Esophageal Diseases

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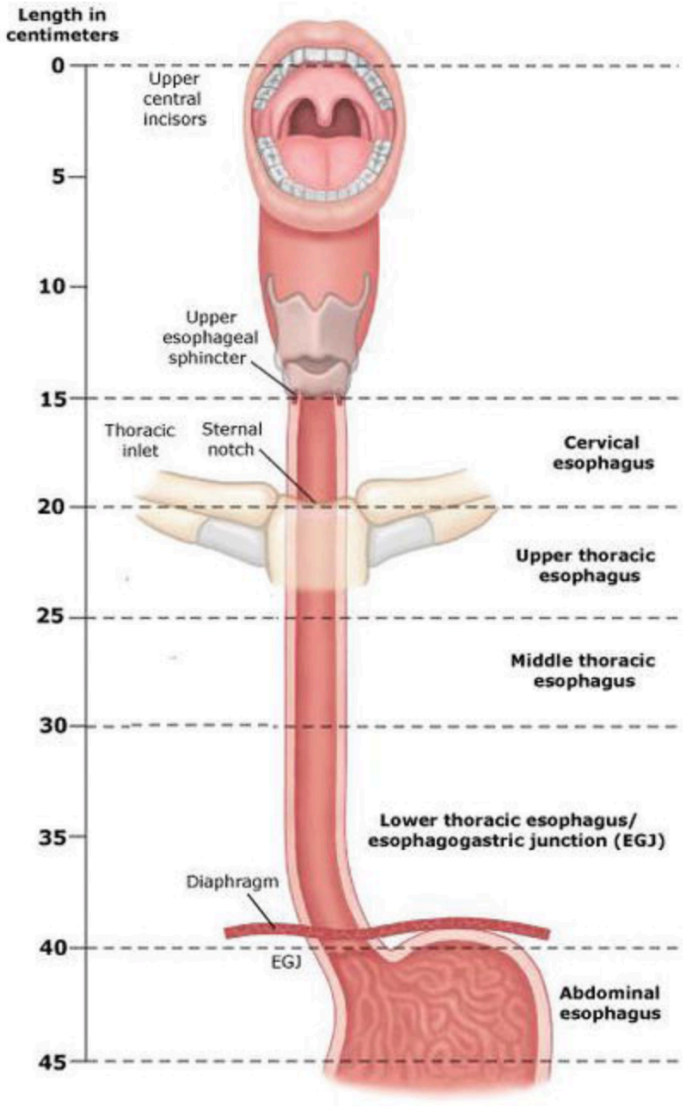
Surgical Anatomy

- Fibromuscular tube (25cm) long.
- Posterior mediastinum.
- 4cm of esophagus below the diaphragm.
- Lined by squamous epithelium.



Esophageal Diseases

- ***Benign:***
  - Esophageal motility disorder
  - Esophageal diverticula
  - Benign esophageal tumors
  - GERD and Hiatus Hernia
  - Esophageal perforation
  - Caustic Injury
  - Esophageal cancer

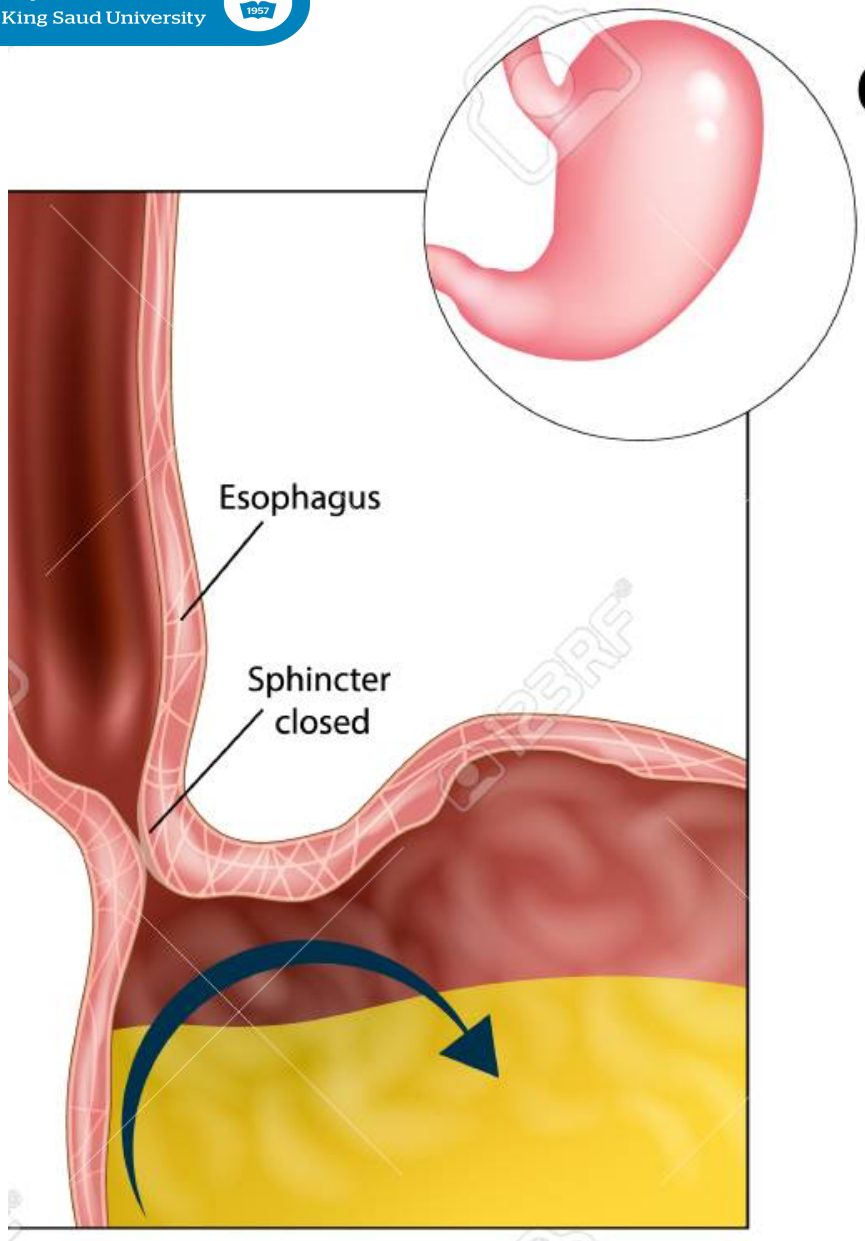
- ***Malignant*** ➔ Esophageal cancer

GASTROESOPHAGEAL REFLUX DISEASE (GERD)

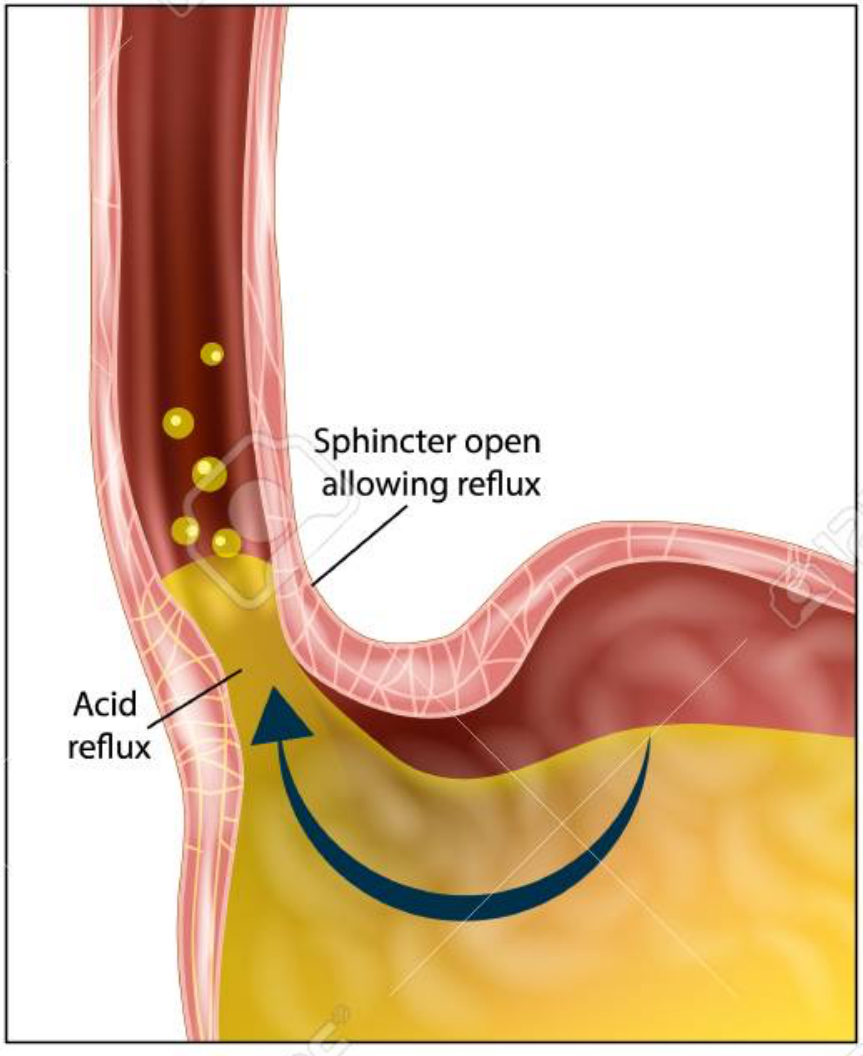
LES has primary role of preventing reflux of the gastric contents into the esophagus.

When LES has too low pressure to prevent the reflux of gastric contents from entering the esophagus → GERD.

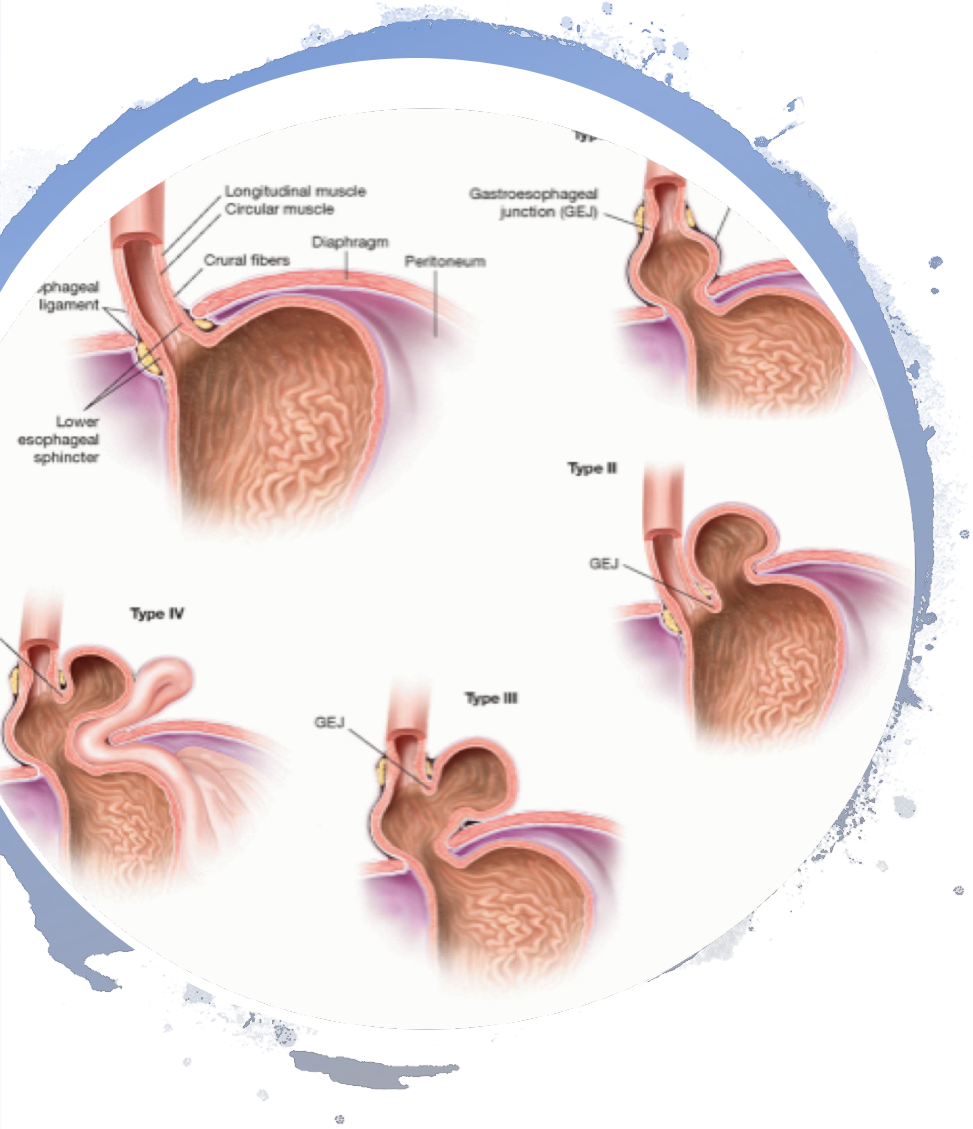
GASTROESOPHAGEAL REFLUX DISEASE



Healthy stomach



Gerd stomach



GASTROESOPHAGEAL REFLUX DISEASE

- **GERD is often associated with a hiatal hernia**

GASTROESOPHAGEAL REFLUX DISEASE

- **Definition :**
- **Symptoms OR mucosal damage produced by the abnormal reflux of gastric contents into the esophagus**
- **Often chronic and relapsing**
- **May see complications of GERD in patients who lack typical symptoms**

GASTROESOPHAGEAL REFLUX DISEASE

- ***Epidemiology*** :
  - **About 44% of the US adult population have heartburn at least once a month**
  - **14% of Americans have symptoms weekly**
  - **7% have symptoms daily**

Clinical Presentations of GERD

- Classic GERD
- Extra esophageal/Atypical GERD
- Complicated GERD

Clinical Presentations of GERD

- ***Classic GERD:***
- Substernal burning and or regurgitation
- Postprandial pain
- Aggravated by change of position
- Prompt relief by antacid

Extra-esophageal Manifestations of GERD

Pulmonary

- Asthma
- Aspiration pneumonia
- Chronic bronchitis
- Pulmonary fibrosis

Other

- Chest pain
- Dental erosion

ENT

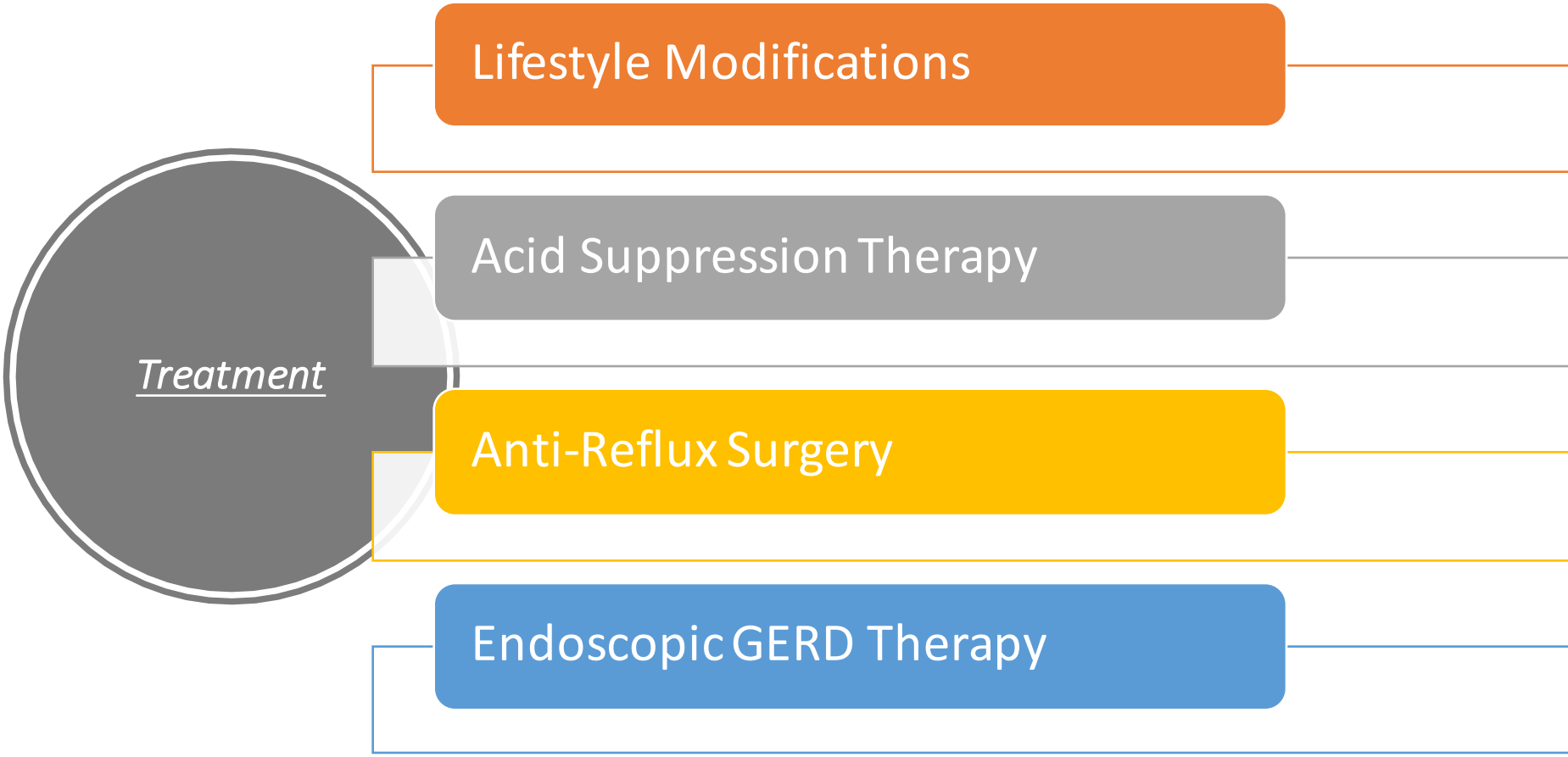
- Hoarseness
- Laryngitis
- Pharyngitis
- Chronic cough
- Globus sensation
- Dysphonia
- Sinusitis
- Subglottic stenosis
- Laryngeal cancer

Diagnostic Tests for GERD

- Barium swallow
- Endoscopy
- Ambulatory pH monitoring
- Esophageal manometry

Clinical Presentations of GERD

- Symptoms of Complicated GERD :
  - Dysphagia
    - Difficulty swallowing: food sticks or hangs up
  - Odynophagia
    - Retrosternal pain with swallowing
  - Bleeding


Treatment
Lifestyle Modifications
Acid Suppression Therapy
Anti-Reflux Surgery
Endoscopic GERD Therapy

Treatment

- Lifestyle Modifications
  - Elevate head of bed 4-6 inches
  - Avoid eating within 2-3 hours of bedtime
  - Lose weight if overweight
  - Stop smoking
  - Modify diet
    - Eat more frequent but smaller meals
    - Avoid fatty/fried food, peppermint, chocolate, alcohol, carbonated beverages, coffee and tea
  - OTC medications prn

Acid Suppression Therapy for GERD

H_2-Receptor Antagonists (H_2RAs)

- Cimetidine (Tagamet®)
- Ranitidine (Zantac®)
- Famotidine (Pepcid®)
- Nizatidine (Axid®)

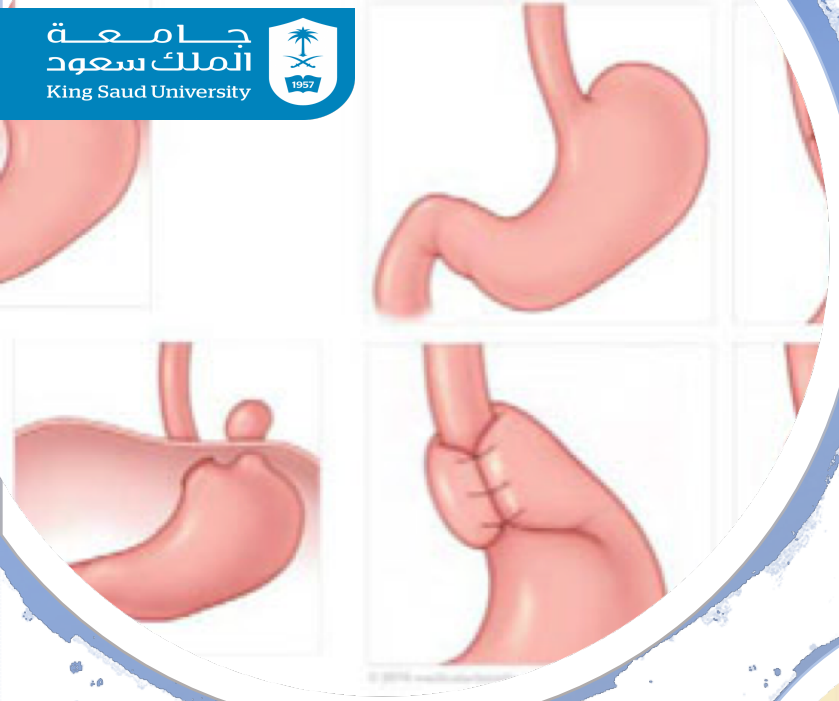
Proton Pump Inhibitors (PPIs)

- Omeprazole (Prilosec®)
- Lansoprazole (Prevacid®)
- Rabeprazole (Aciphex®)
- Pantoprazole (Protonix®)
- Esomeprazole (Nexium®)

- ***Indication for Surgery:***
  - Failed medical therapy.
  - Patient desire.
  - Complications of GERD (e.g. Barrett's esophagus; grade III or IV esophagitis).
  - Medical complications attributable to a large hiatal hernia. (e.g. bleeding, dysphagia).
  - ”Atypical" symptoms and reflux documented on 24 hour pH monitoring

Endoscopic GERD Therapy

- Stretta procedure radiofrequency heating of GE junction
- Endoscopic plication TIF
- Enteryx



Surgical treatment

- Fundoplication
- LINX®
- EndoStim

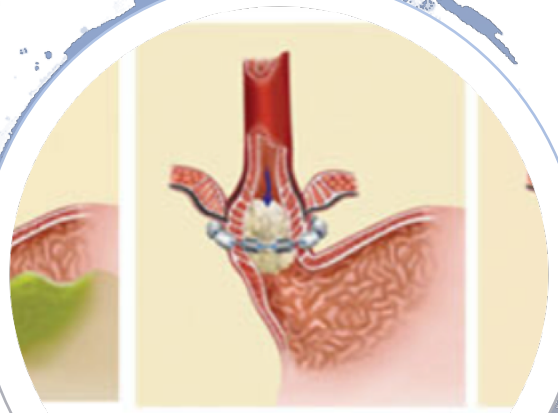
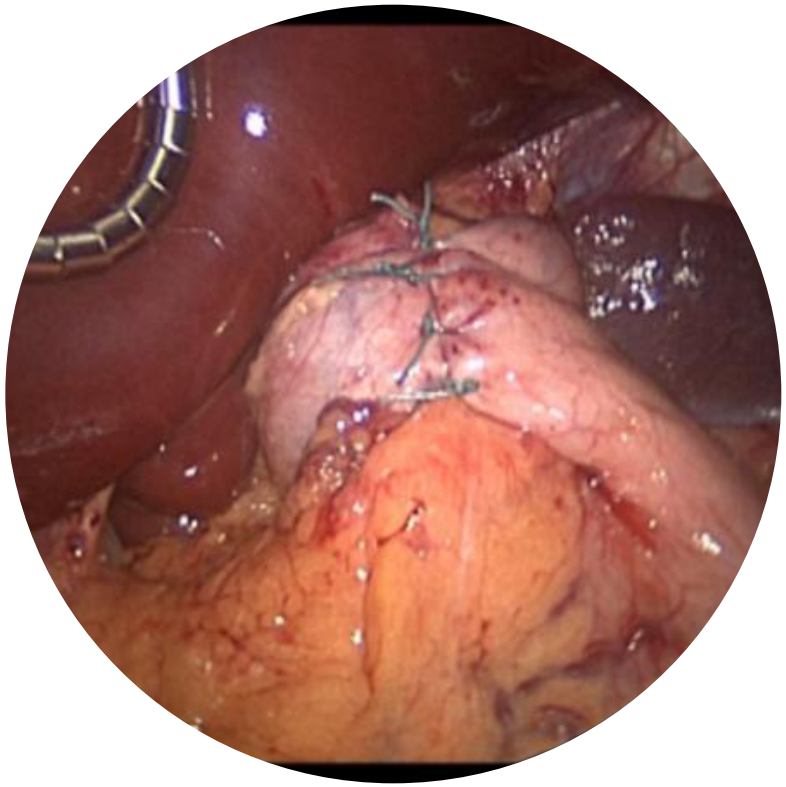
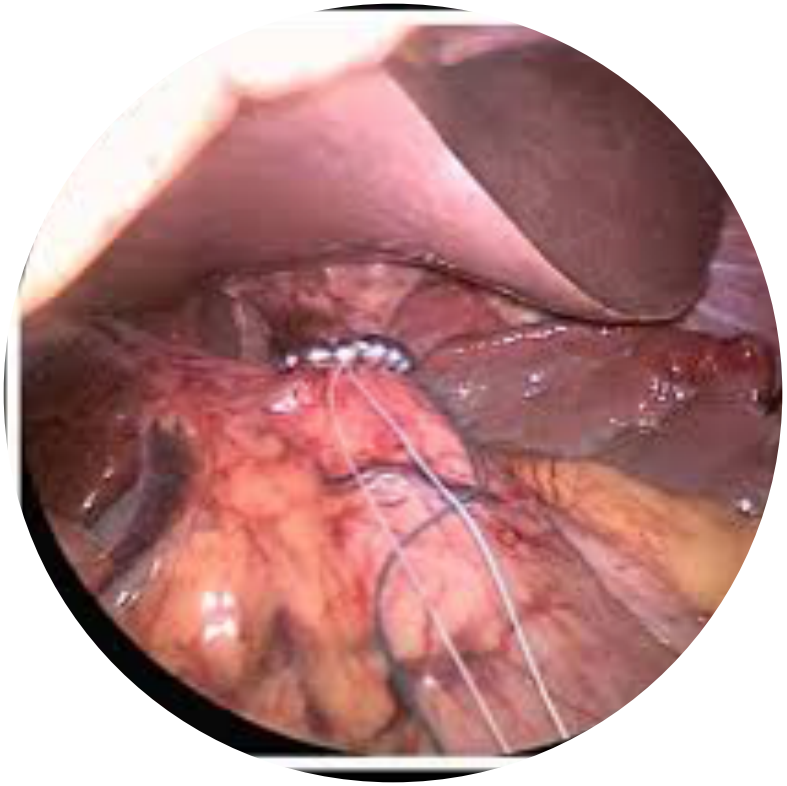


Figure 2. The LINX system is designed to expand to allow for normal swallowing.





Achalasia

- An uncommon disease of esophageal motility disorder.
- It is characterized by degeneration of the myenteric neurons that innervate LES and esophageal body

❖ ***Pathogenesis:***

Primary vs. secondary

Primary:

autoimmune ?

Viral ?

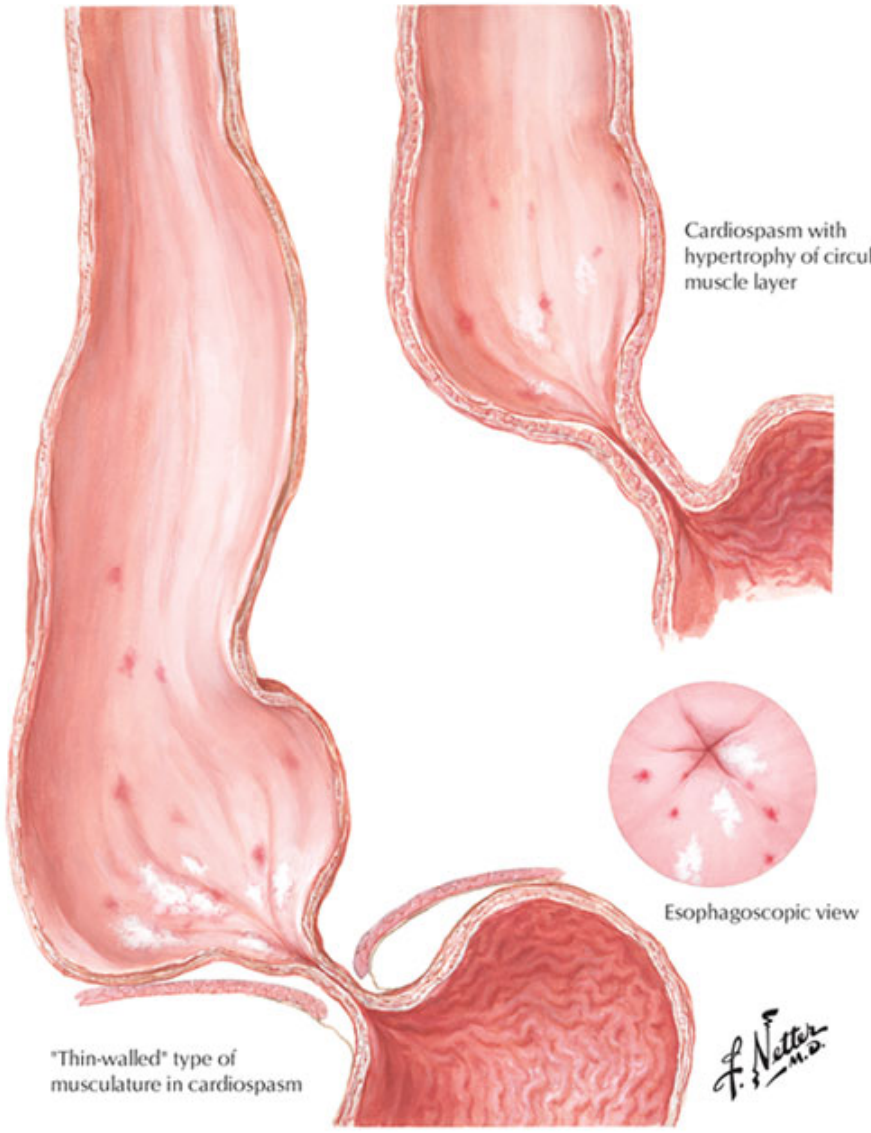
Gentics ?

Secondary:

- **Chagas' disease is a parasitic infection caused by *Trypanosoma cruzi* which can cause *secondary achalasia***

Pseudoachalasia

- **The most concerning secondary etiology is cancer ➔which can present as achalasia through mechanical obstruction of the GEJ.**



Clinical features

2 peaks. Age of 20s and 60s

Equal male-to-female distribution.

-Dysphagia:

Most common presenting symptom.

Both liquids and solids

Clinical features

-Regurgitation:

second most common symptom

occurring in 60% of patients.

Nocturnal regurgitation lead to night time cough and aspiration.

Clinical features

- Chest pain : 20% to 60% of patients.

- Heartburn : 30% of Achalasia patients).

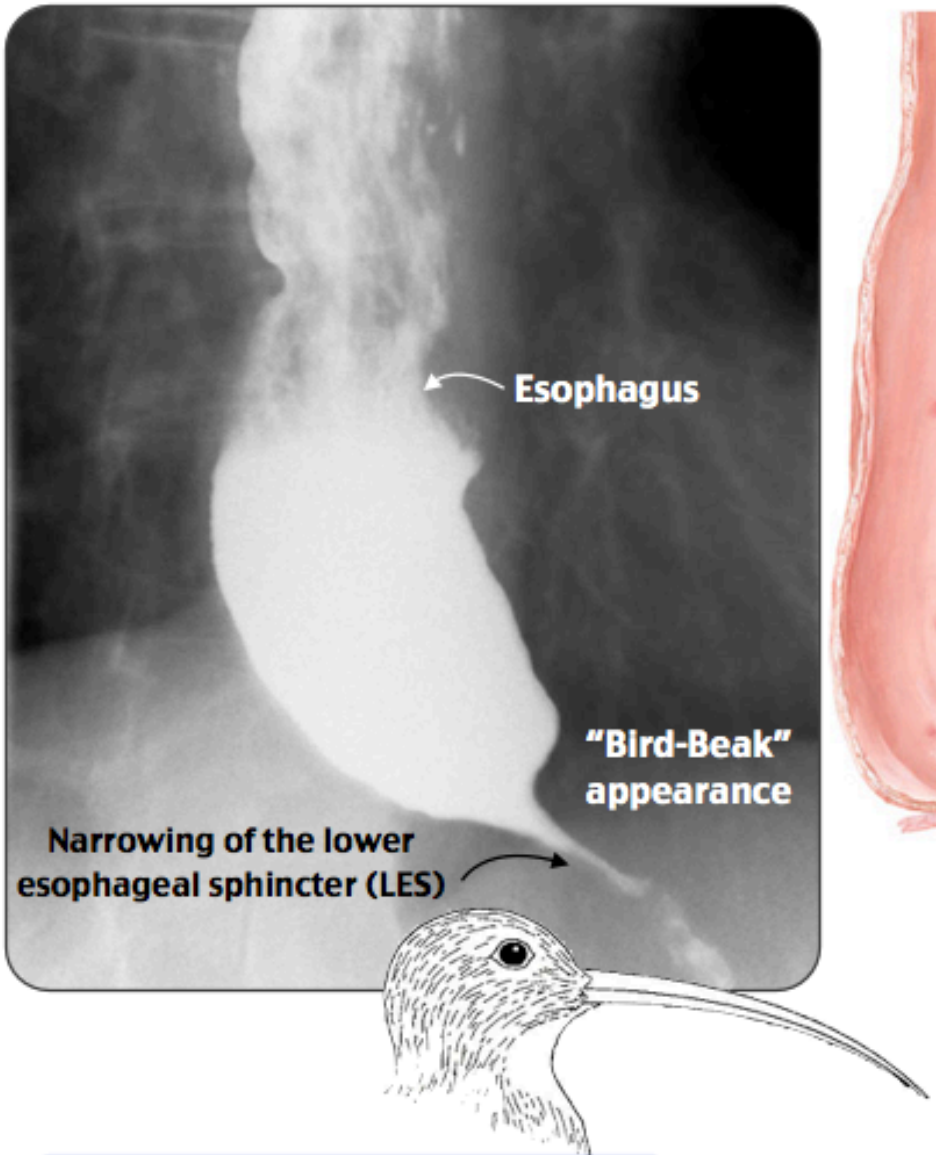
may be related to direct irritation of the esophageal lining by retained content , or acidic by-products of bacterial metabolism of retained food.

Diagnosis

CXR may show air-fluid level.

Barium swallow quite dilated, and an air-fluid level.

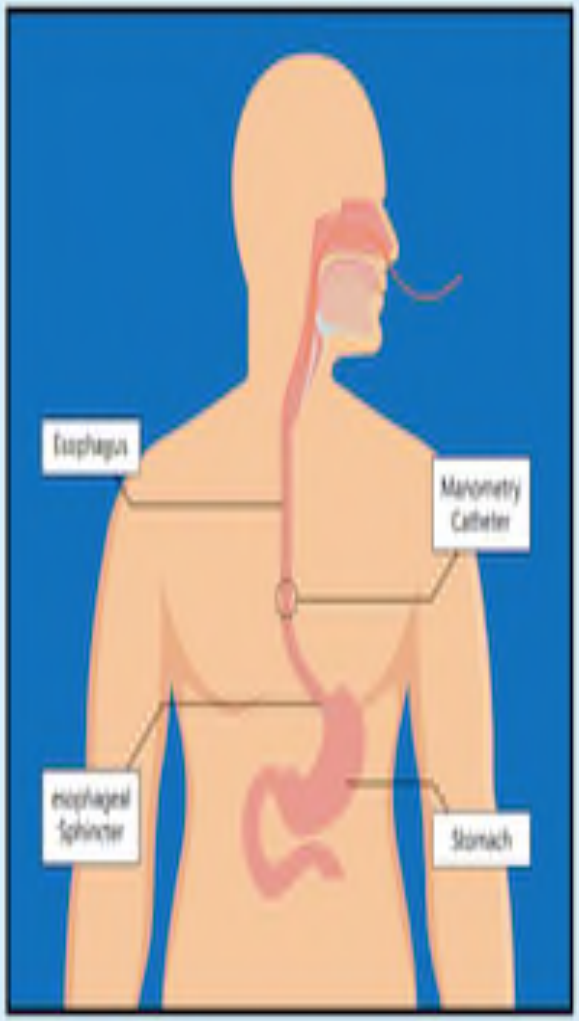
The classic finding is a gradual tapering at the end of the Esophagus, similar to a bird's beak.

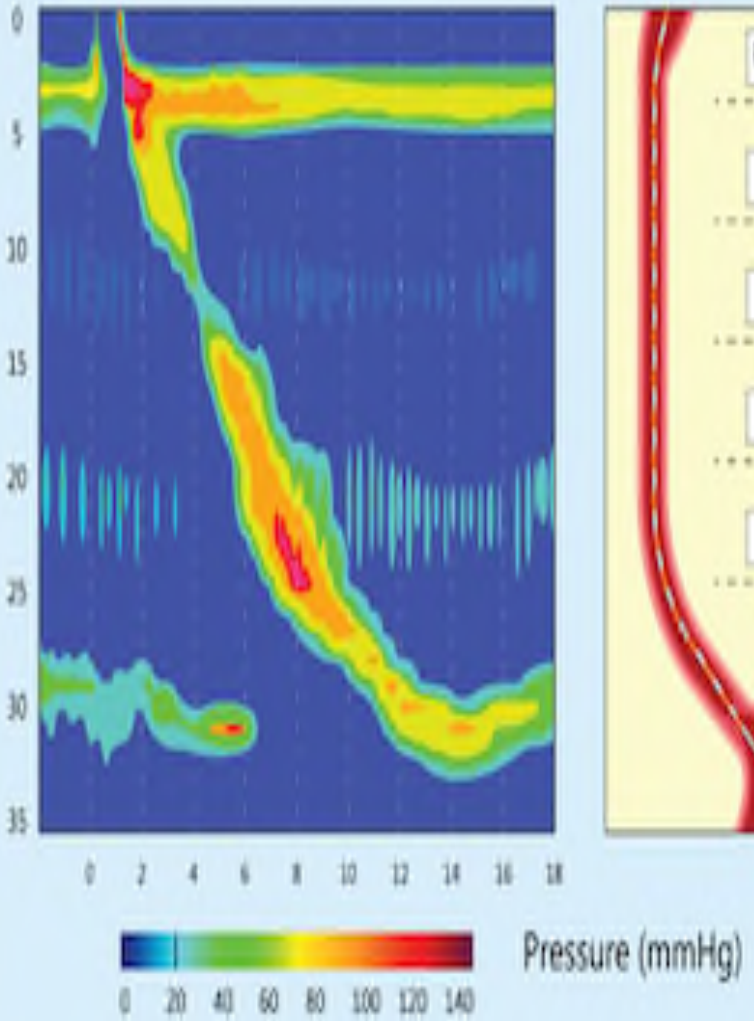

Esophagus
"Bird-Beak" appearance
Narrowing of the lower esophageal sphincter (LES)

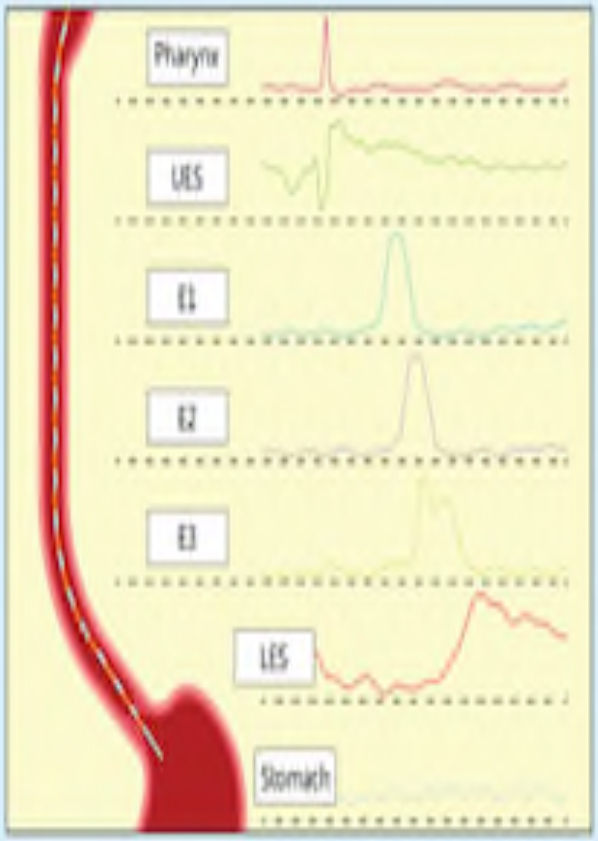
Upper endoscopy:

- Role out other causes of dysphagia.
- Dilated esophagus with retained food particles.

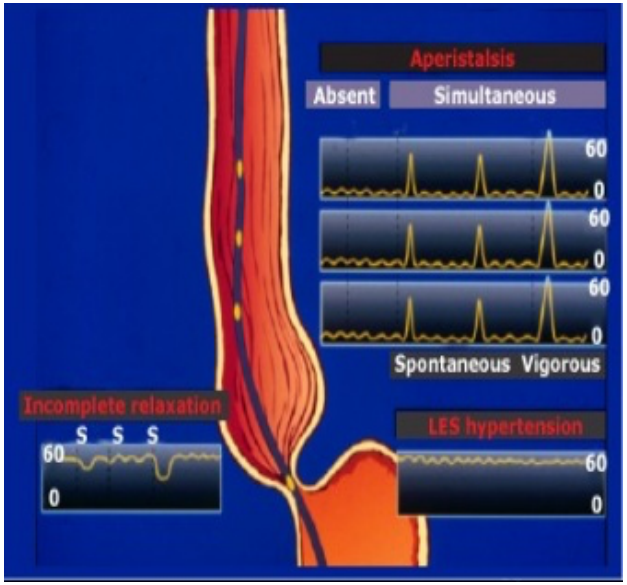
Esophageal Manometry







Esophageal manometry



- **Aperistalsis of the esophageal body.**
- **Incomplete or absent LES relaxation with swallowing.**
- **Hypertensive LES.**

Diagnosis

- **Manometric variants of achalasia exist**
  - **The best known is vigorous achalasia**
  - **defined by the presence of normal to high amplitude esophageal body contractions in the presence of a non-relaxing LES**

- **Manometric variants of achalasia exist**
  - **vigorous achalasia may represent an early stage of achalasia**

Treatment

Goal of treatment is Symptom relief.

Treatment options:

- Medical therapy
- Botulinum toxin injection
- Pneumatic dilation
- Surgical myotomy

Pharmacological Therapy

- **Decrease the LES pressure by causing smooth muscle relaxation.**

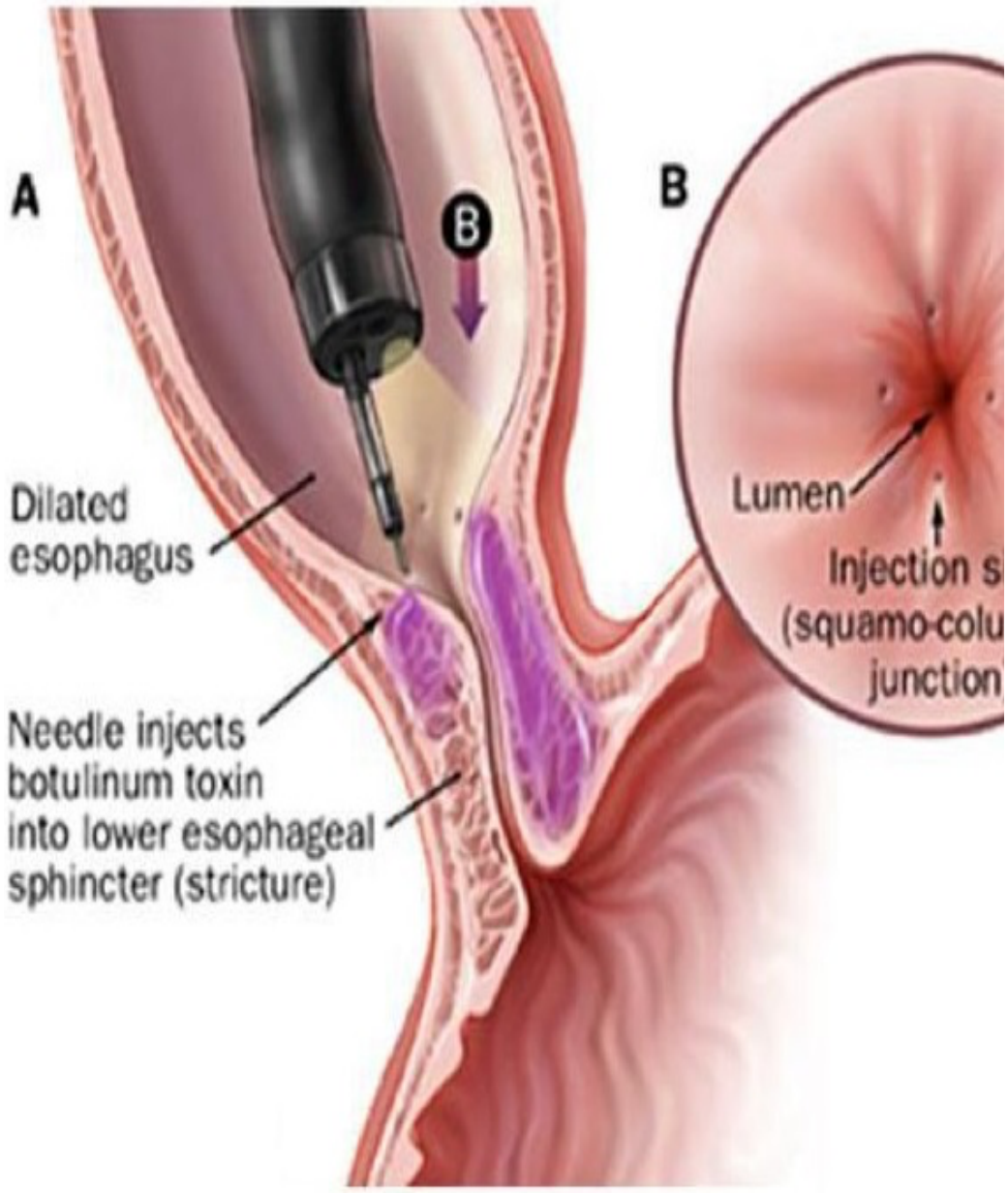
- **Inconvenient as they are :**
- **Partially effective.**
- **frequently associated with side effects.**

- **It is reserved for patients who are awaiting or unable to tolerate more invasive treatment modalities**

- ***Nitrates*** were first recognized as an effective treatment of achalasia
    - their systemic vasodilatory effects and headaches limit their tolerability by patients

- ***Calcium channel blockers*** have a better side-effect profile when compared with nitrates
    - 30% of patients report adverse side effects including peripheral edema, hypotension, and headache.

Botulinum Toxin



Reserved for use in patients:

- **Not candidates for more invasive treatments.**

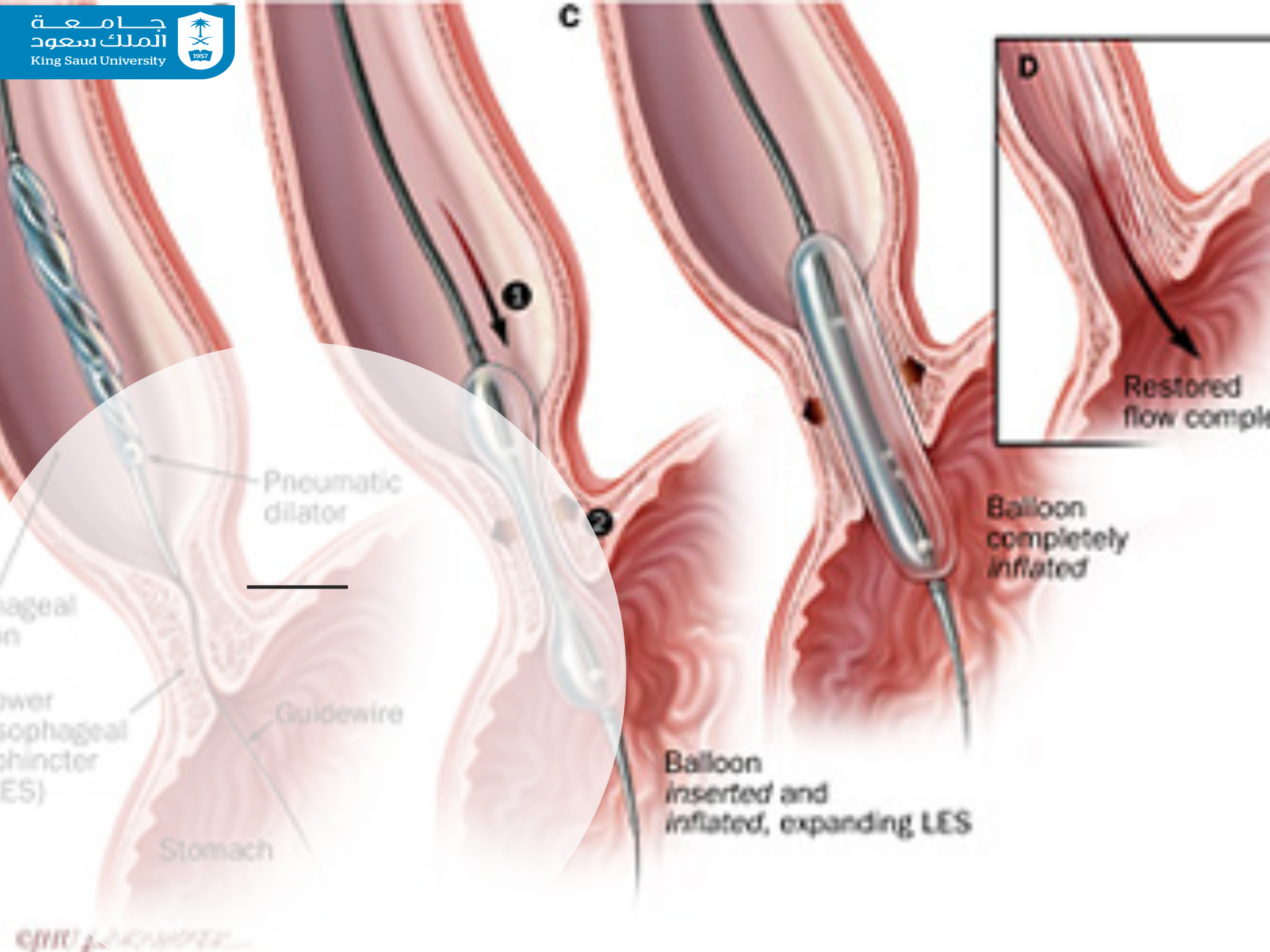
- **Refuse surgical intervention.**

<u>Efficacy up to 6-12 months</u>

Pneumatic Dilation

- A balloon is insufflated at the level of the gastroesophageal junction to rupture the muscle fibre.
- Success rate 70-80%.
- 50% will require more than 1 dilation session.

- Complications of pneumatic dilation exist
  - Gastroesophageal reflux 25-35%
  - Esophageal perforation 3 %

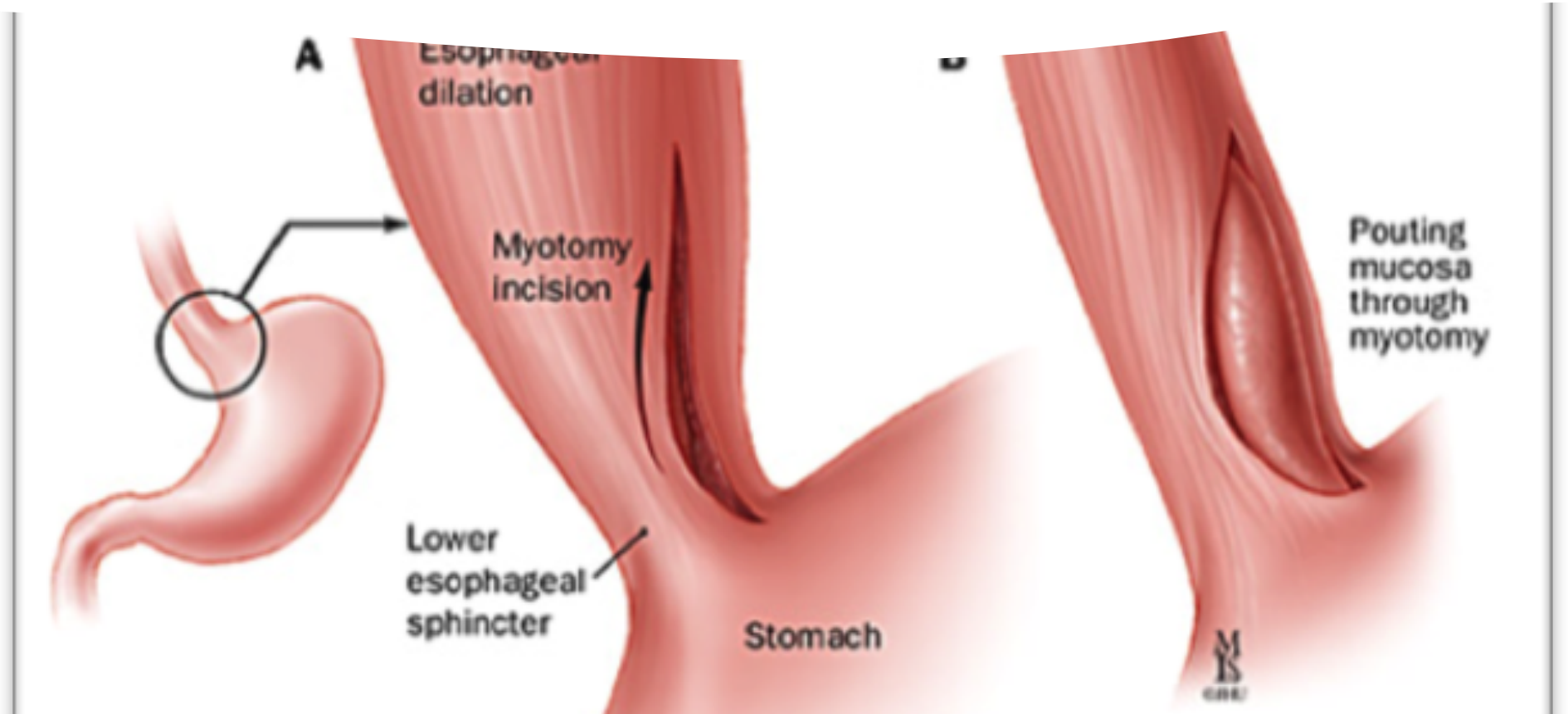


Surgical Therapy

- Has success rates in excess of 90%.
- Acid exposure is a known complication of surgical intervention for achalasia.

Surgical Therapy

- Laparoscopic Heller's myotomy demonstrated excellent results, with 98% of patients reporting symptomatic improvement.



Complications of Achalasia

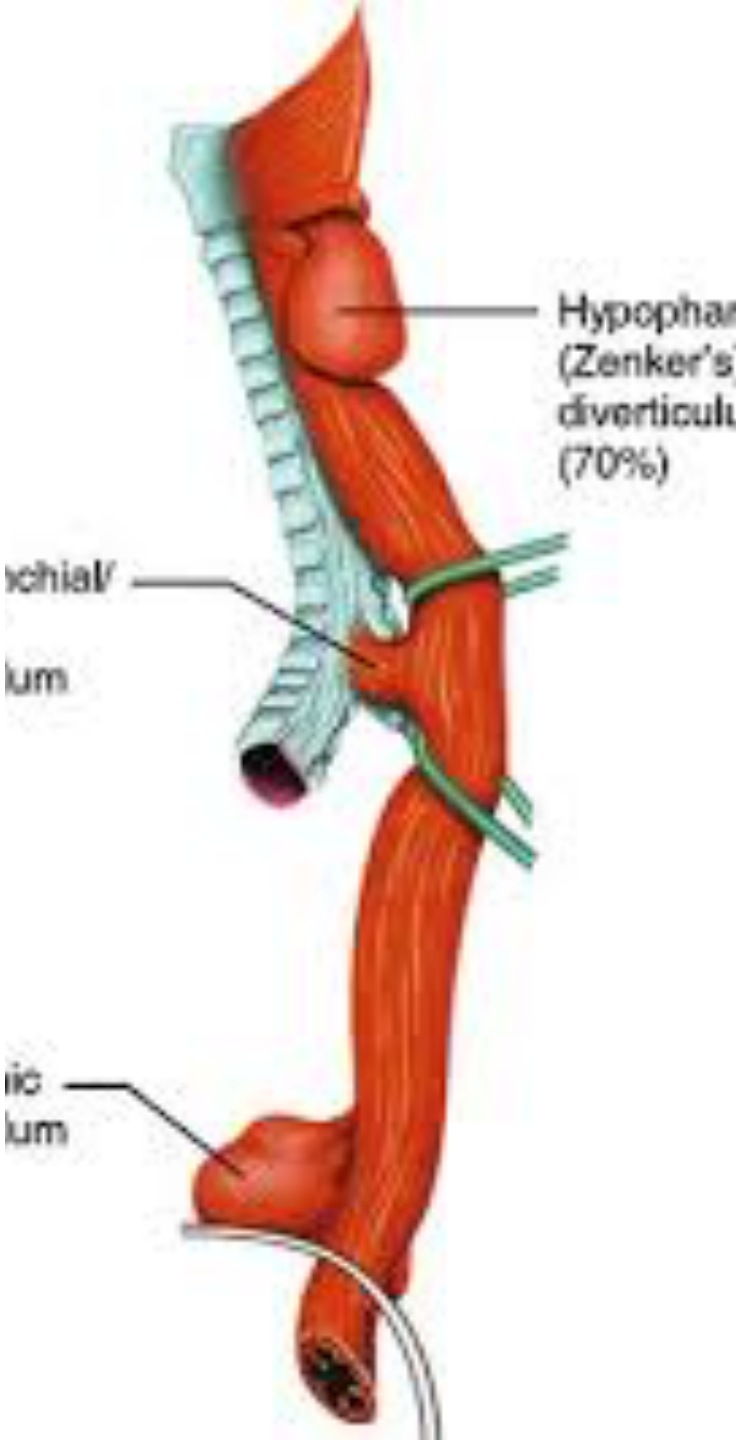
The primary complications of achalasia are related to the functional obstruction ➔ progressive malnutrition and aspiration.

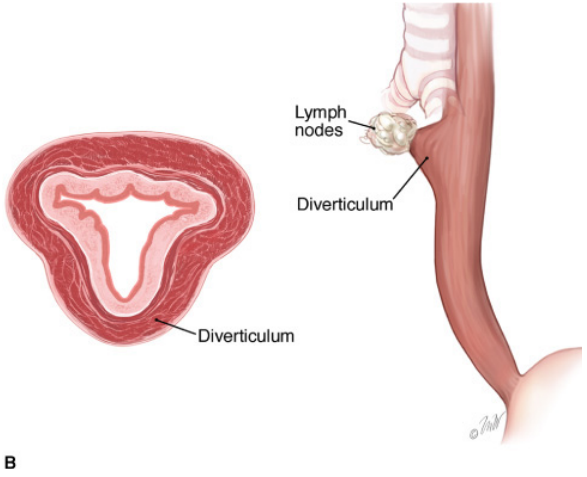
Uncommon but important secondary complications of achalasia include:

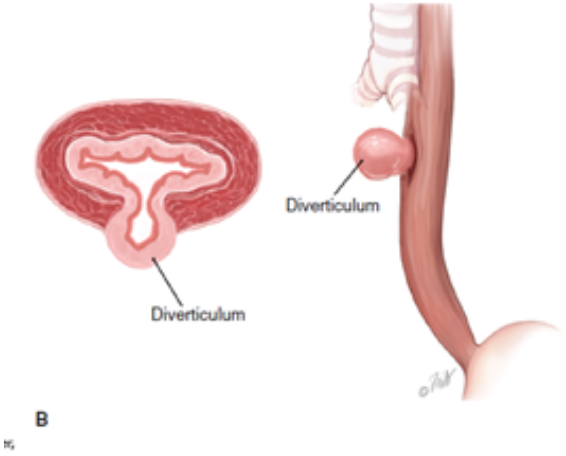
- Epiphrenic diverticula.
- Esophageal squamous cell carcinoma.

Esophageal Diverticula

- **The three most common sites of occurrence are:**
- **Pharyngoesophageal (Zenker's).**
- **Parabronchial (midesophageal).**
- **Epiphrenic.**







Esophageal Diverticula

- ***True diverticula*** **involve all layers of the esophageal wall, including mucosa, sub-mucosa, and muscularis.**

- ***False diverticulum*** **consists of mucosa and submucosa only.**

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Esophageal Diverticula

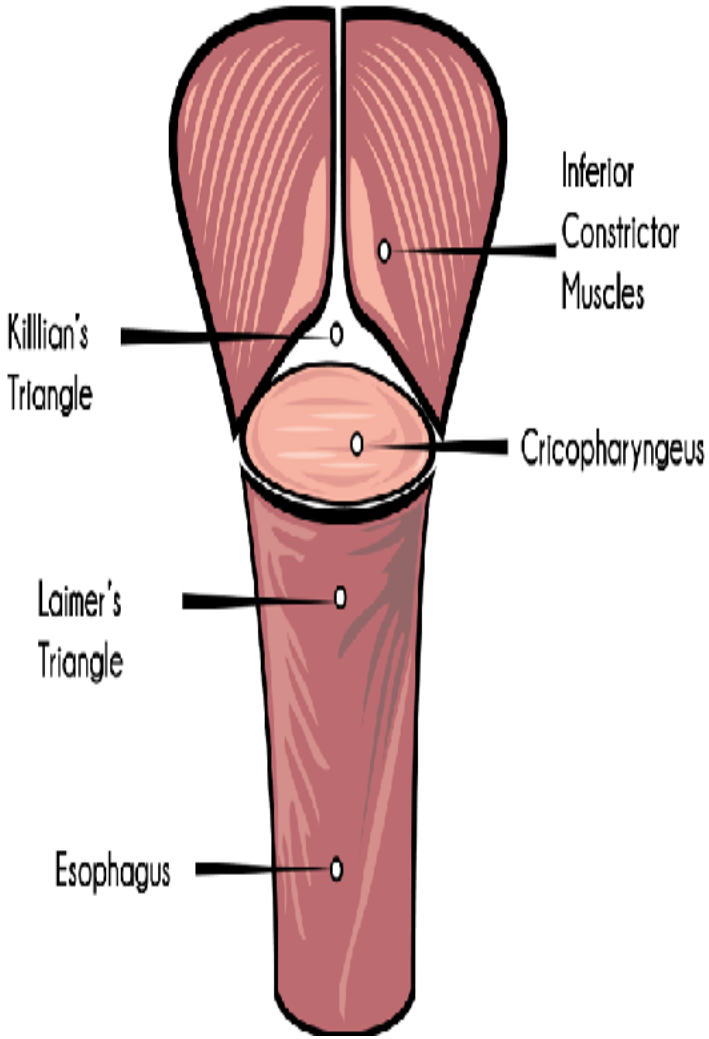
False (Pulsion) diverticula:

Zenker's diverticulum

Epiphrenic diverticulum

True (Traction) diverticula :

external inflammatory mediastinal lymph nodes adhering to the esophagus



Pharyngoesophageal (Zenker's) Diverticulum

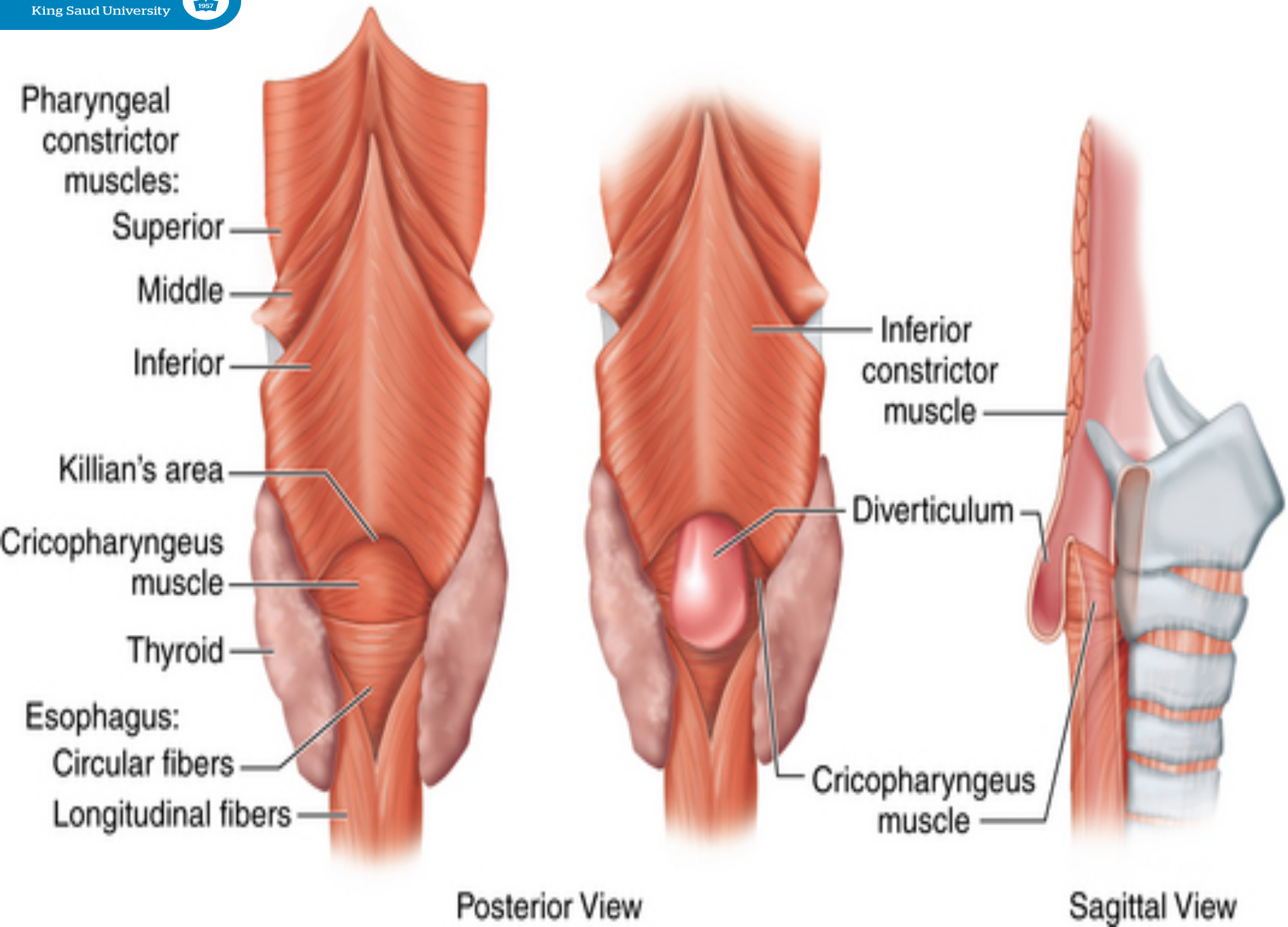
Is the most common esophageal diverticulum found today.

Disease of elderly (7th decade of life).

Herniation at Killian's triangle between:

- **Oblique fibers of the thyro-pharyngeus muscle**
- **Horizontal fibers of the crico-pharyngeus muscle.**

Zenker's Diverticulum Anatomy



Symptoms and Diagnosis

Commonly, patients complain of a sticking in the throat.

Nagging cough, excessive salivation, and intermittent dysphagia often are signs of progressive disease.

As the sac increases in size, regurgitation of foul-smelling, undigested material is common

Symptoms and Diagnosis

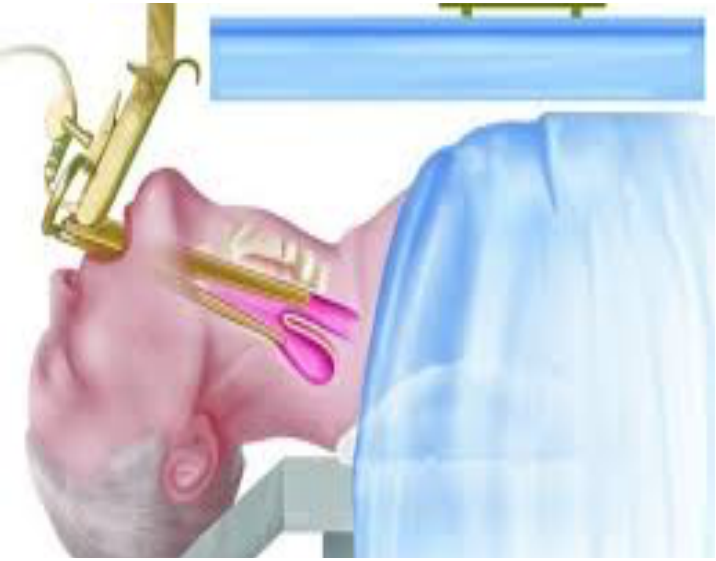
- **Halitosis, voice changes, retro-sternal pain, and respiratory infections are especially common in the elderly population**

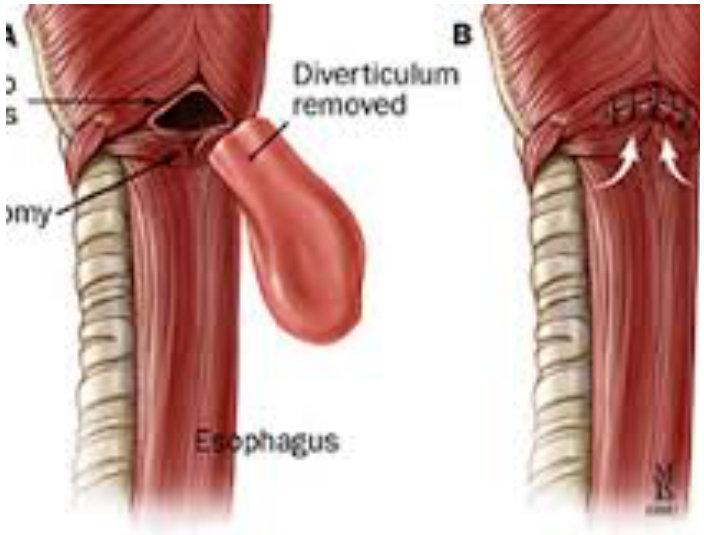
- ***The most serious complication*** **from an untreated Zenker's diverticulum is aspiration pneumonia or lung abscess**

Diagnosis

- **Diagnosis is made by *barium esophagram.***

- **Neither esophageal manometry nor endoscopy are needed to make a diagnosis of Zenker's diverticulum.**





Treatment

- Surgical or endoscopic repair of a is the gold standard of treatment.

Barrett's Esophagus

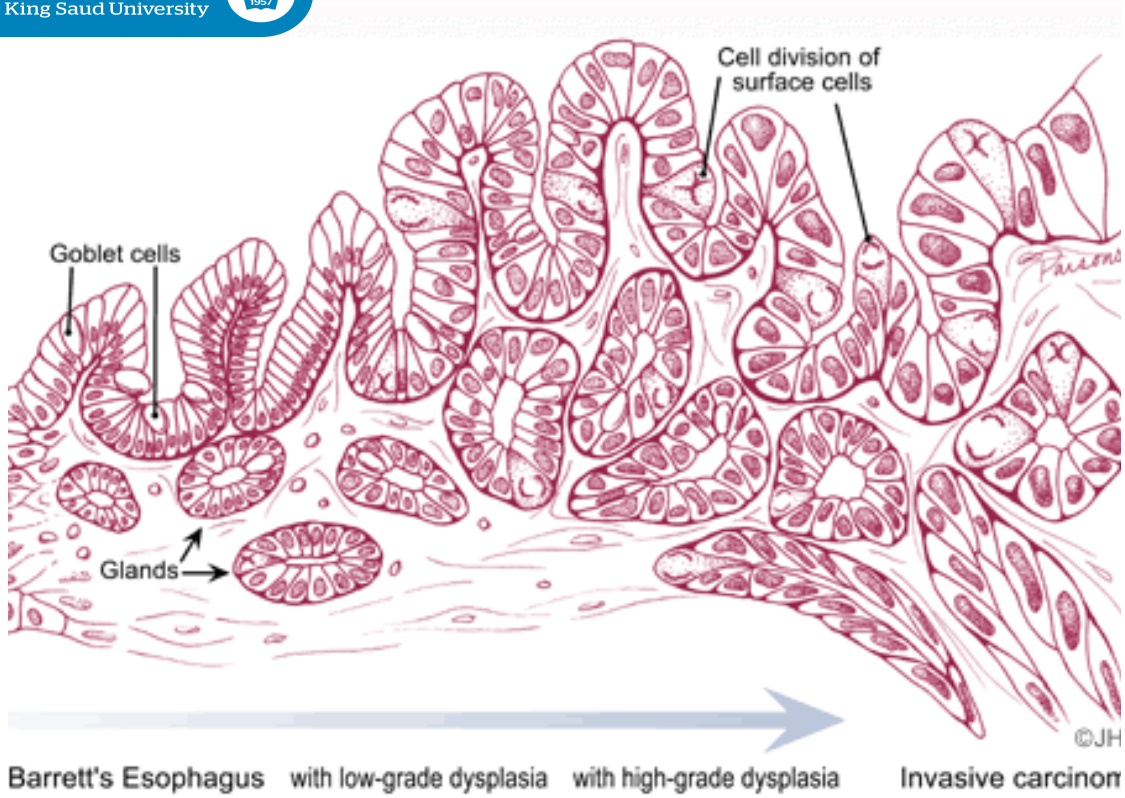
- Squamous epithelium that normally lines the distal esophagus replaced by columnar epithelium.
- Chronic gastro-esophageal reflux is the factor that both injures the squamous epithelium and promotes repair through columnar metaplasia

Barrett's Esophagus

- Although these metaplastic cells may be more resistant to injury from reflux, they also are more prone to malignancy.
- 10 % of patients with GERD develop Barrett's esophagus.

Clinical significance

- **the 40-fold increase in risk for developing esophageal carcinoma in patients with Barrett's esophagus**



Barrett's Esophagus

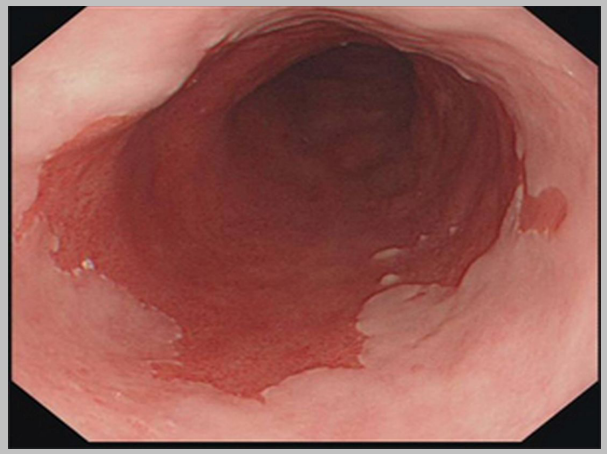
- With continued exposure to the reflux disease, metaplastic cells undergo cellular transformation to low- and high-grade dysplasia.

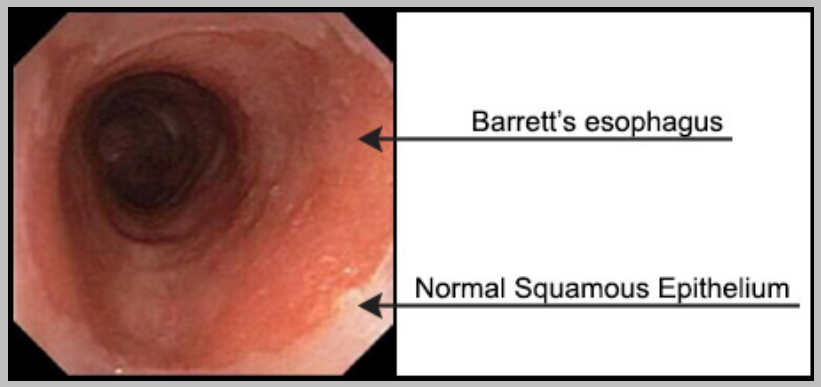
Symptoms and Diagnosis

- **Many patients harboring intestinal metaplasia in their distal esophagus are asymptomatic.**
- **Most patients present with symptoms of GERD.**

Diagnosis

- **The diagnosis of BE is made by endoscopy and pathology.**



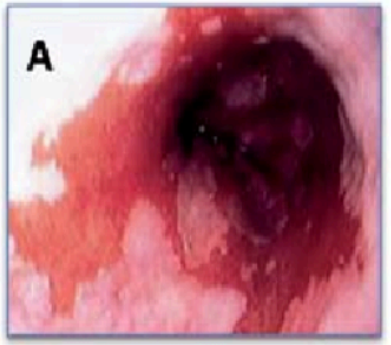


Treatment

- Yearly surveillance endoscopy is recommended in all patients with a diagnosis of Barrett's esophagus.
- Patients undergoing surveillance are placed on acid suppression medication and monitored for changes in their reflux symptoms.

Diagnosis of Barrett's esophagus

- **A. Columnar type mucosa in the distal esophagus on endoscopy**
- **B. Specialized intestinal metaplasia on histology**



No dysplasia → Repeat endoscopy in 6–12 months to confirm lack of dysplasia → Endoscopy every 3 years

Low-grade dysplasia → Repeat endoscopy in 6 months and ask an expert pathologist to confirm low-grade dysplasia → Endoscopy every 12 months
Endoscopic ablation

High-grade dysplasia → Ask two expert pathologists to confirm high-grade dysplasia
Consider new imaging techniques → Endoscopy every 3 months with biopsy every 1 cm
Endoscopic or surgical treatment

Treatment

- The benefits of anti-reflux surgery in patients with Barrett's esophagus.
- Surgery renders the LES competent and restores the barrier to reflux
- Studies have demonstrated regression of metaplasia to normal mucosa up to 57% of the time in patients who have undergone antireflux surgery

Treatment

- Esophageal resection for Barrett's esophagus is recommended only for patients in whom high-grade dysplasia is found
- Pathologic data on surgical specimens demonstrate a 40% risk for adenocarcinoma within a focus of high-grade dysplasia

Caustic Injury

- Best cure for this condition is prevention.
- In children, ingestion of caustic materials is accidental and tends to be in small quantities.
- In teenagers and adults, however, ingestion usually is suicide attempts, and much larger quantities of caustic liquids are consumed

Caustic Injury

Alkali ingestion is more common than acid ingestion because of its lack of immediate symptoms

Alkali ingestion are much more devastating and almost always lead to significant destruction of the esophagus

Table 41-3 -- Three Phases of Tissue Injury From Alkali Ingestion

PHASE	TISSUE INJURY	ONSET	DURATION	INFLAMMATORY RESPONSE
1	Acute necrosis	1-4 days	1-4 days	Coagulation of intracellular proteins
				Inflammation
2	Ulceration and granulation	3-5 days	3-12 days	Tissue sloughing
				Granulation of ulcerated tissue bed
3	Cicatrization and scarring	3 weeks	1-6 months	Adhesion formation
				Scarring

Symptoms and Diagnosis

- 1st Stage ➔ patients may complain of oral and substernal pain, hyper salivation, odynophagia and dysphagia, hematemesis, and vomiting.
- 2nd Stage ➔ these symptoms may disappear only to see dysphagia reappear as fibrosis and scarring begin to narrow the esophagus throughout stage three

Symptoms and Diagnosis

- Symptoms of respiratory distress, such as hoarseness, stridor, and dyspnea, suggest upper airway edema and are usually worse with acid ingestion.
- Pain in the back and chest may indicate a perforation of the mediastinal esophagus, whereas abdominal pain may indicate abdominal visceral perforation

Symptoms and Diagnosis

- **Diagnosis is initiated with a physical exam specifically evaluating the mouth, airway, chest, and abdomen**
- **Careful inspection of the lips, palate, pharynx, and larynx is done**
- **The abdomen is examined for signs of perforation**

Symptoms and Diagnosis

- **Early endoscopy is recommended 12 to 24 hours after ingestion to identify the grade of the burn.**
- **Serial chest and abdominal radiographs are indicated to follow patients with questionable chest and abdominal exams**

Table 41-4 -- Endoscopic Grading and Treatment of Corrosive Esophageal and Gastı

DEGREE OF BURN	ENDOSCOPIC EVALUATION	TREATMENT
First degree	Mucosal hyperemia	48-hr observation
	Edema	Acid suppression
Second degree	Limited hemorrhage	Aggressive IV resuscitation
	Exudates	IV antibiotics
	Ulceration	Acid suppression
	Pseudomembrane formation	
Third degree	Mucosal sloughing	Inhaled steroids
	Deep ulcerations	Fiberopticintubation(ifneeded)
	Massive hemorrhage	
	Complete luminal obstruction	
	Charring	
	Perforation	

Treatment

- Management of the acute phase is aimed at limiting and identifying the extent of the injury.
- It begins with neutralization of the ingested substance.
- Alkalis (including lye) are neutralized with half-strength vinegar or citrus juice.

Treatment

- Acids are neutralized with milk, egg whites, or antacids.
- Emetics and sodium bicarbonate need to be avoided because they can increase the chance of perforation.

Treatment

- **First-Degree Burn :**
  - **48 hours of observation is indicated**
  - **Oral nutrition can be resumed when a patient can painlessly swallow saliva**
  - **A repeat endoscopy and barium esophago-gram are done in follow-up at intervals of 1, 2, and 8 months**

Treatment

- **Second- and Third-Degree Burns :**
  - **Resuscitation is aggressively pursued**
  - **The patient is monitored in the intensive care unit**
  - **kept (NPO) with IV fluids. IV antibiotics and a proton pump inhibitor are started**
  - **Fiber optic intubation may be needed and must be available**

Benign esophageal tumor

Benign Esophageal Tumors and Cysts

- Benign tumors are rare (< 1 %)
- Classified in two groups
  - Mucosal
  - Extramucosal (intramural)
- More useful classification:
  - 60% of benign neoplasms are leiomyomas
  - 20% are cysts
  - 5% are polyps
  - Others (< 2 percent)

Esophageal Cysts

- Arise as diverticula of the embryonic foregut
- ¾ of this cyst present in childhood
- 60% are located along the right side of the esophagus
- 60% present in the first year of life with either respiratory or esophageal symptoms
- Cyst found in the upper third of the esophagus present in infancy while lower third lesions present later in childhood

Pedunculated Intraluminal Tumors (Polyps)

- Rare.
- Occur in older men and may cause intermittent dysphagia
- Easily missed with barium swallow and esophagoscopy

Leiomyoma

- **60% of all benign esophageal tumors.**
- **Found in men slightly more than women**
- **Present in the 4th and 5th decades**
- **They are found in the distal two thirds of the esophagus more than 80% of the time**

Leiomyoma

They are usually solitary and remain intramural, causing symptoms as they enlarge.

Recently, they have been classified as a gastrointestinal stromal tumor (GIST)

GIST tumors are the most common mesenchymal tumors of the gastrointestinal tract and can be benign or malignant

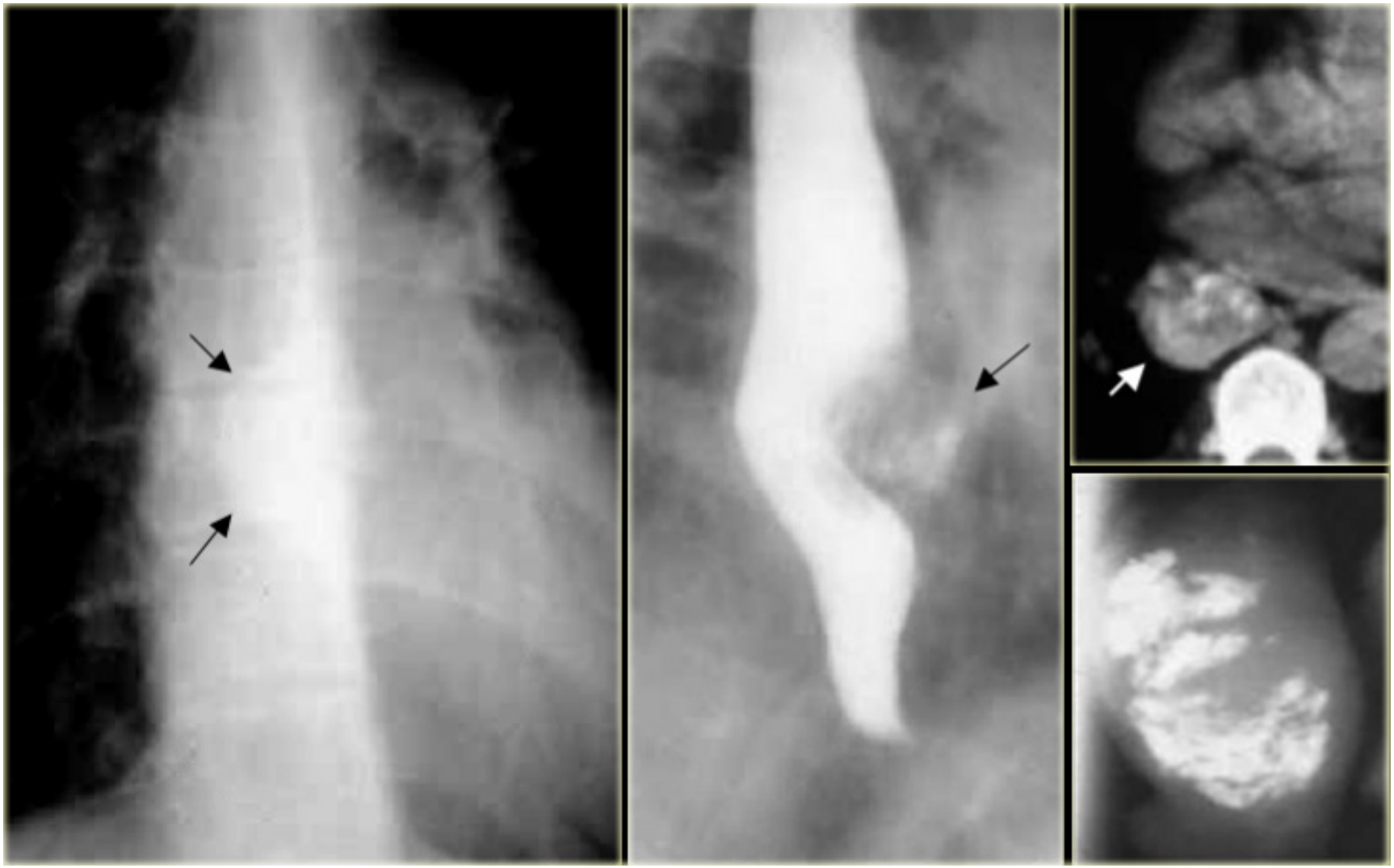
Leiomyoma

Nearly all GIST tumors occur from mutations of the c-*KIT* oncogene, which codes for the expression of c-*KIT* (CD117).

All leiomyomas are benign with malignant transformation being rare

Symptoms and Diagnosis

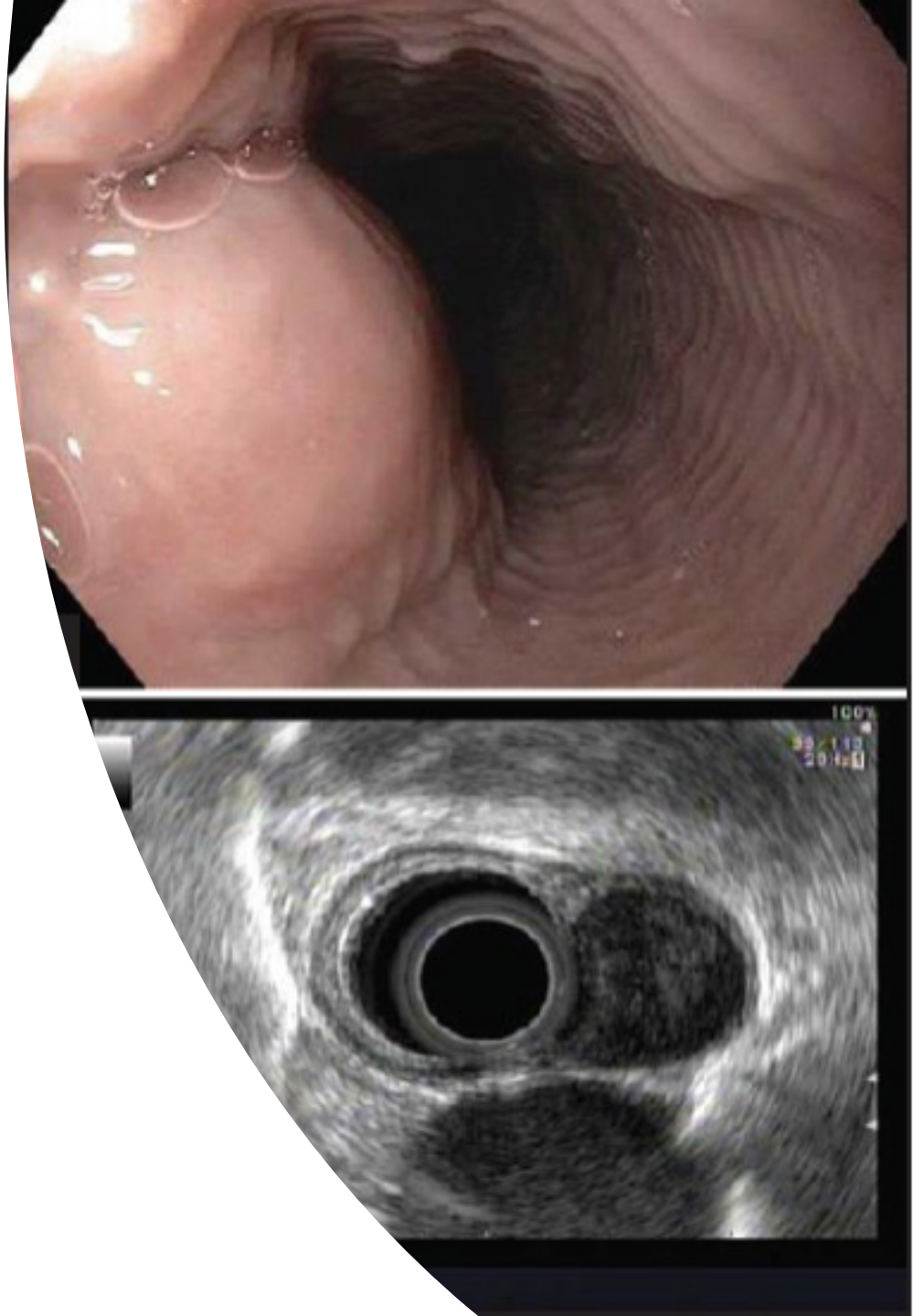
- **Many leiomyomas are asymptomatic.**
- **Dysphagia and pain are the most common symptoms and can result from even the smallest tumors**
- **Barium esophagram ➔ a leiomyoma has a characteristic appearance.**



Leiomyoma

- **Endoscopy ➔**
- **Extrinsic compression**
- **Overlying mucosa is intact.**

- **Endoscopic ultrasound (EUS) ➔ hypoechoic mass in the submucosa or muscularis propria**



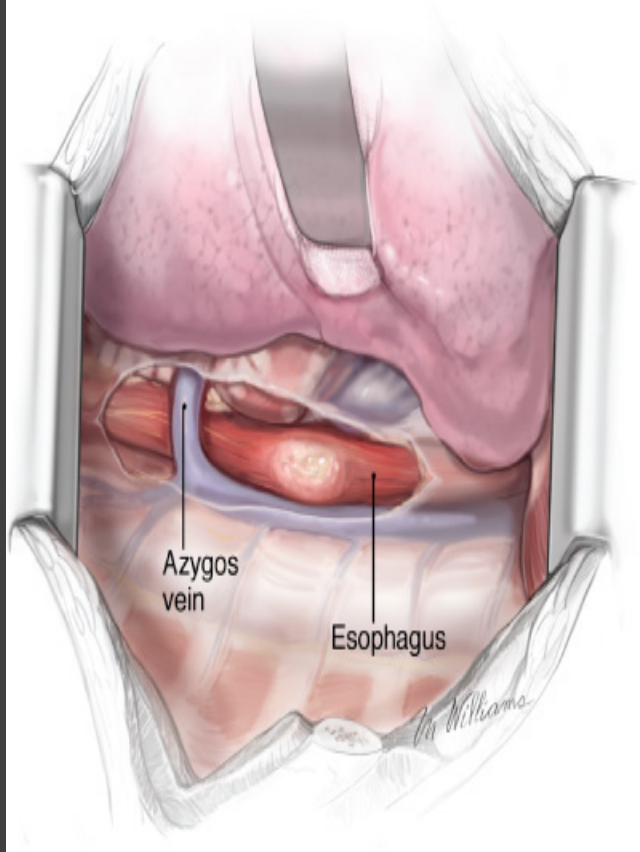
Treatment

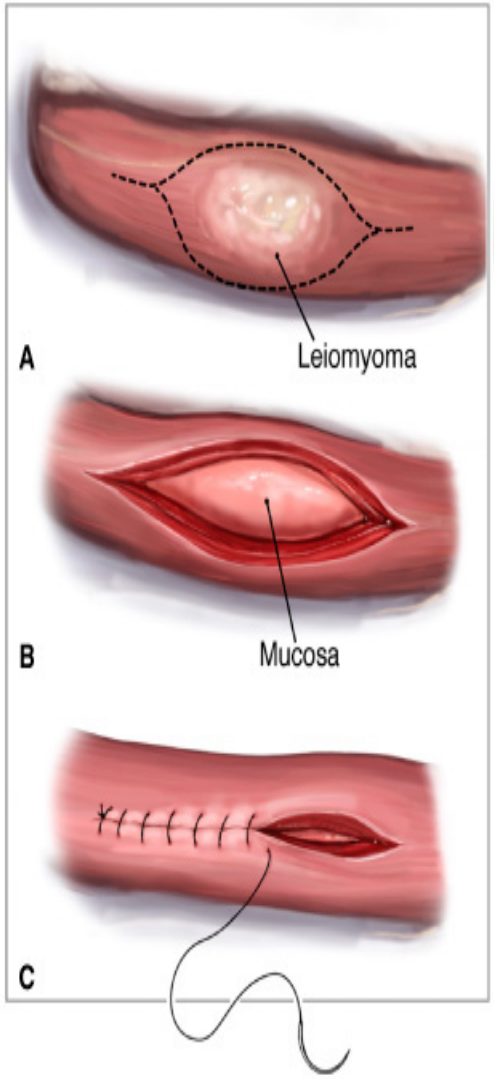
Leiomyomas are slow-growing tumors with rare malignant potential that will continue to grow and become progressively symptomatic with time.

Although observation is acceptable in patients with small (<2 cm) asymptomatic tumors or other significant co morbid conditions, in most patients, surgical resection is advocated

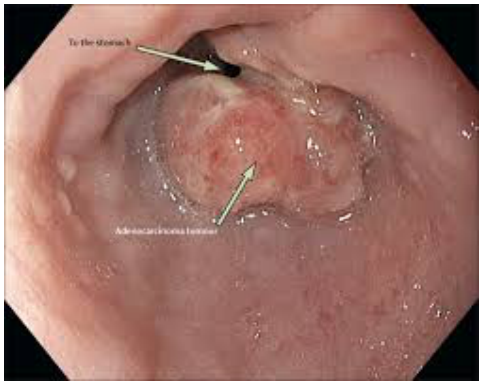
Treatment

- **Surgical enucleation of the tumor remains the standard of care.**
- **Success in relieving dysphagia approaches 100%**





Source: Sugarbaker DJ, Bueno R, Krasna MJ, Mentzer SJ, Zellos L: *Adult Chest Surgery:* http://www.accesssurgery.com

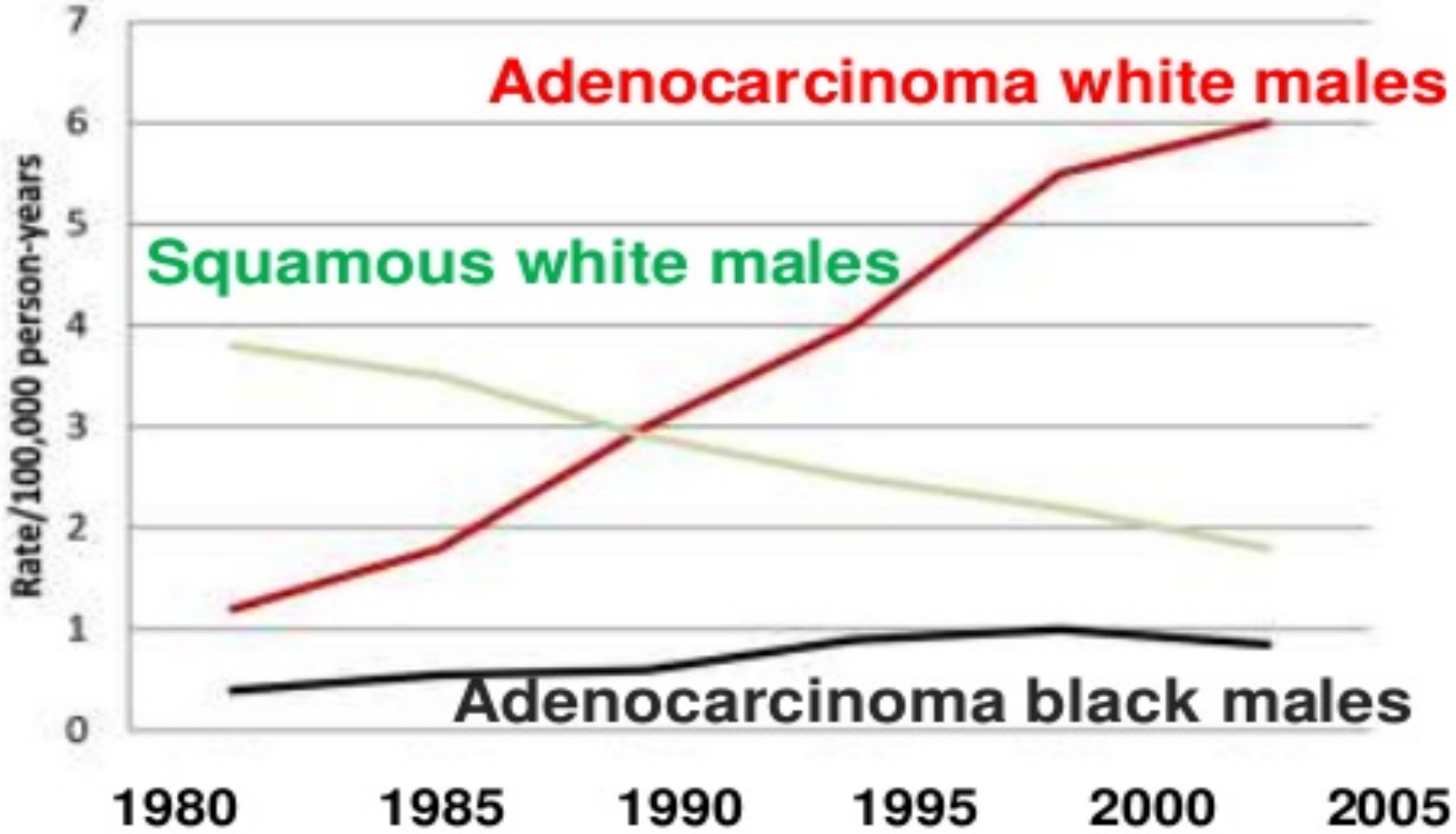


Malignant esophageal tumor

2 common types :

- Squamous cell
- Adneocarcinoma
- Others

Trends in the US for Esophagus Cancer



Squamous cell cancer

- **Arise from the squamous mucosa that is native to the esophagus.**
- **Found in the upper and middle third of the esophagus 70% of the time**
- **Smoking and alcohol both increase the risk for f cancers by 5-fold. Combined**

Food additives, including nitrosamines found in pickled and smoked foods, long-term ingestion of hot liquids.

Caustic ingestion, achalasia, bulimia, tylosis (an inherited autosomal dominant trait), Plummer-Vinson syndrome, external-beam radiation, and esophageal diverticula all have known associations with squamous cell cancer.

CARCINOMA OF THE ESOPHAGUS

- Survival based on the stage of the disease ➔
- 5 year survival
- 70% with polypoid lesions.
- 15% with advanced tumors.

Esophageal adenocarcinoma

- **There are a number of factors that are responsible for this shift in cell type:**
  - **Western diet ➔ obesity**
  - **Increasing incidence of GERD**
- **Intake of caffeine, fats, and acidic and spicy foods all lead to decreased tone in the LES and an increase in reflux ➔ increase risk of Barrett's metaplasia.**

Symptoms

- Early-stage cancers may be asymptomatic or mimic symptoms of GERD.
- Most patients with esophageal cancer present with dysphagia and weight loss.
- Because of the distensibility of the esophagus, a mass can obstruct two thirds of the lumen before symptoms of dysphagia are noted

Symptoms

- Choking, coughing, and aspiration from a tracheo-esophageal fistula, as well as hoarseness and vocal cord paralysis from direct invasion into the recurrent laryngeal nerve, are ominous signs of advanced disease.

- Systemic metastases to liver, bone, and lung can present with jaundice, excessive pain, and respiratory symptoms.

Diagnosis

- There are a plethora of modalities available to diagnose and stage esophageal cancer.
- Radiologic tests, endoscopic procedures, and minimally invasive surgical techniques all add value to a solid staging workup in a patient with esophageal cancer.

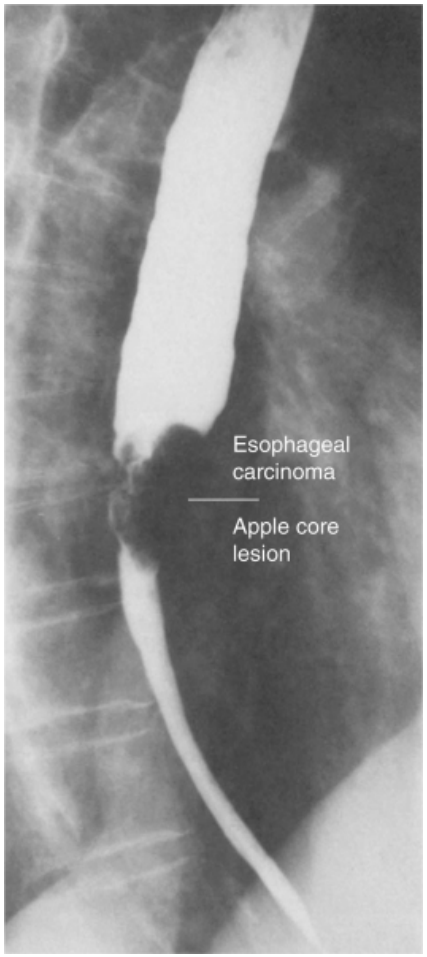
Esophagram

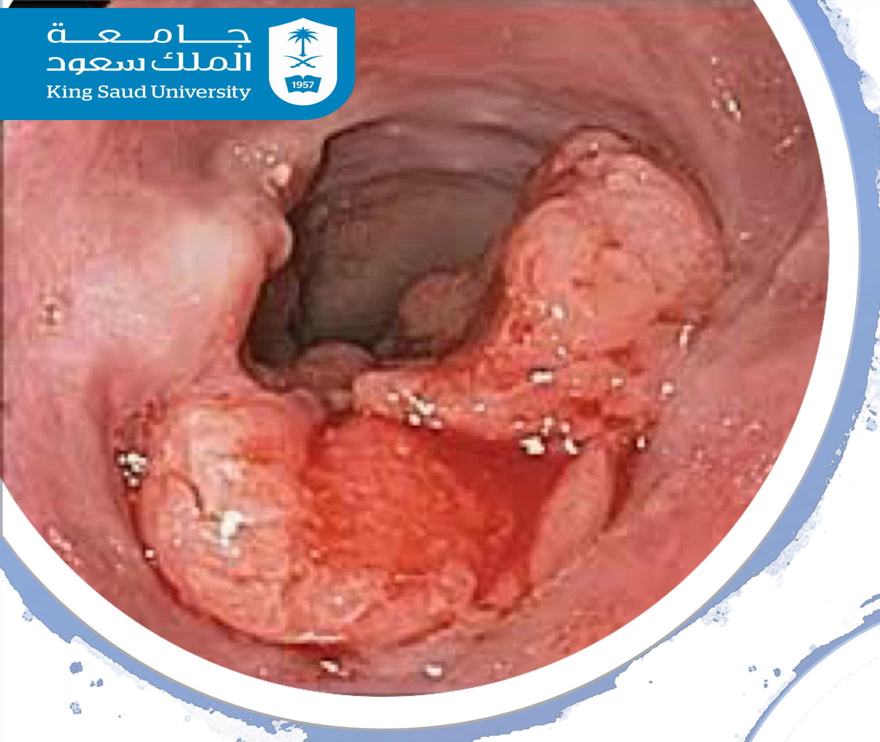
- **A barium esophagram is recommended for any patient presenting with dysphagia**

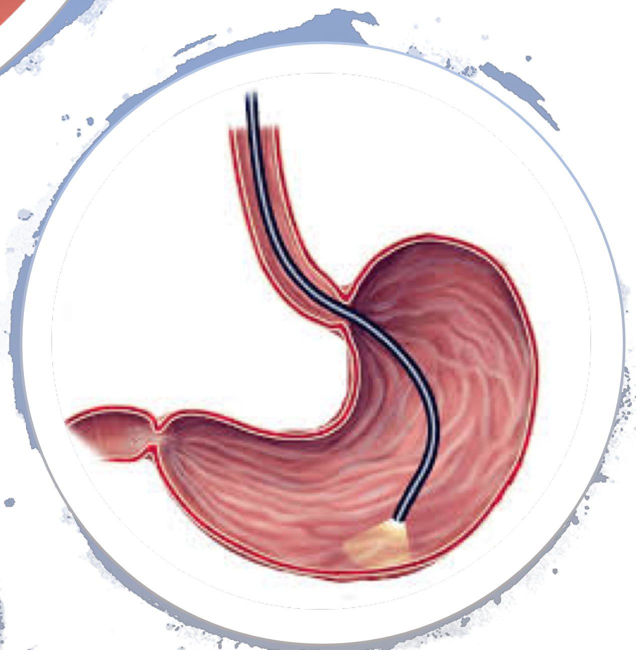
- **Differentiate:**
- **Intra-luminal vs. intramural lesions**
- **Intrinsic vs. extrinsic compression**

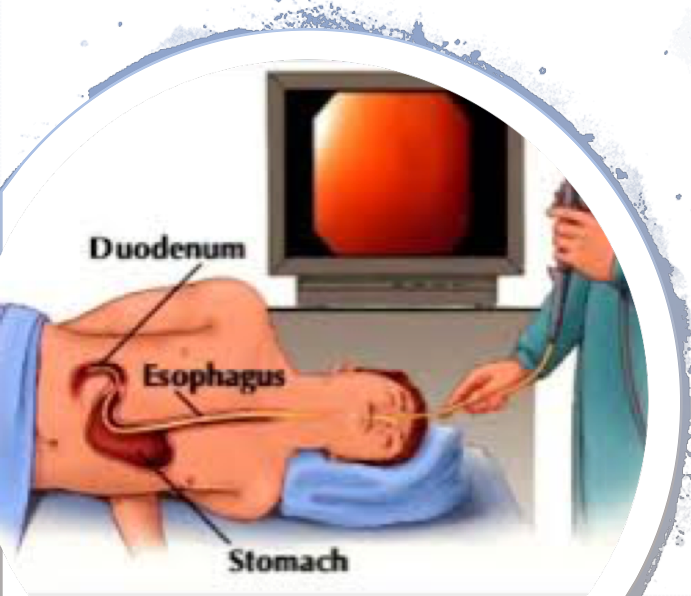
Esophagram

- **The classic finding of an apple-core lesion in patients with esophageal cancer is recognized easily**


Esophageal carcinoma
Apple core lesion





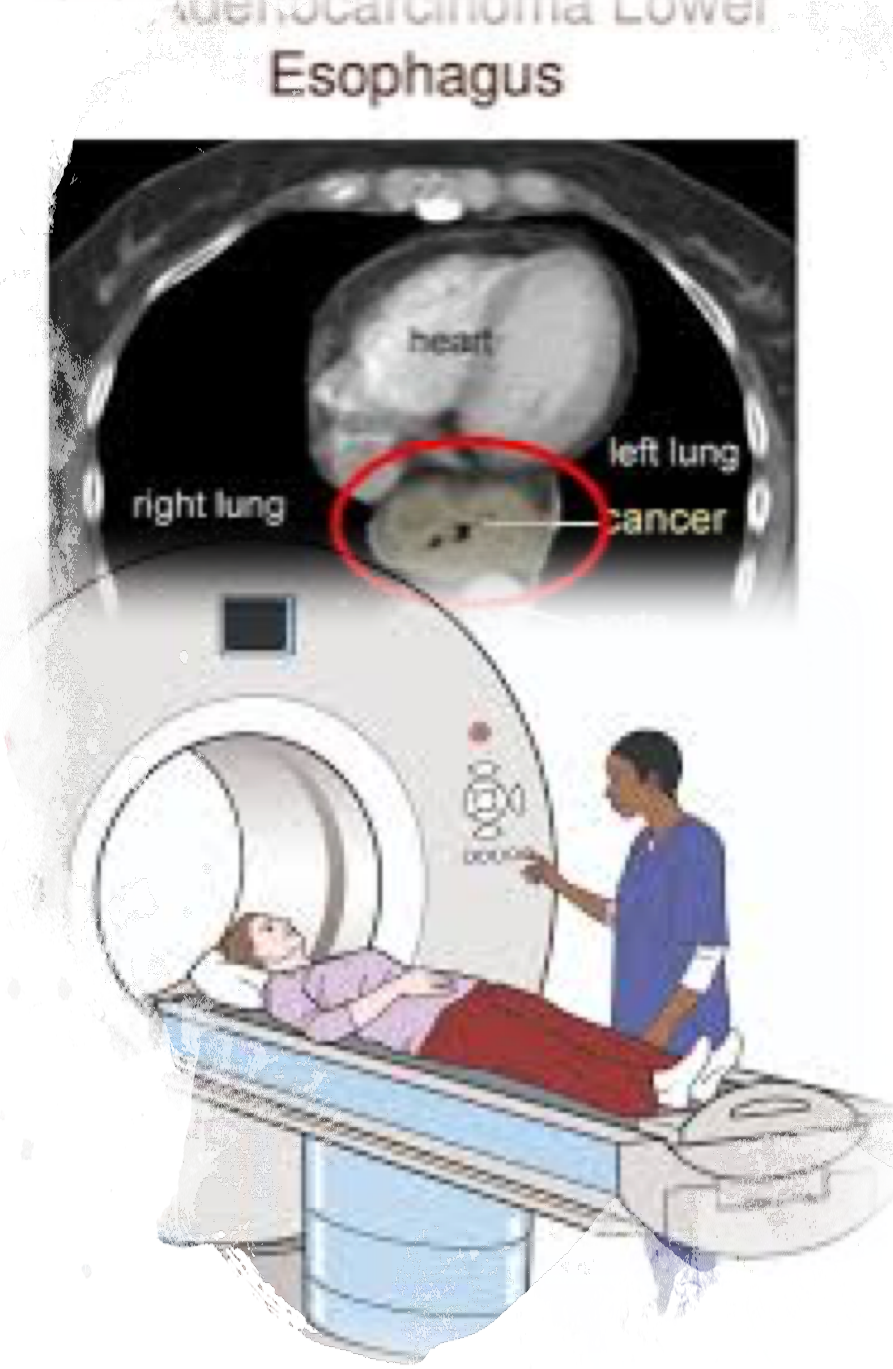


Endoscopy

- Allow:
- Direct visualization
- Determine the location of the tumor
- Biopsies.

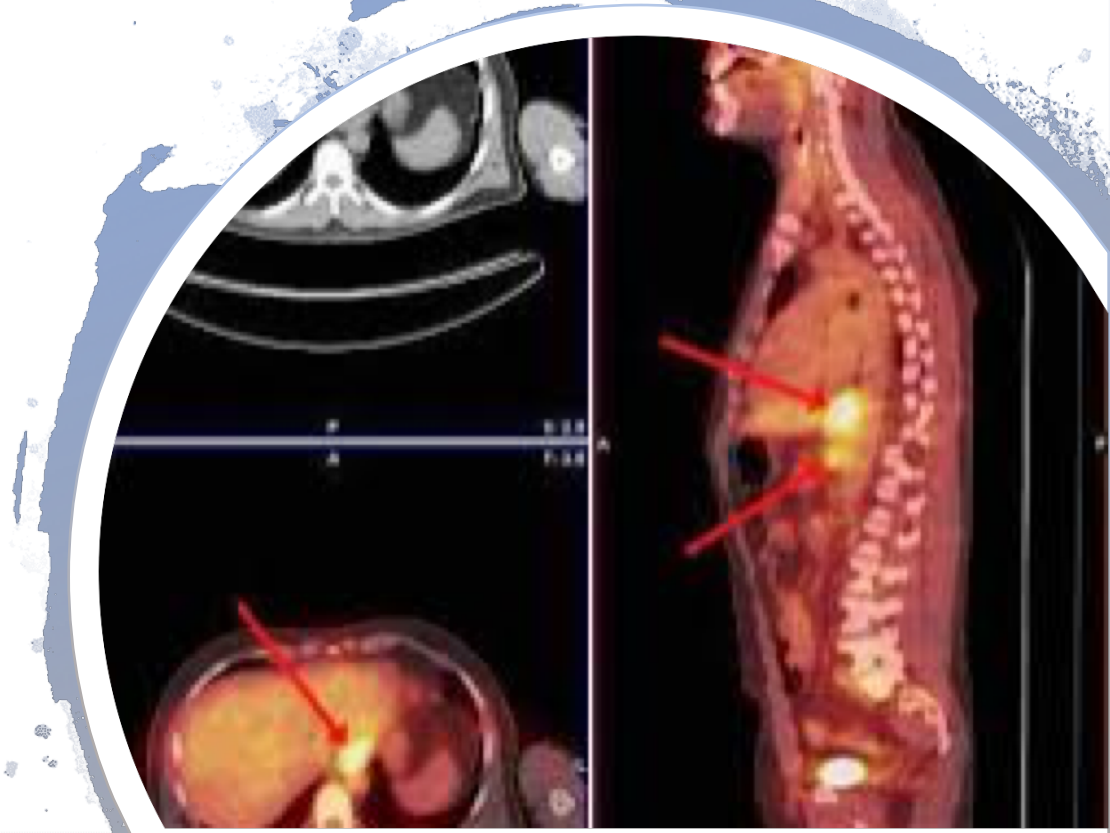
Computed Tomography

- **CT scan of the chest and abdomen and pelvis :**
- **assess the length of the tumor**
- **thickness of the esophagus and stomach**
- **regional lymph node status**
- **distant disease to the liver and lungs**

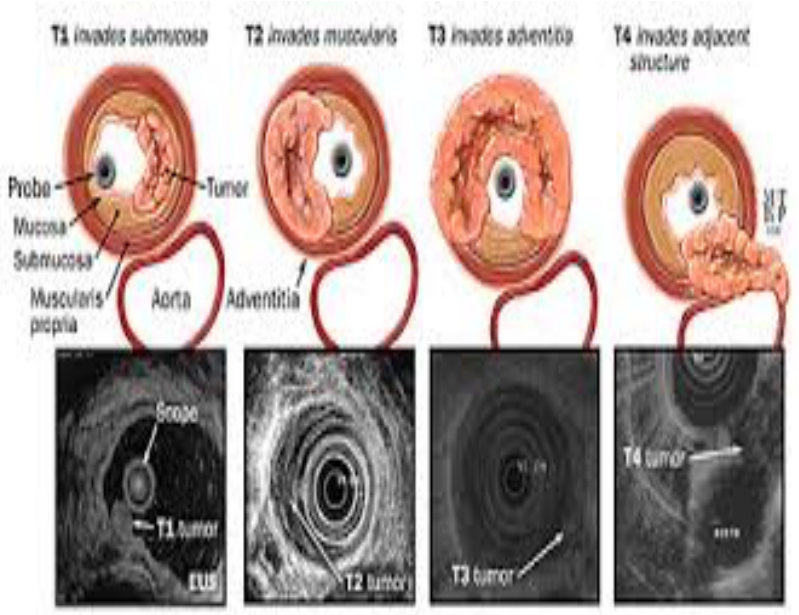


Positron Emission Tomography

- ***PET scan evaluates:***
- **The primary mass**
- **Regional lymph nodes**
- **Distant metastasis**



Endoscopic Ultrasound



- **EUS is the most critical component of esophageal cancer staging.**
- **Determine:**
- **Invasion depth of the tumor**
- **Lymph nodes involvement.**

Treatment

- Depend on the clinical stage.
- Usually combined multimodality therapy.
- Chemothepary + radiotherapy +/- surgery