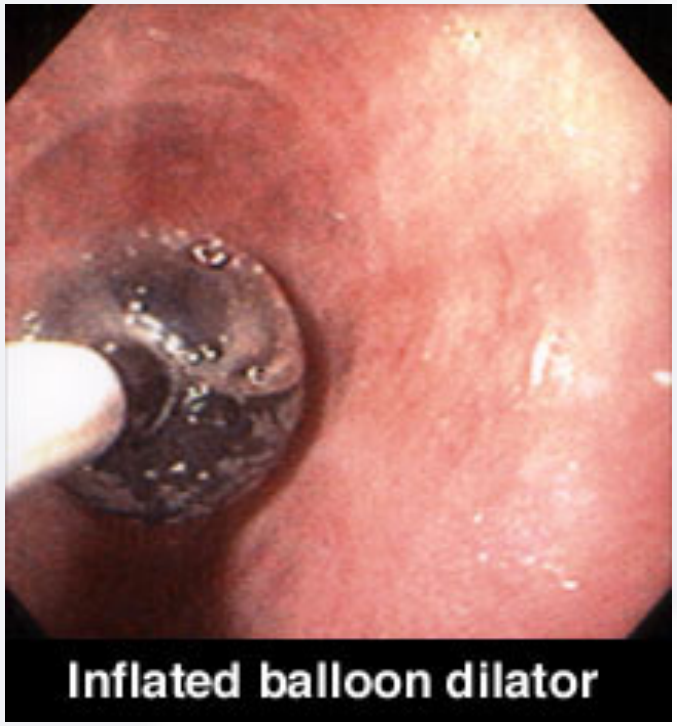


Figure 3. Pneumatic dilator sizes 3.0 cm (bottom), 3.5 cm (middle), and 4.0 cm (top) used in treating patients with achalasia. Graded approach of starting with the smaller 3.0-cm balloon and progressing to the larger sizes if failed therapy is recommended in all except younger male patients in whom initial approach with 3.5-cm balloon may be used.

Pneumatic Dilation



Inflated balloon dilator

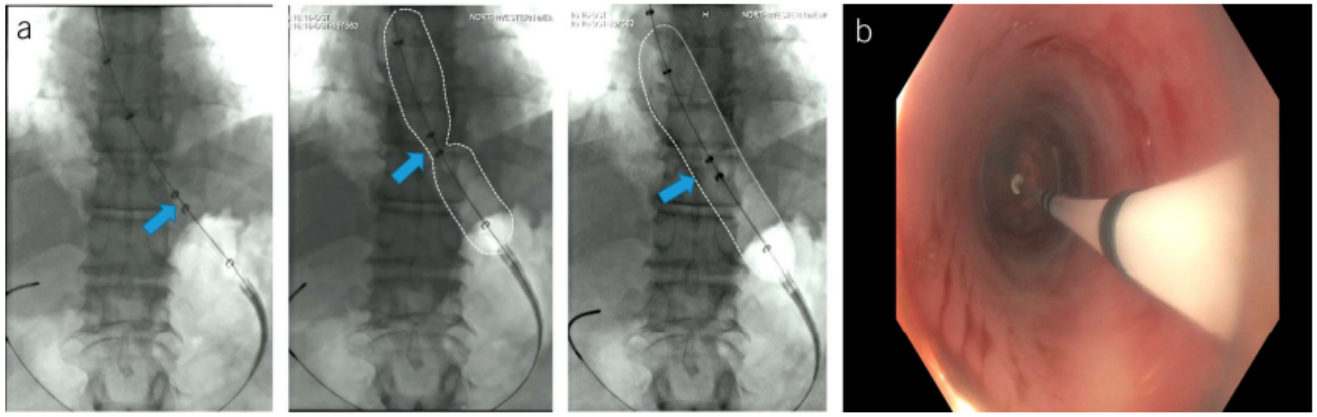


Figure 4. (**a**) Fluoroscopic image of using a 3.0-cm pneumatic dilator in a patient with achalasia showing balloon positioning of 2 rings (middle of balloon; blue arrow) at the esophagogastric junction with subsequent dilation to obliterate the balloon waist. (**b**) Pneumatic dilation via direct endoscopic approach showing positioning of the 2 rings at the esophagogastric junction during endoscopy with inflation of the balloon to the maximum pressure of 13 mm Hg.

Best outcomes after pneumatic dilation :

- Those with narrow esophageal diameter

- Those with a type II pattern by high-resolution manometry

Post-procedure complications :

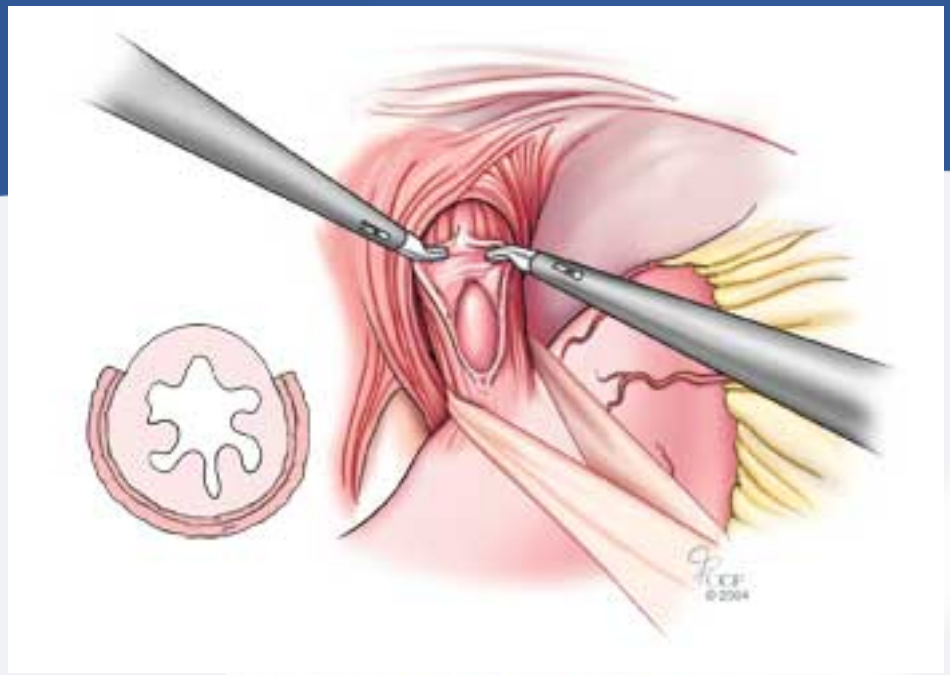
perforation (approximately 2%)
Heartburn (15 to 35%)

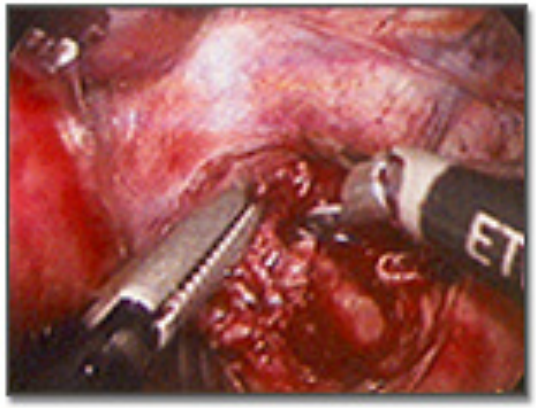
For patients who elect to undergo pneumatic dilation, we suggest surgical myotomy if symptoms persist despite three attempts at pneumatic dilation

Achalasia

Surgical myotomy

- The most common surgical procedure for achalasia is laparoscopic Heller myotomy, with an antireflux procedure (partial fundoplication); good to excellent results are reported in 62–100% of cases. Occasionally.
- patients with advanced disease fail to respond to pneumatic dilatation or Heller myotomy.
- In such refractory cases, esophageal resection with gastric pull-up or interposition of a segment of transverse colon may be the only option other than gastrostomy feeding.

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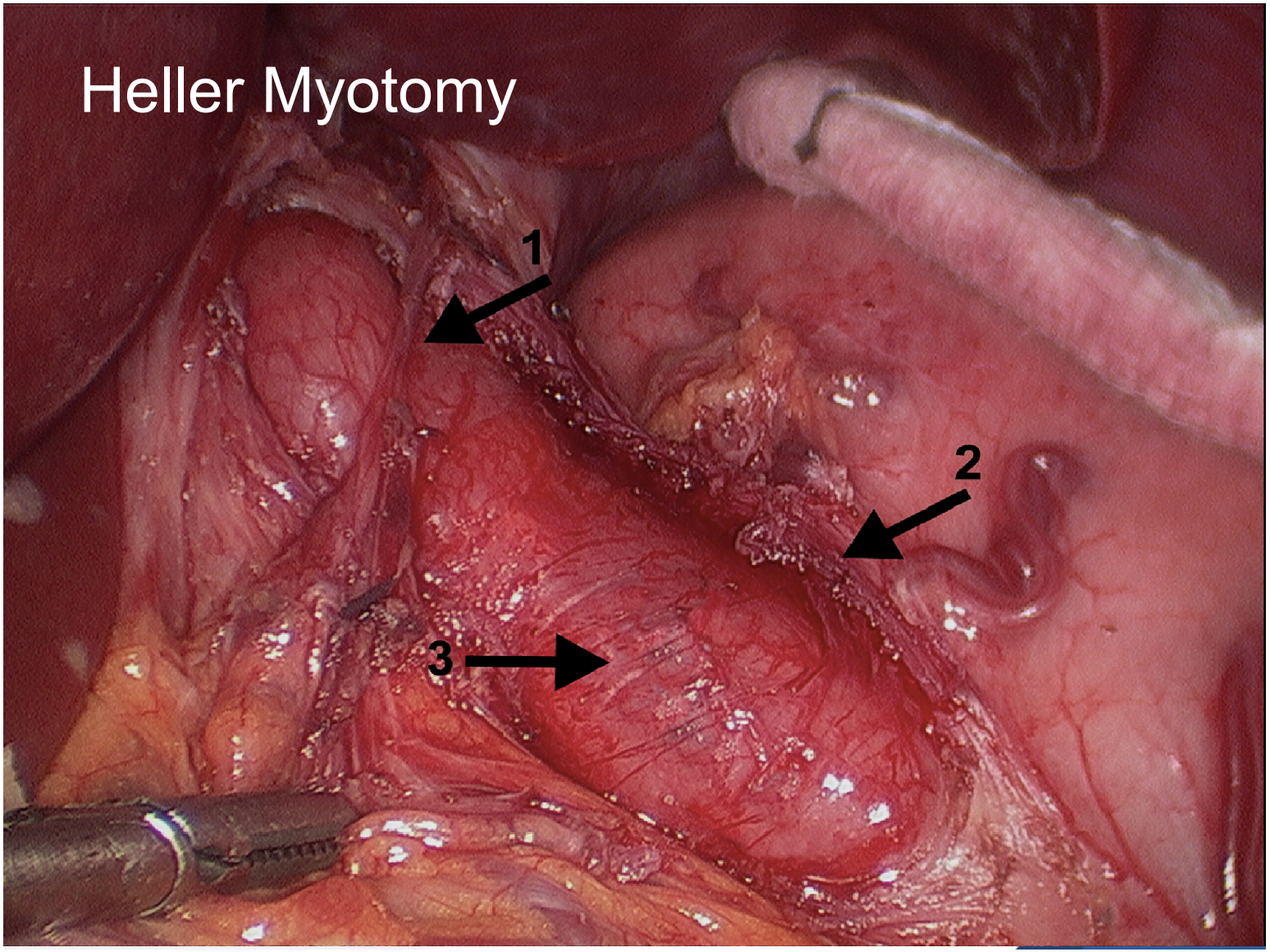

Heller Myotomy
1
2
3

Peroral endoscopic myotomy (POEM)

(POEM): a form of natural orifice transluminal endoscopic surgery (NOTES)

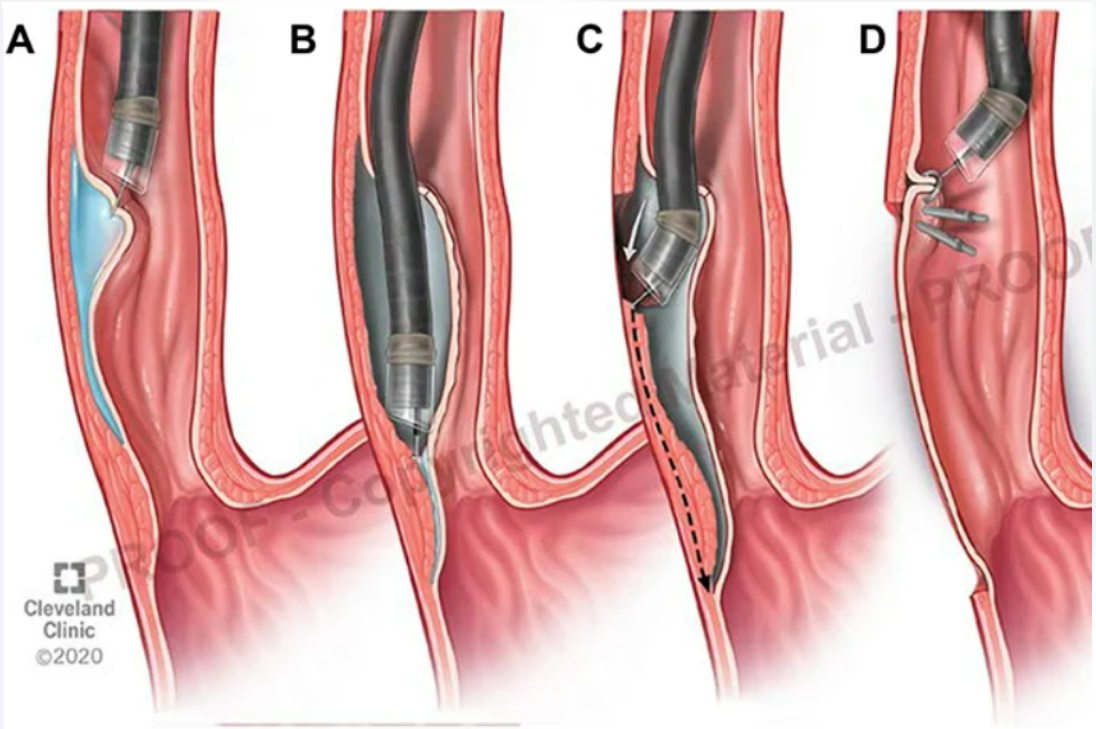
Is an endoscopic method for performing myotomy of the LES .During POEM, the endoscopist makes an incision in the esophageal mucosa, and the endoscope is passed through that incision into the esophageal submucosa, creating a submucosal tunnel that is extended distally into the gastric cardia.

POEM is an effective submucosal endoscopic technique for performing myotomy of the LES and more proximal esophageal muscle.

In addition, good results for POEM have been reported in patients with achalasia conditions that often do not respond well to conventional therapies, such as *type III (spastic)* achalasia and "end-stage" achalasia (markedly dilated, sigmoid esophagus), and in patients *who have failed prior endoscopic and surgical achalasia treatments*.

The role of POEM in the treatment of achalasia continues to evolve, although there is a consensus that POEM is the procedure of choice for the treatment of type III achalasia

POEM



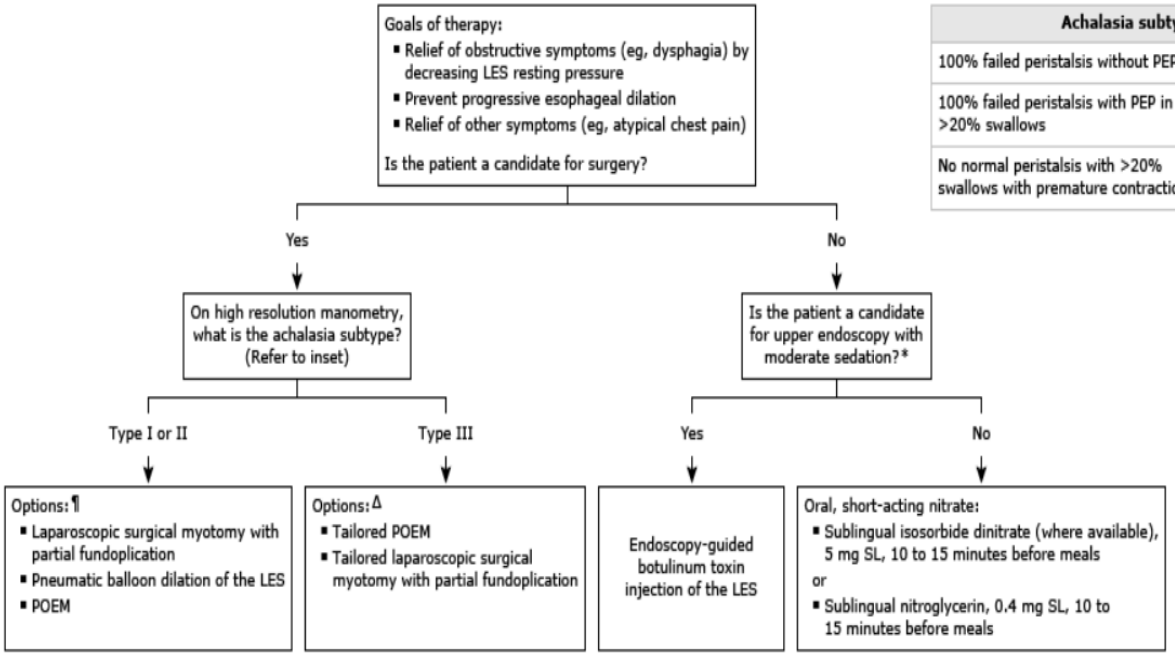
Peroral Endoscopic Myotomy as a Novel Treatment for Achalasia: Patient | CEG

POEM

Per-oral endoscopic myotomy for achalasia: An American

Visit

Approach to initial management of adults with achalasia



Achalasia subtypes	
100% failed peristalsis without PEP	Type I achalasia
100% failed peristalsis with PEP in >20% swallows	Type II achalasia
No normal peristalsis with >20% swallows with premature contractions	Type III achalasia

Options for therapy include mechanical disruption of the LES (eg, myotomy or pneumatic dilation) or pharmacologic reduction in LES pressure (eg, botulinum toxin injection or oral nitrate). Mechanical disruption of the LES has been associated with more durable results. In addition to efficacy, the choice of intervention is informed by patient characteristics, patient preferences, institutional expertise, and safety. Refer to UpToDate content on management of achalasia for additional details.

PEP: panesophageal pressurization; LES: lower esophageal sphincter; ASA: American Society of Anesthesiologists; GERD: gastroesophageal reflux disease; POEM: peroral endoscopic myotomy; SL: sublingual.

* The preprocedure evaluation includes a medical history, anesthesia-directed physical examination, and assessing ASA physical status. Refer to UpToDate content on anesthesia for gastrointestinal endoscopy for details.

¶ Higher rates of GERD are reported after POEM than either laparoscopic myotomy or pneumatic dilation. This may be a consideration for younger

Achalasia

Recommendations

Either graded pneumatic dilation (PD) or laparoscopic surgical myotomy with a partial fundoplication are recommended as initial therapy for the treatment of achalasia in those fit and willing to undergo surgery

2. PD and surgical myotomy should be performed in high-volume centers of excellence

3. The choice of initial therapy should be guided by patients ' age, gender, preference, and local institutional expertise

4. Botulinum toxin therapy is recommended in patients who are not good candidates for more definitive therapy with PD or surgical myotomy

5. Pharmacologic therapy for achalasia is recommended for patients who are unwilling or cannot undergo definitive treatment with either PD or surgical myotomy and have failed botulinum toxin therapy

Spastic Motility Disorders of the Esophagus

Spastic Motility Disorders of the Esophagus

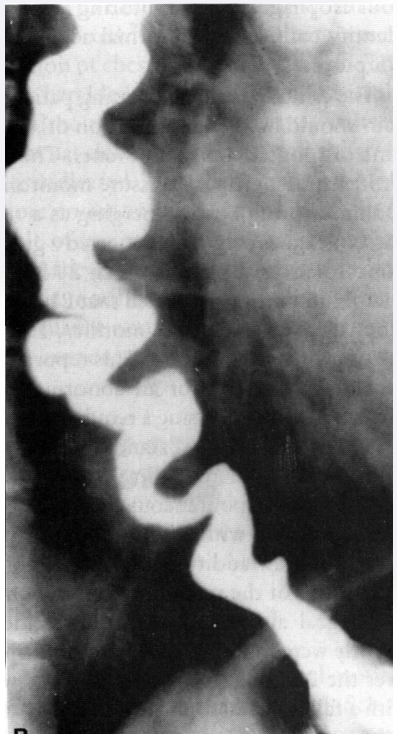
- Diffuse Esophageal Spasm
- Nutcracker Esophagus
- Hypertensive LES
- Nonspecific Esophageal Dysmotility

Clinical Presentation

- Dysphagia to solids and liquids
  - intermittent and non-progressive
  - present in 30-60%, more prevalent in DES (in most studies)
- Chest Pain
  - swallowing is not necessarily impaired
  - can mimic cardiac chest pain
- Pyrosis (20%)
- IBS symptoms (>50%)
- Symptoms and Manometry correlate poorly

Diffuse Esophageal Spasm

- frequent non-peristaltic contractions
  - simultaneous onset (or too rapid propagation) of contractions in two or more recording leads



Diffuse Esophageal Spasm (Des)

- DES is manifested by :
- episodes of dysphagia and chest pain attributable to abnormal esophageal contractions with normal deglutitive LES relaxation

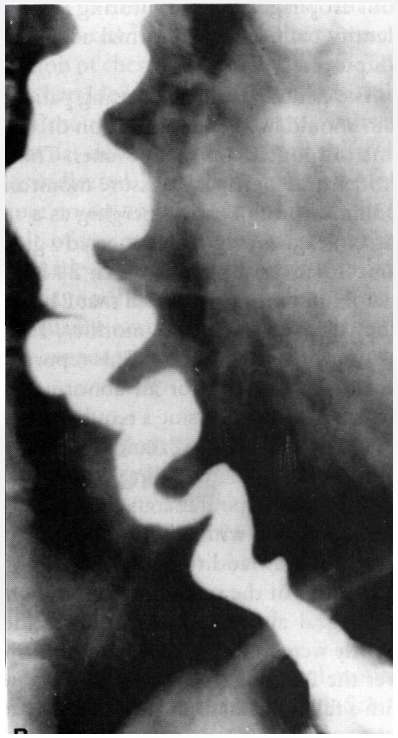
Diffuse Esophageal Spasm (Des)

➢ The pain mimic angina pectoris.

- Features suggesting an esophageal, as opposed to a cardiac, etiology of chest pain include :
- (1) prolonged, nonexertional pain;
- (2) pain that interrupts sleep;
- (3) meal-related pain;
- (4) relief with antacids; and
- (5) additional accompanying esophageal symptoms such as heartburn, dysphagia, or regurgitation. However, even these characteristics occasionally exhibit overlap with cardiac pain..

Diffuse Esophageal Spasm (Des)

- Radiographically, a "corkscrew esophagus," "rosary bead esophagus," pseudodiverticula, or curling can be indicative of DES, but these are also found with spastic achalasia



Diffuse Esophageal Spasm (Des)

- Manometrically,
- uncoordinated ("spastic") activity in the distal esophagus, spontaneous and repetitive contractions, or high amplitude and prolonged contractions.

- For patients with manometric criteria consistent with DES:

we *exclude secondary causes* of esophageal dysmotility including :

- ➢ opioid-induced dysmotility,
- ➢ eosinophilic esophagitis,
- ➢ impaired esophagogastric junction opening (esophagogastric junction outflow obstruction or mechanical obstruction).

Diffuse Esophageal Spasm (Des)

Treatment

- The goal of initial therapy is to control (GERD) symptoms.
- For patients with DES and who have GERD symptoms, we suggest initiating (PPI) therapy. The PPI is typically started with a standard dose taken twice daily .

➢ For patients who have either no GERD or well-controlled GERD, we suggest a trial of pharmacologic therapy to relax smooth muscle .

- Treatment options include an antispasmodic agent (eg, hyoscyamine 0.25 mg three times daily as needed), a nitrate (eg, isosorbide dinitrate 5 to 10 mg twice to three times daily as needed), a calcium channel blocker (eg, nifedipine, extended release, 30 mg daily), or phosphodiesterase inhibitor (sildenafil 25 to 50 mg daily).

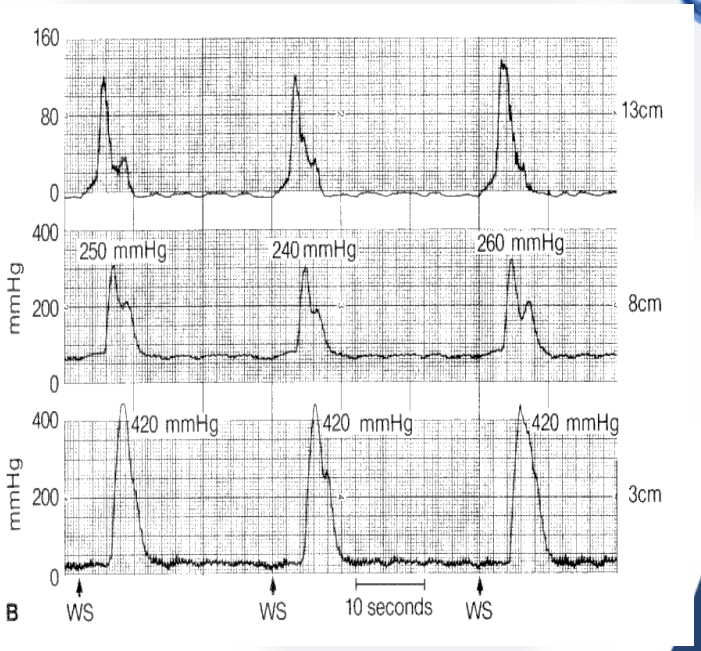
Diffuse Esophageal Spasm (Des)

Treatment

- If therapy to relax esophageal smooth muscle is not effective or is not well tolerated, we begin a low-dose TCA (eg, nortriptyline 10 to 25 mg at bedtime), especially for patients *with pain-predominant* symptoms.
- For patients who do not have symptom relief or who do not tolerate pharmacologic therapy, endoscopy-guided botulinum toxin injection into the distal esophagus is a subsequent therapeutic option.
- For selected patients (eg, those with complications of esophageal dysmotility), myotomy (ie, POEM or surgical myotomy) is an alternative intervention because data suggest that myotomy was associated with improvement in symptoms and manometric features.

Nutcracker Esophagus

- high pressure peristaltic contractions
- avg pressure in 10 wet swallows is >180 mm Hg
- 33% have long duration contractions (>6 sec)
- may inter-convert with DES



Hypertensive LES

- high LES pressure
  - >45 mm Hg
- normal peristalsis
- often overlaps with other motility disorders

Nonspecific Esophageal Dysmotility

- abnormal motility pattern
- fits in no other category
  - non-peristalsis in 20-30% of wet swallows
  - low pressure waves (<30 mm Hg)
  - prolonged contractions

Diagnosis of Spastic Motility Disorders of the Esophagus

- Manometry
- Barium Esophagram
- Endoscopy
- PH monitoring

Spastic Motility Disorders of the Esophagus

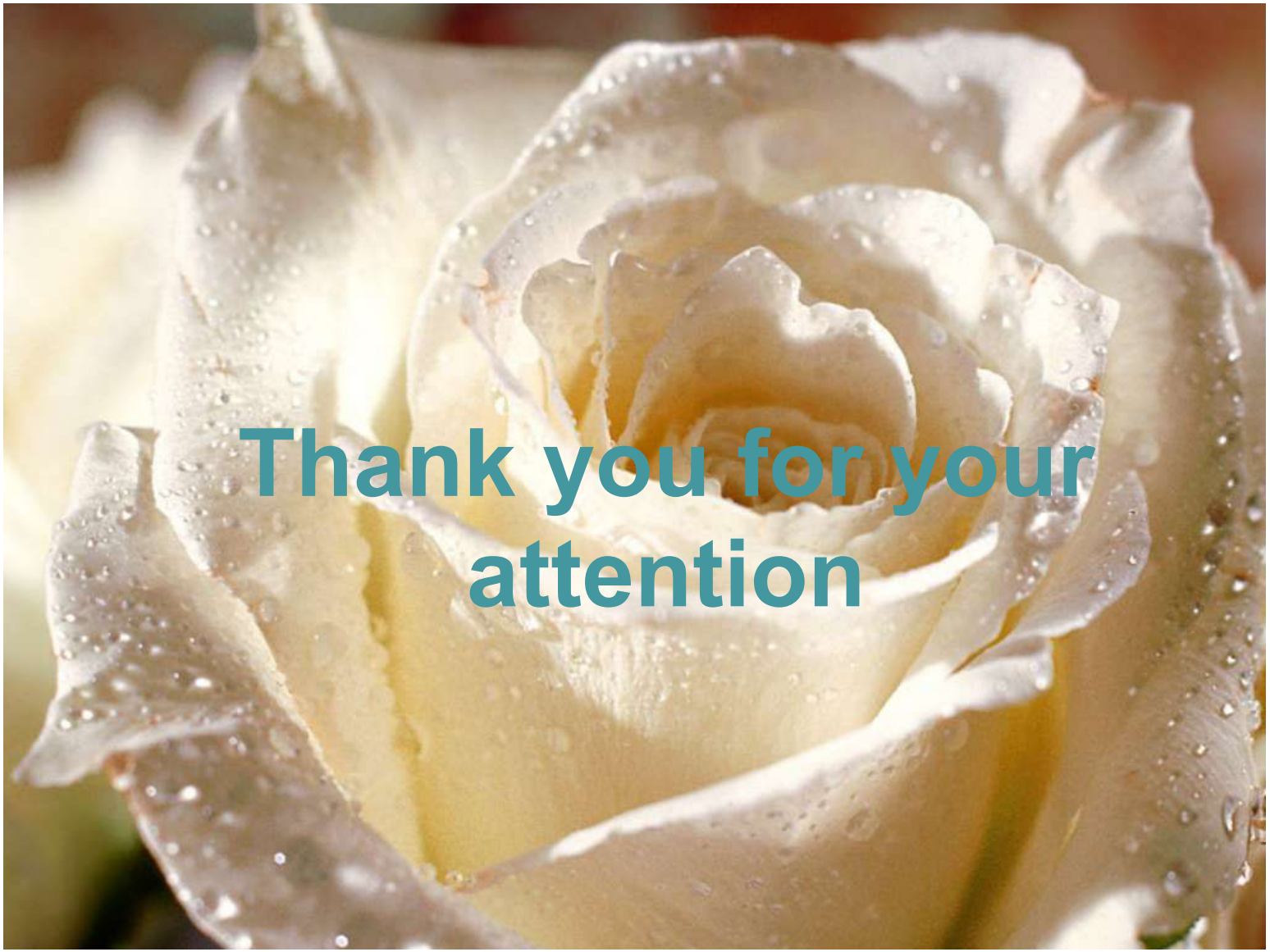
- treatment
  - reassurance
  - nitrates, anticholinergics, hydralazine - *all unproven*
  - calcium channel blockers - *too few data with negative controlled studies in chest pain*
  - psychotropic drugs – *trazodone, imipramine and setraline effective in controlled studies*
  - dilation - *anecdotal reports, probable placebo effect*

Hypomotilty Disorders

- **primary (idiopathic)**
  - aging produces gradual decrease in contraction strength
  - reflux patients have varying degrees of hypomotility
    - more common in patients with atypical reflux symptoms
    - usually persists after reflux therapy
  - defined as
    - low contraction wave pressures (<30 mm Hg)
    - incomplete peristalsis in 30% or > of wet swallows

Hypomotilty Disorders

- **secondary**
  - scleroderma
    - in >75% of patients
    - progressive, resulting in aperistalsis in smooth-muscle region
    - incompetent LES with reflux
  - other “connective tissue diseases”
    - CREST
    - polymyositis & dermatomyositis
  - diabetes
    - 60% with neuropathy have abnormal motility on testing
  - other
    - hypothyroidism, alcoholism, amyloidosis


Thank you for your attention