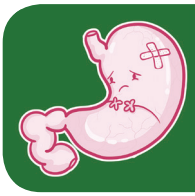

NOTES
ESOPHAGEAL DISEASE

GENERALLY, WHAT IS IT?

PATHOLOGY & CAUSES

- Pathologies of the esophagus
- Esophageal motility disorders
  - Diseases interfering with correct function of esophagus' various muscular components

CAUSES

- Infections, autoimmune disease, anatomical defects, irritative processes

SIGNS & SYMPTOMS

- Difficulty/pain while swallowing, especially spasm-type pain
- Difficulty with food regurgitation

DIAGNOSIS

- Individual history/clinical features, esophagogastroduodenoscopy (EGD), barium swallow X-ray, esophageal manometry, endoscopic biopsy

TREATMENT

- See individual diseases

ACHALASIA

osms.it/achalasia

PATHOLOGY & CAUSES

- Esophageal smooth muscle fibres fail to relax → lower esophageal sphincter remains closed/fails to open
- AKA esophageal achalasia, achalasia cardiae, cardiospasm, esophageal aperistalsis
- Progressive degeneration of ganglion cells in myenteric plexus within esophageal wall → lower esophageal sphincter fails to relax → loss of peristalsis in distal esophagus
- Involves smooth muscle layer of esophageal, lower esophageal sphincters
- Affected individual lacks nonadrenergic, noncholinergic, inhibitory ganglion cells → imbalanced excitation and relaxation → incomplete lower esophageal sphincter relaxation, increased lower esophageal tone, lack of esophageal peristalsis

CAUSES

- Likely caused by underlying autoimmune process triggered by previous viral infection/ genetic predisposition/ neurodegenerative disease/other infective process

Primary achalasia (most common)

- No known underlying cause → failure of distal esophageal inhibitory neurons

Secondary achalasia

- Esophageal cancer
- Chagas disease
  - Protozoan infection due to *Trypanosoma cruzi* → loss of intramural ganglion cells → aperistalsis, incomplete lower esophageal sphincter relaxation

SIGNS & SYMPTOMS

- Dysphagia to solids/liquids, odynophagia (rarely), heartburn unresponsive to proton pump inhibitor therapy, symptoms worsen progressivelys, regurgitation of undigested food, substernal chest pain, hiccups
- Weight loss
- Coughing while lying horizontally, aspiration of food → recurrent pulmonary complications

DIAGNOSIS

DIAGNOSTIC IMAGING

Barium swallow X-ray and continuous fluoroscopy

- Normal peristalsis not seen
- Acute tapering at lower esophageal sphincter
- Narrowing of gastroesophageal junction (bird's beak/rat's tail appearance)
- Dilated esophagus above narrowing
- Air-fluid margin over barium column due to lack of peristalsis

Esophageal endoscopy with or without endoscopic ultrasound

- May appear normal
- Unusually increased resistance to passage of endoscope through esophagogastric junction
- Retained food in esophagus on upper endoscopy

Endoscopic biopsy

- Hypertrophic musculature
- Absence of specific nerve cells within myenteric plexus

OTHER DIAGNOSTICS

Esophageal manometry

- Lower esophageal sphincter fails to relax upon wet swallow (< 75% relaxation)
- Lower esophageal pressure
  - Normal < 26mmHg
  - Achalasia > 100mmHg
  - Nutcracker achalasia > 200mmHg
- Aperistalsis in esophageal body
- Relative increase in intraesophageal pressure vs. intragastric pressure

TREATMENT

MEDICATIONS

- Calcium channel blockers for mild to moderate disease
- Nitrates effective before dilatation occurs
- Antimuscarinic agents (rarely effective)
- Proton pump inhibitors (after surgery/pneumatic dilatation) to prevent reflux damage

SURGERY

Laparoscopic Heller myotomy

- Esophageal dilatation via surgical cleaving of muscle
- Only cut through outer muscle layers (those failing to relax), leaving inner mucosal layer intact

Endoscopic myotomy

- Peroral endoscopic myotomy, minimally invasive → incision made through esophageal mucosa, innermost circular muscle layer divided and extended through lower esophageal sphincter, 2cm/0.8in into gastric muscle

OTHER INTERVENTIONS

- Eat slowly, chew well, drink plenty of water with meals, avoid eating near bedtime, raise head off bed when sleeping with pillows (promotes emptying of esophagus with gravity)
- Avoid foods that aggravate reflux → ketchup, citrus, chocolate, caffeine

Botox injection

- Paralyze muscle keeping lower esophageal sphincter shut (causes scarring of sphincter → may complicate later myotomy)

Pneumatic dilatation

- Muscle fibres stretched/torn by forceful inflation of balloon placed in lower esophageal sphincter
- Lowers basal lower esophageal tone by disruption of muscular ring

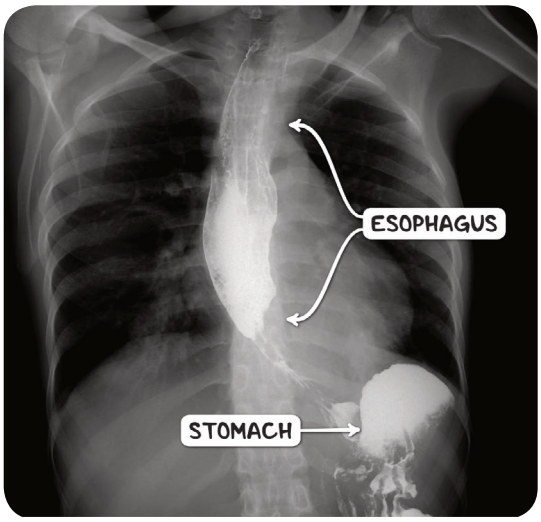

Figure 30.1 A barium swallow demonstrating the bird's beak sign in achalasia. The proximal esophagus is dilated.

BARRETT'S ESOPHAGUS

osms.it/barretts-esophagus

PATHOLOGY & CAUSES

- Premalignant condition; metaplasia of cells lining lower esophagus
- Normal stratified squamous epithelium → simple columnar epithelium, goblet cells (usually native to lower gastrointestinal tract)
- Chronic acid exposure → reflux esophagitis (chronic irritation) → metaplasia
- Bile acids → intestinal differentiation → promotes cancer growth

TYPES

- If z-line and gastroesophageal junction coincide → intestinal metaplasia at gastroesophageal junction
  - Associated with *Helicobacter pylori*

Long-segment Barrett's

- Distance between z-line and gastroesophageal junction > 3cm/1.2in
  - Associated with more severe reflux
  - Upright/supine reflux
  - Significantly more likely to develop adenocarcinoma

Short-segment Barrett's

- Distance between z-line and gastroesophageal junction < 3cm/1.2in
  - Greater prevalence
  - Shorter history of heartburn
  - Usually asymptomatic
  - Predominantly upright reflux
  - Less mucosa involved → lower incidence of dysplasia

RISK FACTORS

- Bulimia
- Central obesity
- Previous chemical damage to esophageal epithelium (e.g. swallowing lye)
- Smoking
- Hiatal hernia

COMPLICATIONS

- Esophageal adenocarcinoma

SIGNS & SYMPTOMS

- Often asymptomatic
- Same as reflux, not (initial) cancerous changes
- Frequent, prolonged heartburn, dysphagia, hematemesis, epigastric pain, weight loss (due to painful eating)

DIAGNOSIS

DIAGNOSTIC IMAGING

Esophagogastroduodenoscopy

- Fiber optic camera inserted via mouth → examine and biopsy esophagus, stomach, duodenum

LAB RESULTS

Biopsy

- Specimen from esophagogastroduodenoscopy must contain goblet cells → "intestinal metaplasia" → marker for progression of metaplasia to dysplasia → adenocarcinoma
- Immunohistochemical staining assists in diagnosis
- Biopsy classification
  - Nondysplastic
  - Low-grade dysplasia
  - High-grade dysplasia
  - Frank carcinoma

OTHER DIAGNOSTICS

Screening

- Biological males, > 60 years old, long standing reflux, life expectancy > five years
- Anyone with diagnosis of Barrett's esophagus

Esophageal pH studies

- Establish efficacy of proton pump inhibitor treatment

TREATMENT

MEDICATIONS

Proton pump inhibitors

- E.g. omeprazole; manage acid reflux

Chemoprevention

- Nondysplastic/low-grade lesion
  - Aspirin, NSAIDS → inhibition of cyclooxygenase (COX-1 & 2) may protect against progression of disease

SURGERY

Treatment of dysplastic lesions

- Endoscopic mucosal resection, surgical removal of esophagus, radiation therapy, systemic chemotherapy

OTHER INTERVENTIONS

Annual endoscopic observation

- For nondysplastic/low-grade lesions

Management of acid reflux

- Avoid/reduce intake of foods known to worsen reflux: chocolate, coffee, tea, peppermint, alcohol, fatty/spicy/acidic foods

Treatment of dysplastic lesions

- Radiofrequency ablation
  - Electrical current used to destroy small regions of tissue
- Spray cryotherapy
  - Liquid nitrogen spray applied to small region of tissue → freezing → tissue death
- Photodynamic therapy
  - Chemical photosensitizer → cytotoxicity when stimulated by certain frequency of light

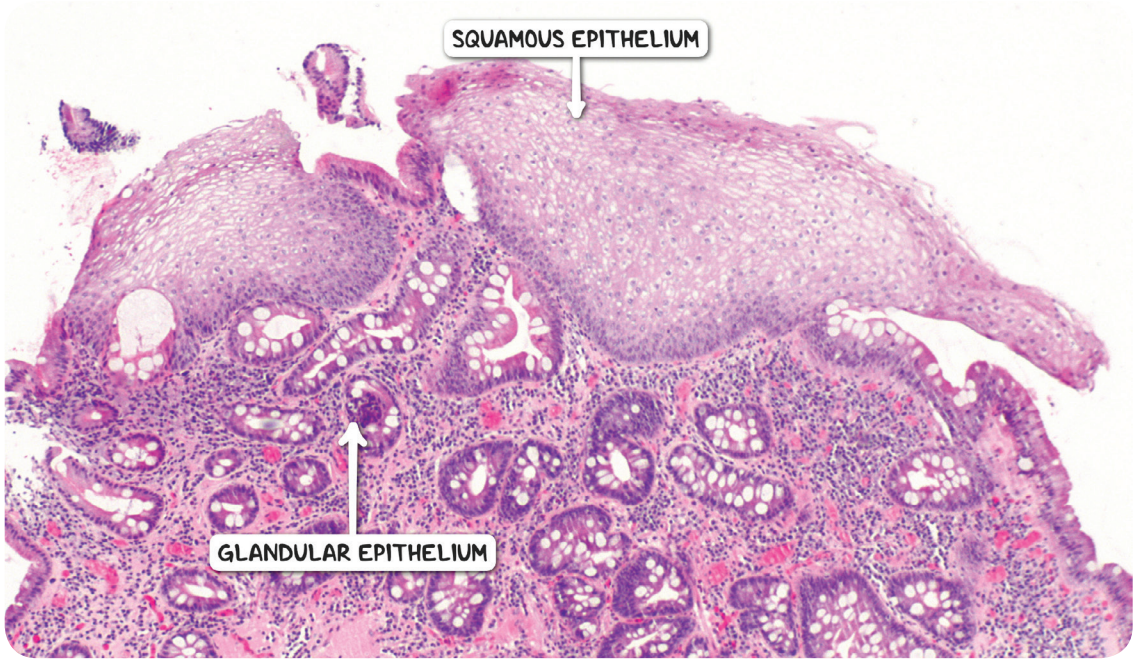

Figure 30.2 Histological appearance of the squamocolumnar junction in a case of Barrett's esophagus. The underlying glandular epithelium contains goblet cells, indicating intestinal metaplasia.

BOERHAAVE SYNDROME

osms.it/boerhaave-syndrome

PATHOLOGY & CAUSES

- Rupture through esophagus caused by increased intraesophageal pressure and negative intrathoracic pressure
- Vomiting / retching → unrelaxed esophagus, closed glottis → increase in esophageal pressure, slight drop in intrathoracic pressure → spontaneous rupture of esophageal wall → contamination of mediastinum with gastric contents → chemical mediastinitis
  - Tears commonly occur at left posterolateral aspect (distal esophagus), just above esophageal hiatus of diaphragm
  - Can be fatal without treatment → sepsis
  - Chemical mediastinitis → mediastinal necrosis → rupture of overlying pleura → contamination of pleural cavity → pleural effusion
  - Effort rupture of cervical esophagus → localized cervical perforation
  - Spread of contamination slow due to attachments of esophagus to prevertebral fascia
- Usually occurs in anatomically normal esophagi

RISK FACTORS

- Caustic ingestion, pill/medication esophagitis, eosinophilic esophagitis, Barrett's esophagus, infectious ulcers, stricture dilatation

SIGNS & SYMPTOMS

- Severe vomiting → profound retrosternal chest pain (may radiate to left shoulder) or abdominal pain
  - Followed by painful swallowing (odynophagia), tachypnea, dyspnea, cyanosis, fever, shock
- *Mackler's triad:* chest pain, vomiting, subcutaneous emphysema
- *Hamman's sign:* crunching/rasping sound, synchronous with heartbeat:
  - Heard over precordium, left lateral position
  - Caused by mediastinal emphysema
- *Cervical perforation:* neck pain, difficulty swallowing (dysphagia), difficulty speaking (dysphonia), tenderness of sternocleidomastoid
- *Intra-abdominal perforation:* epigastric pain (may radiate to left shoulder), back pain, inability to lie supine, acute abdomen pain

DIAGNOSIS

- Non-specific symptoms → diagnostic delay, poor outcome
- Physical examination often unhelpful; history important

DIAGNOSTIC IMAGING

Chest X-ray

- *Early:* free mediastinal air
- *Hours to days later:* pleural effusion, pneumothorax, widened mediastinum, subcutaneous emphysema

Chest CT scan

- Esophageal wall edema/thickening, extraesophageal air, periesophageal fluid, mediastinal widening, pneumothorax

Fluoroscopy

- Water soluble contrast (gastrografin) esophagram → location and extent of extravasation of contrast
- Barium sulfate common contrast material, but spillage into mediastinal and pleural spaces → inflammatory response → fibrosis

Endoscopy avoided

- May extend tear, introduce air into mediastinum

LAB RESULTS

- Hemoglobin and hematocrit
- Assess severity of initial bleeding
- Pleural effusion fluid may be high in amylase (saliva), low pH
- Leukocytosis

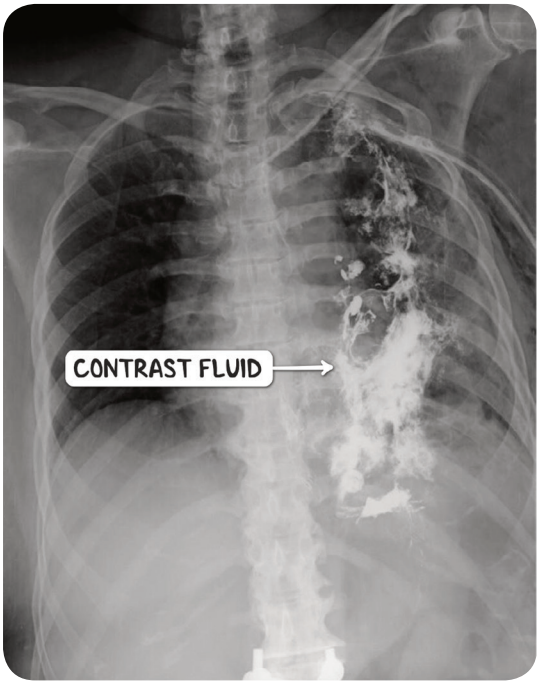

Figure 30.3 A contrast swallow in an individual with Boerhaave's syndrome. The contrast has leaked into and accumulated in the thoracic cavity.

TREATMENT

MEDICATIONS

- IV proton pump inhibitor → reduce acidity, irritation
- Prophylactic antibiotic therapy

SURGERY

- Debride infected/necrotic tissue, repair defect/resection of defect/diversion

OTHER INTERVENTIONS

- Parenteral/enteral (jejunostomy/PEG tube) nutritional support

DIFFUSE ESOPHAGEAL SPASM

osms.it/esophageal-spasm

PATHOLOGY & CAUSES

- Esophageal motility disorder characterized by repetitive, non-peristaltic, spontaneous contractions of the distal esophageal smooth muscle
- Sphincter function = normal

CAUSES

- Cause relatively unknown
- Uncontrolled brain signals and extremely hot/cold beverages can trigger disease

COMPLICATIONS

- Leads to difficulty swallowing, impaired advancement of food and/or regurgitation

SIGNS & SYMPTOMS

- Intermittent dysphagia
- Atypical chest pain that mimics cardiac chest pain; may radiate to jaw, arms, back
- Food regurgitation relatively uncommon

DIAGNOSIS

DIAGNOSTIC IMAGING

Barium swallow x-ray (upper GI)

- "Corkscrew" appearance is characteristic

Endoscopy

- Exclude heart disease, mechanical intraluminal obstruction

OTHER DIAGNOSTICS

24-hour esophageal manometry

- Shows uncoordinated esophageal contractions of normal amplitude

TREATMENT

- No cure

MEDICATIONS

- Nitrates, calcium channel blockers, and/or botulinum toxin injections to lower esophageal muscle; used to decrease spasms
- Antidepressants, anti-anxiety medications

SURGERY

- Surgical esophagomyotomy rarely considered

GASTROESOPHAGEAL REFLUX DISEASE (GERD)

osms.it/gastroesophageal-reflux

PATHOLOGY & CAUSES

- AKA acid reflux
- Failure of lower esophageal sphincter → poor closure/inappropriate relaxation (poor tone) of lower esophageal sphincter → stomach contents re-enter esophagus
- Commonly associated with decreased esophageal motility, gastric outlet obstruction, hiatal hernia

RISK FACTORS

- Obesity, pregnancy, smoking, hiatal hernia
- Medications
  - Antihistamines, calcium channel blockers, antidepressants, hypnotics, glucocorticoids
- Zollinger–Ellison syndrome, high blood calcium (increased gastrin production), scleroderma/systemic sclerosis (esophageal dysmotility)
- Visceroptosis

COMPLICATIONS

- Esophagitis, esophageal strictures, Barrett's esophagus (premalignant condition), esophageal adenocarcinoma, laryngitis, chronic cough, pulmonary fibrosis, earache, asthma, recurrent pneumonia

SIGNS & SYMPTOMS

- Acid taste in mouth, heartburn, retrosternal chest pain, early satiety, regurgitation, odynophagia, increased salivation, postprandial nausea and vomiting, sore throat, sensation of lump in throat, coughing, wheezing
  - Often felt shortly after eating meals (worse after large meals/when lying down)
- Halitosis, tooth decay

DIAGNOSIS

- Can be diagnosed based on clinical symptoms, history alone

DIAGNOSTIC IMAGING

Endoscopy

- Used when therapeutic response poor/concerning symptoms present (dysphagia, anemia, blood in stool, wheezing, weight loss, voice changes)

Upper GI series X-rays with barium contrast

- Useful to identify complications
- Early stages of reflux esophagitis: granular nodular appearance of mucosa in distal third of esophagus with numerous ill-defined 1–3mm lucencies
- Shallow ulcers and erosions
  - Collections of barium in distal esophagus near gastroesophageal junction
  - Identify stricture (tapered area of concentric narrowing in distal esophagus)

LAB RESULTS

- 24-hour esophageal pH monitoring in lower esophagus

Biopsy

- Edema, basal hyperplasia (non-specific inflammation)
- Lymphocytic inflammation (non-specific)
- Neutrophilic inflammation (reflux/*Helicobacter* gastritis)
- Eosinophilic inflammation (usually reflux, if > 20 eosinophils per high-power field extending beyond distal esophagus, more like eosinophilic esophagitis)
- Elongation of papillae
- Goblet cell intestinal metaplasia
- Thinning of squamous cell layer
- Dysplasia
- Carcinoma

OTHER DIAGNOSTICS

- Esophageal manometry (excludes motility disorder)
- Short term trial of proton-pump inhibitors

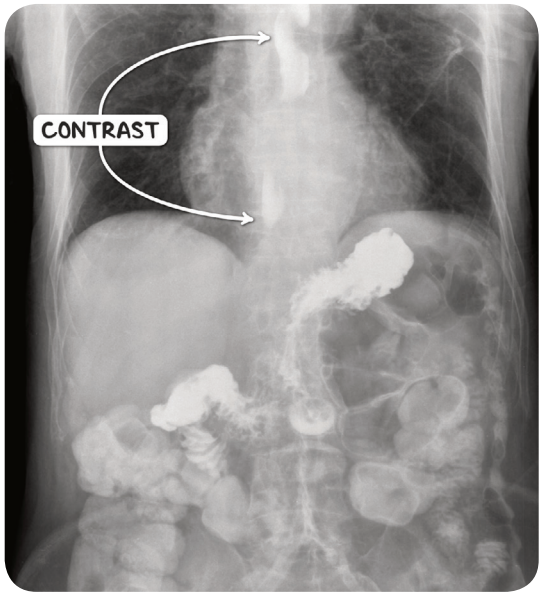

Figure 30.5 A contrast X-ray demonstrating gastroesophageal reflux. The contrast medium was injected percutaneously into the stomach and has migrated into the esophagus.

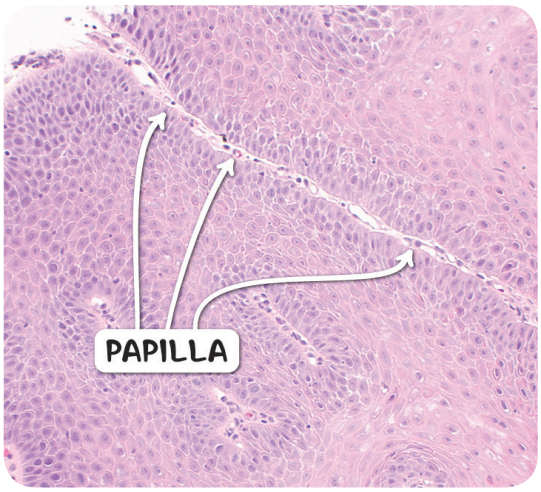

Figure 30.4 The histological appearance of the squamous-lined esophagus in a case of reflux. The papillae become elongated and there is overgrowth of the basal cells (darker blue) known as basal cell hyperplasia.

TREATMENT

MEDICATIONS

- Antacids neutralise acidity of gastric secretions
- H2 receptor blockers decrease acidification of gastric secretions
- Proton pump inhibitors decrease acidification of gastric secretions
- Prokinetics strengthen lower esophageal sphincter (LES), causing stomach contents to empty faster
- Baclofen (GABAB agonist)
  - Inhibits transient LES relaxations, particularly in postprandial period
  - Modestly effective, but rarely used due to frequent dosing requirements

Surface agents and alginates

- Sucralfate (aluminium sucrose sulfate)
  - Adheres to mucosal surface → promotes healing, protects from peptic injury
- Sodium alginate
  - Polysaccharide derived from seaweed → forms a viscous gum that floats within stomach → reduced postprandial acid pocket in proximal stomach

SURGERY

Nissen fundoplication

- Upper part of stomach wrapped around lower esophageal sphincter → strengthens sphincter, prevents acid reflux

Transoral incisionless fundoplication

- Similar procedure to Nissen fundoplication, performed transorally with endoscope

LINX reflux management system

- Titanium beads with magnetic cores wrapped around weak native lower esophageal sphincter → attractive force between beads closing sphincter → force of peristaltic wave of caused by swallowing can transiently open beads

OTHER INTERVENTIONS

Lifestyle modifications

- Avoid lying down within three hours after eating, wedge pillow when sleeping to elevate head, weight loss, avoid certain foods (coffee, alcohol, chocolate, fatty/acidic/spicy foods), smoking cessation, moderate exercise

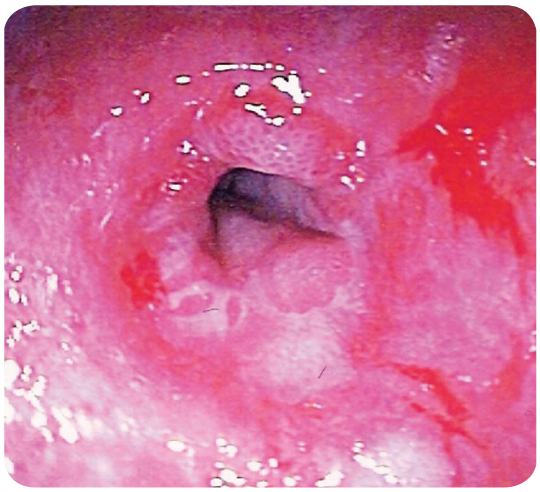

Figure 30.6 An endoscopic view of an esophageal stricture, a potential consequence of severe, long-standing reflux.

MALLORY–WEISS SYNDROME

osms.it/mallory-weiss

PATHOLOGY & CAUSES

- Severe vomiting → sudden increase in intra-abdominal pressure → partial thickness laceration at gastroesophageal junction → bleeding from mucosa
- Also called gastroesophageal laceration syndrome
- Laceration known as "Mallory–Weiss tear", involves mucosa and submucosa, not muscular layer

CAUSES

- Vomiting, straining, coughing, seizures, blunt abdominal injury, nasogastric tube placement, gastroscopy

RISK FACTORS

- Alcoholism, bulimia, food poisoning, hiatal hernia, NSAID abuse, biological male sex (80%), hyperemesis gravidarum (severe morning sickness in pregnancy)

SIGNS & SYMPTOMS

- Hematemesis after episode of violent retching/vomiting
- Melena
- Bleeding associated symptoms may cease after 24–48 hours
- Epigastric, back pain
- Signs of hemodynamic instability
  - Resting tachycardia, hypotension

DIAGNOSIS

DIAGNOSTIC IMAGING

Endoscopy

- Tears appear as red longitudinal breaks in mucosa, may be covered by clot

LAB RESULTS

- Hemoglobin, hematocrit (assess severity of initial bleeding)

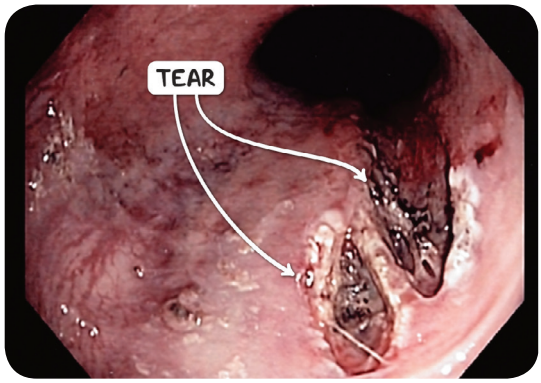

Figure 30.7 Endoscopic appearance of a Mallory-Weiss tear.

TREATMENT

- In absence of comorbidities (esp. portal vein hypertension), significant healing occurs in first 24–48 hours

MEDICATIONS

Supportive (persistent bleeding uncommon)

- Acid suppression
  - IV proton pump inhibitor
- If nausea and vomiting persistent
  - Antiemetics

SURGERY

Endoscopy (for spurting/oozing tears)

- Cauterization, hemoclips (hemostasis of small defects), endoscopic band ligation (with or without epinephrine injection), arterial embolization

PLUMMER–VINSON SYNDROME

osms.it/plummer-vinson

PATHOLOGY & CAUSES

- Triad of iron deficiency anemia, dysphagia, cervical esophageal web
- AKA Paterson–Brown–Kelly syndrome, sideropenic dysphagia
- Premalignant disease

CAUSES

- Exact cause unknown, likely connected to genetic factors, nutritional deficiencies

RISK FACTORS

- Postmenopause

COMPLICATIONS

- Esophageal/pharyngeal squamous cell carcinoma

SIGNS & SYMPTOMS

- Esophageal signs and symptoms
  - Esophageal webs, difficult/painful swallowing, Plummer–Vinson syndrome at upper end of esophagus, Schatzki ring lower end of esophagus
- Iron deficiency signs and symptoms
  - Glossitis, cheilosis, angular stomatitis, koilonychia, splenomegaly, dizziness, pallor, dyspnea

DIAGNOSIS

- Presence of esophageal web in individual with iron deficiency anemia

DIAGNOSTIC IMAGING

Barium esophagography, videofluoroscopy, esophagogastroduodenoscopy

- Esophageal web

LAB RESULTS

- Anemia
  - Complete blood cell count, peripheral blood smear, iron study

TREATMENT

MEDICATIONS

- Iron supplementation, folate, vitamin B12 → correct iron deficiency anemia

SURGERY

- Mechanical widening of esophagus

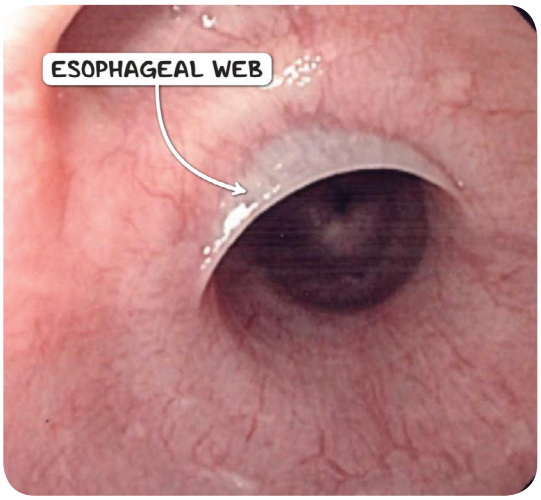

Figure 30.8 An endoscopic view of an esophageal web which is usually associated with Plummer-Vinson syndrome.

ZENKER'S DIVERTICULUM

osms.it/zenkers

PATHOLOGY & CAUSES

- Diverticulum (outpouching) of pharyngeal mucosa through Killian's triangle (area of muscular weakness), between transverse fibres of cricopharyngeus muscle and oblique fibres of lower inferior constrictor muscle
- AKA pharyngoesophageal diverticulum, pharyngeal pouch, hypopharyngeal diverticulum
- Pseudodiverticulum
  - Does not involve all layers of esophageal wall → contains mucosa, submucosa

CAUSES

- Uncoordinated swallowing, impaired relaxation and swallowing, impaired relaxation and spasm of cricopharyngeus muscle → increased pressures in distal pharynx → excessive lower pharyngeal pressures → diverticulum formation

RISK FACTORS

- Biological male > 60 years old

SIGNS & SYMPTOMS

- May be asymptomatic
- Difficulty swallowing, sense of lump in throat, cervical webs
- Food trapping
  - Regurgitation, cough, halitosis, infection

DIAGNOSIS

DIAGNOSTIC IMAGING

Barium swallow

- Distinct outpouching visible

Upper gastrointestinal endoscopy

- Pouch visualized

CT scan with oral contrast

- Distinct outpouching visible

TREATMENT

- Small/asymptomatic diverticula do not require treatment

SURGERY

- Neck surgery → cricopharyngeal myotomy, diverticulopexy

OTHER INTERVENTIONS

- Non-surgical endoscopic technique
- Endoscopic stapling
- Endoscopic laser

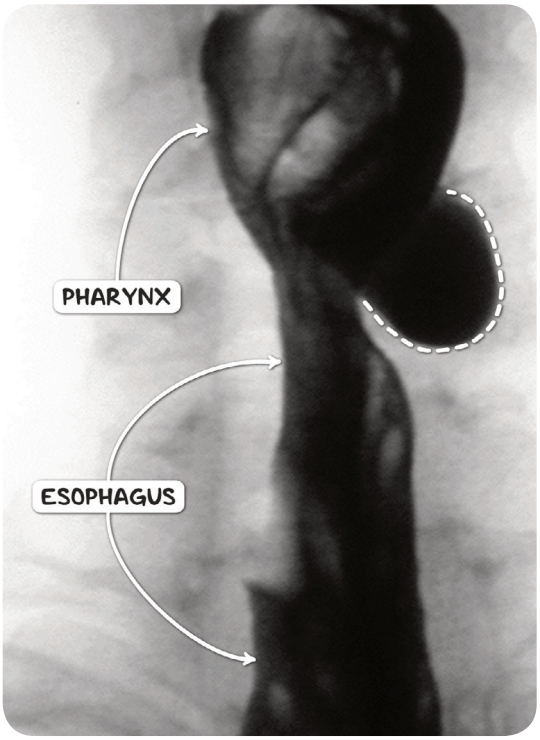

Figure 30.9 A barium swallow demonstrating a Zenker's diverticulum, outlined on the right of the image.