Diseases of the esophagus 1

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Diseases that affect the esophagus

- 1. Obstruction: mechanical or functional.
- 2. vascular diseases: varices.
- 3. Inflammation: esophagitis.
- 4. Tumours.

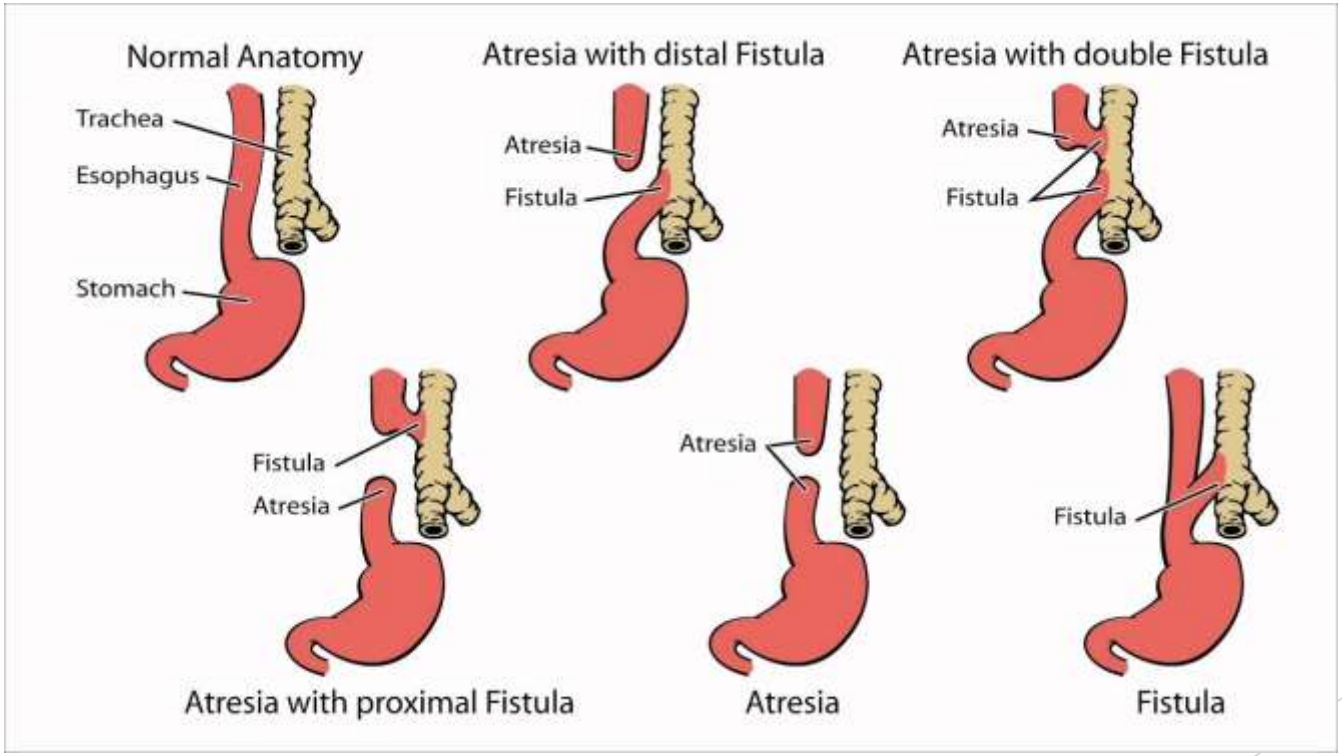
Mechanical Obstruction

- Congenital or acquired.

- Examples:
- Atresia
- Fistulas
- Duplications
- Agenesis (v rare)
- Stenosis.

Atresia

- Thin, noncanalized cord replaces a segment of esophagus.
- Most common location: at or near the tracheal bifurcation
- +- fistula (upper or lower esophageal pouches to a bronchus or trachea).


Normal Anatomy
Trachea
Esophagus
Stomach
Atresia with distal Fistula
Atresia
Fistula
Atresia with double Fistula
Atresia
Fistula
Fistula
Atresia
Atresia with proximal Fistula
Atresia
Atresia
Fistula
Fistula

Clinical presentation:

- Shortly after birth: regurgitation during feeding
- Needs prompt surgical correction (rejoin).

- Complications if w/ fistula:
- Aspiration
- Suffocation
- Pneumonia
- Severe fluid and electrolyte imbalances.

Esophageal stenosis

- Acquired>>>Congenital.
- Fibrous thickening of the submucosa & atrophy of the muscularis propria.
- Due to inflammation and scarring

- Causes:
- Chronic GERD.
- Irradiation
- Ingestion of caustic agents

Clinical presentation

- Progressive dysphagia
- Difficulty eating solids that progresses to problems with liquids.

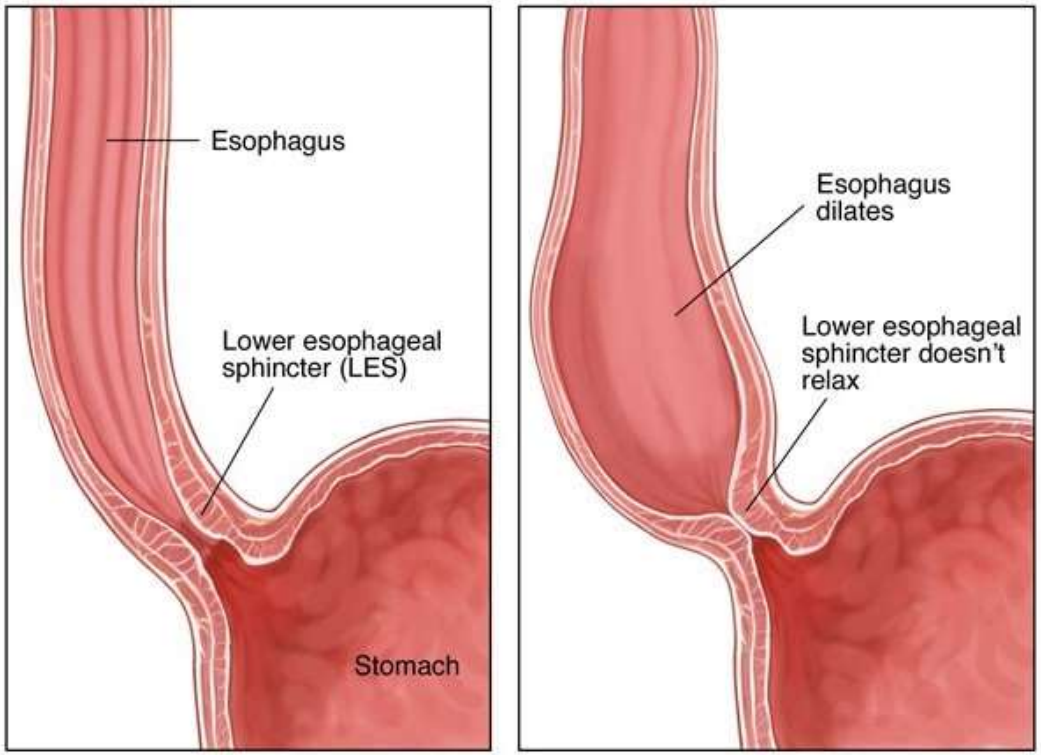
Functional Obstruction

- Efficient delivery of food and fluids to the stomach requires coordinated waves of peristaltic contractions.
- Esophageal dysmotility: discoordinated peristalsis or spasm of the muscularis.
- Achalasia: the most important cause.

Achalasia

- Triad:
- Incomplete LES relaxation
- Increased LES tone
- Esophageal aperistalsis.

- Primary >>>secondary.



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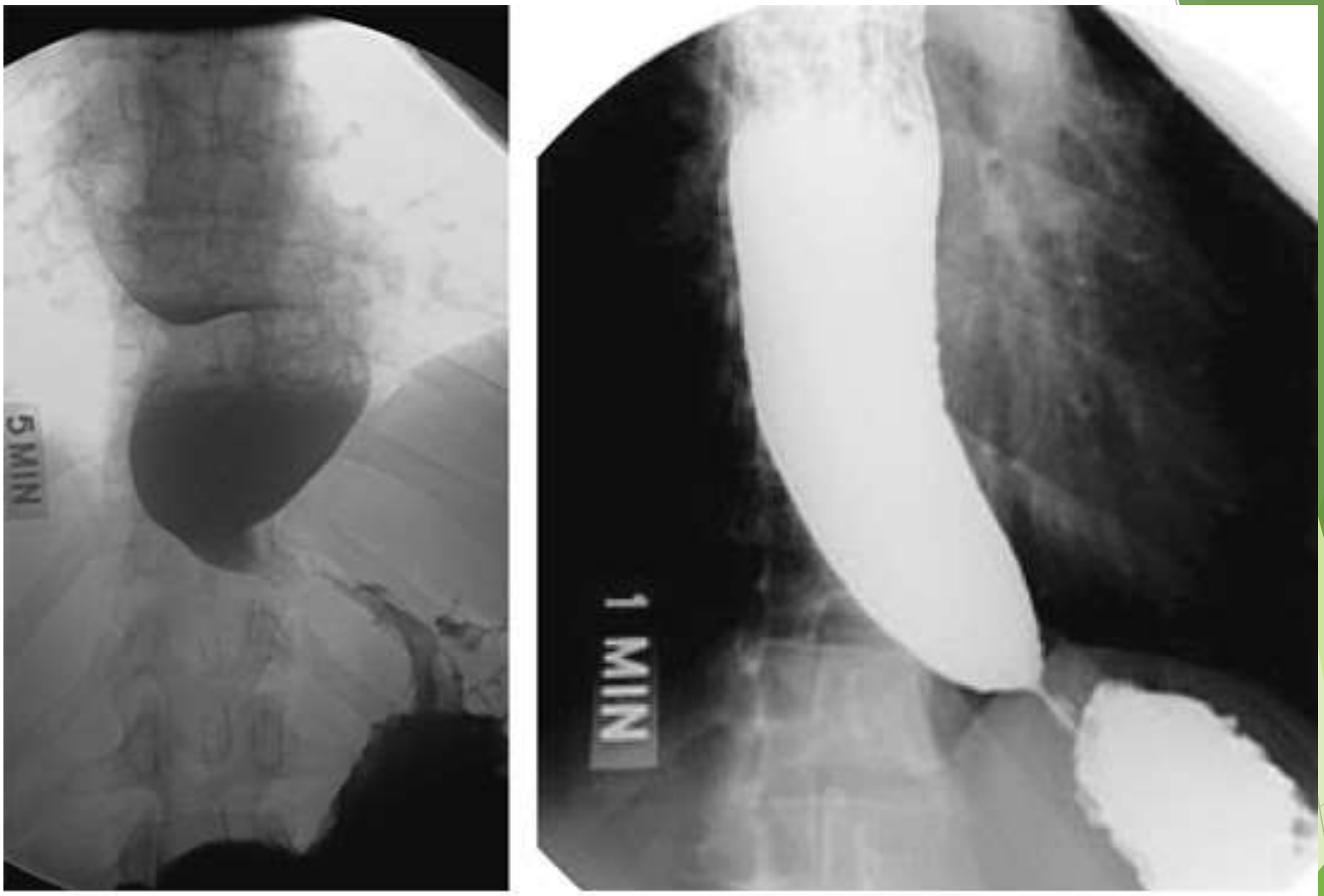


Source: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson JL, Loscalzo J: *Harrison's Principles of Internal Medicine, 18th Edition*: www.accessmedicine.com

Primary achalasia

- Failure of distal esophageal inhibitory neurons.
- Idiopathic
- Most common

Secondary achalasia

- Degenerative changes in neural innervation
- Intrinsic
- Vagus nerve

- **Chagas disease**, Trypanosoma cruzi infection>>destruction of the myenteric plexus>> failure of LES relaxation>> esophageal dilatation.

Clinical presentation

- Difficulty in swallowing
- Regurgitation
- Sometimes chest pain.

Vascular diseases: Esophageal Varices

- Tortuous dilated veins within the submucosa of the distal esophagus and proximal stomach.
- Diagnosis by: endoscopy or angiography.

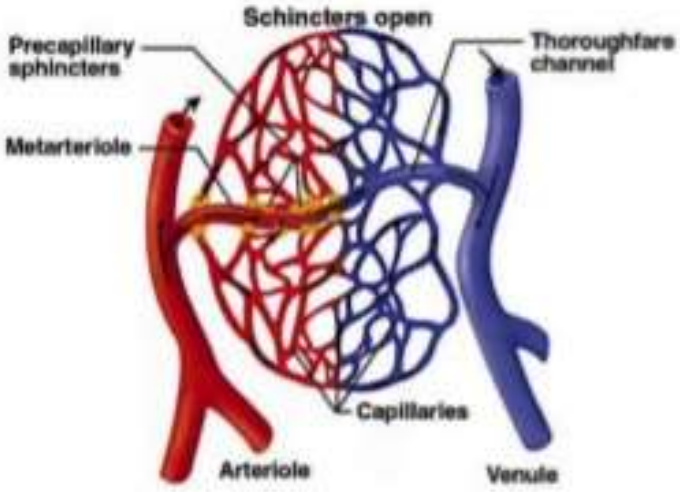
Medpics - UCSD School of Medicine

Pathogenesis:

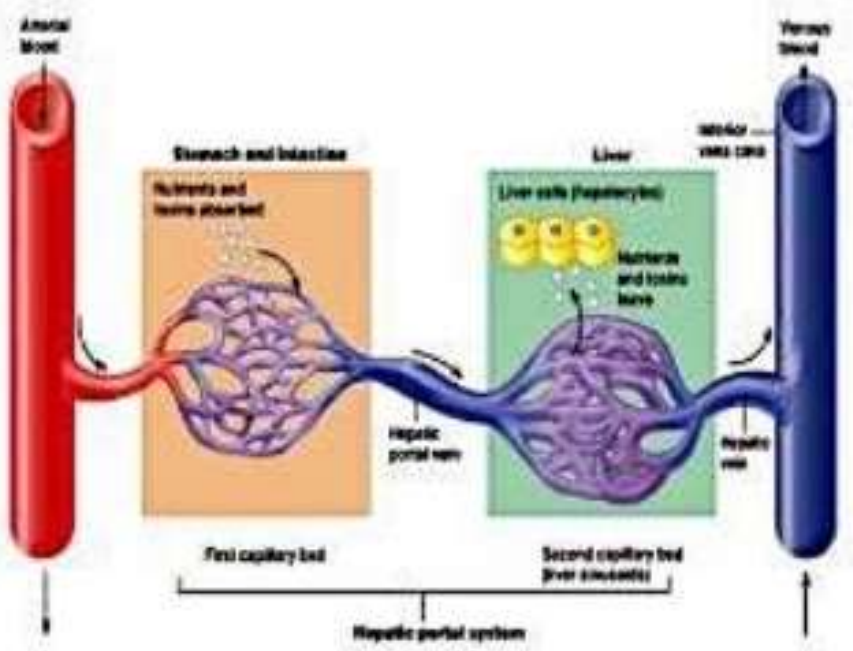
- **Portal circulation**: blood from GIT>>portal vein>>liver (detoxification)>>inferior vena cava.
- Diseases that impede portal blood flow >> portal hypertension >>esophageal varices.
- Distal esophagus : site of Porto-systemic anastomosis.
- **Portal hypertension**>>collateral channels in distal esophagus>>shunt of blood from portal to systemic circulation>>dilated collaterals in distal esophagus>>varices

Portal system

Usual circulation



Portal circulation



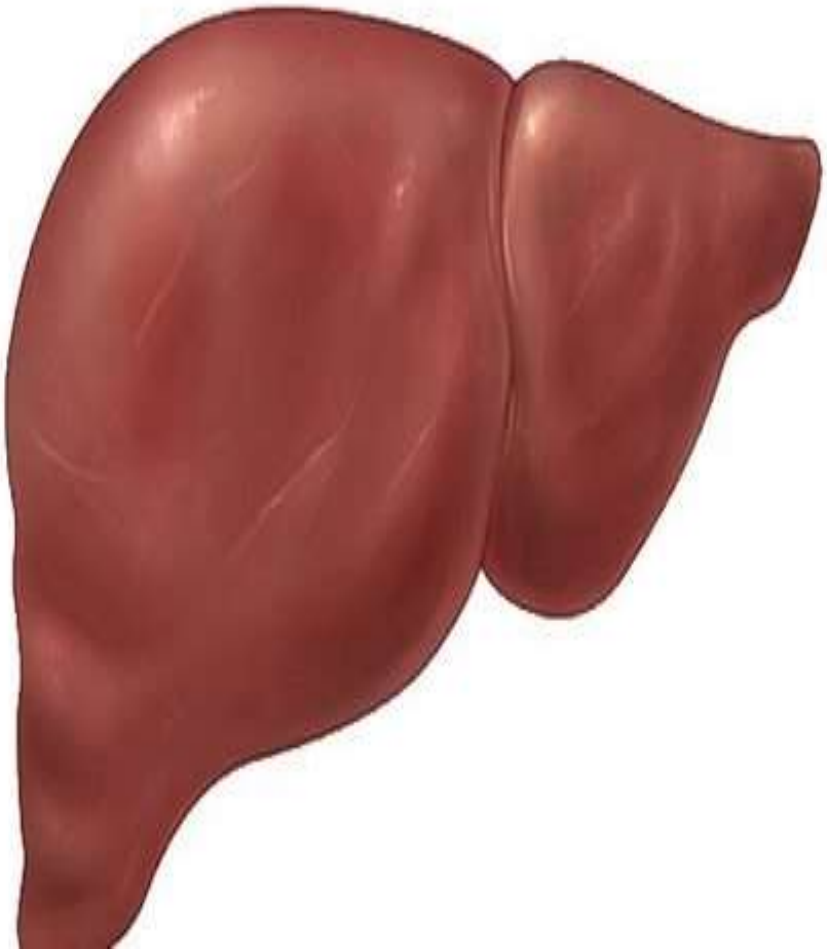
https://www.slideshare.net/rongon28us/hepatic-portal-vein-and-portocaval-anatomosis

Causes of portal hypertension

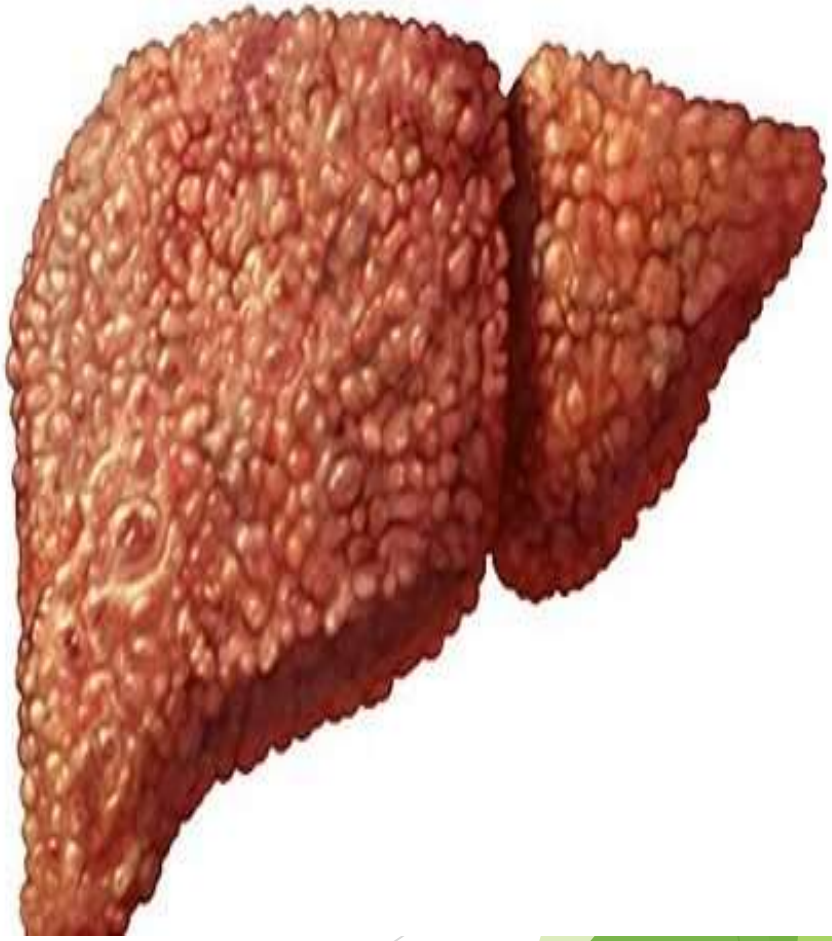
- Cirrhosis is most common

  Alcoholic liver disease.
- Hepatic schistosomiasis 2nd most common worldwide.

Normal Liver



Liver with Cirrhosis



http://www.researchintoasthma.com/7-random-facts-about-liver-cirrhosis.html

Clinical Features

- Often asymptomatic.
- Rupture leads to massive hematemesis and death.
- 50% of patients die from the first bleed despite interventions.
- Death due to: hemorrhage, hepatic come, and hypovolemic shock
- Rebleeding in 20%.

ESOPHAGITIS

- Esophageal Lacerations.
- Mucosal Injury
- Infections
- Reflux Esophagitis
- Eosinophilic Esophagitis

Esophageal Lacerations

- Mallory weiss tears are most common
- Due to : severe retching or forceful prolonged vomiting
- Present with hematemesis.
- vomiting>>stretching>>>tear.

- Linear lacerations
- longitudinally oriented
- Cross the GEJ.
- Superficial
- Heal quickly , no surgical intervention

Chemical Esophagitis

- **Damage to esophageal mucosa by irritants**
- Alcohol,
- Corrosive acids or alkalis
- Excessively hot fluids
- Heavy smoking
- Medicinal pills (doxycycline and bisphosphonates)
- Iatragenic (chemotx, radiotx , GVHD)

Clinical symptoms & morphology

- Ulceration and acute inflammation.

- Only self-limited pain, odynophagia (pain with swallowing).
- Hemorrhage, stricture, or perforation in severe cases

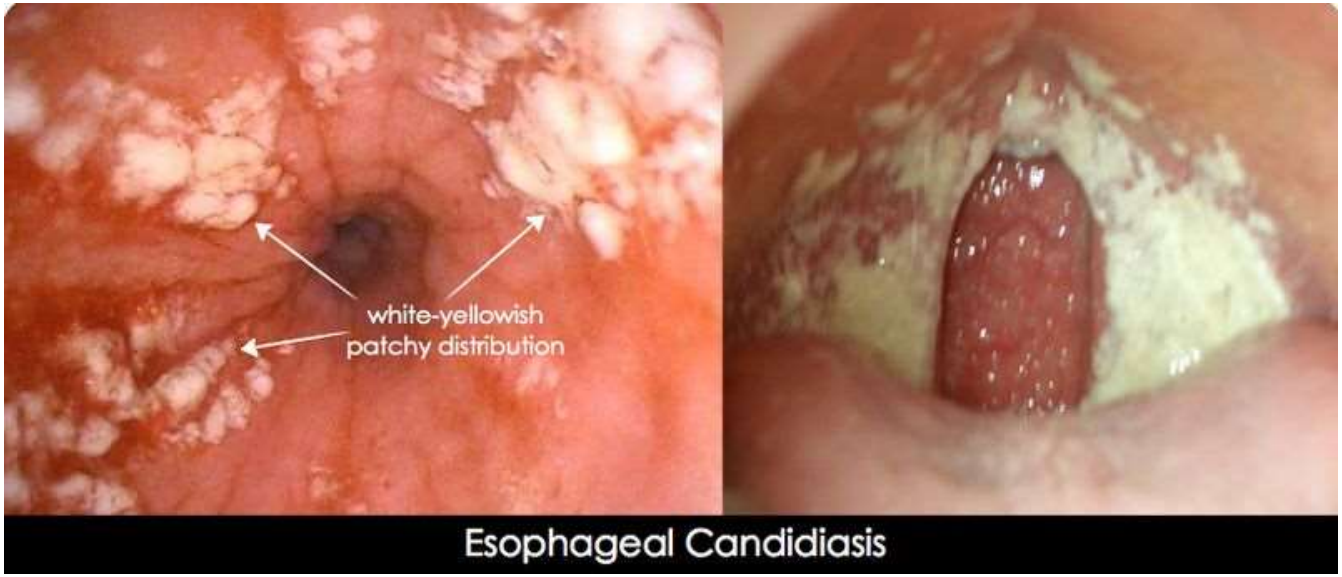
Infectious esophagitis

- Mostly in debilitated or immunosuppressed.

- Viral (HSV, CMV)
- Fungal (candida >>> mucormycosis & aspergillosis)
- Bacterial: 10%.

- Candidiasis :

- Adherent.
- Gray-white pseudomembranes
- Composed of matted fungal hyphae and inflammatory cells



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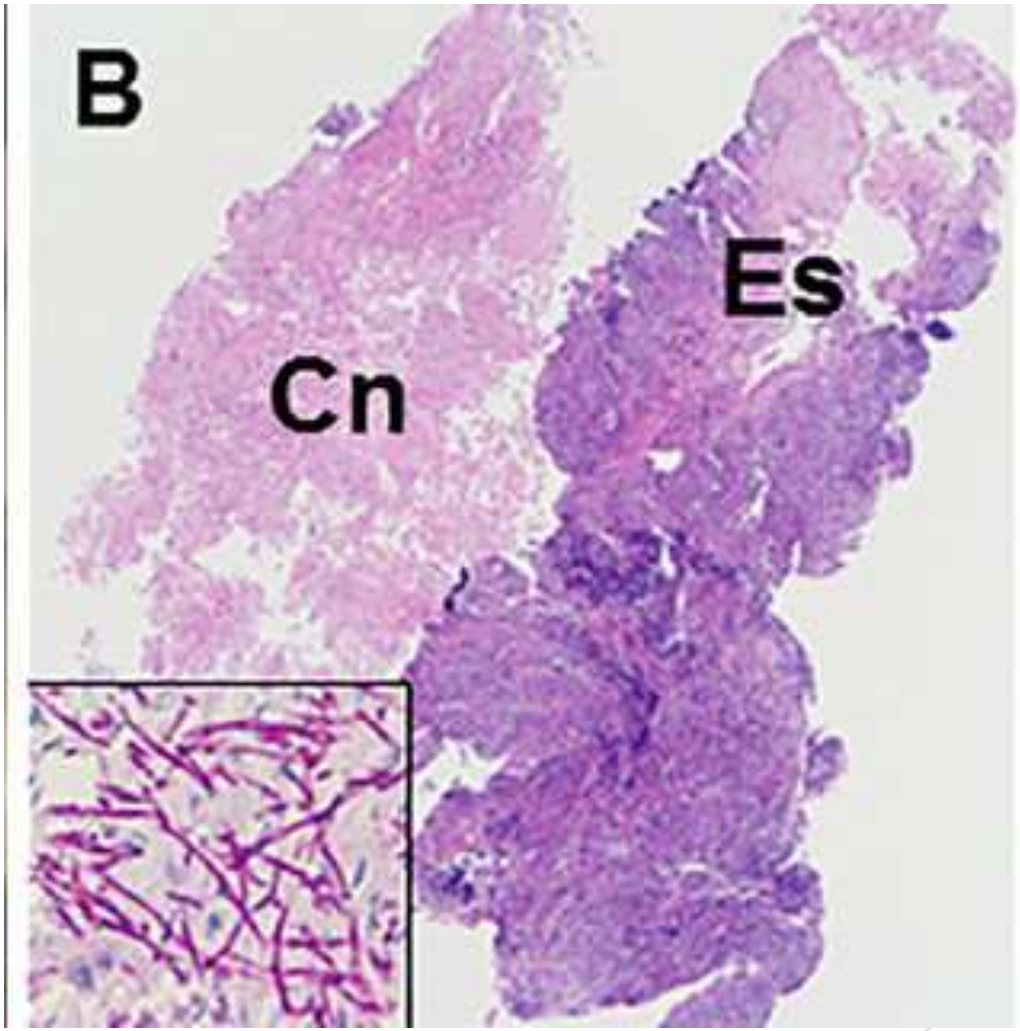
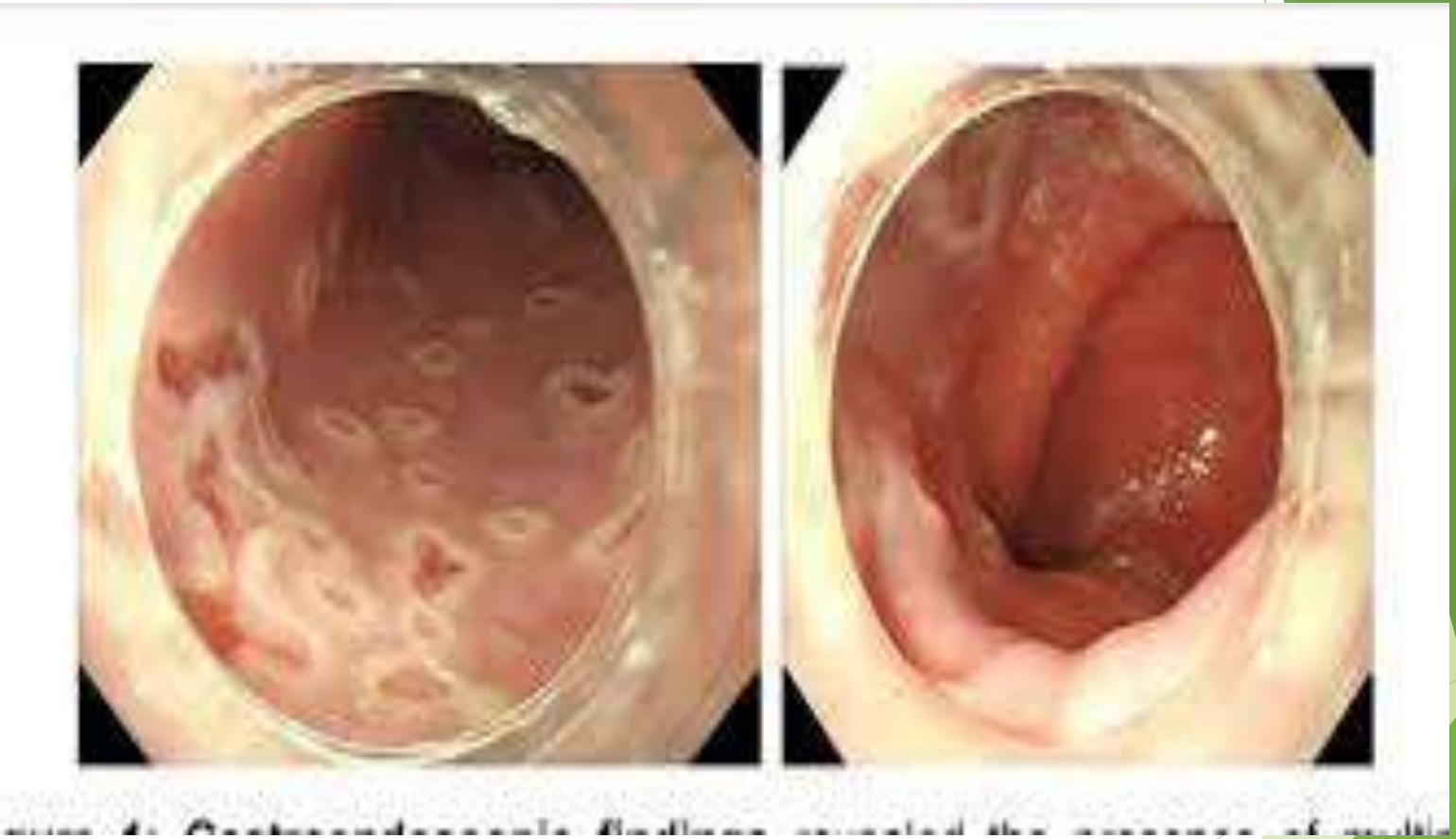
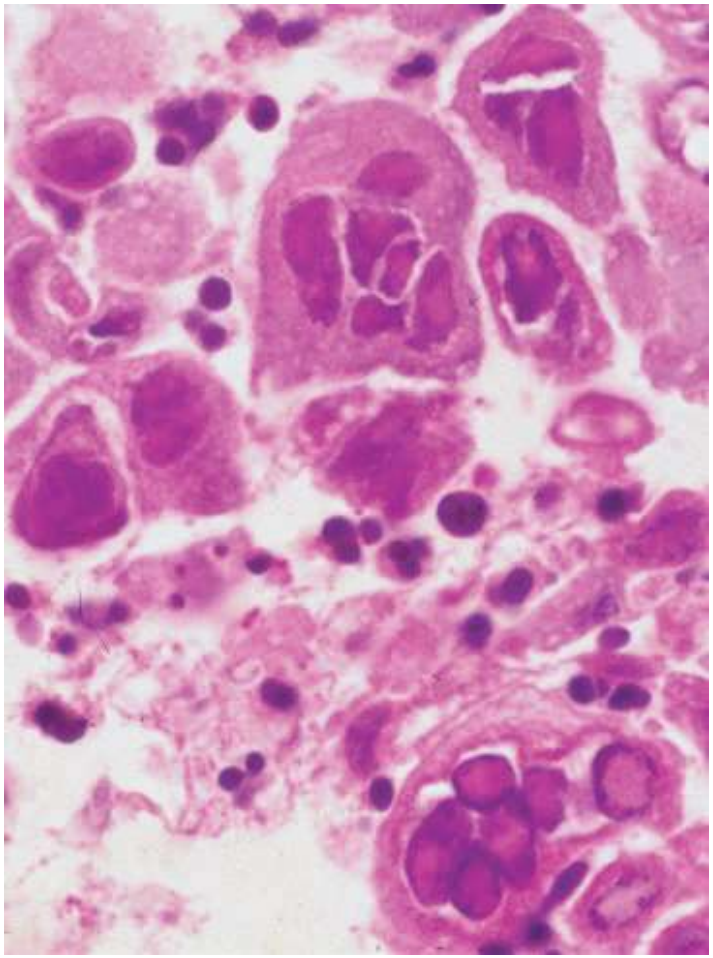


www.researchgate.net/publication/285369734_Esophageal_Candidiasis_as_the_Initial_Manifestation_of_Acute_Myeloid_Leukemia

- Herpes viruses
- Punched-out ulcers
- Histopathologic:
- Nuclear viral inclusions
- Degenerating epithelial cells ulcer edge
- Multinucleated epithelial cells.



Semantic Scholar



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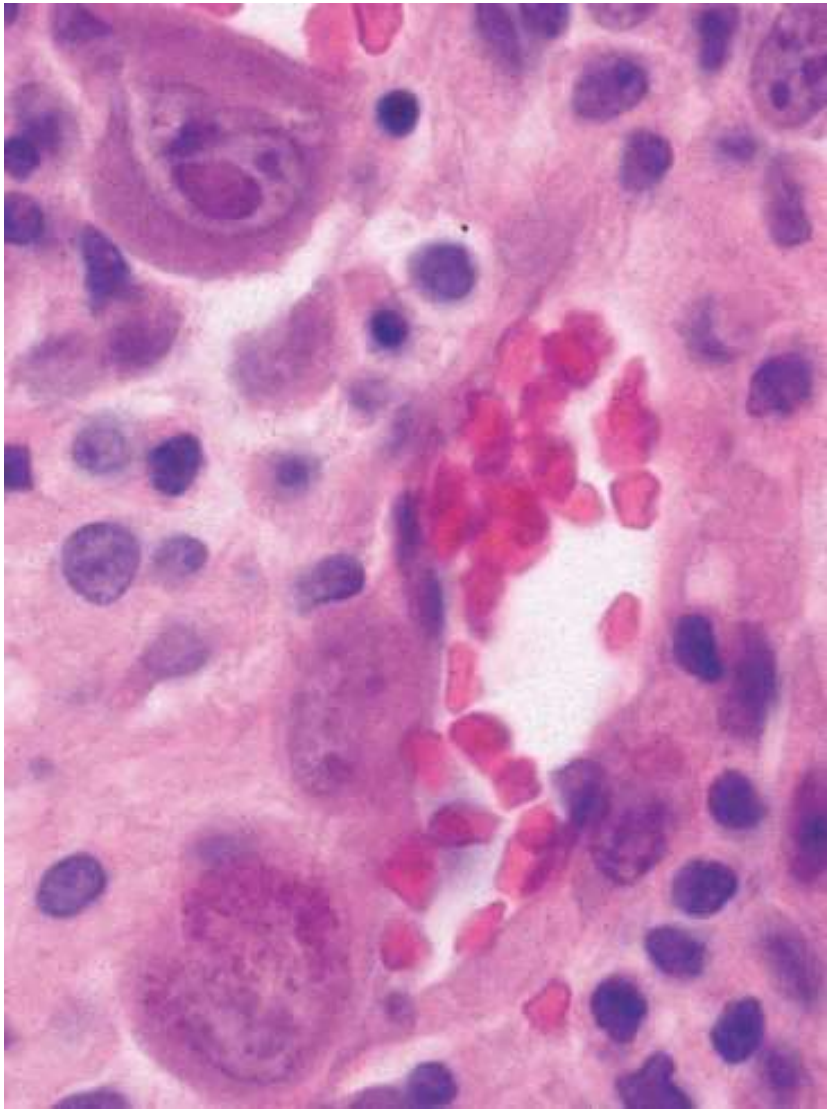
- CMV :
- Shallower ulcerations.
- Biopsy: nuclear and cytoplasmic inclusions in capillary endothelium and stromal cells. Megalo. cells



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Reflux Esophagitis

- Reflux of gastric contents into the lower esophagus
- Most frequent cause of esophagitis
- Most common complaint by patients
- Gastroesophageal reflux disease, GERD

- Squamous epithelium is sensitive to acids
- Protective forces: mucin and bicarbonate, high LES tone

Pathogenesis

- Decreased lower esophageal sphincter tone

  (alcohol, tobacco, CNS depressants)

- Increase abdominal pressure

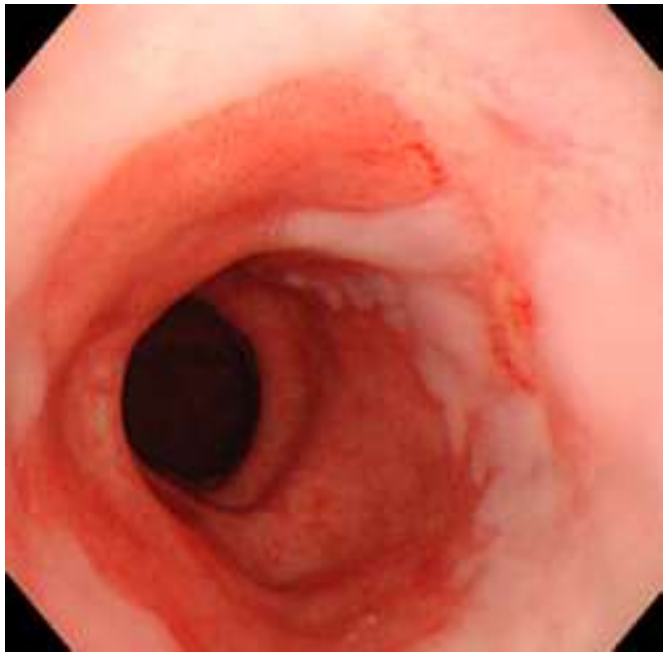
  (obesity,, pregnancy, hiatal hernia, delayed gastric emptying, and increased gastric volume)

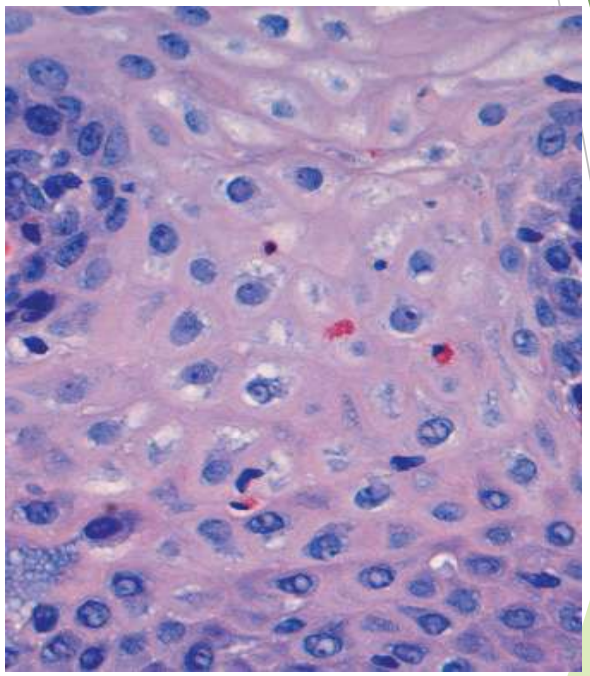
- Idiopathic!!

MORPHOLOGY

- Macroscopy (endoscopy)
- Depends on severity (Unremarkable, Simple hyperemia (red)

- Microscopic:
- Eosinophils infiltration
- Followed by neutrophils (more severe).
- Basal zone hyperplasia
- Elongation of lamina propria papillae





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Clinical Features

- Most common over 40 years.
- May occur in infants and children
- Heartburn , dysphagia,
- Regurgitation of sour-tasting gastric contents
- Rarely: Severe chest pain, mistaken for heart disease

- Tx: proton pump inhibitors

Complications

- Esophageal ulceration
- Hematemesis
- Melena
- Strictures
- Barrett esophagus (precursor of Ca.)

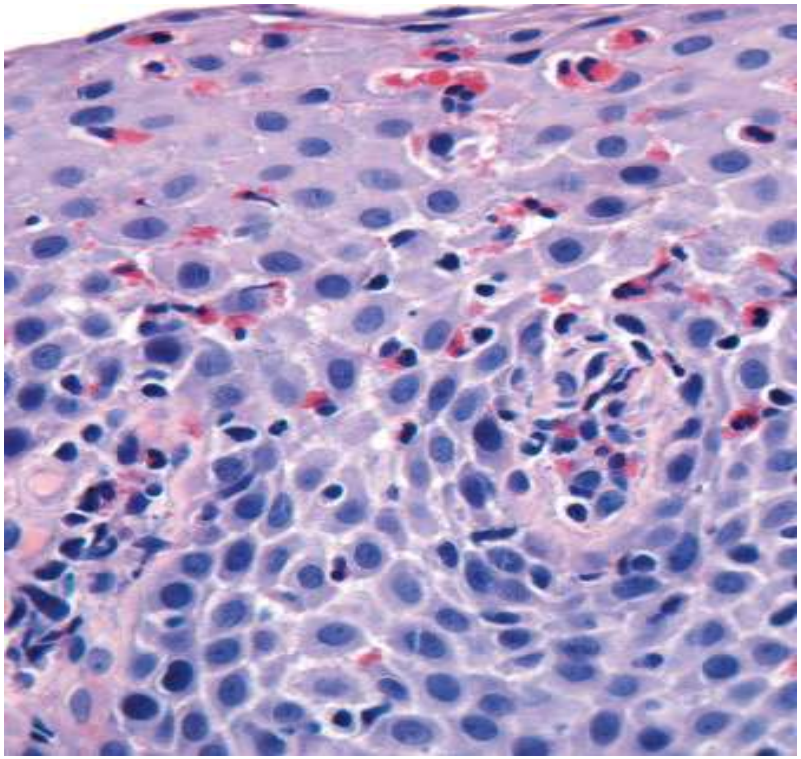
Eosinophilic Esophagitis

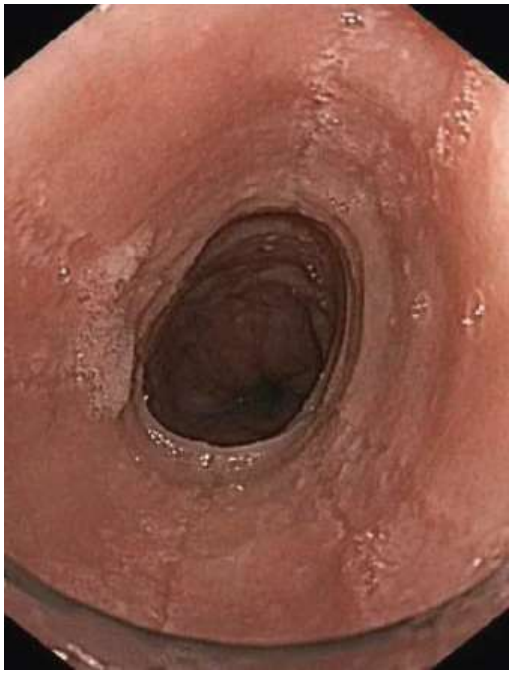
- Chronic immune mediated disorder

- Symptoms:
- Food impaction and dysphagia in adults
- Feeding intolerance or GERD-like symptoms in children

- Endoscopy:
- Rings in the upper and mid esophagus.

- Microscopic:
- Numerous eosinophils w/n epithelium
- Far from the GEJ.





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- Most patients are: atopic (atopic dermatitis, allergic rhinitis, asthma) or modest peripheral eosinophilia.

- Tx:
- Dietary restrictions(cow milk and soy products)
- Topical or systemic corticosteroids.
- Refractory to PPIs.

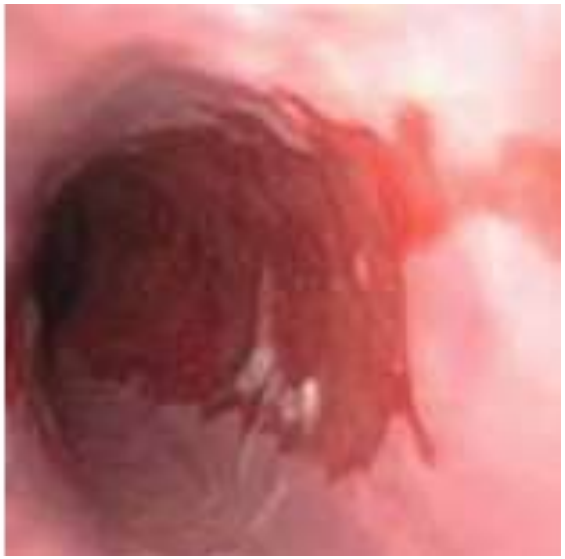
Barrett Esophagus

- Complication of chronic GERD
- Intestinal metaplasia within the esophageal squamous mucosa.
- 10% of individuals with symptomatic GERD
- Males>>females, 40-60 yrs
- **Direct precursor of esophageal adenocarcinoma**
- **Metaplasia >> 0.2-1% /year >>dysplasia>> adenocarcinoma**.

MORPHOLOGY

- Endoscopy:
- Red tongues extending upward from the GEJ.

- Histology:
- Gastric or **intestinal metaplasia**
- Presence of goblet cells
- +-Dysplasia : low-grade or high-grade
- Intramucosal carcinoma: invasion into the lamina propria.



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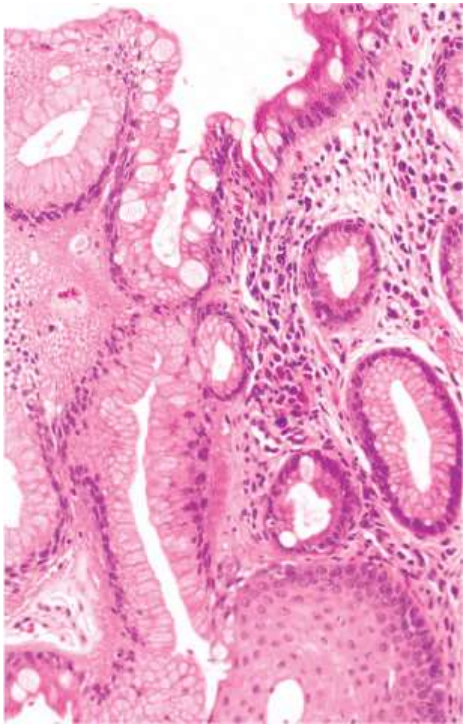

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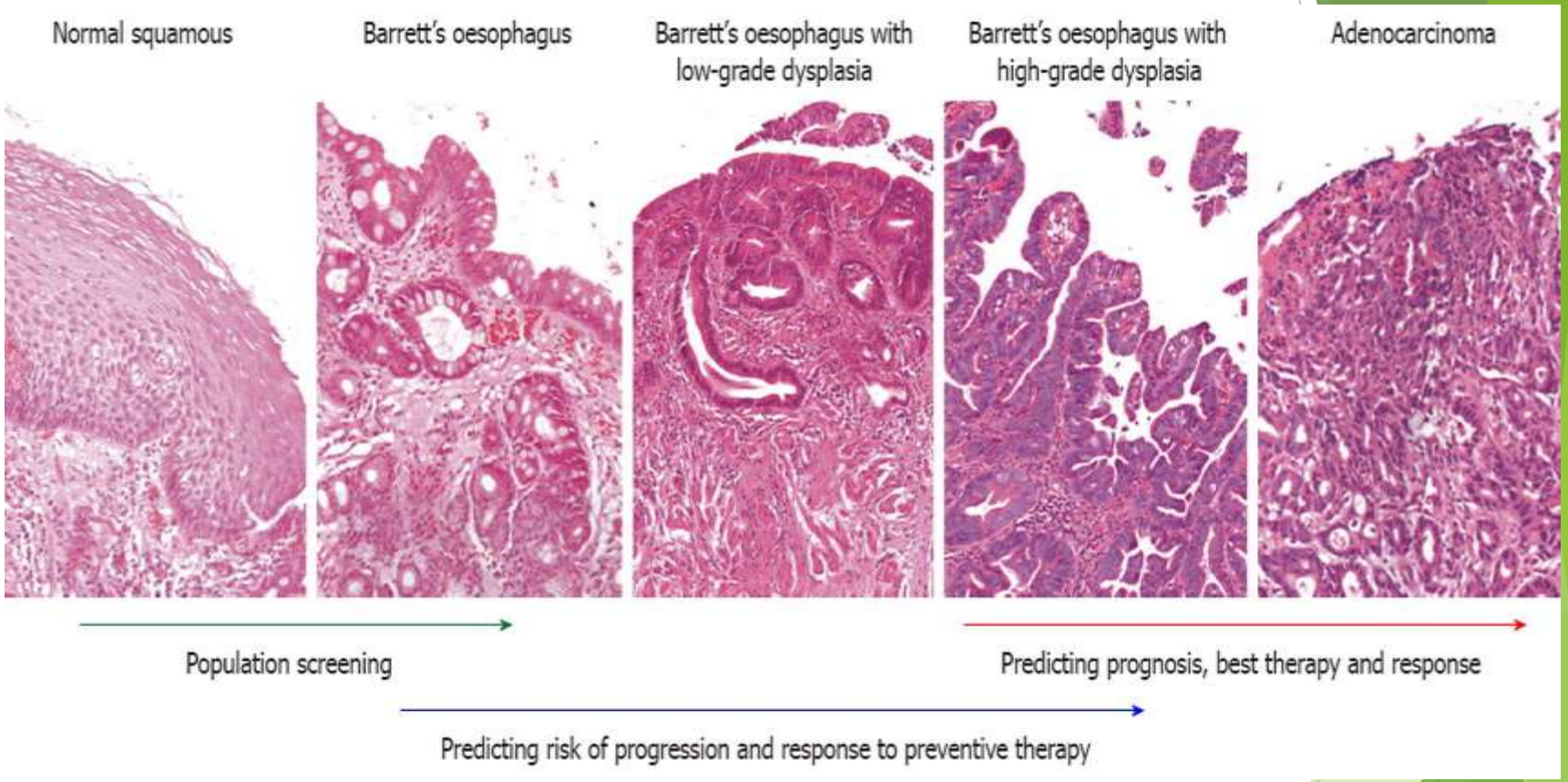

Management of Barrett

- Periodic surveillance endoscopy with biopsy to screen for dysplasia.
- High grade dysplasia & intramucosal carcinoma needs interventions.

ESOPHAGEAL TUMORS

- Squamous cell carcinoma (most common worldwide)
- Adenocarcinoma (on the rise, half of cases)

Adenocarcinoma

- Background of Barrett esophagus and long-standing GERD.
- Risk factors: dysplasia associated Barrett, smoking, obesity, radioTx.
- Male : female (7:1)
- Geographic & racial variation (developed countries)

Pathogenesis

- From Barrett>>dysplasia>>adenocarcinoma
- Acquisition of genetic and epigenetic changes.
- Chromosomal abnormalities and TP53 mutation.

MORPHOLOGY

- Distal third.
- Early: flat or raised patches
- Later: exophytic infiltrative masses

- Microscopy:
- Forms glands and mucin.

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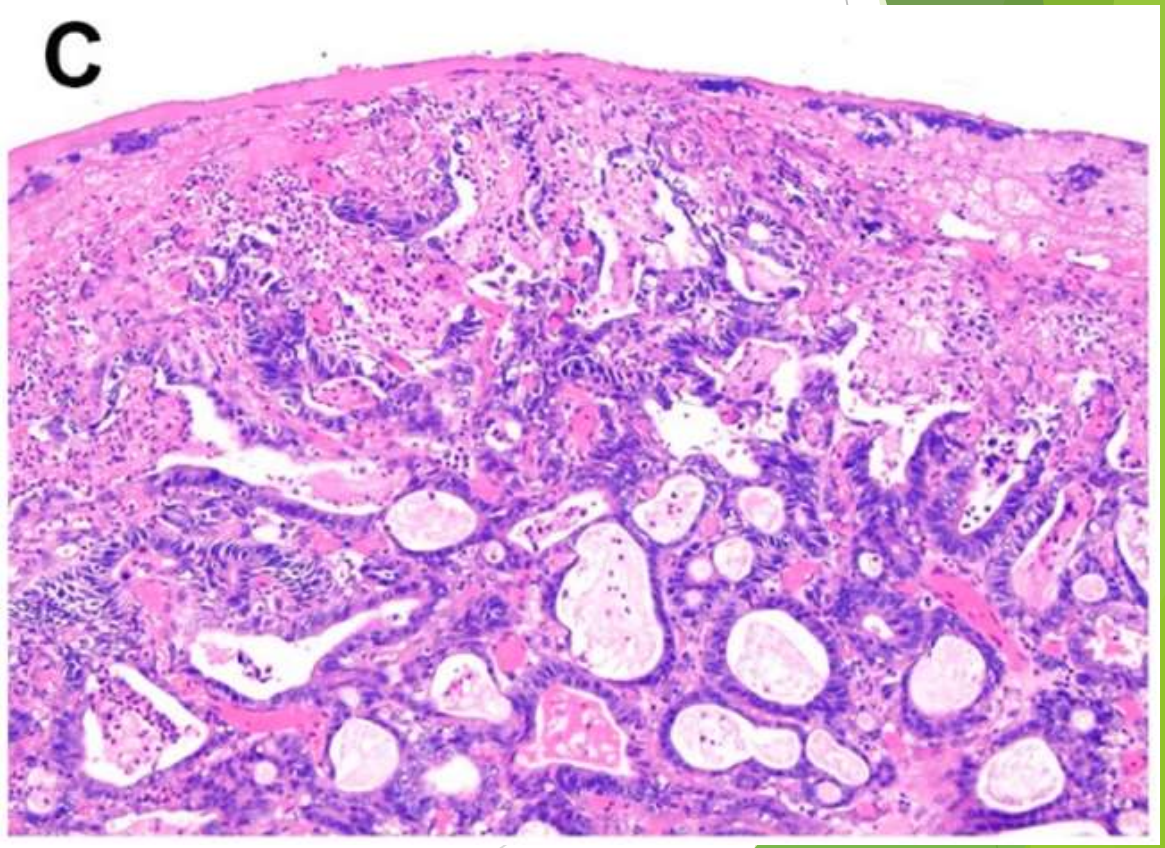


Clinical Features

- Pain or difficulty swallowing
- Progressive weight loss
- Chest pain
- Vomiting.
- Advanced stage at diagnosis: 5-year survival <25%.
- Early stage: 5-year survival 80%

Squamous Cell Carcinoma

- Male : female (4:1)
- Underdeveloped countries.

- **Risk factors:**
- Alcohol
- Tobacco use
- Poverty
- Caustic injury
- Achalasia .
- Plummer-Vinson syndrome
- Frequent consumption of very hot beverages
- Previous radiation Tx .

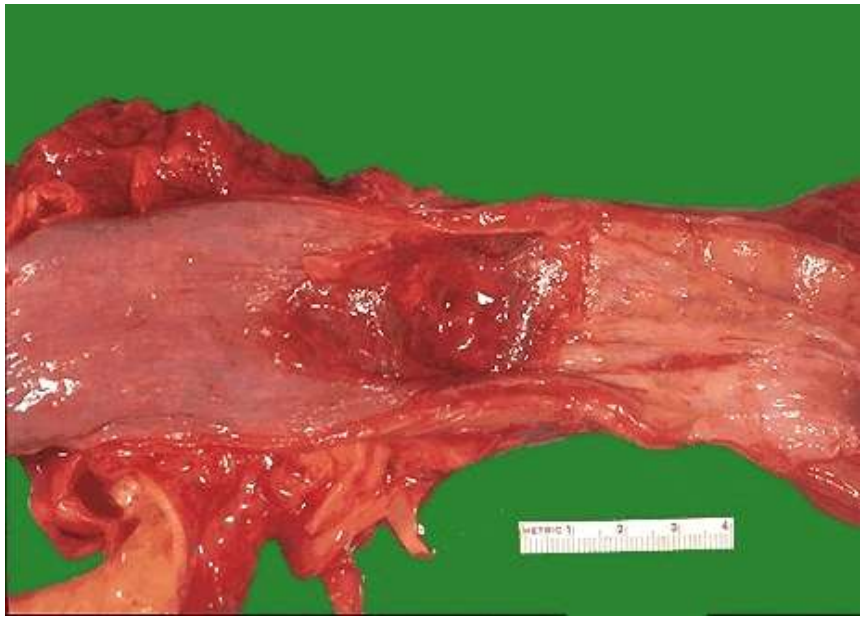
Pathogenesis

- In western : alcohol and tobacco use.
- Other areas: polycyclic hydrocarbons, nitrosamines, fungus-contaminated foods
- HPV infection implemented in high risk regions.

MORPHOLOGY

- Middle third (50% of cases)
- Polypoid, ulcerated, or infiltrative.
- Wall thickening, lumen narrowing
- Invade surrounding structures (bronchi, mediastinum, pericardium, aorta).

Mid esophagus



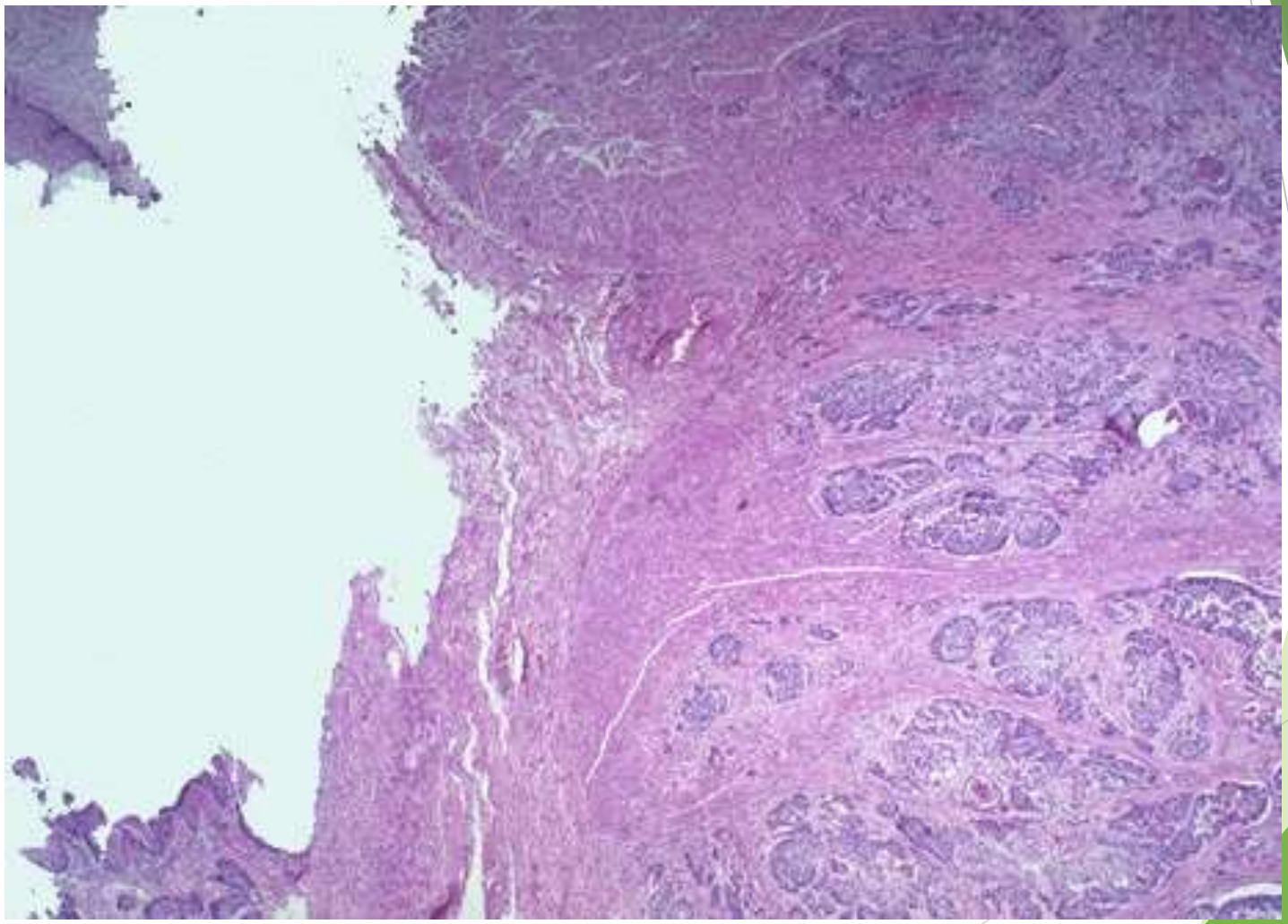
Microscopy:

- Pre-invasive: Squamous dysplasia & CIS.
- Well to moderately differentiated invasive SCC.
- Intramural tumor nodules
- Lymph node metastases :
- Upper 1/3: cervical LNs
- Middle 1/3: mediastinalparatracheal, and tracheobronchial LNs.
- Lower 1/3: gastric and celiac LNs.

Clinical Features

- Dysphagia
- Odynophagia
- Obstruction
- Weight loss and debilitation
- Impaired nutrition & tumor associated cachexia
- Hemorrhage and sepsis if ulcerated.
- Aspiration via a tracheoesophageal fistula
- Dismal Px: 5 year survival <9%

Invasive SCC



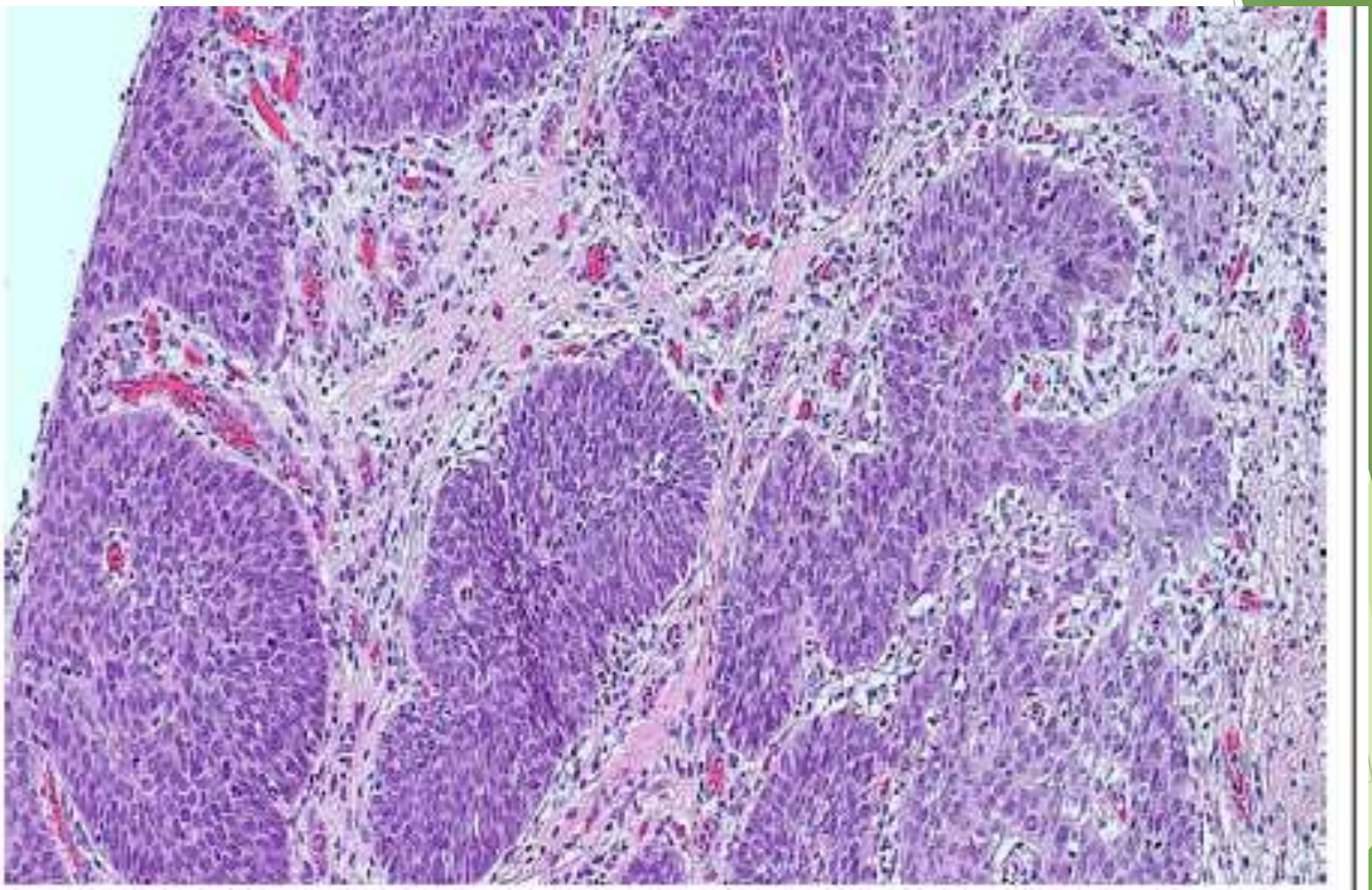


Figure 4: Squamous cell carcinoma of the esophagus with focal invasion into the muscularis mucosa and associated desmoplastic response.