Diseases of esophagus
Esophageal Obstruction

• Structural (mechanical) obstruction

• Functional obstruction (disruption of the coordinated waves of peristaltic contractions)
  -------- Esophageal dysmotility
• Esophageal dysmotility:
  - Nutcracker esophagus:
  - Diffuse esophageal spasm:
    - Lower esophageal sphincter dysfunction/hypertensive lower esophageal sphincter
• **Nutcracker esophagus:**

- High-amplitude contractions of the distal esophagus—contractions proceed in a coordinated manner
- dysphagia, chest pain
What is Nutcracker Esophagus or Hypertensive Peristalsis?

It is a benign condition and one of the motility disorders of the esophagus where the patient has contractions in the smooth muscles of the esophagus, which occur for excessive duration or amplitude.

For More Information:
Visit: www.epainassist.com
esophageal motility study (esophageal manometry)

Esophageal motility studies involve pressure measurements of the esophagus after a patient takes a wet (fluid-containing) or dry (solid-containing) swallow. Measurements are usually taken at various points in the esophagus.
<table>
<thead>
<tr>
<th></th>
<th>Normal*</th>
<th>High</th>
<th>Low</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lower esophageal sphincter pressure (mm Hg)</td>
<td>24 ± 10</td>
<td>&gt;45</td>
<td>&lt;10</td>
</tr>
<tr>
<td>Contraction amplitude† (mm Hg)</td>
<td>99 ± 40</td>
<td>&gt;180</td>
<td>&lt;30</td>
</tr>
<tr>
<td>Contraction duration† (sec)</td>
<td>3.9 ± 0.9</td>
<td>&gt;7</td>
<td>&lt;1.3</td>
</tr>
</tbody>
</table>

*Mean ± standard deviation.
†Mean of measurements at 3 cm and 8 cm above the LES.
• Diagram of esophageal motility study in nutcracker esophagus: The disorder shows peristalsis with high-pressure esophageal contractions exceeding 180 mmHg and contractile waves with a long duration exceeding 6 sec.
• **Diffuse esophageal spasm:**
  - repetitive, simultaneous contractions of the distal esophageal smooth muscle — uncoordinated contractions of the esophagus, contractions that are of normal amplitude
  - Dysphagia, chest pain
• Esophageal manometry tracing demonstrates diffuse esophageal spasm. Note the multiple uncoordinated contractions in the third tracing from the distal esophagus.
• Corkscrew appearance of the esophagus, Barium swallow
Lower esophageal sphincter dysfunction:
- such as high resting pressure or incomplete relaxation.
- Termed hypertensive lower esophageal sphincter in absence of altered patterns of esophageal contraction that seen in disorders described above.
esophageal dysmotility may result in development of diverticulae:

- **Epiphrenic diverticulum** (above the lower esophageal sphincter)

- **Zenker diverticulum/pharyngoesophageal diverticulum** (above the upper esophageal sphincter) if large ---- accumulation of food ------ mass, regurgitation and halitosis.
Mechanical obstruction:
presents as progressive dysphagia that begins with inability to swallow solids - liquids.
can be caused by:
1- strictures/stenosis (chronic gastro-esophageal reflux, irradiation, caustic injury, cancer)
2- Cancer
3- Esophageal mucosal webs:
   - idiopathic
   - Paterson-Brown-Kelly or Plummer-Vinson syndrome -- webs with iron-deficiency anemia, glossitis, and cheilosis
   - gastroesophageal reflux
   - chronic graft-versus-host disease
   - blistering skin diseases
4- Esophageal rings, or Schatzki rings
Achalasia

- It is an esophageal motility disorder.
- characterized by the triad of:
  1- incomplete LES relaxation upon swallow (<75%),
  2- increased LES tone ( >100 mm),
  3- aperistalsis of the esophagus.

Measured by Manometry ( esophageal motility study)

- Result in functional esophageal obstruction
- Symptoms: dysphagia, chest pain
- Bird's beak" appearance and "megaesophagus," typical in achalasia
Primary achalasia:
- The cause is unknown
- is the result of distal esophageal **inhibitory ganglion cell degeneration**.
- Degenerative changes in the extraesophageal vagus nerve or the dorsal motor nucleus of the vagus may also occur.

Secondary achalasia
- may arise in **Chagas disease**, in which Trypanosoma cruzi infection causes destruction of the myenteric plexus.
- Other causes: diabetic autonomic neuropathy; infiltrative disorders such as malignancy, amyloidosis, or sarcoidosis; lesions of dorsal motor nuclei, particularly polio or surgical ablation; may also be driven by immune-mediated destruction of inhibitory esophageal neurons
• Treatment modalities for both primary and secondary achalasia aim to overcome the obstruction, and include laparoscopic myotomy and pneumatic balloon dilatation. Botulinum neurotoxin (Botox) injection, to inhibit LES cholinergic neurons, can also be effective.
Lacerations

Mallory-Weiss tears:
- mucosal tears at the gastroesophageal junction
- Most often associated with severe vomiting secondary to acute alcohol intoxication

Boerhaave syndrome:
transmural tear and rupture of the distal esophagus----- lethal
Esophagitis

- Chemical and Infectious Esophagitis
- Reflux Esophagitis
- Eosinophilic Esophagitis
Reflux Esophagitis

- Reflux of gastric contents into the lower esophagus is the most frequent cause of esophagitis.
- The associated clinical condition is termed gastroesophageal reflux disease (GERD).

- Pathogenesis:
  - The most common cause of gastroesophageal reflux is **transient lower esophageal sphincter relaxation mediated via vagal pathways**.
  - Conditions that decrease lower esophageal sphincter tone or increase abdominal pressure and contribute to GERD include: alcohol and tobacco use, obesity, central nervous system depressants, pregnancy, **hiatal hernia**, delayed gastric emptying, increased gastric volume, coughing, and straining.
  - Reflux of gastric juices is central to the development of mucosal injury in GERD. In severe cases, reflux of bile from the duodenum may exacerbate the damage.
Figure 17-5 Esophagitis. A, Reflux esophagitis with scattered intraepithelial eosinophils and mild basal zone expansion. B, Eosinophilic esophagitis is characterized by numerous intraepithelial eosinophils. Abnormal squamous maturation is also apparent.
• Clinical Features:
- GERD is most common in individuals older than age 40 but also occurs in infants and children.
- The most frequent clinical symptoms are heartburn, dysphagia, and regurgitation of sour-tasting gastric contents.
- chest pain
- Complications: ulceration, hematemesis, melena, stricture development, and Barrett esophagus.
- Treatment: proton pump inhibitors, H2 histamine receptor antagonists
• Hiatal hernia:
  - can give rise to symptoms, such as heartburn and regurgitation of gastric juices, **that are similar to those of GERD**.
  - It is characterized by separation of the diaphragmatic crura and protrusion of the stomach into the thorax through the resulting gap.
  - Congenital hiatal hernias are recognized in infants and children, but many are acquired in later life.
Esophageal Varices

• Venous blood from the GI tract passes through the liver, via the portal vein, before returning to the heart.
• Diseases that impede this flow cause portal hypertension and can lead to the development of esophageal varices
• Pathogenesis:
- Portal hypertension results in the development of **collateral channels** at sites where the portal and caval systems communicate.
- These collateral veins allow some drainage to occur, but at the same time they lead to development of congested subepithelial and submucosal venous plexi within the distal esophagus and proximal stomach. These vessels, termed **varices**
- Develop in the vast majority of **cirrhotic patients**, most commonly in association with alcoholic liver disease. Worldwide, hepatic schistosomiasis is the second most common cause.
Figure 17-6 Esophageal varices. A, Although no longer used as a diagnostic approach, this angiogram demonstrates several tortuous esophageal varices. B, Collapsed varices are present in this postmortem specimen corresponding to the angiogram in A. The polypoid areas represent previous sites of variceal hemorrhage that have been ligated with bands. C, Dilated varices beneath intact squamous mucosa.
Clinical Features:
- 25-40% of patients with cirrhosis develop variceal bleeding.
- **Variceal hemorrhage is an emergency** that can be treated medically by inducing splanchnic vasoconstriction or endoscopically by sclerotherapy (injection of thrombotic agents), balloon tamponade, or ligation.
- Despite these interventions, 30% or more of patients with variceal hemorrhage die as a direct consequence of hemorrhage such as hypovolemic shock, hepatic coma, or other complications.
- more than 50% of patients who survive a first variceal bleed have recurrent hemorrhage within 1 year, and this carries a mortality rate similar to that of the first episode.
• Risk factors for hemorrhage, including:
  large varices
  Elevated hepatic venous pressure gradient
  previous bleeding
  advanced liver disease

These patients treated prophylactically with beta-blockers to reduce portal blood flow and with endoscopic variceal ligation.

it is important to recognize that cirrhosis patients with small varices that have never bled are at relatively low risk for bleeding and death.
Barrett Esophagus

• Barrett esophagus is a complication of chronic GERD that is characterized by intestinal metaplasia within the esophageal squamous mucosa.
• occur in as many as 10% of individuals with symptomatic GERD.
• most common in white males and typically presents between 40 and 60 years of age.
• The greatest concern in Barrett esophagus is that it confers an increased risk of esophageal adenocarcinoma.
• The presence of dysplasia, a preinvasive change, is associated with prolonged symptoms, longer segment length, increased patient age, and Caucasian race.
Figure 17-7 Barrett esophagus. **A**, Normal gastroesophageal junction. **B**, Barrett esophagus. Note the small islands of residual pale squamous mucosa within the Barrett mucosa. **C**, Histologic appearance of the gastroesophageal junction in Barrett esophagus. Note the transition between esophageal squamous mucosa (left) and Barrett metaplasia, with abundant metaplastic goblet cells (right).
Figure 17-8 Dysplasia in Barrett esophagus. A, Abrupt transition from Barrett metaplasia to low-grade dysplasia (arrow). Note the nuclear stratification and hyperchromasia. B, Architectural irregularities, including gland-within-gland, or cribriform, profiles in high-grade dysplasia.
• Clinical Features:

Barrett esophagus can only be identified thorough endoscopy and biopsy, which are usually prompted by GERD symptoms.
Esophageal Tumors

• The vast majority of esophageal cancers fall into one of two types:
  1- adenocarcinoma
  2- squamous cell carcinoma

Squamous cell carcinoma is more common worldwide.
Adenocarcinoma

- Most esophageal adenocarcinomas arise from Barrett esophagus.
- increased rates of esophageal adenocarcinoma

**risk factors:**
- gastroesophageal reflux/Barrett esophagus
- tobacco use
- Exposure to radiation

risk is reduced by:
- diets rich in fresh fruits and vegetables.
- Some serotypes of Helicobacter pylori
• occurs most frequently in Caucasians and shows a strong gender bias, being sevenfold more common in men.

• **Pathogenesis**: Molecular studies suggest that the progression of Barrett esophagus to adenocarcinoma occurs over an extended period through the stepwise acquisition of genetic and epigenetic changes.
Clinical Features:
pain or difficulty in swallowing, progressive weight loss, hematemesis, chest pain, or vomiting. Occasionally discovered in evaluation of GERD or surveillance of Barrett esophagus.

As a result of the advanced stage at diagnosis, overall 5-year survival is less than 25%.
Squamous Cell Carcinoma

- Risk factors include:
  - alcohol and tobacco use (polycyclic hydrocarbons, nitrosamines)
  - Poverty
  - caustic esophageal injury
  - Achalasia
  - Plummer-Vinson syndrome
  - diets that are deficient in fruits or vegetables
  - frequent consumption of very hot beverages
  - Previous radiation to the mediastinum
  - HPV infection has also been implicated in esophageal squamous cell carcinoma in high-risk areas
  - Fungus contaminated foods

Esophageal squamous cell carcinoma is nearly eight-fold more common in African Americans than Caucasians, a striking risk disparity that reflects differences in rates of alcohol and tobacco use as well as other poorly understood factors.

It occurs in adults older than age 45 and affects males four times more frequently than females.
Clinical Features:
- The onset of esophageal squamous cell carcinoma is insidious and it most commonly presents with dysphagia, odynophagia (pain on swallowing), or obstruction.
- Weight loss
- Hemorrhage and sepsis may accompany tumor ulceration,
- Occasionally, the first symptoms are caused by aspiration of food via a tracheoesophageal fistula.
- The overall 5-year survival rate in the United States remains less than 20%, and varies by tumor stage and patient age, race, and gender.
Figure 17-9 Esophageal cancer. A, Adenocarcinoma usually occurs distally and, as in this case, often involves the gastric cardia. B, Squamous cell carcinoma is most frequently found in the mid-esophagus, where it commonly causes strictures.
Figure 17-10 Esophageal cancer. A, Esophageal adenocarcinoma organized into back-to-back glands. B, Squamous cell carcinoma composed of nests of malignant cells that partially recapitulate the organization of squamous epithelium.