Diseases of mediastinum
ACHALASIA

A primary esophageal motility disorder characterized by the absence of esophageal peristalsis and impaired relaxation of the lower esophageal sphincter (LES) in response to swallowing.

Pathophysiology:
- LES pressure and relaxation are regulated by excitatory (e.g., acetylcholine, substance P) and inhibitory (e.g., nitric oxide, vasoactive intestinal peptide) neurotransmitters. Persons with achalasia lack nonadrenergic, noncholinergic, inhibitory ganglion cells, causing an imbalance in excitatory and inhibitory neurotransmission.
# Signs and symptoms

- Dysphagia (most common)
- Regurgitation
- Chest pain
- Heartburn
- Weight loss

## Eckardt score, 1992

<table>
<thead>
<tr>
<th>Weight loss (kg)</th>
<th>Dysphagia</th>
<th>Retrosternal pain</th>
<th>Regurgitation</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>1</td>
<td>Occasional</td>
<td>Occasional</td>
<td>Occasional</td>
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<tr>
<td>2</td>
<td>Daily</td>
<td>Daily</td>
<td>Daily</td>
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<tr>
<td>3</td>
<td>Each meal</td>
<td>Each meal</td>
<td>Each meal</td>
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</tbody>
</table>

## Treatment success evaluation

<table>
<thead>
<tr>
<th>Score</th>
<th>Symptom</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Remission</td>
</tr>
<tr>
<td>I</td>
<td>Remission</td>
</tr>
<tr>
<td>II</td>
<td>Treatment failure</td>
</tr>
<tr>
<td>III</td>
<td>Treatment failure</td>
</tr>
</tbody>
</table>
WORKUP

- **CHEST X-RAY**
- **ENDOSCOPY**
- **CT**
- **ESOPHAGEAL PRESSURE TOPOGRAPHY**
Based on dilation of esophagus on esophagram

- Grade 1: <4 cm
- Grade 2: 4-6 cm
- Grade 3: >6 cm
WORKUP

- CHEST X-RAY
- ENDOSCOPY
- CT
- ESOPHAGEAL PRESSURE TOPOGRAPHY

TREATMENT APPROACHES

- **Drugs**
  - Atropin, ganglioblockers, calcium channel blockers, nitrates (not perfect)
- **Intrasphincteric injection of botulinum toxin**
- **Pneumatic dilatation**
- **Surgery:**
  - *Heller Procedure*,
    - Laparoscopy/laparotomy
  - Esophagus extirpation with plastics.
INTRASPHINCTERIC INJECTION OF BOTULINUM TOXIN
POEM: Peroral endoscopic myotomy
Pharmacotherapy plays a very limited role in the treatment of achalasic patients and should be used in very early stages of the disease, temporarily prior to more definitive treatments, or for patients who fail or are not candidates for other treatment modalities (++++, strong).

Laparoscopic myotomy with partial fundoplication provides superior and longer-lasting symptom relief with low morbidity for patients with achalasia compared with other treatment modalities and should be considered the procedure of choice to treat achalasia. (++++, strong).

- The optimal type of fundoplication is debated (posterior vs. anterior), but partial fundoplication should be favored over total fundoplication, as it is associated with decreased dysphagia rates and similar reflux control (++, weak).
- The length of the esophageal myotomy should be at least 4 cm on the esophagus and 1-2 cm on the stomach (+, weak).

Botulinum toxin injection can be administered safely, but its effectiveness is limited especially in the long term. It should be reserved for patients who are poor candidates for other more effective treatment options such as surgery or dilation (++++, strong).

Endoscopic dilation is the most effective for dysphagia relief in patients with achalasia but is also associated with the highest risk of complications. It should be considered in selected patients who refuse surgery or are poor operative candidates (++++, strong).

Esophageal stents cannot be recommended for the treatment of achalasia (++, strong).
Hiatal hernia (HH) is a protrusion of any abdominal structure other than the esophagus into the thoracic cavity through a widening of the hiatus of the diaphragm.

Cephalad migration of the gastroesophageal junction may result from weakening of the phrenoesophageal ligament. Depletion of elastin fibers leads to stretching of the ligament and proximal displacement of the gastroesophageal junction. Most cases of hiatal hernia are acquired rather than congenital, though familial clustering has been reported and in a very small number of cases, multifactorial inheritance may play a part.
Hiatal hernias are more common in Western countries. The frequency of hiatus hernia increases with age, from 10% in patients younger than 40 years to 70% in patients older than 70 years.

- 50% - asymptomatic,
- 25% - need PPI
- 15% - need surgery.

GERD – typical complication

- USA, EU – 10-20% have GERD, 80% of them have decreased life quality


Main surgical aspects:

- Big and giant HH:
  - Best type of procedure is not established,
  - high frequency of unsatisfactory results.

- Severe complications need urgent surgery:
  - Cameron lesion with bleeding or stricture,
    - severe anemia, which is refractory for treatment.
  - Gastric volvulus and incarceration lead to necrosis.
CLASSIFICATION

- IDC – 10:
  - K44.0 DIAPHRAGMATIC HERNIA WITH OBSTRUCTION, WITHOUT GANGRENE
  - K44.1 DIAPHRAGMATIC HERNIA WITH GANGRENE
  - K44.9 DIAPHRAGMATIC HERNIA WITHOUT OBSTRUCTION OR GANGRENE
**SAGES CLASSIFICATION**

- **Type I hernias (95%)** are sliding hiatal hernias, where the gastroesophageal junction migrates above the diaphragm. The stomach remains in its usual longitudinal alignment and the fundus remains below the gastroesophageal junction.
  - GERD (and Barrett’s esophagus) is common

- **Type II hernias** are pure paraesophageal hernias (PEH); the gastroesophageal junction remains in its normal anatomic position but a portion of the fundus herniates through the diaphragmatic hiatus adjacent to the esophagus.

- **Type III hernias** are a combination of Types I and II, with both the gastroesophageal junction and the fundus herniating through the hiatus. The fundus lies above the gastroesophageal junction.

- **Type IV hiatal hernias** are characterized by the presence of a structure other than stomach, such as the omentum, colon or small bowel within the hernia sac.

- **Types II-IV- paraesophageal hernias** (III - 90%, II - uncommon),
  - Have hernia sac,
  - Phrenoesophageal ligament is relatively intact.

- **Giant HH** (III and IV types) - ½ of stomach or more protrudes to mediastinum.
**DIAGNOSIS**

- Basic for surgery:
  - Chest X-ray,
  - EGDS
  - X-ray scopy with BaSo₄ (better than EGDS for size evaluation)
- CT – if deeper investigation needed only (e.g. Gastric volvulus; Type IV hernia).

- **GL SAGES:** Hiatal hernia can be diagnosed by various modalities. Only investigations which will alter the clinical management of the patient should be performed (+++-strong).
TYPE I HH

- Evaluation of esophageal and gastric mucosa
  - Esophagitis, Barrett’s esophagus;
- Size and type of hernia (X-ray is better)
TYPE II-IV HH

- Evaluation of esophageal and gastric mucosa
  - Esophagitis, Barrett’s esophagus, ulcer lesions;
- Size and type of hernia (X-ray is better)
Plain chest radiographs may identify soft tissue opacity with or without an air fluid level within the chest. A retrocardiac air-fluid level on chest x-ray is pathognomonic for a paraesophageal hiatal hernia. Visceral gas may be seen in cases of intestinal herniation.
CONTRAST STUDIES

- **Barium** is the contrast agent most frequently reported in the literature as used for this purpose.

- Given the increased aspiration risk of patients with paraesophageal hernias presenting with acute gastric outlet obstruction, **ionic water soluble contrast** should be generally avoided due to the risk of aspiration pneumonitis.

- Size and mobility of HH,

- Cardia localization and presence of short esophagus
COMPUTED TOMOGRAPHY

- May be useful in an urgent situation for patients with suspected complications from a volvulized paraesophageal hernia. The hernia site and any herniated organs within the chest cavity are clearly visualized in most cases.
- Also useful for recurrent HH, in case of mesh migration.
**ADDITIONAL METHODS**

- *Esophageal manometry* can demonstrate the level of the diaphragmatic crura, the respiratory inversion point and the location of the lower esophageal sphincter.

- **pH testing** has limited relevance in the diagnosis of a hiatal hernia, but is critical to identify the presence of increased esophageal acid exposure in patients with sliding hiatal hernias that might benefit from antireflux surgery.
INDICATIONS FOR SURGERY (SAGES)

• Repair of a type I hernia in the absence of reflux disease is not necessary (+++, strong)

• Surgery only indicated if:
  1. No effect from PPI;
  2. Constant need in PPI;
  3. PPI are contraindicated.

• All symptomatic paraesophageal hiatal hernias should be repaired (++++, strong), particularly those with acute obstructive symptoms or which have undergone volvulus.
ACCESS TYPE

- **Laparoscopic hiatal hernia repair** is as effective as open transabdominal repair, with a reduced rate of perioperative morbidity and with shorter hospital stays. It is the preferred approach for the majority of hiatal hernias (+++++, strong)

- **Open conversion** is occasionally necessary for reasons such as bleeding, splenic injury or dense adhesions, and it is important that surgeons taking these on as laparoscopic procedures are comfortable with an open repair should conversion become necessary.

- **Open conversion – urgent surgery:**
  - Laparotomy of left-side thoracotomy;
  - Also useful for evaluation of gastric necrosis size in case of volvulus.
Heller procedure (laparoscopy)
MESH USAGE

- The **use of mesh** for reinforcement of large hiatal hernia repairs leads to decreased short term recurrence rates (**+++**, **strong**)
- There is inadequate long-term data on which to base a recommendation **either for or against** the use of mesh at the hiatus
URGENT SURGERY

INDICATIONS:
- volvulus,
- incarceration,
- bleeding
GASTRIC VOLVULUS

• Rare complication for III and IV types of HH
• Can occur in abdominal or in thoracic cavity

**Borchardt triad:**
• severe sudden epigastric pain
• intractable retching without vomiting
• inability to pass a nasogastric tube

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Paraesophageal Hernia and Gastric Volvulus
K. Conley Coleman and Daniel Grabo

• **Indications for urgent surgery:**
  • Signs of obstruction
  • Bleeding, perforation
  • Respiratory distress

• **Procedures (GL SAGES: ++++, strong):**
  • Laparoscopy should be performed first, open conversion—in case of technical difficulties.
HH IN HIGH RISK PATIENTS

- Hernia reduction with **gastropexy alone** and no hiatal repair may be a safe alternative in high-risk patients but may be associated with high recurrence rates (++, weak). Formal repair is preferred (+++, strong).

- **Crurorrhaphy** is better than gastropexy alone.
ACUTE MEDIASTINITIS
Mediastinum extends from the posterior aspect of the sternum to the anterior surface of the vertebral bodies and includes the paravertebral sulci. The mediastinum is limited bilaterally by the mediastinal parietal pleura and extends from the diaphragm inferiorly to the level of the thoracic inlet superiorly.

- **Mediastinum contents:**
  - Superior: aortic arch and great arteries, right and left brachiocephalic veins, left superior intercostal vein, upper half of superior vena cava, phrenic, vagus and cardiac nerves, cardiac plexus, left recurrent laryngeal nerve, lymph nodes brachiocephalic, tracheobranchial, paratracheal, trachea, esophagus, thoracic duct, thymus.
  - Inferior:
    - Anterior: thymus, lymphnodes and may contain the portions of a retrosternal thyroid;
    - Middle: pericardium, heart, great vessels joining the heart, ascending aorta, pulmonary trunk, right pulmonary artery, left pulmonary artery, the lower half of the superior vena cava, both main bronchi, phrenic nerves, cardiac plexus, tracheobranchial lymph nodes.
    - Posterior: esophagus, descending aorta, thoracic trunk, thoracic spinal ganglion, sympathetic trunk and major branches, rami communicantes of intercostal nerves, phrenic nerves, vagus nerve.
  - Organs are surrounded with connective fatty tissue.
ACUTE MEDIASTINITIS

- **Mediastinitis** — acute infectious inflammation of mediastinal connetive tissue. Mortality 10-67%.

- **Main causes**
  - Dissemination of oropharyngeal infections
  - Esophageal perforation,
  - Postoperative (after sternotomy).

- **Rare causes:**
  - Pleural empyema, pancreatitis, subdiaphragmatic abscess, osteomyelitis of breastbone, hematogenic infections.

- **Classification**
  1. **Origin:** primary/secondary
  2. **Inflammation type:** abscess, phlegmon.
  3. **Localization:** upper, lower (anterior and posterior), total.
  4. **Type of exudate:** serous, purulent, ichorous, etc.
  5. **Causative agent:** Staphylococcus spp., E.coli, Klebsiella spp., etc.
  6. **Clinical course type:** acute, chronic.
  7. **Complications type:** pleural effusion/empyema, pericarditis, osteomyelitis, etc.
SIGNS AND SYMPTOMS

- **Depends on:**
  - localization of primary infection focus
  - quickness of disease progression

- **Typical but NOT specific signs:**
  - Tachycardia and hyperthermia
    - fulminant form – septic shock, death during 24-72 hours.

- **Chest pain – common but not constant sign:**
  - Anterior localization – behind breastbone, increases during head tilting,
  - Posterior localization – deep pain between shoulder blades, increases during processes spinous percussion, long back muscles rigidity
  - Superior localization – the same as anterior but with irradiation to upper extremities
  - Pain during swallowing and breathing – involvement of mediastinal pleura.

- **Dysphagia, crepttation in the upper thoracic aperture – esophagus perforation.**
- **Dyspnea – Compression of trachea and bronchi.**
- **Voice changes – recurrence nerves involvement.**
- **Constant hiccup – phrenic nerve involvement.**
- **Horner’s syndrome (ptosis, miosis, enophthalmos) – sympathetic trunk involvement.**
- **Face/neck cyanosis, eye noise, neck veins dilation** – compression of upper cava vein.
WORKUP

- Life history – not significant, but attention should be paid to:
  - Oropharyngeal infections,
  - Medical procedures on esophagus,
  - Cardiosurgery procedures.

- Lab test - not significant, but
  - Pay attention to WBC count and CRP

- Visualization methods are the most informative, especially:
  - Plain chest X-ray
  - CT with intravenous enhancement.

- For proper early antibiotic treatment:
  - Microbiology tests for exudate
  - Blood microbiology tests (can be even MORE informative)
PLAIN CHEST X-RAY

- enlargement of mediastinum (relatively often)
- pneumomediastinum (relatively rare)
COMPUTED TOMOGRAPHY

- Mediastinum enlargement (100%)
- Free air (57.5%)
- Fluid in mediastinum (55%)
- Pleural effusion (85%)
- Hydropericardium (27.5%)
Findings from nuclear medicine scans/radioactive WBC scans involving labeled WBCs are reported to have very high specificity; however, few studies have been performed on postoperative patients.
ANATOMY BACKGROUND: SPACES OF THE NECK

1. Visceral space
2. Carotid space
3. Retropharyngeal space
4. Posterior cervical space
5. Perivertebral space
ODONTOGENIC DESCENDING NECROTIZING MEDIASTINITIS

- Ludwig’s angina (progressive submaxillary cellulitis of the floor of the mouth) with dissemination to superior mediastinum
  - As a rule visceral, carotid and posterior cervical spaces are affected
- Before antibiotics invention ~ 30% of all mediastinitis, nowadays ~5%

**Presentation (K.R. Cardenas-Malta et al. 2005):**
- High body temperature (100%),
- Dysphagia (80%),
- Dyspnoe (80%),
- Chest pain (60%),
- Orthopnoe (40%),
- Tachycardia (20%).

**In case of anaerobic causative agent:**
- Gas production with emphysema and compression (upper v. cava syndrome)
- Destruction of visceral pleura and empyema

**Types of clinical course:**
- Acute mediastinitis (infection spreads gradually):
  - First focus of infection (tooth mainly) – floor of the mouth – deep neck spaces – superior mediastinum
- Fulminant mediastinitis:
  - Quick spread of infection to mediastinal and neck spaces
  - Very rapid deterioration with septic shock.
DESCENDING NECROTIZING MEDIASTINITIS FROM PRIMARY PHARYNGEAL FOCUS

- **Peritonsillar abscess** - infection spreads to visceral or retropharyngeal spaces:
  - Dull voice.
  - Severe pain in the throat, unilateral, especially during swallowing
  - Trismus
  - Enlargement of pharyngeal wall with dislocation of uvula
  - Edema under the lower jaw
SURGERY IN DESCENDING NECROTIZING MEDIASTINITIS

- Debridement, sanation and control over infection source
  - Superior mediastinitis – transcevical access and drainage,
    - Videoassisted thoracoscopy (VATS);
  - Below carina tracheae – cervical and transthoracic access and drainage
- Surgery must follow the established diagnosis during 24 hours
SURGERY IN DESCENDING NECROTIZING MEDIASTINITIS
Boerhaave's syndrome

In 1724, Hermann Boerhaave, a Dutch physician and professor of clinical medicine, first described spontaneous rupture of the esophagus, which typically occurs after forceful emesis.

It occurs in 15-20% of all esophageal perforations

- **Mackler triade:**
  - Vomiting with recently eaten food
  - Subcutaneous emphysema,
  - Severe cutting pain in the chest or epigastrium. It appears during vomiting and intensifies during swallowing

- **Pathogenesis:**
  - It is a result of a sudden rise in intraluminal esophageal pressure produced during vomiting
  - Transmural rupture.
  - The most common anatomic location is at the left posterolateral wall of the lower third of the esophagus, 2-3 (3-6) cm proximal to the gastroesophageal junction.
  - Chemical mediastinitis caused by gastric contents contaminating the mediastinal cavity,
  - Left-sided empyema is common due to damage of mediastinal pleura during rupture.
WORKUP

- Chest X-ray,
- Fluoroscopy with water soluble dye
- CT
- EGDS (in unclear cases ONLY)
Boerhaave's syndrome: SUGERY

- Vast infected area in the mediastinum causes severe patient condition including sepsis
  - Thoracotomy: defect suturing or resection and fistula; sanation, drainage
  - Gastrostomy: feeding and decompression
75% of perforations are iatrogenic:
- Balloon dilatation of strictures (risk of perforation 4-17%),
- Diagnostic EGDS – insufficient experience, tumors, diverticula, strictures, etc.
- Coagulation/sclerotherapy during hemostasis:
- Stent placement,
- Intubation of trachea; nasogastric tube placement,
- Foreign bodies evacuation.

Clinical signs can be both good manifested or not clear
- Pain
  - Cervical part perforations – neck pain, which accelerates with movements
  - Thoracic perforations – chest pain.
- High body temperature and tachycardia,
- Subcutaneous emphysema in aperture thoracica superior.

Pathogenesis:
- Small defects in strictures, which consist of sclerotic tissues,
- No aggressive gastric content,
- Infection focus locates in mediastinum. Mediastinal pleura is rarely affected.
SURGERY FOR TREATMENT OF ESOPHAGEAL PERFORATIONS

- **Stable patients with no severe symptoms. They have:**
  - Abscess, which drains into esophagus lumen
  - Small iatrogenic perforations, which were early diagnosed
    - Medical treatment with careful surveillance
      - Antibiotics
      - Nasogastric tube
      - Endoscopic placement of drainage to the cavity of lesion/VAC therapy.
POST-STERNOTOMY MEDIASTINITIS

- Post-sternotomy mediastinitis is one of severe complications in cardiosurgery:
  - post-sternotomy form of mediastinitis is the most common one in developed countries,
  - Majority appears after cardiac surgery:
    - After heart or lung transplantation – 2.5-7.5% of cases
    - After other cardiac surgical procedures – 0.5-4.5% of cases

- The definition of mediastinitis requires at least one of the following criteria:
  - Patient has organisms cultured from mediastinal tissue or fluid.
  - Patient has evidence of mediastinitis on gross anatomical or histopathological examination.
  - Fever (>38°C), chest pain or sternal instability.
    - And at least one of the following:
      - Purulent drainage from mediastinal area;
      - Mediastinal widening on imaging.

- Types of post-sternotomy mediastinitis:
  - superficial — dermis and subcutaneous tissue
  - deep — infection reaches under the sternum and the anterior mediastinum.
  - Causative agents – skin bacteria, Staphylococcus aureus nasal carriage.
POST-STERNOTOMY MEDIASTINITIS: TREATMENT TACTICS

• Surgical strategies
  • Open – revision with open dressings;
  • Closed – primary closure, closed irrigation;
  • Negative pressure wound therapy (NPWT; VAC) with changes of polyurethane foam every 48-72 hours – optimal method (IB). Reconstruction with vascularized soft tissue flaps (e.g. omentum, pectoral muscle) can be made afterwards.
**MEDIASTINITIS TREATMENT**

- **Prompt surgical management is the main principle of effective treatment**
  - Debridement and source control matter
  - Simultaneous work of surgical teams:
    - Mediastinum – thoracic surgeon
    - Head and neck – oral and maxillofacial surgeon or otolaryngologist
    - Esophagus perforation – general (abdominal) surgeon.

- **Empiric antibiotic therapy:**
  - Esophagus perforation/neck and odontogenic infections:
    - Causative agents – Staphylococcus spp., Anaerobes, Gram negative bacteria
    - Main groups:
      - Carbapenemes or piperacillin/tazobactam
      - Cephalosporines IV (Cefepime) + metronidazole/clindamycin.
    - Additionally:
      - Suspicious for MRSA – vancomycin or daptomycin.

- Post-sternotomy mediastinitis:
  - Causative agents – nosocomial bacteria (MRSA, Enterococcus spp., Gram negative bacteria);
    - Main groups (MRSA, Streptococcus, Enterococcus spp.)
      - Vancomycin or daptomycin.
    - Additionally (Gram negative bacteria incl. P. aeruginosa):
      - Carbapenemes or piperacillin/tazobactam
      - Cephalosporines IV (Cefepime) + metronidazole/clindamycin.
MORTALITY IN MEDIASTINITIS

- Esophagus perforation - 15-20%,
- Descending necrotizing mediastinitis - 10-20%,
- Post sternotomy mediastinitis – <5%.
  - **Early surgical intervention (<24 h) decreases mortality remarkably.**
    - Reconvalescents have increased death risk (59% during 10 years) in comparison with people, who did not suffer from this disease.