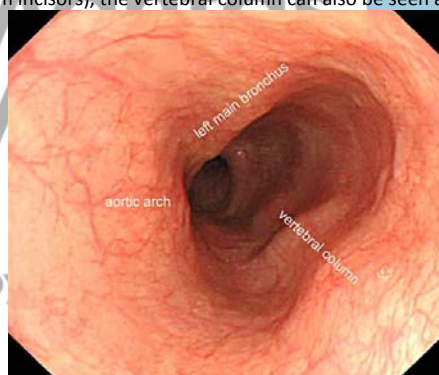


A&P

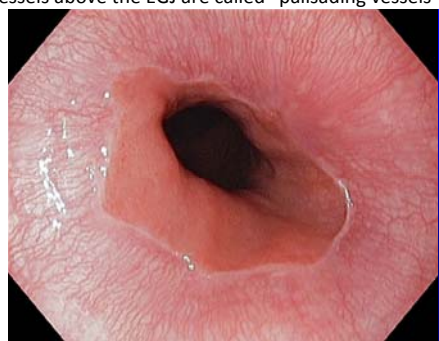
- Embryology
 - Endodermal tube (the entire GI tract) develops a bud near the base of the mouth which extends forming the trachea and then a septum closes the connection
- Pharynx
 - Saliva: alpha-amylase, lingual lipase, mucus to lubricate, antimicrobials (lysozyme/lactoferrin), IgA
 - Major ducts: parotid, submaxillary, sublingual vs Minor Ducts: submucosal
 - Taste: fungiform papillae (ant 2/3) vs vallate papillae () vs foliate papillae (lateral)
 - Smell: 1000 receptors w/ odorants able to bind various combination of receptors
- Histology
 - Mucosa (non-keratinized stratified squamous epithelium)
 - Submucosa (contains glands that secrete lubricants thru tortuous collecting ducts)
 - <5% striated, 1/3 mixed, 2/3 smooth



- Anatomy
 - Lies w/in posterior mediastinum behind heart/trachea but in front of aorta on the L side, you can see external indentations from pulsating aortic arch (23cm from incisors), L main bronchus (25cm from incisors), pulsating L atria (30cm from incisors), the vertebral column can also be seen along the length of the posterior esophagus

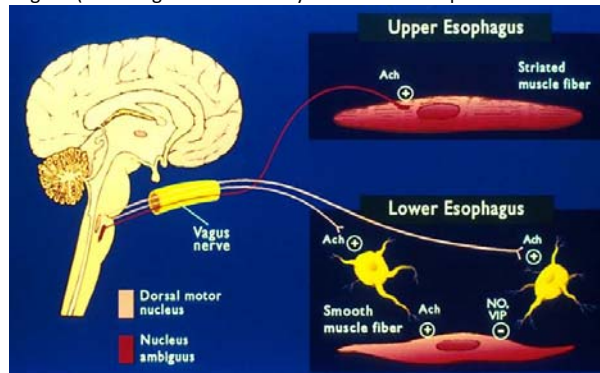


- Blood/Lymphatic Supply/Drainage: upper (thyroid artery branches and SVC and Cervical LNs), middle (bronchial/aorta artery branches and azygous vein and Mediastinal LNs), lower (left gastric, left inferior phrenic, splenic artery branches and portal vein and Celiac/Gastric LNs) very robust with multiple sources hence ischemia is rare, the longitudinal fine mucosal vessels above the EGJ are called "palisading vessels" are normal and not esophagitis

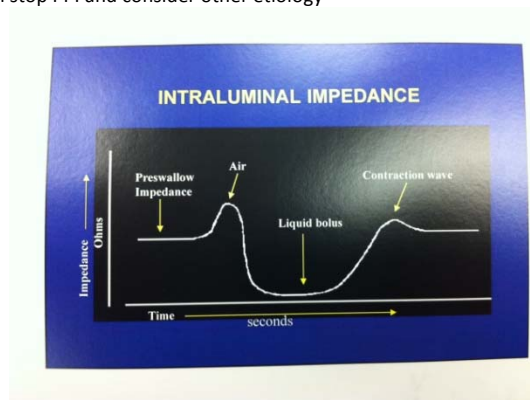


- Innervation: all via Vagus nerve, mixed with nerves that interact w/ heart/lungs hence Sx can be mixed

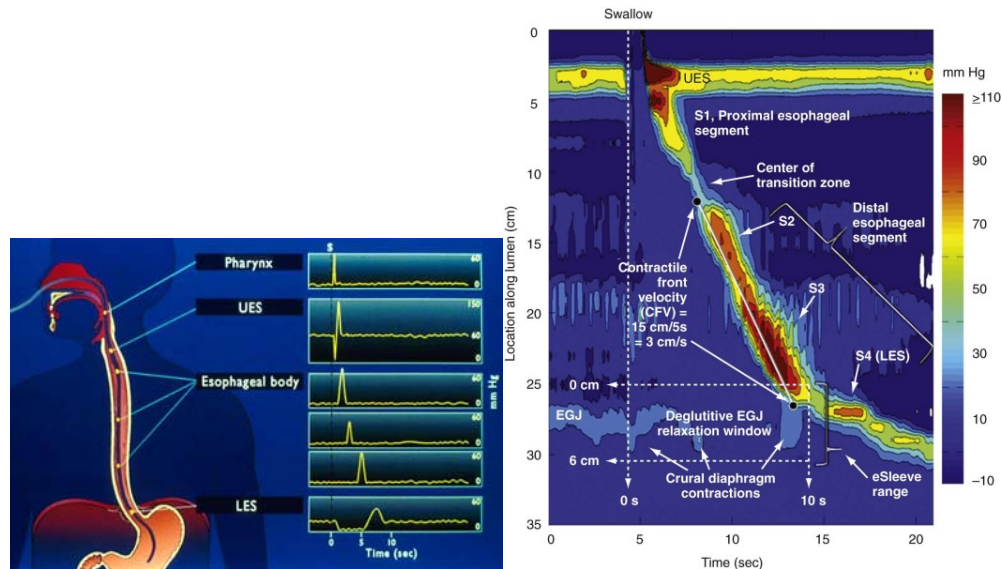
- Voluntary Deglutition via Frontal Cortex & Dorsal Motor Nucleus from Pharynx to Upper Esophagus then Involuntary Peristalsis from Upper Esophagus to Lower Esophagus via Swallowing Center Medulla/Pons Nuclear Ambiguus (there is great redundancy such that if one part fails another part picks up)



- Physiology (Three Functional Zones) [two different values based on type of manometry: conventional vs high resolution]
 - Pharynx (0mmHg)
 - (1) UES
 - Parameters
 - Resting Pressure: 100mmHg
 - Relaxation Pressure: ~0mmHg after which there is sharp and brief rebound
 - (2) Body
 - Parameters (smooth muscle portion)
 - Resting Pressure: -5mmHg
 - Peristaltic Pressure: 30-180mmHg aka Distal Esophageal Amplitude (DEA) vs <5000mmHg/cm/sec aka Distal Contractile Integral (DCI)
 - Peristaltic Velocity: 1-6 vs 2-8cm/sec aka Contractile Front Velocity (CFV)
 - Aboral/Antegrade vs Retrograde Peristalsis
 - Primary Peristalsis: initiated by swallow, pushes bolus down
 - Secondary Peristalsis: initiated by residual food, clears debris/reflux down
 - Tertiary Peristalsis: reported by radiologist but there is NO physiologic correlate
 - (3) LES
 - Parameters
 - Resting Pressure: 10-45 vs 10-35mmHg
 - Relaxation Pressure: <8 vs <15mmHg
 - Stomach (5mmHg, to know that the manometer is in the stomach there should be increased pressure as the pt takes a deep breath as the diaphragm contracts increasing abdominal pressure)
 - Multichannel Intraluminal Impedance (MII)
 - reflects the differences in resistance (Ohms) to an electric current b/t two electrodes in the presence of different mediums (eg. liquid → high [ion] → high conductivity → low impedance vs gas) thus it can tell you the composition of reflux
 - If multiple catheters then you can determine if it is liquid that is being reflux or liquid that is passing antegrade
 - Tells you if there is reflux and if correlates w/ Sx
 - If linked w/ a pH probe and tells you if it is acidic vs non-acidic reflux
 - If pt has Sx from acid reflux then surgery vs if pt has Sx from non-acid reflux then add carafate and consider surgery vs if pt has Sx not from reflux then stop PPI and consider other etiology



- **Ambulatory Lower Esophageal 24hr pH Monitoring**
 - NEVER DONE AS A PRIMARY DIAGNOSTIC TEST
 - Placement
 - Either on meds or off x7d for PPIs, x3d for H2B, x6hrs for AA depending on what you are doing it for, tell pts to live a normal life (if prior to surgery then do off PPI)
 - Probe is passed nasally and then placed 5cm above manometrically determined LES, probe is connected to a data recorder, pH is monitored Q4-6sec and pt activated by Sx/meals/body position change, 24hr study, do this if you are concurrently doing manometry
 - NB a wireless version (BRAVO) that attaches via suction at 6cm above LES and records for 48hr but does not have MII capabilities
 - Have nurses calibrate BRAVO, do exam, determine GEJ, take scope out, feed BRAVO down mouth using finger to determined distance, repass scope to confirm placement in esophagus, make sure probe still at same distance, place scope over shoulder, place suction tube on handle and suck for 30sec, then push down on blue plunger and hold for 5sec, then bring plunger back up just a fraction then with thumb rotate it to fully bring plunger up and release the probe, then take mechanism out, relook with scope and be careful to not knock off the probe, 48hr study, falls off after 5d
 - NB some recommend placing a proximal BRAVO to exclude acidic food ingestion not included in diary
 - Complications: perforation, CP or sensation of foreign body
 - Results (NI pH 5.0-6.8)
 - Definition of reflux is pH <4.0 as this is when pepsin is active (acid is most injurious when pepsin is present) 98.5% of normal pts have pH >4
 - Definition of GERD is not clear, many parameters are measured including total time, upright/supine periods, duration of longest episode, correlation w/ Sx, etc
 - NB if the pH drops to 0 or 1 then rises to 7 then it likely fell into the stomach and then entered the duodenum
 - Scores (symptom calculations are not great w/ a sensitivity of 30-70% and specificity 70-80%, most look at the first two)
 - **“The 4 Rule”**: when pH <4.0 for >4.2/5.3% of total time for catheter/BRAVO studies = GERD or **Demeester Score**: depends on the machine used by the Baylor ones are >14 = GERD
 - **Symptom Index (SI)**: # of Sx episodes when pH<4 / total # of Sx episodes = >50% = GERD
 - **Symptom Sensitivity Index (SSI)**: # of Sx episodes when pH<4 / total # of reflux episodes = >10% = GERD (this implies that most people have reflux but don't have Sx)
 - **Symptom Association Probability (SAP)**: fancy calculation which tells you the probability that the Sx are from reflux = >95% = GERD
- **High Resolution Manometry vs Conventional Manometry**
 - How? transnasal catheter placed into stomach, 10 wet swallows w/ water >30sec apart, if there is something abnormal it needs to be seen in ≥2 swallows for it to be abnormal
 - Limitations: pt is not sedated, manometry cannot assess longitudinal contractions, cannot assess non-occlusive contractions
 - **Important Points**
 - **cause of most motility problems is not known**
 - **many manometric abnormalities have no apparent physiologic consequences**
 - **even if you Tx the manometric abnormality pts can still have Sx**
 - **the proximal striated and distal smooth muscle esophageal contractile segments are separated by a center of transition zone (first pressure trough) which is where muscle types change**
 - **the LES should be relaxed from the START of the swallow to the very end aka “deglutitive EGJ relaxation window”**



- GER
 - GERD: frequent/abnormal amounts of GER causing Sx (70% of GERD is 2/2 increased TLESR vs 30% of GERD is 2/2 from non-TLESR problems!!!)
 - Defensive Factors
 - Acid Prevention (hiatal hernia affects everything below, esp if non-reducible (gastric rugae remain above diaphragm b/t swallows)
 - LES
 - 4cm in length with SC jxn in middle w/ 2cm (esophageal tissue) above and 2cm (gastric tissue) below diaphragm
 - clasp fibers encircle esophagus medially (high tone and less responsive to ACh) while gastric sling fibers run laterally on the left (less tone and more responsive to ACh)
 - **Transient Lower Esophageal Sphincter Relaxation (TLESR)**
 - relaxations that are not preceded by a swallow
 - last a long time (10-45sec vs <10sec relaxations that occur after a swallow)
 - normally occur 2-6x/hr

	Increases Tone	Decreases Tone
Time	Night	Day
Eating	Fasting	Meals, Swallow, Belching, Vomiting
Food	Protein	Fat, Chocolate, Peppermint
Hormones	GABA, Gastrin, Motilin, Substance P	NO, CCK, Secretin, Cholecystikinin, Somatostatin, VIP
NT	+alpha/cholinergic, -beta	(opposite)
Meds	Histamine, AAs, Reglan	Opiates, CCB, Anesthesia
Lung	Inspiration, Coughing, Valsalva	Expiration

- **Angle of His** (sharp angle b/t esophagus and stomach acting as a valve)
- **Diaphragmatic Right Crura** (provides extrinsic squeeze to the LES during inspiration/coughing/sneezing/straining when the abdominal-thoracic pressure gradient is high)
- **Phrenoesophageal Ligaments** (anchors LES at correct position)
- **Intra-Abdominal Location of LES** (higher pressure)
- **NB in a hiatal hernia the angle of His changes, the diaphragmatic right crura are below the LES and the phrenoesophageal ligaments are lost**
- Acid Resistance
 - Thick (30 Cell) **Epithelial Layer** w/ tight junctions, intracellular buffers, and H+ pump
 - **Increased Blood flow** which removes H+ and brings HCO₃⁻
 - Gravity and Esophageal **Peristalsis** w/ Subsequent Gastric Emptying
 - Salivary/Esophageal Gland **Bicarb** Secretion
- Aggressive Factors (rarely the main cause)
 - Gastric Content (need some pepsin to disrupt mucosal barrier to allow for acid to cause damage)
 - Gastric Volume (delayed gastric emptying)

Approach

- **Glycogenic Acanthosis**

- Benign plaques seen in old pts
- **Oropharyngeal aka Swallow aka Transfer Dysphagia**
 - Etiology
 - Structural Lesion: cancer, post head & neck surgery/radiation, webs/bars/sings/stenosis, diverticula including Zenker's, cricopharyngeal achalasia, abscess, foreign body, thyromegaly, vertebral spur, cervical LAD, vascular anomalies, diffuse idiopathic skeletal hyperostosis, cervical osteophytes
 - NM Disorder: CNS Dz (CVA of brainstem/cerebellum or bilateral hemispheres, if stroke related dysphagia spontaneously improves it does so during the first 2wks therefore wait for PEG after 2wks), PNS Dz, Skeletal Muscle Dz (Cryopharyngeal Dysfunction or spasm or failure to relax, systemic disorders)
 - Other: xerostomia
 - With age there is cerebral atrophy, nerve deterioration, muscle atrophy which collectively affects swallowing but in most cases we can compensate without Sx therefore you need to investigate for other causes
 - Dx: Modified Barium Swallow (MBS) aka Video-Fluoroscopy Swallow Study (VFSS) and if abnormal then Fiberoptic Endoscopic Evaluation of Swallow (FEES)
 - S/S: difficulty swallowing resulting in food getting stuck, sialorrhea, drooling, food spillage, nasopharyngeal regurgitation, tracheobronchial aspiration (coughing, choking, recurrent PNA), slurred or nasal speech, dysarthria, dysphonia, swallowing with gurgling sound, halitosis, relief with repeated swallowing, raising arms, throwing shoulders back, Valsalva
 - Tx: general swallow therapy by ST, difficult to actually Tx therefore the focus should be on how to maintain adequate nutrition
- **Esophageal Dysphagia**
 - Anatomic Problem (S then S+L dysphagia, constant) vs Motility Problem (S+L dysphagia, intermittent, worsening w/ cold and improvement w/ hot liquids) always work-up dysphagia never empirically Tx
 - first test is usually Barium Swallow, why? b/c cheap, there is risk of perforation from EGD/Manometry w/ diverticula or high grade obstruction, info from barium swallow may preclude need for EGD/Manometry, after barium swallow then do EGD (best for anatomic problems, <13mm causes Sx balloon or Bougie dilation) vs Manometry (best for motility problems)
 - always characterize Sx as Intermittent (signifies that the pathology is not always present (ex. DES) or is fixed (ex. esophageal ring), less worrisome) vs Progressive (signifies that that pathology is always present and worsening (ex. cancer, achalasia, scleroderma), more worrisome, some normal pts have abnormal sensory perception leading to the sensation of dysphagia
 - S/S: relief with regurgitation or vomiting
 - it is important to note that studies have demonstrated that the location where food stops as described by the patient is not an accurate clinical guide to determine the level of the lesion but a good general rule is that the pt will point to lesion or above lesion but never below it therefore if pt points to throat then it can be anywhere from throat to lower esophagus but if the pt points lower then the accuracy increases
- **Chest Pain**
 - 1st rule out cardiac disease and other extra-esophageal disease including pancreatoco-biliary (55% of chest pain is not cardiac)
 - 2nd GI work-up (heart and esophagus have same innervation therefore once you r/o cardiac check esophagus)
 - 1st if alarming Sx then EGD but if not then likely atypical GERD therefore trial of PPI BID x2mo
 - 3rd if + response to PPI then try to slowly taper but if no response w/ PPI then possibly hypercontractile
 - 3rd if normal manometry then treat with pain modifying agents
- **Globus Hystericus**
 - "lump in throat", rarely something anatomic, likely heightened sense of esophageal stretch 2/2 psychologic state, 4th most common Sx in somatization disorder, therefore the goal is to just exclude anything serious, causes included GERD, LAD, hypopharyngeal cancer, goiter, Sx often improves during swallow
- **GERD** (if classic Sx then just empirically Tx)
- **Esophagitis (odynophagia)** (always do an EGD)
 - NB you can get odynophagia w/ anatomic/motility problems and you can get dysphagia w/ esophagitis
- **Hiccups aka Singultus**
 - Mechanism: abnormal circuit b/t vagus and CNS and phrenic resulting in quick inhalations that follow abrupt rhythmic involuntary diaphragmatic contractions and glottic closures
 - Bad b/c causes malnutrition, weight loss, fatigue, dehydration, insomnia, wound dehiscence
 - If <2d the likely idiopathic then home remedies to break reflex arc (breathing in bag, drinking from opposite side of glass of cold water, pull tongue, eat a packet of sugar, hold breath, noxious odors, scare) if fails then treat
 - If >2d (persistent) or >2mo (intractable) then treat and explore cause (1) systemic: toxic-metabolic disorder (hypoNa, uremia, EtOH, anesthesia), (2) CNS: CNS lesion or psychogenic (stress, excitement, anxiety, malingering), (3) vagus/phrenic nerve irritation (mass, distension, reflux, sudden changes in gastric temp, alcohol, tobacco, post-op)
 - Tx: anti-dopaminergics 1° Thorazine (chlorpromazine) 25-50mg PO TID if refractory then 25mg IV at 1mg/min x1 (will drop BP) 2° metoclopramide, baclofen, TCAs, AEDs, nifedipine, haloperidol, et al 3° phrenic nerve block or electric stimulator
- **Oral Cavity**
 - Ulcers
 - Aphthous Stomatitis aka Canker Sore (single, sharply demarcated, erythematous margin, shallow, grey based, painful mucosal ulcer of unknown etiology but often are precipitated after trauma as in a teeth bite, Celiac Dz,

nutritional deficiencies, stress/anxiety, food allergies, etc, can also occur in anywhere else in GI tract even rectum!, 25% of people will have an aphthous ulcer at some point in their life, resolved in <2wks, Tx: treat underlying precipitant, analgesics (refer), fluocinonide (Lidex), clobetasol (Temovate), 5% amlexanox paste, MOM, salt water, Sulfuric Acid + Sulfonated Phenolics (Debacterol) or Maltodextrin + Propylene Glycol (Gelclair), thalidomide if occurs in HIV pts, coccidine, cimetidine, azathioprine, intralesional/systemic steroids if refractory)

- Canker Sore = unkeratinized (lateral tongue, mouth floor, labial/buccal mucosa, soft palate, pharynx) vs Fever Blister = keratinized (gingival, hard palate, dorsal tongue)
 - Herpetic Labiogenivostomatitis aka Fever Blister aka Cold Sores, NB you can also get CMV infection
 - Other Infections: Syphilis, Hand-Foot-Mouth Dz (cxsackie A16 infection resulting in vesicular lesions on hand/foot and oral ulcers w/ constitutional Sx, seen in children and young adults, occurs as epidemics, very contagious, self-limited lasting <10d, only symptomatic Tx), Acute Necrotizing Ulcerative Gingivitis aka Trench Mouth aka Vincent's Dz (necrosis and ulceration of interdental papillae 2/2 decreased resistance to infection by normal oral flora resulting in foul-smelling breath and gingival pain, Tx: surgery and abx)
 - CVD: Behcet's, Reiter's
 - Derm: Pemphigus Vulgaris & Bullous Pemphigoid, EM/SJS/TEN
 - GI: Crohn's
 - Other: GVHD
 - Glossitis
 - Mech: inflammation of tongue
 - Etiology: Nutritional deficiencies, Irritants, Drug reaction, Amyloidosis, Sarcoidosis, Radiation, Infections esp Candida, Benign Migratory Glossitis aka Geographic Tongue (benign, patchy glossitis, seen in 2% of population, could represent atypical psoriasis)
 - S/S: glossodynia, hypogeusia/dysgeusia, erythema w/ loss of papilla, erosion, smooth atrophy
 - Tx: correct underlying problem, zinc
 - Black Tongue (2/2 pepto-bismol use)
 - Strawberry Tongue (2/2 Kawasaki Dz or Scarlet Fever)
 - Bald Tongue (atrophy 2/2 nutritional deficiency esp Pellagra/IDA/Pernicious Anemia, xerostomia, glucagonoma)
 - Leukoplakia (white patch that cannot be scraped off and does not respond to antifungals thus get Bx)
 - SCC (2/2 smoking/smokeless tobacco, ventral surface of tongue, floor of mouth, lower lip, soft palate, gingival)
 - Oral Hairy Leukoplakia (on lateral tongue, EBV infection, seen in immunocompromised pts esp HIV, asymptomatic, Tx is elective and includes antivirals, etc)
 - Thrush (white patch that can be scraped off and does respond to antifungals)
 - Erythroplakia (red velvety lesion, DDx: SCC, Kaposi Sarcoma, Bleeding Dyscrasias, Hemangioma, Bacillary Angiomatosis)
 - Fissured Tongue
 - Lichen Planus w/ Wickham's Stria (refer)
 - Dental Caries (Tx: filling then root canal then tooth extraction)
 - Gingivitis (Tx: I&D, abx)
 - Tooth Avulsion (Tx: place back in socket but if not able the place in milk and seek dentist ASAP)
 - Salivary Glands
 - Xerostomia (dry mouth, 2/2 CTD (esp Sjogren's), after radiation Tx, or w/ meds (esp anticholinergics, diuretics, etc, resulting in odynophagia/dysphagia, loss of taste, inflammation, dental caries, candida, sialolith, Tx: improve salivary flow by chewing gum, 1% sodium carboxymethyl cellulose to moisten mouth, cholinergic agonists esp cevimeline (Evoxac)
 - Sialadenitis (inflammation of a salivary gland with mucocele being the most common lesion formed in which blockage or rupture of the salivary gland ducts results in leakage of saliva and subsequent swelling with a blue translucence, due to trauma, autoimmune disorder (Sjogren Syndrome), or viral infection (Mumps)
 - Head & Neck SCC: RFs (tobacco, alcohol, EBG for nasopharyngeal, HPV for naso/oropharyngeal), often present at advanced stage w/ Sx depending on site, almost all are SCC, there is higher r/o of other cancers given RFs is lung/esophageal, Dx via FNA, TNM stage, Tx: surgery, XRT, chemo
 - Gland Cancer: 70% Parotid (20% of which are malignant, 80% are benign and include: Pleomorphic Adenoma aka Mixed Tumors and Warthin Tumor aka Papillary Cystadenoma Lymphamatosum), 10% Submandibular (40% of which are malignant), 20% Sublingual (80% of which are malignant) NB the smaller the gland the greater the chance that the cancer is malignant
- **Foreign Body**
 - RFs: underlying pathology usually in the esophagus
 - Types
 - Adults: food impaction esp meat esp chicken or bone, parts of dentures, endoscopic capsules (1% risk), drug stuffers (small amount of poorly packed drugs w/ purpose of not being caught by police) vs packers (large amount of tightly packed drugs w/ purpose of smuggling), bezoars (indigestible material that accumulates and coalesces in GI tract, phytobezoars = vegetable matter w/ lots of fiber, trichobezoars = hair or any stringy material, pharmacobezoars = meds esp enteric coated, vitamins/minerals, bezoar formation usually requires altered GI anatomy/motility esp gastroparesis, slow growing, pressure ulceration is common)
 - NB often seen in psych and incarcerated pts
 - Children: shiny metal objects (button batteries (very dangerous), coins, safety pins, etc)
 - Always consider the possibility of more than one object

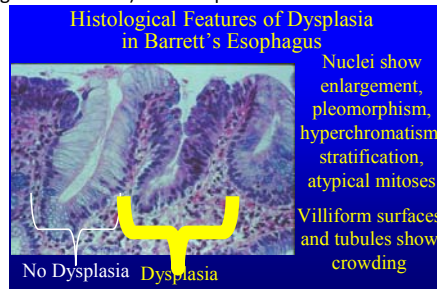
- S/S: odynophagia, dysphagia, choking (can't talk) vs impaction (can talk), inability to handle oral secretion w/ drooling, regurgitation, NB pts are only 35% correct if they say that the object is in their esophagus and more distally it is 0%, check for subcut emphysema and peritonitis
- Complications: most foreign bodies pass but complications do occur including obstruction (esp at sphincter, anatomic areas of acute angulation, prior surgery hence always ask about prior surgery/endoscopy to see if risk higher), perforation, bleeding, fistulization, abscess formation, pressure necrosis if >12-24hr in esophagus
- Dx: PA/L Neck/Chest/Ab xray (only good for radioopaque objects), consider metal detector for coins, avoid oral contrast fluoroscopy/CT studies b/c will interfere with endoscopy, endoscopy is really the best test
- Tx: **if symp or asymp but (1) or (2) or (3) then remove otherwise weekly x-ray and if Sx develop symptoms, complications occur or no movement in 3d then surgery**
 - **(1) object has not reached stomach b/c once in stomach 85% of ingested objects pass spontaneously w/in 7-10d w/o complications, observe stool for passage but if not then check for obstruction at pylorus, duodenal sweep, ICV, anus**
 - **(2) not benign (caustic, sharp, wide >2cm, long >5cm)**
 - **(3) pt has abnormal anatomy from pathology/surgery**
- Endoscopy: always have a plan, intubate, suction before endoscopy, use endoscope w/ overtube (US endoscopy makes an esophageal and gastric one, preload it onto scope and pass both to area of interest and then leave overtube in place while taking scope in and out, allows for multiple passes, safe removal of objects, etc), it's good to practice outside of the body with a similar object to plan on how to grab while inside the body, use US Endoscopy rat tooth forceps, US Endoscopy Talon Grasping Device, various snares/nets/baskets w/ condom hoods, sometimes you can gently push the object into stomach along w/ air insufflation but never forcefully, break apart w/ forceps if possible, b/c impactions are usually 2/2 some pathology blind retrieval behind obstruction should never be done, don't forget to assess for underlying pathology, when removing sharp objects always remember "leading edges perforate while trailing edges do not", if object is above UES then ENT consult
- Medical: smooth muscle relaxants (glucagon, nifedipine, nitroglycerin), gas forming agents to distend lumen (carbonated drinks), never use Adolf's meat tenderizer papain as it can cause tissue necrosis, never use emetics to induce regurgitation as high r/o aspiration
- Pt always needs f/u EGD to assess early and late complications and underlying pathology
- Rectal Foreign Bodies: r/o sexual assault, sedate pt, place lubricated Foley catheter past object b/c pulling it out creates a vacuum making it difficult (the catheter prevents a vacuum from forming), place pt in lithotomy position to allow for gravity and ab pressure, after removal do flexsig to rule out perforation then a CT)
- **Tears & Perforation**
 - **Tears aka "Rents" (mucosal)**
 - Etiology: forceful vomiting/retching (**Mallory-Weiss Tears**) but only a h/o retching is obtained in a <30% of cases
 - Location: at or just below GEJ, 15% multiple
 - S/S: hematemesis
 - Dx: EGD
 - Tx: Stable (room temp water lavage to see if there are any stigmata of rebleed) vs Unstable (thermal, injection, hemoclips, banding) NB in general 90% of pts bleeding stops spontaneously w/o Tx and only 5% rebleed
 - **Perforation (transmural)**
 - Etiology: trauma, iatrogenic after dilation of strictures (50% of all causes), forceful vomiting (**Boerhaave's Syndrome**)
 - Location: left lateral posterior esophagus ~4cm above GEJ
 - S/S: GI (hematemesis), CV (RS CP, Hamman's Sign (mediastinal crunch produced by heart beating against air-filled tissue), HD instability), Pulm (creptitation of neck, left sided pneumothorax and pleural effusion, tachypnea, dyspnea), ID (mediastinitis w/ subsequent sepsis)
 - Dx: CXR (widened mediastinum w/ left sided pneumothorax and pleural effusion), Gastrograffin Esophagogram
 - Tx: Stable/Small/Neck (IVF, NPO, NT suction, BS-abx, PPI, consider stent placement) vs Unstable/Large/Non-Neck (immediate surgical repair, mediastinal debridement, pleural drainage)

GastroEsophageal Reflux Disease (GERD)

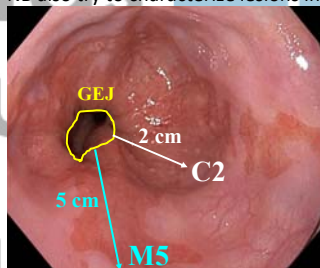
- New
 - Since E-cadherin is a junctional protein that is damaged in GERD one may be able in the future to measure E-cadherin fragment to assess GERD
- Epidemiology
 - 50/15/5% of US heartburn at least x1 per mo/wk/day
 - Age: increases with age and are more likely to develop complications but older pts have less severe Sx
 - M=F except when complications then M>F
 - Equal ethnicity except when complications then W>AA
 - Increasing incidence US b/c of decreasing prevalence of HP (low incidence in Africa/Asia)
 - Some familial clustering but no genetic mechanism discovered
- RFs
 - FHx

- Obesity (VERY IMPORTANT RF!!!, body fat increases intra-abdominal pressure and also increases carcinogenesis)
- Hiatal Hernia
- NO HP infection (but if you find it still Tx HP)
- Pregnancy (refer)
- Scleroderma (refer)
- ZES
- After Heller Myotomy
- Prolonged NGT
- Mechanism
 - Acid/Pepsin damages cells jxns b/t cells increasing permeability allowing for fluid to dilate intracellular space, acid/pepsin then activates nociceptive neurons
 - Recently studies show that nitric oxide (NO) is also causing damage (dietary nitrate (NO₃) in vegetables is absorbed by SI w/ 25% secreted by salivary glands and subsequent mouth bacteria convert it into nitrite (NO₂) and when nitrite encounters acid it is converted to NO which is toxic)
- S/S (asymptomatic esp in elderly)
 - Esophageal
 - Heartburn (1hr postprandial retrosternal rising burning pain exacerbated by lying down on right side and w/ certain foods, relief w/in 5min of use of AA)
 - Atypical CP (even stress testing can induce GERD)
 - Regurgitation
 - Globus Sensation
 - Post-Prandial N/V (very important!!!)
 - Hiccups (reflex to triggered acid reflux)
 - Odynophagia/Dysphagia (2/2 complications including inflammation alone, stricture/ring, dysmotility, cancer)
 - Extra-Esophageal w/ Reflux extending into Larynx/Pharynx
 - General
 - Very controversial as most pts w/ so-called atypical extraesophageal Sx have NO typical esophageal Sx or reflux esophagitis on endoscopy or abnormal pH on pH studies (Why? maybe these structures are more sensitive to acid-pepsin than the esophagus so even a little bit causes damage)
 - Mechanism: direct chemical irritation, vagal reflexes leading to bronchospasm, aspiration
 - Tx: generally requires longer duration and higher doses of PPI compared to standard GERD Tx
 - Laryngopharyngeal Reflux-LPR (classic one, characterized by posterior pharyngeal inflammation, seen by ENT doctors, pts p/w hoarseness)
 - Globus Sensation
 - Waterbrash (reflex salivary hypersecretion) w/ Increased Swallowing
 - Dental Enamel Erosions
 - Gingivitis
 - Hoarseness
 - Throat Clearing
 - Sore throat w/ continued clearing of throat
 - Laryngitis (mucosal thickening, erythema and edema mainly in POSTERIOR larynx)
 - Vocal Cord granulomas/ulcers/inflammation with Dysfunction
 - paroxysmal laryngospasm
 - laryngomalacia
 - recurrent leukoplakia
 - Non-productive nocturnal cough from Esophageal-Tracheobronchial Reflex (due to aspiration of refluxed material or a reflex triggered by acid reflux)
 - Asthma (very controversial, 2/2 heightened vagal response and microaspiration, recent studies indicate that Tx of asymptomatic GERD in poorly controlled asthmatics does NOT improve their asthma Sx as once thought)
 - Sinusitis
 - Recurrent aspiration pneumonia w/ cytology on BAL revealing lipid-laden macrophages
 - Sleep apnea
- Complications (RF: older, white, male 7x, hiatal hernia, obese BMI>30 2x, smoking 2x) NB if any other race you should be cautious attributing complications below w/ GERD
 - Erosive Esophagitis (EE) 30% vs Non-Erosive Reflux Disease (NERD) 70%
 - % above are the results of EGD in pts w/ typical Sx
 - can bleed or perforate and subsequently lead to further complications below
 - Extra-Esophageal Disease
 - Ulcers (5%)
 - always rescope to make sure it is healing in 2mo
 - Stricture & Schatzki Ring (4-20%)
 - Bleeding
 - Barrett's Metaplasia (8-20%)
 - General: esophagus (squamous) then gastric cardia (columnar) and normally the EGJ (beginning of rugal folds) is the same as the SCJ (white-red) but in Barrett's the "SCJ becomes proximal to the EGJ" (nevertheless one cannot reliably differentiate metaplasia from gastric tissue as foveolar cells sometimes look like goblet cells) as

there is conversion of esophageal squamous to specialized intestinal columnar (called "specialized" b/c not just columnar but also the presence of goblet cells, architectural distortion and lamina propria inflammation hence different than gastric mucosa) and this process is called "intestinal metaplasia"



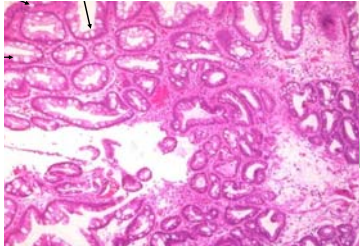
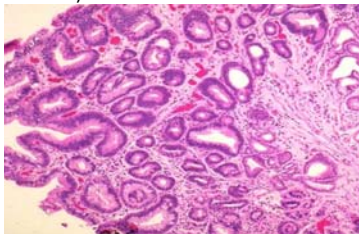
- Epidemiology: 1-2% of pts just undergoing endoscopy for whatever reason, 6-12% of GERD pts that were scoped had Barrett's, 25% of pts w/ Barrett's had no reflux Sx, 95% of pts w/ adenocarcinoma did not know they had Barrett's!!!, usually pt had untreated severe GERD for >10yrs, most cases of Barrett's go undetected for life
- NB length of dz remains the same over time
- S/S: GERD Sx improve b/c innervations have decreased OR otherwise no different Sx
- Complication: 30-125x increased r/o developing adenocarcinoma such that 0.5% of Barrett's pts develop cancer per year, despite the risk of cancer most pts die with Barrett's than from cancer, cancer develops over 20-30yrs
- Endoscopy: pearly esophageal mucosa is replaced w/ red velvety metaplasia, proximal margin is often irregular w/ tongues extending upward, long segment (histologic SC jxn extends >3cm above anatomic EG jxn determined endoscopically) vs short segment vs at EG Jxn (when there is no clear endoscopic recognizable Barrett's and thus the EG jxn is at the SC jxn but specialized intestinal metaplasia is present), pale islands of residual squamous epithelium can be seen, esophagitis can sometimes obscure the sharp margin, complications of stricture/ulcers/nodules can be present
 - Classification Description: Prague C&M Criteria
 - C: length of BE that is circumferential from EGJ
 - M: maximum length of BE from EGJ
 - NB also try to characterize lesions in terms of point on clock where 12 o'clock is anterior



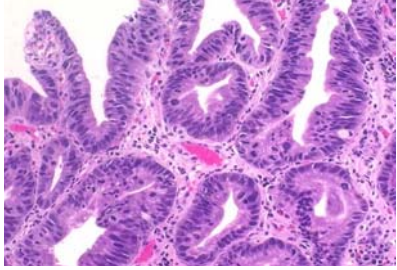
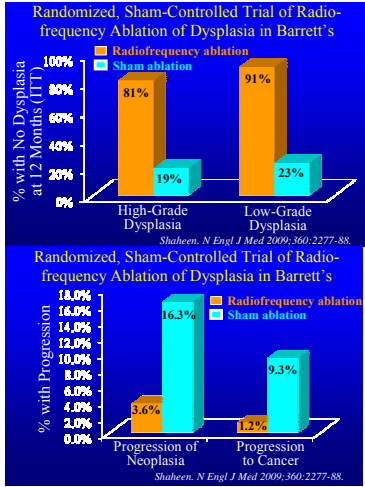
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- It is well known that the longer the BE the higher the r/o EAC
- Barrett's Inspection Time (BIT) >1min per cm of BE
- Diagnostic EMR may be very helpful
- Try to use advanced imaging modalities (refer)
- Seattle Surveillance Protocol
 - very controversial b/c (1) lack of evidence that it improves mortality, (2) the inability to predict who has BE as there is a subgroup of pts who have BE but NO h/o reflux, (3) there are 40million who have GERD but only 10k cases of esophageal cancer even though it is dramatically increasing in incidence the numbers are still very low
 - ASGE
 - one time EGD w/ new imaging modality
 - pt is on PPI (b/c acute inflammation interferes w/ interpretation of BE and if present then increase PPI)
 - Indications: smoking obese white men >55yo with FHx, alarming Sx, frequent (several times per week esp nocturnal Sx), chronic (>5yrs), symptomatic GERD despite PPI x4-8wks, along w/ HH and pt can undergo Tx if HGD/cancer is found (therefore general screening is not recommended)
 - RFs for progression of BE
 - Long segment >3cm (x3)
 - FHx (300x)
 - Age
 - if EGD is negative then no further screening is recommended b/c if pt hadn't developed BE after 5yrs then almost never will in the future

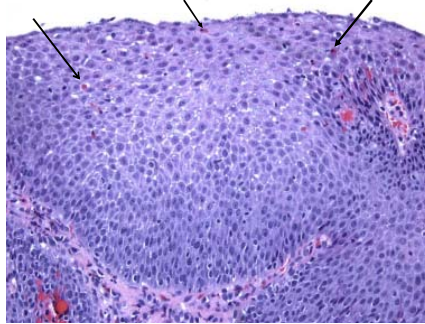
- Biomarkers instead of histologic dysplasia are being studied to assess r/o cancer

<p>Dysplasia 60% regress 10% stable 30% progress Always needs to be confirmed by another pathologist</p>	<p>Surveillance (Hz below is variable b/t societies and controversial) (jumbo forcep 4 quadrant Bx Q1-2cm from GEJ to SCJ and targeted Bx of any other mucosal abnormality) NB other RFs include nodular lesions NB make sure pt can undergo Tx before you survey NB if you lose dysplasia b/c of Tx or just on its own the pt should still undergo surveillance according to the highest degree of dysplasia previously found b/c there is high r/o sampling error</p>	<p>Treatment</p>
<p>No Dysplasia (NDBE) (0.3/2% r/o progression to cancer per yr / ever)</p> <p>NB every year the % seem to be dropping and most recently a 2011 NEJM study reports that r/o AC from BE is even lower than 0.3% more like 0.1% questioning role of further surveillance (maybe just a one time endoscopy to screen and if negative for dysplasia then do nothing else b/c progression of BE to dysplasia and then cancer is actually exceedingly low)</p> 	<p>f/u at 1yr (to ensure you didn't miss LGD) and if no LGD then Q3yrs and if no BE x2 then Q5yrs (continue regardless if there is no more BE found!!!)</p>	<ul style="list-style-type: none"> • Endoscopic Surveillance • Endoscopic Therapy (controversial) <ul style="list-style-type: none"> ○ El-Serag, et al Gastroenterology 2011, RFA of NDBE is like polypectomy in the colon ○ Some do it for NDBE if lots of RFs and for all LGBE • Acid Reduction: PPI (very controversial as the evidence that PPI regresses metaplasia and/or decreases r/o cancer progression is weak nevertheless most initiate therapy), Anti-Reflux Surgery (proven to not be helpful) • Chemoprevention <ul style="list-style-type: none"> ○ Aspirin/NSAID (very controversial and not routinely done, decrease PGE2 which when present increases cellular proliferation) ○ Statins (controversial and not routinely done, mechanism unclear) • Lifestyle Changes: weight loss, stop smoking, eating fruits/vegetables
<p>Low Grade Dysplasia (LGD) (1.6/7% r/o progression to cancer per yr / even)</p> <p>NB Dysplasia = atypia (loss of polarity, irregular nuclei), high N/C ratio, high mitotic activity, architectural disarray (crowding, budding, branching, cribriform, papillary, villiform)</p> 	<p>f/u at 6mo (to ensure you didn't miss HGD/cancer) and if no HGD then Q1yr and if no LGD x2 then Q3yrs</p>	

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<p>High Grade Dysplasia (HGD) (6/22% r/o progression to cancer per yr / ever)</p> <p>NB CIS = ragged BM, single cell infiltration, desmoplastic stromal response</p> 	<p>f/u at 3mo (to ensure you didn't miss cancer) and if no cancer then Q3mo and if no HGD x2 then Q1yr (regardless of Tx)</p> 	<p>Tx is generally based on multiple factors including pt age/comorbidities/preference, type of HGD (unifocal vs multifocal, mucosa irregularity), experience of gastroenterologist/surgeon, etc</p> <p>(1) Endoscopic Therapy</p> <ol style="list-style-type: none"> Multimodality Therapy: EMR of nodules first and then ablative therapy of everything else (refer to cancer for types) Main Study: Shaheen 2009 NEJM compared HALO to sham for both LGD and HGD, results to the left after your treat continue surveillance at Q3-6mo no study has demonstrated that this type of therapy truly decreases the long term r/o cancer, THE BIG PROBLEM IS THE RETURN OF METAPLASIA BELOW NEO SQUAMOUS TISSUE!!! So called "Buried-Barrett's" and you can't even Bx to look for it b/c forceps do not go deep enough, in addition to the fact that you can't confidently ablate all original BE and there is a r/o stricture 5%, bleeding, perforation) <p>(2) Esophagectomy</p>
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- Diagnosis
 - 1st If Hx consistent w/ typical uncomplicated GERD then Empiric Tx (+ if >50% response in 1-2wks)
 - 2nd If Hx consistent w/ complicated GERD then EGD w/ Bx to (1) diagnosis esophagitis, (2) assess degree, (3) assess complications and (4) to exclude other cause for Sx
 - When? alarming Sx (bleeding, weight loss), Sx for >5yrs despite med, >50yo, concern for complications, other Sx to suggest another diagnosis (dysphagia, odynophagia)
 - Normal endoscopy does not rule out reflux as only 50% of pts w/ GERD have visible esophagitis
 - Bx is NOT advocated of endoscopically normal mucosa to exclude or confirm Dx only done to exclude complications or other dz esp EoE
 - Histology
 - Very Mild Intraepithelial lymphocytes/eosinophils (not so much neutrophils which suggests more infectious esophagitis)
 - increased epithelial turnover signs including basal cell hyperplasia (>15% of epithelial thickness) and increased height of rete pegs (>2/3 epithelial thickness)
 - tight junction damage resulting in intraepithelial fluid which dilates the interepithelial spaces
 - ballooning of epithelium as the accumulate glycogen in their cytoplasm

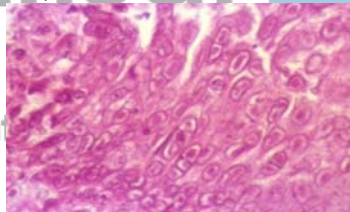


Classification	Grade	Description
Los Angeles	A	Mucosal Breaks <5mm long
	B	Mucosal Breaks >5mm long but not continuous
	C	Continuous Mucosal Breaks <75%

- NB sometimes pts with severe disease have a shortened esophagus creating a problem during surgery but a **Collis Lengthening Procedure** creates a 3-5cm neoesophagus from proximal stomach allowing the fundoplication to be placed in the abdomen under minimal tension
- Complications
 - <1% mortality 25% morbidity
 - Acute: spleen injury, esophageal perforation, pneumothorax
 - Chronic
 - (1) gas-bloating syndrome
 - (2) dysphagia w/ psuedoachalasia
 - (3) diarrhea (mechanism unclear)
 - If GERD Sx persists after 1yr then consider displaced fundoplication or damage to Vagus nerve
- Effect
 - Reduces Sx in 90% of pts
 - 7% require repeat surgery and 60% returned to regular use of medications
 - Decreased need to manage complications (aka reduced need to dilate stricture) except for Barrett's which does NOT reverse or AC which does not have a decreased risk
 - Studies indicate surgery is superior to antacids, H2B, and prokinetics but NOT PPIs
- Endoscopy (sham controlled studies are poor and/or show no benefit hence not advocated)
 - Stretta radiofrequency thermal energy ablation of the LES resulting in hypertrophy and fibrosis and thus narrowing of the distal esophageal lumen
 - EndoCinch submucosal sewing of gastric cardiac folds together (gastroplication) along the lesser curvature thus accentuating the angle of His
 - Plicator full thickness sewing of...
 - Enteryx biopolymer is injected into the LES
 - Gatekeeper biopolymer is implanted into the LES

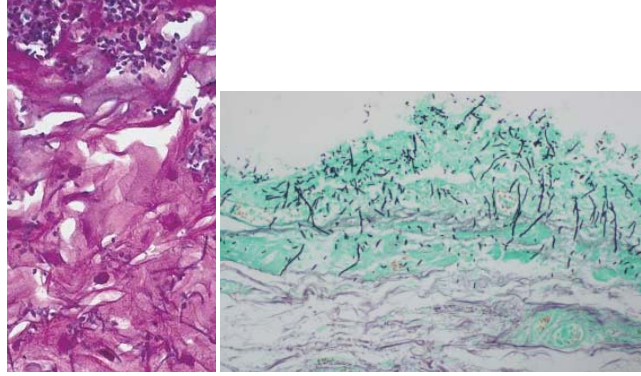
Esophagitis

- GERD Esophagitis (refer)
- Eosinophilic Esophagitis (refer to Eosinophilic Disorders, actually causes more dysphagia than odynophagia b/c chronic and w/ structural changes)
- Infectious Esophagitis
 - RFs: immunocompromised states (HIV, DM, etc), meds (Abx, topical/systemic steroids, PPI/H2B), anatomic/motility disorders, advanced age, alcoholism, cancer, adrenal deficiency
 - Non-Immunocompromised (HIV CD4 <500)
 - Viral (Cx is more sensitive than cytopathology)
 - HSV (mod inflammation w/ vesicles and small volcano ulcers, Bx side b/c HSV infects epithelial cells)

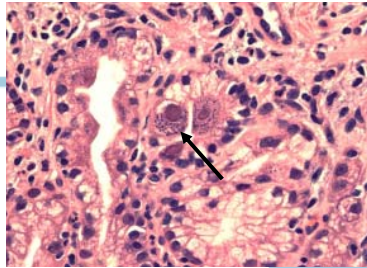


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- Fungal
 - Candida aka Monilial (20% of healthy adults are colonized and thus are asymptomatic, when the host is impaired then mucosal invasion occurs, complication: intramural psuedodiverticulosis, Px: Fluconazole 100mg PO QD or 150mg PO Qwk)
 - if pt has thrush, mild dysphagia and no odynophagia then empirically Tx for Candida alone but if no thrush or severe dysphagia or odynophagia is present or no improvement after fluconazole then do EGD to look for other causes
 - Bx: budding yeast w/ psuedohyphae or GMS stain
 - 1° Fluconazole 200mg PO x1 then 100mg PO Qd x10d
 - 2° Nystatin Swish&Swallow 100,000U/mL 4mL PO QID x10d, Clotrimazole 10mg troche dissolve in mouth slowly do not chew/swallow PO 5x/d x10d



- Parasite
 - Trypanosoma cruzi (Chagas Disease = Achalasia)
- Immunocompromised (HIV CD4 <50)
 - Viral
 - CMV (severe inflammation w/ large linear/serpiginous ulcers, 30% of pts also CMV infection in other parts of GI tract, Bx base b/c CMV infects endothelial cells)



- HIV (giant ulcers, Tx w/ steroids/thalidomide)
- VZV
- EBV
- HPV
- Fungal
 - Endemic Mycosis
 - Mucormycosis
 - Turoloopsis
- Parasite
 - Crypto
 - Leishmania
- Bacteria
 - TB (primarily infecting the esophagus or extension from mediastinum w/ sinus tracts, extrinsic compression and ulceration)
 - Tertiary Syphilis

- **Radiation Esophagitis**

- **Caustic Esophagitis**

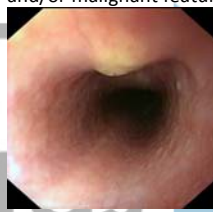
- Epidemiology
 - suicidal/intoxicated/mentally-ill adults (larger volume and don't stop when it hits the mouth therefore more GI than pharynx damage)
 - inquisitive children (opposite)
- Types
 - **Acidic** (immediate pain hence often not swallowed, bad odor/taste)
 - Eg (UN-common household items): HCl, Oxalic Acid, Nitric Acid, Phosphoric Acid, Sulfuric Acid, et al used in cleaners
 - Mechanism: **coagulative necrosis** (clumping of cell membrane → shallow penetration)
 - stomach>esophagus damage
 - **Alkaline** (delayed pain hence swallowed, tasteless/odorless)
 - Eg (common household items): NH₃ (Ammonia) & NaOH (Lye) used in cleaning product, batteries (not only objects causing obstruction but they also contain alkalotic agents and toxic metals like mercury)
 - NB b/f 1960s lye was a crystal (much more difficult to swallow resulting pharynx damage as little was swallowed) vs after 1960s lye was a liquid (swallowed much more easily resulting in more GI injury)

- (a) increase GERD (eg. anticholinergics)
 - (3) Unknown (many meds)
 - Other Factors: contact time, pill coating, etc
 - S/S
 - acute severe radiating retrosternal chest pain (always w/o a cardiac/pulmonary events)
 - odynophagia but dysphagia is uncommon
 - sometimes dehydration, fever, weight loss, hematemesis, worsening GERD may occur
 - Sx often awaken the pt in the middle of the night as pts often take pills before sleeping
 - Sx increase during the first 3-4d
 - Dx
 - **Usually occurs at around ~25cm but why?**
 - **(1) This is the transition zone from striated to smooth muscle where pressure the esophagus cannot generate pressure (refer to manometry)**
 - **(2) This is where there is extrinsic compression by vessels/bronchus/spine**
 - Clinical but endoscopy may be needed if unclear, severe Sx, prolonged Sx, complications (one or more pinpoint to circumferential discrete ulcers to diffuse esophagitis w/ pill fragments in ulcers, complications of fistula, perforation, hemorrhage, stricture, etc, typically dz occurs in mid esophagus just above indentation of aortic arch or L mainstem bronchus unless an underlying anatomic/motility disorder is present)
 - Meds
 - Anti-Bacterials: doxycycline/tetracyclins (most common b/c of its frequency rather than its propensity for injury), clindamycin, penicillin, etc (the other abx are rare)
 - Anti-Virals: Zalcitibine, Zidovudine, Nelfonavir
 - Bisphosphonates: Alendronate (highest risk, similar to quinidine), Etidronate, Pamidronate, Risedronate (lowest risk) (Bx show polarizable crystals and multinucleated giant cells)
 - Chemo: Dactinomycin, Bleomycin, Cytarabine, Danorubicin, 5-FU, Methotrexate, Vincristine
 - NSAIDs: ASA, Naproxen, Ibuprofen
 - Vitamins/Minerals: KCL, iron, Ascorbic Acid, MVI
 - Other: Kayexalate, Theophylline, Quinidine (unique in that it is very injurious causing formation of exudates that are thick enough to appear as a filling defect on esophagram)
 - Variceal Sclerotherapy: causes (1) gross structural injury 2/2 necrosis/ulceration w/ high r/o strictures and (2) dysmotility, hence not used anymore
 - Tx
 - No specific Tx that alters the course of injury
 - Aimed at symptom control w/ local anesthetics (viscous lidocaine) or systemic analgesics (opiates)
 - Prevent superimposed injury from GERD w/ PPI but no data demonstrate that hastens symptomatic/pathologic improvement
 - Discontinue offending agent and any other potentially caustic agents
 - IVF/TPN if severe odynophagia that prohibits PO intake
 - Most pts have resolution of Sx in 2-3wks
 - In general tell pts to take pills w/ >8oz of water, remain upright for >30min after ingestion, consider alternative meds or routes (liquid), that future rechallenge w/ the same med is controversial except bisphosphonates which clearly cause recurrent injury
 - Dilation in the future
- **Other: Bullous Skin Disease, SJS, Behcets**
- **Esophagitis Dissecans Superficialis**
 - Epidemiology: rare but likely unrecognized
 - Etiology: unknown
 - S/S: mucosa is vomited up, rarely dysphagia/odynophagia
 - Endoscopy: large superficial fragments of detached/sloughed mucosa (similar to bullae) w/ MINIMAL inflammation
 - Tx: none
 - Complications: despite dramatic appearance it is actually benign w/o lasting esophageal pathology

Structural Problem

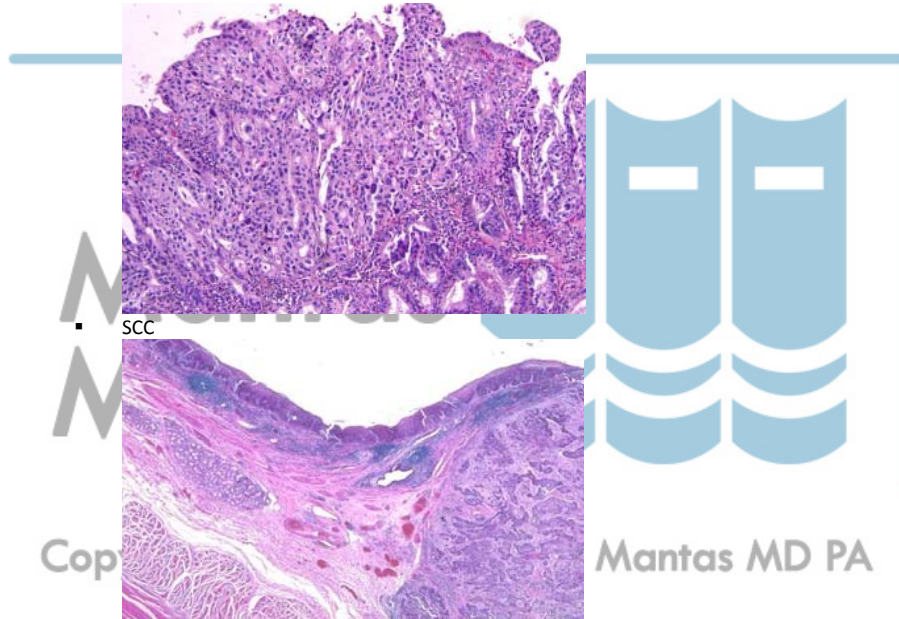
- NB congenital dz of the esophagus is actually common 1/4000 births
- External Changes
 - adjacent structures may indent esophagus specifically aortic arch, left mainstem bronchus, left atrium (enlarged w/ CHF), diaphragm
 - degenerative arthritis of the spine forming spurs
 - adhesions can develop s/p thoracotomy
- **Esophageal Atresia (EA) and Tracheo Esophageal Fistula (TEF)** (most common congenital abnormality)
 - 1/?
 - 5% EA alone ve 95% associated w/ TEF
 - Failure to recanalize = EA vs Failure of the lung bud to separate = TEF
 - most common (~90%) is the blind proximal esophagus and fistula connecting distal esophagus w/ trachea
 - if blind proximal esophagus then fetal polyhydramnios and infant excessive oral secretions w/ aspiration
 - if fistula connecting distal esophagus w/ trachea then chemical pneumonitis

- Dx: in utero polyhydramnios, inability to place NGT w/ coiling on CXR
- Tx: surgery w/ direct anastomosis or colonic interposition
- NB pts often have other associated problems esp CV defects including the VACTERL syndrome (Vertebral, Anal, Cardiac, Tracheal, Esophageal, Renal, Limb) seen in premature infants
- If pt survives Tx they are at high risk for severe GERD and its complications early in adulthood
- **Congenital Esophageal Stenosis**
 - 1/25000
 - Varies from 2-20cm in length, usually lying w/o middle/lower 1/3 of esophagus
 - Can be associated w/ EA/TEF
 - Always do EGD to Bx and determine if cause is secondary or if one of three primary processes: (1) tracheobronchial remnants suggesting that there was incomplete separation of endoderm tube from lung bud or (2) fibromuscular hypertrophy 2/2 myenteric plexus damage, (3) membranous diaphragm
 - In most cases they are mild presenting w/ dysphagia and regurgitation during childhood as more solid food is ingested
 - Tx: dilation or surgery
- **Esophageal Duplication Cyst**
 - 1/8000
 - Epithelial lined outpouching producing cystic/tubular structures
 - Cyst (round structure) much more common than tubular structures (long tube that parallels the lumen communicating at neither, either or both ends)
 - In the R posterior inferior mediastinum
 - S/S: asymptomatic or compression of tracheobronchial tree or esophagus
 - Dx: submucosal mass on esophagram, mediastinal mass on CXR
 - Tx: surgical excision to r/o a cystic neoplasm
- **Intrathoracic Vascular Anomalies**
 - 2% of the population, rarely do they produce Sx (dysphagia lusoria = "trick of nature") even if they compress the esophagus or tracheobronchial tree, b/c so common even if present you should explore other causes of Sx if the pt is symptomatic
 - **Aberrant R Subclavian Artery** specifically refers to the R subclavian artery aberrantly arising from L aortic arch and coursing to the right and posterior/anterior to the esophagus compressing it, Dx: diagonal pencil indentation at T3/4 on esophagram, during endoscopy the R radial pulse may diminish as you inflate the esophagus
 - Tx: if you decide to Tx reanastomosis to the ascending aorta is done
- **Heterotopic Gastric Mucosa aka Inlet Patch**
 - Epidemiology
 - overall incidence & prevalence is unclear but autopsy series suggest a prevalence of 4.5% and endoscopy series suggest a prevalence of 10%
 - natural history including frequency of symptoms & complications is poorly understood
 - Endoscopy
 - Location: cervical esophagus below UES
 - sometimes they are missed during intubation of EGD as the scope is quickly passed after it overcomes the resistance of UES (most of the ones I have seen have during removal of the scope)
 - Morphology: highly variable
 - microscopic foci to macroscopic islands of velvety red mucosa amid the lighter pink-colored mucosa of normal esophagus
 - single/multiple
 - extend transversally/circumferentially
 - depressed/flat/raised
 - sometimes only the complications are present (eg. stricture)
 - coexisting adjacent esophagitis is common
 - Histology
 - Oxyntic Mucosa
 - Parietal Cells (secrete HCl & IF) demonstrated by low pH proximally with normal pH distally on 24hr ambulatory pH monitoring
 - Chief Cells (secrete pepsin)
 - Surface/Neck Cells (secrete mucin)
 - Complications
 - HP Infection
 - only colonized when infection exists in the stomach
 - an interesting hypothesis is that inlets serve as a reservoir for oral-to-oral transmission and a niche where abx might have difficulty reaching
 - Gastritis w/ Subsequent Esophagitis → Tracheoesophageal Fistula, GIB, Perforation, Stricture, Barrett's and Adenocarcinoma
 - Plummer-Vinson Syndrome (US) vs Paterson-Brown-Kelly Syndrome (UK)
 - Upper Esophageal Webs + IDA + Koilonychia (spoon-shaped fingernails) + Angular Cheilitis w/ Glossitis
 - NB 10% r/o SCC in oropharynx/esophagus

- NB mainly seen in middle aged women
 - Non-Specific Oropharyngeal Sx (globus sensation, sore throat, chronic cough, hoarseness, chronic throat clearing) VERY CONTROVERSIAL
 - Tx
 - Asymptomatic Pts: no Tx, however it is argued whether Bx is necessary to confirm clinical dx and once confirmed to r/o malignant transformation furthermore surveillance is still controversial
 - Symptomatic Pts w/o Morphologic Change: thermal ablation and PPI can result in replacement of patch w/ normal esophageal tissue
 - Symptomatic Pts w/ Morphologic Change: dilation
 - Dysplasia: very unclear, thermal ablation, close surveillance, surgery
 - Cancer: surgery
- Tumors
 - **Benign Non-Epithelial Tumors** (rare)
 - Benign Sarcomas
 - Granular Cell Tumor
 - Def: benign tumor derived from Schwann cells
 - Locations: tongue (40%), skin (30%), breast (15%), GI (5% w/ 1/3 in esophagus)
 - S/S: asymptomatic or non-specific Sx depending on location
 - Complications: malignant conversion (RFs: relapse, rapid growth, focal necrosis, cell atypia or high mitotic activity)
 - Dx: EGD (small firm sessile submucosal nodules), EUS (refer), Path (large infiltrating cords of fusiform cells w/ hyperchromatic nucleus and eosinophilic granular cytoplasm)
 - Tx: conservative if <1cm and asymptomatic then EGD Q1-2yrs but if it grows and/or pt develops Sx then resection either w/ EMR/ESD unless >2cm, attached to muscular layer and/or malignant features then surgery
 - 
 - GISTs (round submucosal mass, usually distal 1/3 of distal esophagus)
 - Fibrovascular Polyp (large polyp, upper 1/3 of esophagus, usually asymptomatic but occasionally bizarre Sx of polyp regurgitation w/ asphyxiation)
 - Hamartoma
 - Hemangioma
 - Lipoma (increasing Hz as you move down the GI tract thus rare in esophagus)
 - **Malignant Non-Epithelial Tumors** (rare)
 - Mets (breast cancer, et al, signified by compression w/o mucosal disruption)
 - Lymphoma (more common in immunocompromised pts esp HIV/AIDS)
 - Malignant Sarcomas: Leiomyosarcoma, Rhabdomyosarcoma
 - **Benign Epithelial Tumors** (rare)
 - Squamous Papilloma (single small white sessile/polypoid polyp in proximal esophagus, histology shows finger-like projections of hyperplastic squamous epithelium w/o inflammation, thought to be 2/2 GERD or HPV but not entirely clear)
 - Inflammatory Fibroid PseudoPolyps aka Eosinophilic Granulomas (occurring in distal esophagus at EG jxn, thought to be 2/2 GERD)
 - Adenoma
 - **Malignant Epithelial Tumors** (AC/SCC very common others are very rare)
 - Adenocarcinoma (refer below)
 - Squamous Cell Carcinoma (refer below)
 - Verrucous Carcinoma (slow growing & mildly malignant thus favorable prognosis, exophytic papillary mass)
 - Carcinosarcomas (large polypoid lesions, often multiple)
 - Small Cell Carcinoma (most common extrapulmonary site, highly malignant)
 - Primary Melanoma
 - Mets (melanoma)
- SCC/AC (although very different in their in epidemiology/RFs/biology both SCC/AC have similar Dx/Staging/Tx)
 - **SCC** (1° middle/upper third)
 - Epidemiology
 - Most common type world-wide
 - 1/1,000 in high risk areas (Central Asia from Iran to China)
 - 1/2,000 in intermediate risk areas (Africa, South America)
 - 1/10,000 in low risk areas (Rest of the World including US and Europe esp Blacks)

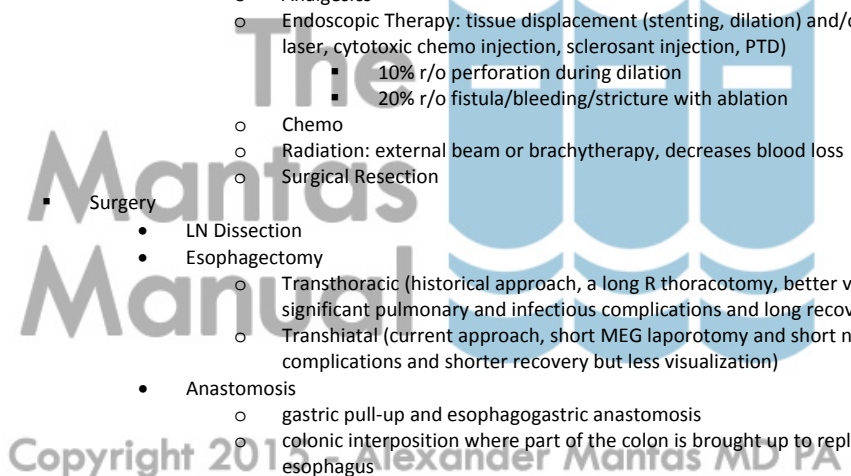
- NB geographic variation is believed to be 2/2 genetics, diet, environmental exposure, alcohol/tobacco use
- RFs (black males)
 - EtOH & Any Tobacco Containing Product (main RFs in low risk areas)
 - Diet (main RFs in high risk areas)
 - Betel Nuts (Asia, looks like nutmeg, cut into small pieces and lime paste is used to soften it up and this paste is rolled into a betel leaf and chewed, creates a red like paste, has a peppery taste)
 - Nitrates (refer to gastric cancer)
 - Vitamin/Mineral Deficiencies (however no single deficiency is directly causal)
 - Hydrocarbons given off from coal
 - Drinking very hot liquids (tea/coffee, esp in Turkey)
 - Chewing sunflower seeds
 - Caustic Ingestion
 - Red Meat
 - Other
 - SCC of Head & Neck (high r/o synchronous/metachronous lesions)
 - Infection (HPV and rarely EBV)
 - s/p partial gastrectomy (controversial)
 - Disorders
 - FHx (central China but no susceptibility gene identified)
 - Long Standing Achalasia (refer)
 - Plummer-Vinson Syndrome (refer)
 - Tylosis w/ Esophageal SCC = Howell Evans Syndrome
 - Tylosis = Hyperkeratosis (thickening) of Palms/Soles
 - Only Type A Tylosis has an associated risk of esophageal SCC and rarely other malignancies (gastric, larynx, lung, tongue, tonsil, breast, uterus)
 - AD Full Penetrance
 - Locus: 17q23-25 → Protein: Cytoglobulin (fxn unknown, this same mutation is interestingly found in some sporadic SCC)
 - Risk of 40-92% depending on the kindred studied
 - SCC usually in distal esophagus
 - 10 genealogies documented in US/UK/Germany
 - Qyr EGD and if any precancerous lesions identified esophagectomy is recommended (some have recommended prophylactic esophagectomy by age 45yo)
- Adenocarcinoma (50% of esophageal cancer and increasing and is actually the most rapidly growing cancers in the US)
 - only distal third (very difficult to distinguish cardiac AC vs junction AC vs esophageal AC)
 - RFs (white males)
 - Barrett's Metaplasia RFs (refer)
 - NB why increasing? suspected that b/c of effective Tx of HP there is overall more gastric acid output b/c HP inhibits gastric acid
 - Smoking (not alcohol)
 - BMI
 - Scleroderma
- S/S
 - S then S+L Dysphagia
 - Weight loss
 - Anorexia
 - Odynophagia (a later finding suggesting ulceration)
 - Chest Pain (a later finding suggesting neuromediastinal invasion)
 - Hematemesis (a later finding suggesting ulceration or aorto-esophageal fistula)
 - Tracheoesophageal Fistulas (very poor prognosis w/ median survival of 1-4mo)
 - Aspiration
 - Pseudoachalasia (when infiltrates the LES)
 - Acrokeratosis Paraneoplastica aka Bazex's Syndrome
 - symmetric psoriasiform dermatosis affecting hands, feet, ears, nails
 - usually manifests shortly b/f the dx of cancer of the aero digestive tract
 - paraneoplastic process (cross-reactivity b/t tumor Ag and epidermal BM Ag)
 - Tx of the cancer leads to resolution of skin lesions
 - PEx: chronically ill (weight loss, muscle wasting, etc) otherwise unremarkable unless gross metastatic dz
 - Labs: chronically ill (hypoalbuminemia, ACOD, etc) otherwise unremarkable unless gross metastatic dz
- Screening & Surveillance

- no evidence that general screening of low risk populations decrease mortality HOWEVER surveillance of high risk populations MAY decrease mortality THOUGH controversial b/c of sampling error, variability in the interpretation of dysplasia, et al
- AC: Barrett's Metaplasia (refer) vs SCC: Tylosis (refer), Caustic Ingestion (refer), Fanconi's Anemia (?)
- Dx
 - EGD w/ chromoendoscopy (endophytic mass but one can also see just submucosal infiltration w/ no appreciable mass) w/ Bx/BrushCytology (for histologic diagnosis)
 - Staging Approach
 - 1st: CT-H/C/A/P to r/o mets and clear unresectability aka Stage IV Dz
 - 2nd: if CT negative then EUS to assess T staging (ACTUALLY EMR IS A BETTER T STAGING APPROACH THEN EUS) and to guide FNA of LNs for N staging (more accurate for T staging (85%, more accurate the deeper the cancer) than N staging (75%) which is better than CT (can miss local invasion and LNs))
 - Other: PET is becoming helpful
 - Important Points
 - AC are not as locally invasive as SCC rather they tend to metastatically spread
 - In general most esophageal cancers have advanced dz at time of dx b/c the esophagus has rich lymphovascular supply and lacks a serosal lining
 - Tumors confined to mucosa/submucosa/muscle have 3/30/60% chance of LN involvement
 - 90% of pts selected for curative surgery have BM micromets
 - Adeno



- Tx
 - Endoscopic
 - Treatment (elimination of metaplasia and in situ can result in squamous mucosa regeneration)
 - Endoscopic Mucosal Resection (EMR)
 - Ablation
 - Cryo: Liquid Nitrogen Cryotherapy
 - Procedure: cool down system, do EGD exam, pass 0.037in Savary wire into stomach, remove scope, place 20F suction tube over wire into stomach w/ black mark at GEJ (liquid nitrogen expands 700x), tape over forehead and then attach to cryo machine, place clear plastic cap over tip of scope w/ soft end out, place metal introducer onto blue cap on scope, pass cryo catheter thru scope w/ end of tip right beyond cap, turn suction on and begin cryotherapy of area (once area begins to turn white (frosted) turn on timer and Tx for 20sec further then let thaw for 2min then turn off timer), Tx same site 3 times, defrost system before pulling out catheter
 - Pt will be in pain afterwards therefore give some dilaudid in post-op area and at home give Lortab 7.5/500mg per 15mL elixir (5mL PO Q2hr prn pain dispense 250mL), Mylanta/Lidocaine 2% elixir

- (10mL PO Q30min prn pain dispense 250mL), Carafate 1g/10mL solution (10mL PO Q6hrs before each meal)
- Repeat 1mo later
- NB don't if significant esophagitis b/c gas can track thru mucosal breaks
- NB always palpate stomach to make sure not getting distended
- Thermal: Radiofrequency Ablation (RFA): BARRX Halo RF Thermal Ablation, Nd:YAG Laser, MPEC, APBC
- Cytotoxic: PhotoDynamic Therapy (PTD) NOT DONE ANYMORE
 - intravenous injection of sodium porfimer (US, deep effect, more skin complication) or oral dose of aminolevulinic acid (Europe, shallow effect, little skin complication) which selectively accumulates in tumor cells and when exposed to a specific wavelength (~red) of laser delivered close x12min to the lesion the hematoporphyrin excites and produces radicals which kill the cell, high cost,
 - Chronic Complications: cutaneous photosensitivity (hematoporphyrin accumulates in the skin, 46%), strictures (38%), tracheoesophageal fistulas
 - Acute Complications: chest pain (32%), odynophagia (21%), N, F, leukocytosis, pleural effusion
- Palliation
 - nutritional support w/ J-feeding tube (no gastric placement b/c of complications w/ GERD w/ distal cancer and difficult to do if a gastric pull-up is performed)
 - Analgesics
 - Endoscopic Therapy: tissue displacement (stenting, dilation) and/or ablation (thermal, laser, cytotoxic chemo injection, sclerosant injection, PTD)
 - 10% r/o perforation during dilation
 - 20% r/o fistula/bleeding/stricture with ablation
 - Chemo
 - Radiation: external beam or brachytherapy, decreases blood loss
 - Surgical Resection
- Surgery
 - LN Dissection
 - Esophagectomy
 - Transthoracic (historical approach, a long R thoracotomy, better visualization but significant pulmonary and infectious complications and long recovery)
 - Transhiatal (current approach, short MEG laparotomy and short neck incision, less complications and shorter recovery but less visualization)
 - Anastomosis
 - gastric pull-up and esophagogastric anastomosis
 - colonic interposition where part of the colon is brought up to replace the missing esophagus
 - M&M: presently morbidity ~20-40% (anastomotic strictures, leaks, chronic aspiration, infection, chylothorax) and mortality ~3-10%
- Radiation/Chemo
 - Generally AC less responsive than SCC to chemo/radiation
 - Cisplatin, 5-FU, Epirubicin, Paclitaxel, Irinotecan
 - Neoadjuvant chemoradiation is controversial
 - Pro: Tx metastatic dz, downstage dz, better tolerated than post-op chemoradiation
 - Con: increase post-op complications, delay surgery



Stage	5yr Surv (for any given stage AC tends to do better than SCC)	TNM (developed by the Japanese Committee for Registration of Esophageal Carcinoma and thus more applicable to SCC than AC)	Tx
0		TisN0M0 = Carcinoma In Situ	Early Stage (5%) = Curative Intent
I	60%	T1N0M0 = Mucosa + Submucosa <ul style="list-style-type: none"> • T1a: Muscularis Mucosae • T1b: Submucosa • NB this distinction is a new concept b/c the lymphatic system drains submucosal "fluid" but not mucosal "fluid" (this superficial lymphatic system is unique in esophagus and is not 	

Stage HGD: EMR (nodular) vs RFA (not nodular)
 Stage 0/1a: EMR (nodular) vs Cryo (not nodular)
 b/c Cryo is deeper than RFA

Stage Ib/IIA: Neoadjuvant Chemo/Radiation +

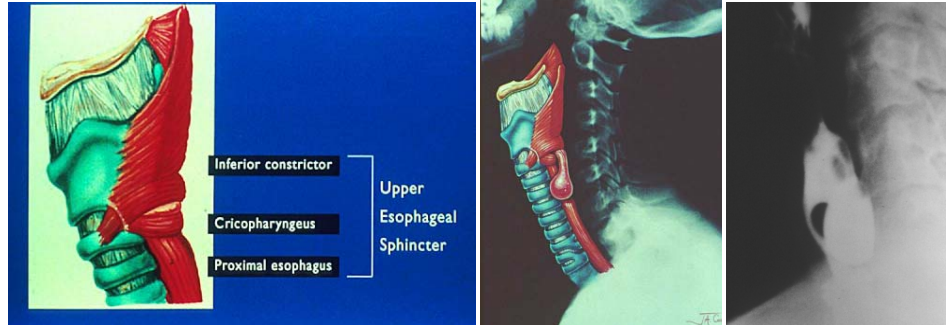
			<p>seen in other parts of the GI tract) = mucosal cancer metastasizes to LNs <2% of the time (but remember not 0%) WHILE submucosal cancer metastasizes to LNs >20% of the time</p>	Surgery + Adjuvant Chemo/Radiation
II	A	31%	T2N0M0 = Muscularis Propria	<p>Regionally/Locally Advanced & Metastatic (95%) = Palliative Intent</p> <p>Stage IIB: Neoadjuvant Chemo/Radiation then Surgery then Adjuvant Chemo/Radiation</p>
	B		<p>T3N0M0 = Adventitia</p> <p>T1-2N1M0</p> <ul style="list-style-type: none"> 1-2 Regional LNs <ul style="list-style-type: none"> Upper Regional LNs: Supraclavicular Lower Regional LNs: Paraesophageal, Sub Carinal, Para Aortic 	
III	A	20%	<p>T1-2N2M0</p> <ul style="list-style-type: none"> 3-6 Regional LNs <p>T3N1M0</p> <p>T4aN0M0</p> <ul style="list-style-type: none"> T4a: Resectable Adjacent Structures (pericardium, pleura, diaphragm) 	<p>Stage III/IV: palliation of dysphagia, bleeding, esophagorespiratory fistulas, improve nutrition, improve overall quality of life</p>
	B		T3N1-2M0	
	C		<p>T4aN1-2M0</p> <p>T4bN#M0</p> <ul style="list-style-type: none"> T4b: Unresectable Adjacent Structures (aorta, spine, trachea, etc) <p>T4anyN3M0</p> <ul style="list-style-type: none"> ≥7 Regional LNs 	
IV		4% usually 12-18mos	<p>T#N#M1</p> <ul style="list-style-type: none"> M1a: Non-Regional LNs <ul style="list-style-type: none"> Upper Non-Regional LNs: Cervical Lower Non-Regional LNs: Celiac M1b: Organs <ul style="list-style-type: none"> Liver, Lung, Bone, Brain 	

- Foreign Bodies (refer)
- Diverticula

- Pharyngeal

- Zenker's Diverticulum

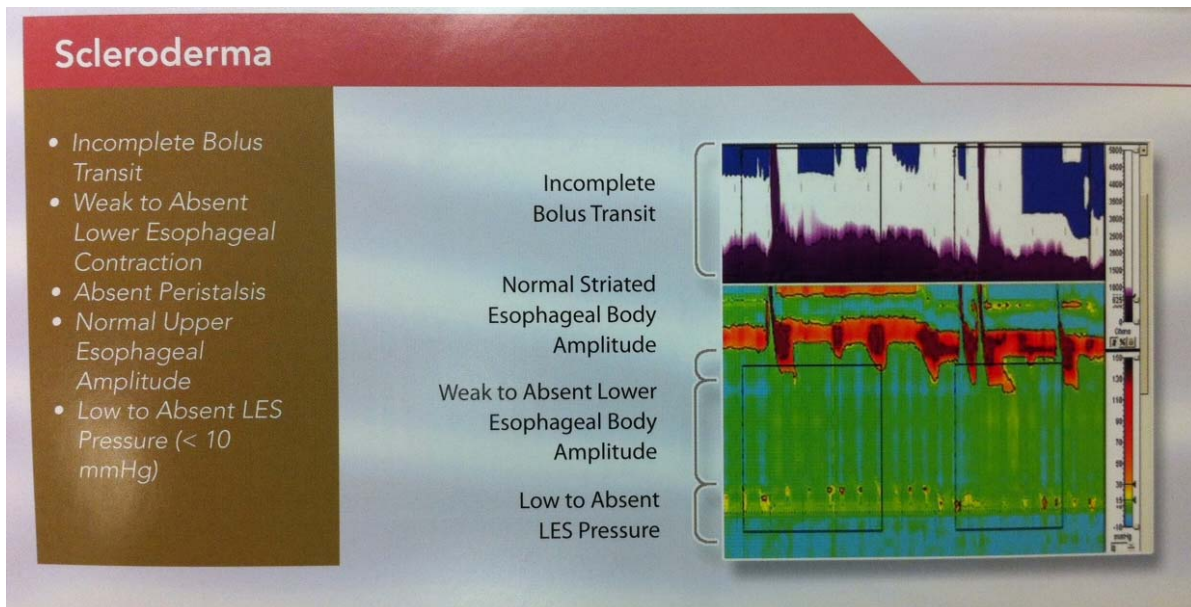
- Pt: old male
- Epidemiology: 1% of esophagrams!!!
- Mech: after years of swallowing some people develop fibrosis at cricopharyngeus → impaired cricopharyngeal compliance → high pressure during swallowing must go somewhere → diverticulum specifically at Killian's dehiscence (an intrinsically weak triangular area b/t inferior pharyngeal constrictors and cricopharyngeus in the posterior pharynx)
- S/S: usually asymptomatic but if symptomatic then dysphagia, regurgitation, choking, voice changes, halitosis, weight loss, lump on side of throat, "gurgling in the neck"
- Complications: aspiration, lodging of meds, difficulty intubating or figuring which lumen is esophagus/diverticulum during endoscopy w/ perforation, ulceration, SCC
- Tx: only if symptomatic then surgical diverticulectomy/diverticulopexy (if >5cm and young/healthy) vs endoscopic diverticulotomy (<5cm or old/comorbidities)



- Esophageal
 - **Epiphrenic/Traction Diverticulum**
 - Mech: Epiphrenic Diverticula (lower 1/3) 2/2 dysmotility vs Traction Diverticula (middle 1/3) 2/2 dysmotility also (but it was originally believed to be 2/2 mediastinal inflammation from infection (esp TB) or cancer resulting in scarring retraction of the esophagus)
 - Complications: esophagobronchial fistulas, perforation
 - S/S: usually asymptomatic but if symptomatic then dysphagia, regurgitation, reflex, weight loss, chest discomfort
 - Dx: b/c 2/2 dysmotility manometry should be check
 - Tx: only if symptomatic then surgical inversion or resection w/ myotomy
 - **Intramural Pseudo Diverticulosis**
 - Epidemiology: 1% of barium swallows!!!
 - Mech: dilation of submucosal glands often associated w/ states causing chronic esophageal inflammation esp Candidiasis and corrosive ingestion, appear as multiple tiny flask shaped outpouchings <5mm in size on barium swallow, often missed on endoscopy but can look like little divots
 - S/S: usually asymptomatic, ? increased r/o SCC
 - Tx: no treatment except at underlying inflammatory problem
- **Rings & Webs & Strictures** (all are likely due 2/2 GERD in fact PPI decreases size but can be genetic in some cases, Tx: dilation)
 - **Rings** (completely circular structures located at the vestibule aka LES)
 - **Type A/Muscular/Proximal Rings:** vary rare, older pts, located at proximal LES, BROAD (~5mm) band made of mucosa/submucosa/hypertrophied muscle, usually asymptomatic, likely 2/2 dysmotility
 - **Type B/Mucosal/Distal Rings:** seen in 4% of EGDs, younger pts, located at distal LES, THIN (~2mm) membrane made of only mucosa/submucosa w/ esophageal squamous tissue above and gastric columnar tissue below, if symptomatic then called "Schatzki Ring", likely 2/2 to reflux b/c almost all have a hiatal hernia, usually asymptomatic
 - **NB Corrugated Ringed Esophagus Syndrome** (multiple concentric evenly spaced rings, now likely EoE)
 - **Webs** (not circular but partial (sparing posterior side) thin mucosal fold)
 - **Regular Web:** can occur anywhere along the length of the esophagus but usually anterior wall of upper esophagus, often missed on endoscopy
 - **NB Plummer-Vinson or Paterson-Kelly-Brown Syndrome** (refer)
 - **Strictures** (smooth walled tapered circumferential narrowing not a distinct structure like ring/web, usually in lower esophagus esp at EGJ as they move up clinician should be more concerned about Barrett's or cancer, regardless always bx to be sure)
 - Benign (peptic, post-sclerotherapy/PDT, radiation, caustic, pill induced, epidermolysis bullosa, EE, anastomotic) vs Malignant (if tight stricture use brush cytology)
 - Simple (symmetric/concentric/non-angulated, traversable via endoscope, single, short <2cm) vs Complex (opposite)
 - Etiology: GERD or occurring 1-2wks after caustic ingestion or after any other cause of esophagitis
 - Proximal (eosinophilic) vs distal (GERD)
 - S/S: usually asymptomatic except when diameter is <13mm then causing dysphagia, NB sometimes if distal can manifest as a decrease in reflux symptoms b/c the ring/stricture acts as a barrier to further reflux, sometimes regurgitation
 - Tx: if symptomatic (refer)
- Extrinsic Process that can affect the esophagus
 - Diffuse Idiopathic Skeletal Hyperostosis
 - Retropharyngeal Abscess
 - Posterior Mediastinal Mass
 - Thyroid Enlargement
 - Aneurysm
 - Cervical Osteophytes
 - Tumors
 - LAD

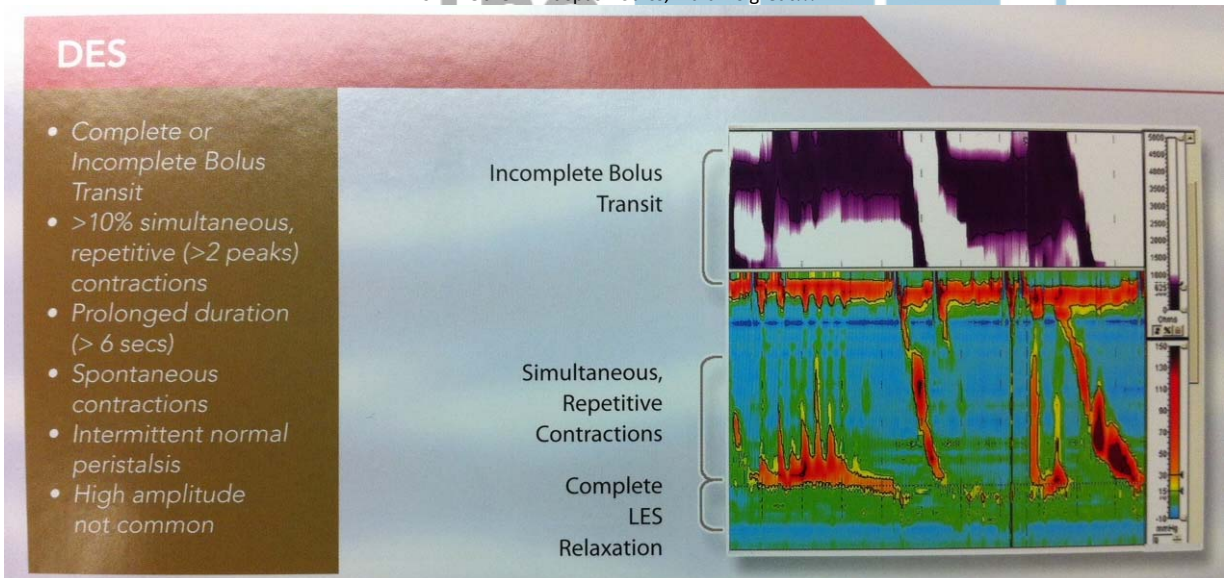
Smooth Motility Problem

- Classifications
 - Spechler & Castell Classification (most disorders fall predominantly into one category but there is considerable overlap)
 - Chicago Classification (newer classification system that may be adopted but uses more complex parameters)
- Hypomotility w/ Normal EGJ
 - Etiology
 - **Idiopathic**
 - **Chronic Atypical GERD** (not heartburn but pulm Sx, Sx do not improve after medical Tx but surgery may help)
 - **Aging Presby-Esophagus**
 - **CTD esp Scleroderma** (most common!!!, refer, aperistalsis + weak to no LES pressure)
 - **DM Autonomic Neuropathy** (60% of DM pts who have neuropathy but often under reported by pts)
 - **Amyloidosis**
 - **Hypothyroidism**
 - **Alcoholism**
 - **CIPO**
 - **MS**
 - Classes (can have one or both)
 - **Hypotensive Peristalsis aka Ineffective Esophageal Motility (IEM) → Absent Peristalsis**
 - **Hypotensive LES**
 - S/S: results in severe GERD, increased r/o pill esophagitis, food bolus

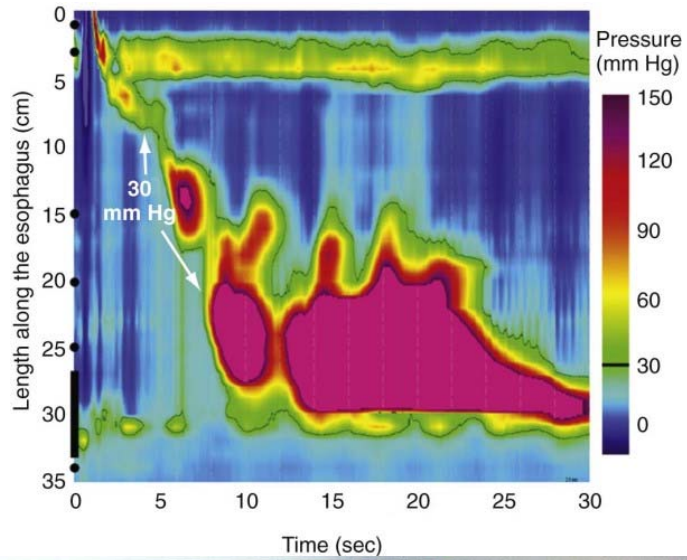


- Eumotility but Uncoordinated w/ Normal EGJ
 - **Diffuse or Segmental Distal Esophageal Spasm (DES)**
 - Etiology
 - Idiopathic
 - Hypersensitive response to acetylcholine and esophageal luminal contents (however no clear neuromuscular pathology is seen unlike in achalasia)
 - Epidemiology
 - 40yo
 - F>M
 - Mechanism
 - Spontaneous non-peristaltic contractions of simultaneously multiple segments of the esophagus preventing appropriate advancement of food bolus
 - Often precipitated by cold or carbonated liquids and occult reflux
 - Unlike in achalasia these pts sometimes have normal periods of peristalsis
 - LES functions normally
 - Clinical Features
 - 1° CP (85%) that mimics angina but more severe, a dull residual discomfort persists after the severe episode abates (unlike cardiac pain), swallowing is not impaired during pain episode, episodes lasts minutes to hours, cp rarely correlates w/ motor dysfunction (eg. pt can have cork-screw w/o Sx and vice versa) suggesting that CP reflects sensory disturbances and psychologic characteristics!!!

- 2° Solid & Liquid Dysphagia (45%) does not correlate w/ cp, usually not as severe/progressive as other motility disorders w/o complications like weight loss, regurgitation, et al
- Other: heartburn, underlying psych dz (anxiety/depression/somatization), functional bowel disease
- **Diagnosis**
 - Always rule out cardiac dz and reflux (DES and CAD/GERD can even co-exist) w/ stress test and pH probe
 - Esophagram: after swallow there is “Cork-Screw” “Rosary Bead” “Curling” appearance to body of esophagus w/ tertiary contractions
 - EGD: mainly used to r/o other causes of dysphagia but in general it is normal
 - Manometry: true diagnostic test ($\geq 20\%$ water swallows produce simultaneous (defined as a **velocity above ULN) non-peristaltic** (unlike nutcracker) contractions restricted to the smooth muscle portion that are otherwise normal in amplitude/duration/Hz/morphology (sometimes they are abnormal) it’s just that they occur simultaneously, you can also see hypertensive LES, contractions can be spontaneous/repetitive/multi-peaked
 - NB these findings are intermittent as pt can have normal esophagus later on differentiating it from achalasia which is constant and progressive
- **Treatment**
 - R/o other causes for CP (CAD, GERD, etc)
 - Lifestyle Changes
 - Avoid stress and cold/carbonated liquids
 - Medical Therapy
 - If GERD then PPI otherwise note helpful
 - TCAs (esp Imipramine), SSRIs (esp Sertraline), SMs (esp Trazodone) appear to diminish visceral sensitivity
 - Smooth Muscle Relaxants w/ Short and Long Acting Nitrates w/ Isosorbide/Sildenafil, CCBs w/ Diltiazem, et al
 - **Other: Antispasmodics, Librax is great!!!**



- Hypermotility w/ Normal EGJ
 - **Non-Spastic Disorders**
 - General
 - Controversial
 - These disorders are non-progressive or fatal and Tx is directed at Sx reduction
 - **Hypertensive LES**
 - Define: high resting LES w/ or w/o high residual LES after relaxation
 - **Nutcracker/Jackhammer Esophagus**
 - Define: spontaneous high pressure (DEA or DCI above ULN) **peristaltic (unlike DES)** contractions



Nutcracker Esophagus

- Complete Bolus Transit
- Peristaltic or Fast Antegrade
- High Esophageal Amplitude with Prolonged Duration
- Normal or High LES Pressure
- Complete or Incomplete LES Relaxation

Complete Bolus Transit

UES

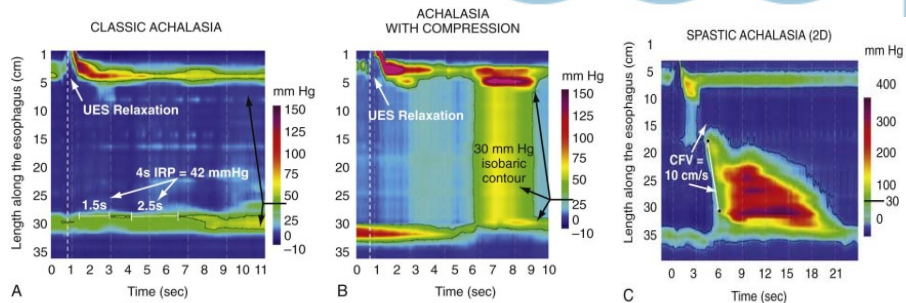
High Esophageal Body Amplitude

Incomplete LES Relaxation

- Mixture of Stuff but specifically Impaired EGJ Relaxation
 - **Achalasia**
 - Epidemiology
 - Any age from childhood to adulthood but most pts are b/t 30-60yo (based on the cause)
 - Incidence 0.5-1/100,00 per year, Prevalence 7.9-12.6/10,00
 - Familial clustering has been noted in some cases
 - No gender/race predilection
 - Etiology
 - Primary aka Idiopathic
 - Theories: Infection (HSV, Measles, JCV, etc), Autoimmune (anti-neural Ab, etc)
 - Secondary (NB "Pseudo-Achalasia" should be used for the secondary causes in which there is not so much nerve damage but rather another process like tumor presence, infiltration, etc)
 - Distal Esophageal / Upper Gastric Tumor or Mets (breast)
 - tumor physically encircles the distal esophagus and/or tumor infiltrates the neural plexus
 - always consider in an elderly pt w/ a SHORT h/o achalasia and B symptoms
 - LES is tight in achalasia but one should still pass it w/ an endoscope but if you can't then cancer should be considered
 - Closely retroflex and Bx and suspicious lesions but many times these cancers are infiltrating such that 20% of EGD will miss cancer hence some use EUS or do random Bx
 - Chagas Dz from *Trypanosoma cruzi*

- Vector: Triatominae spp (called “kissing bug” b/c these insect fall from the ceilings at night and bite people’s faces while defecating near wound site, subsequent scratching of wound allows entry parasite into body) NB cases of blood transfusion have been documented
 - Geography: Tropical Americas
 - S/S: acute flu-like illness w/ localized swelling at site of entry (eg. Romaña’s Sign = eyelid swelling where parasite is rubbed into eye) and if not treated then 30% will developed chronic infection several years later affecting GI tract (megaesophagus/intestine/colon), heart (dilated cardiomyopathy), CNS (peripheral neuropathy)
 - Mech: amastigotes collect in neurons of target organs causing cell death
 - Dx: blood smear and various immunoassays
 - Px: no vaccine just insecticides in paints and mosquito nets
 - Tx: antiparasitics helpful more in acute than in chronic illness, CCB/nitrates is very effective in esophageal disease
 - Familial: 4A aka Allgrove’s Syndrome (Achalasia + Alacrima aka defective tear production + ACTH insufficiency + Autonomic abnormalities)
 - Gene: AR Mutation of Type II Keratin Gene (12q13)
 - Protein: ALADIN (fxn unknown)
 - Infiltrative Disorders (Amyloidosis, Sarcoidosis)
 - Paraneoplastic Syndromes w/ Anti-Hu/PCA/Yo (SCLC, et al)
 - Myopathies (CIPO, Muscular Dystrophy, et al)
 - Neurodegenerative Disorders (Parkinson’s, Hereditary Cerebellar Ataxia, et al)
 - Post Anti-Reflux Surgery
 - Mechanism
 - Degeneration (loss of neurons #, loss of myelin, disrupted axon membrane, T-cell/eosinophil/mast cell infiltrate) of the NO producing inhibitory neurons (ACh producing stimulatory neurons are spared) in the myenteric plexus (Wallerian degeneration can extend into the Vagus and Dorsal Root Ganglion can be seen and rarely the primary lesion could involve these more proximal neurons) that innervate esophageal smooth muscle resulting in...
 - During rest: increased LES resting tone
 - During swallow: failed/incomplete LES relaxation AND disorganized or a- peristalsis
 - NB there is some evidence of achalasia affecting stomach, pylorus, sphincter of Oddi esp if more proximal nerves are affected
 - S/S (Sx are typically present for 4-6 yrs before a diagnosis is made, Eckardt Score System is used to assess clinical changes following Tx)
 - S+L Dysphagia and pts often try contortions to help food move down (eg. raising arms over head, straightening back, standing erect, et al)
 - Food gets stuck resulting in eventual delayed Regurgitation of undigested food (eg. often waking up with dinner in their mouth pts wake up with dinner in their mouth)
 - Chest Pain & GERD (food that retains in esophagus will ferment producing lactic acid creating reflux like Sx, anti-secretory therapy does not work, some have tried giving pancreatic enzymes to digest retained food but the esophagus is also digested, these Sx often occur early in course of dz where imaging is nl hence these pts are often sent for anti-reflux surgery and that is why manometry is always checked b/f surgery!!!)
 - Complications
 - Stasis leading Erosive/Candidiasis/Pill Esophagitis
 - Aspiration Pneumonia
 - Weight Loss
 - SCC
 - 2/2 long standing stasis which leads to prolonged contact of noxious agents
 - SCC but in distal middle 1/3 mainly in the dilated portions so typically pts do not manifest symptoms of obstruction
 - 0.3% r/o at 20yrs from Sx onset (very low but increased compared to general population)
 - ASGE recommends surveillance after 15yrs of untreated achalasia
 - Diagnosis
 - 1st Esophagram
 - normal → (EARLY)
 - “Bird’s Beak” LES constriction, dilation of distal esophagus w/ epiphrenic diverticulae, retained food/saliva that rises to the top of barium forming a distinct air-food-barium level →
 - esophagus rotates to the right and a right angle is formed between the distal esophagus and LES forming a “Sigmoid Configuration” (LATE)

- NB CXR (widened mediastinum and absence of gastric air bubble b/c air cannot easily get into stomach from esophagus)
- 2nd EGD
 - even though you can make a dx w/ esophagram it is important to do an EGD to rule out pseudo-achalasia esp adeno at EGJ as a cause of achalasia and to rule out SCC as a complication of achalasia
 - Keep these pts on clear liquid diet for 2d and NPO for 1d prior
 - Corresponding findings on esophagram can be seen
 - LES does not open spontaneously to allow effortless passage into the stomach but unlike obstructions caused by neoplasms or strictures the contracted LES of achalasia usually can be traversed easily with gentle pressure of the endoscope
 - Dilated esophagus
 - Retained food occasionally w/ esophagitis
- 3rd Manometry (true diagnostic test, findings are in the order of importance such that ~100% have aperistalsis, 60% have incomplete LES relaxation, 30% have hypertensive LES, only 20% have all three findings)
 - (1) aperistalsis in the smooth muscle body of the esophagus or simultaneous non-peristaltic (you can't calculate velocity b/c so fast) isobaric low amplitude (<40mmHg) esophageal contractions after WS
 - NB in Vigorous/Spastic Achalasia a WS produces stronger contractions that may look like DES BUT achalasia pt never have normal peristaltic waves unlike in DES
 - NB resting intraesophageal pressure is higher than intragastric pressure (the reverse is normal)
 - NB during swallow the pressure increases and the pressures are identical across the length of the lower body aka isobaric
 - NB esophageal shortening w/ elevation of the LES
 - NB peristaltic failure can be seen in disorders that cause hypomotility (refer above) BUT resting LES pressure is low and LES relaxation is complete
 - (2) Incomplete LES relaxation after WS
 - NB occasionally relaxation is complete but still impaired b/c delayed onset or shortened duration
 - (3) Hypertensive LES
 - Three Types
 - Type I Classic (25%, ½ respond to therapy)
 - Type II Compression (50%, ~100% respond to therapy)
 - Type III Spastic (25%, ¼ respond to therapy)



▪ Treatment

- Lifestyle Changes: reduce bite size, chew food well, eat slowly, drink liquids w/ meals, stay upright after you eat so that gravity helps with propulsion, avoid eating w/in 2-3hrs of bedtime, keep HOBE
- Approach
 - **Low Surgical Risk (ideal): 1° Surgery (90% response at 1yr) 2° Dilation (75% response at 1yr)**
 - NB never dilate if pt is NOT a surgical candidate b/c if they perforate you have to operate
 - **High Surgical Risk (elderly, abnl anatomy, etc) or Unwilling to Have Surgery → Botox (60% response at 1yr) and if fails then Medical (50% response at 1yr)**
- Types
 - Goal: reduce LES pressure so as to promote effective esophageal emptying (no Tx reliable restores function to the body)

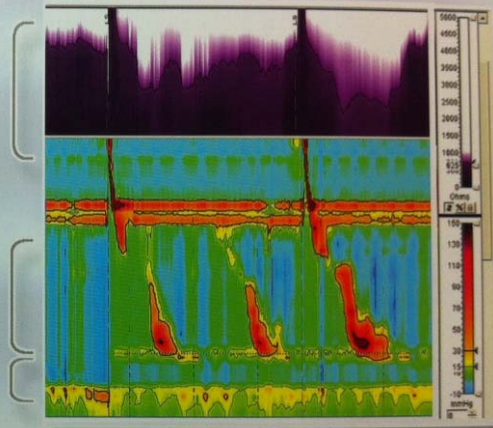
- Medical (inconvenient, not very effective, many SEs, tachyphylaxis, not give PO b/c it will just sit in the esophagus therefore give SL)
 - Block Smooth Muscle (Nitrates are better than CCBs but have significant SEs)
 - Isosorbide Dinitrate 5-20mg SL 15min before each meal
 - Nifedipine IR 10-30mg SL 15min before each meal
 - Block cGMP
 - Viagra
 - NB Block Parasympathetic Nerves from innervating muscle and contracting them: Antimuscarinics (not really used)
- BoTox (*Clostridium botulinum* Toxin)
 - Mech: binds presynaptic cholinergic neurons, internalizes and irreversibly interferes w/ acetylcholine release by preventing NT vesicle from fusing w/ axonal membrane
 - How? comes in a stock vial of 100U dry BoTox, add 5mL of NS to rehydrate to create a 20U/mL solution (must use w/in 4hrs), inject 1mL using a 5mm-sclerotherapy needle into the LES 1cm above Z-line at four quadrants deep into muscle not mucosal or submucosal
 - Effectiveness: 85% effective but 50% recur w/in 6mo b/c of regeneration of affected receptors, older pts and those with “vigorous esophagus” do better, expensive
 - SEs: acute chest pain and rash, long term LES fibrosis
 - F/U: effect lasts ~6mo in 50% of pts thus most need repeat Tx thereafter b/c new axon sprouts appear
- Dilation
 - History: first described by Sir Thomas Willis in 1672 using whale bone
 - Mech: partially tear LES
 - Factors that indicate that dilation will be more effective: older pt, longer duration of Sx, females
 - Before: clear liquids for 1-3d, NPO after MN
 - How? determine LES, mark w/ paper clip under fluoro, pass wire, feed balloon over it w/ waist at fluoroscopically determined LES, pneumatic dilation w/ 30mm balloon, hold dilation for 1min, always perform a post dilation gastrograffin esophagram, watch pts for 6hrs, can increase >4wks apart to 35mm and then 40mm (after three dilations surgery should be strongly indicated)
 - Complications: perforation 3-6% (RFs: sigmoid, previous myotomy, epiphrenic diverticula, large hiatal hernia), GERD 2%, mortality 0.2%
- Surgical
 - History: first described by Ernst Heller in 1913
 - How? laparoscopic thoracic or abdominal Heller myotomy (circular layer of LES muscle is divided down to mucosa and extends 1cm down into stomach and 3cm up into esophagus) ± “loose” antireflux surgery is also always done b/c pts often have GERD after LES disruption
 - NB esophagectomy is reserved for pts w/ megaesophagus (>8cm) or low LES pressure
 - Complications: dysphagia 20% (2/2 incomplete myotomy, stricture, too tight anti-reflux surgery, paraesophageal hernia therefore check post-op esophagram and EGD), GERD 10% (particularly bad b/c the esophageal body is atonic), mortality 0.3%

Hiatal Hernia

- Double High Pressure Zone
- May have Normal LES Pressure & Relaxation
- May have short Esophageal Length

Bolus Transit: May or may not be Complete UES

Normal Esophageal Body Amplitude. May have Short Esophageal Length Double High Pressure Zone



The
Mantas
Manual



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