1. (Amy) Tell us about radiologic testing for dysphagia. What is the difference between a barium swallow and a modified barium swallow; what are indications for each?

Radiologic studies should be performed based upon the associated history.

Plain Films: Helpful in diagnosing epiglottitis, retropharyngeal abscess, radiopaque foreign body. Inexpensive and fast. Does not detect mechanics or mucosal features.

Bolus Scintigraphy: Radioactive isotope with short half-life mixed with single swallow bolus. Gamma camera counts radioactive particles. Able to quantify fraction of bolus aspirated. Not widely available and does not test different bolus consistencies.


Barium Esophagram: Cup of barium swallowed and followed fluoroscopically to stomach. Images are on plain film; not a dynamic study. Good anatomic detail, widely available. Involves radiation exposure. May be difficult in bedridden tests. Ideal for detection of structural disorders (stricture, diverticulum).

Modified Barium Swallow: Different consistencies of material are swallowed in the presence of a speech pathologist and radiologist. The patient is upright and given puree, liquid, and/or solid barium (e.g. solid bolus to evaluate obstruction, liquid bolus if concern for aspiration). The images are recorded using fluoroscopy. For the oral/pharyngeal portion, a mouth full of liquid barium is given and held for 10 seconds prior to swallowing. Volume of the swallow, leakage into the nasal cavity, and entry into the laryngeal vestibule can all be seen in this portion of the exam. If contrast enters the vestibule and the patient coughs immediately, the entry may only have been incidental. However, entry into the larynx without the patient's awareness is abnormal. "Penetration" involves entry of contrast into the larynx during swallowing and is distinguished from "aspiration", which involves entry of contrast into the airway before or after swallowing. The pharyngoesophageal portion of the exam which can identify anatomic weakness or an inability to relax at the end of the pharyngeal stage. Finally, the esophageal segment is performed with the patient both erect and supine to test the adequacy of peristalsis. The modified barium swallow is a comprehensive test that evaluates all phases of swallowing and gives good anatomic detail. It is useful in testing the ability to protect the airway during swallowing. Disadvantages include radiation exposure, difficulty in scheduling/协调, logistics in bedridden patients, and the inability to directly assess sensation.

2. (Deya) List common congenital malformations of the esophagus and treatment for each.

1. Esophageal atresia & TEF

   a) Classification: classified based on presence of atresia & relation of fistula to location of atresia (see figure)

   b) Associations: commonly a/w other embryologic abnormalities, referred to by the acronym VACTERL (vertebral, anal, cardiac, tracheoesophageal, renal, & limb anomalies). 50% chance of having one of these associated anomalies, cardiac most common. Also commonly a/w chronic esophageal dysmotility 2/2 decreased number of Auerbach plexuses, & impairment of pulmonary development 2/2 pressure on the trachea & drainage of amniotic fluid out of the pulmonary tree.

   c) Sx’s, signs: often asymptomatic at birth

      • can then present with excessive drooling, feeding intolerance, aspiration

      • TEF w/o EA often diagnosed at later stage with chronic upper respiratory symptoms & chocking, repeated pneumonias, or asthmatic symptoms

      • can have gastric distention resulting from the passage of air from the trachea to the distal esophagus & chemical tracheobronchitis 2/2 reflux into trachea (type 1)

   d) Dx:

      • Curled NGT in proximal esophagus

      • AXR: TEF to distal esophagus → gas in stomach & small bowel. Gasless abdomen → suggests EA w/o TEF or proximal fistula

      • May also perform barium study, bronchoscopy, & esophagoscopy for further information

      • Echo: to r/o cardiac anomalies & determine side aortic arch for surgical approach (L vs R thoracotomy)

      • R/o rest of VACTERL including renal ultrasound.

   e) Treatment:

      • Historically the Waterston classification has been used for risk evaluation & determination of surgical timing. Category A (birth weight > 5.5 pounds) → prompt surgical correction. Category B (4.0–5.5 pounds or p/w PNA) → short-term delay
of surgical intervention with G-tube prior to repair. **Category C** (< 4.0 pounds or severe PNA & congenital anomaly) → staged repair. **Currently, most children, with the exception of the most ill infants, undergo early complete repair.**

- **Surgery:** performed through lateral thoracotomy. Long gaps b/w esophageal pouches → may attempt serial stretching of proximal segment, proximal circumferential or proximal spiral esophagomyotomies, or esophageal replacement with interposition of colon. Complications of surgery include anastomotic leak, anastomotic stricture.

**f) Prognosis:** lethal if not corrected.Pts with VACTERL association have poorer prognosis. Current survival rate of postsurgical repair is reported to be > 90%.

2. **Laryngotracheoesophageal Cleft** - rare defect related to esophageal atresia & tracheoesophageal fistula occurring in the midline b/w the trachea & the esophagus. Can be minimal or extend down past the carina. Symptoms range from chronic cough to respiratory distress. Diagnosed by rigid bronchoscopy. Severe cases require operative repair involving a right anterolateral cervical approach with lateral pharyngotomy to expose the defect.

3. **Esophageal Stenosis** - rare congenital anomaly. Anatomically, can be tracheal elements in the wall of the esophagus or a mucosal web. Usually present later in life with dysphagia with solids. Diagnosed by barium swallow & esophagoscopy. Dilatation effective for patients with only muscular stenosis. Segmental resection may be required for rigid defects as is found with cartilaginous remnants.

4. **Achalasia** - insufficient LES relaxation and loss of esophageal peristalsis. May be hereditary, degenerative, infectious or autoimmune. Pathologic changes in myenteric plexuses and patchy lymphocytic and eosinophilic infiltrates. Sxs include dysphagia to solids then liquids, regurgitation, and chest pain. Diagnosed by barium swallow → loss of peristalsis in upper 2/3 of esophagus. May see esophageal dilation with smooth tapering of lower esophagus leading to closed LES → “bird beak.” May also use esophageal manometry for diagnosis. All pts should have upper endoscopy to t/o pseudoachalasia 2/2 tumor at GE junction. Treatment consists of graded pneumatic dilation (risk of perforation) and surgical myotomy (antero Heller’s myotomy of LES). Recently, Botox injections into LES have shown promise but have high recurrence rates (used for high risk pts).

5. **Esophageal diverticula** (see question 7)

3. (Rosow) Management of esophageal foreign bodies. How do you manage sharp objects? When are esophageal foreign bodies an emergency?

   - **Patients in unstable condition**
     - Patients with airway compromise; drooling; inability to tolerate fluids; or evidence of sepsis, perforation, or active bleeding.
     - Treatment includes airway management as indicated, followed by imaging (if time allows), then urgent endoscopic removal.
     - Patients who have ingested button batteries are in unstable condition. The presence of a button battery in the esophagus is a medical emergency because necrosis of the esophageal wall may occur within hours. These button batteries must be expeditiously removed. Button batteries in the stomach can be allowed to pass but must be monitored radiographically to observe for disruption of the battery. Follow-up radiographs are needed in 24-48 hours. If the battery is still in the stomach, endoscopic removal is indicated.
     - Patients in stable condition
   - Radiographically localize radiopaque objects. If the foreign body is sharp, elongated (>5 cm in esophagus), or multiple in number, the patient should undergo endoscopy. Sharp objects, such as pins, razor blades, toothpicks, and chicken/fish bones, should be removed endoscopically on an urgent basis because up to 35% of these sharp objects perforate the bowel wall if not removed. Most smaller, sharp foreign bodies, such as straight pins, are believed to transit the GI tract without difficulty, as the peristaltic action carries the blunt end first; however, some recommend endoscopic removal for these as well. If the foreign body is smooth or blunt, consider the following modalities:
     - Endoscopy
     - Foley catheter removal
     - Bougie
     - Sphincter relaxation if lodged at LES.

   - For patients whose history strongly suggests an ingestion of a nonopaque foreign body such as a plastic object, toothpick, or aluminum soda can tab, consider CT scanning and refer for endoscopy. When the history is less clear about the definitive swallowing of a nonradiopaque foreign body, obtain CT scanning and refer for endoscopy if the foreign body is localized in the esophagus.

   - Smooth foreign bodies, such as coins or marbles, almost always transit the GI tract without any difficulties. Coins lodged in the distal esophagus of healthy children spontaneously pass into the stomach in up to 60-80% of cases, usually within several hours of presentation.

4. (Kathy) Diagnosis and management options of GERD. Explain Demeester scoring.

   - **Clinical diagnosis** (accurate in ~70% of patients) is made based on:
     - Esophageal sx- heartburn, regurgitation, belching, atypical chest pain, or dysphagia.
     - Laryngeal/Respiratory sx- hoarseness, cough, sore throat, globus sensation, wheezing.
Dec 20: Esophageal Disorders/Dysphasia (updated 08/06)

Two mechanisms have been postulated for GERD-induced respiratory symptoms: 1) Vagal reflex arc resulting in bronchoconstriction; 2) Microaspiration into the tracheobronchial tree. ENT abnormalities such as laryngitis are instead secondary direct damage of upward extent of acid.

**Diagnostic Tests:**

- **Barium Swallow** - Detects anatomic abnormality such as presence of hiatal hernia or stricture. Not diagnostic of GERD, however, since a hiatal hernia of reflux of barium can be present in patients who do not have GERD.

- **Endoscopy (EGD)** - Detects mainly complications of GERD (esophagitis, Barett esophagus, and stricture), and excludes other pathology (esophageal, gastric, or duodenal). Its value is limited as only 50% of patients with GERD have esophagitis.

- **Esophageal Manometry** - Provides information about LES, including resting pressure, length, and relaxation, as well as about the quality of esophageal peristalsis. In about 40% of patients, the LES pressure and peristalsis are normal. This is also essential for proper placement of pH probe.

- **pH Probe** - This is the most reliable test of GERD, with sensitivity and specificity of about 92%. Indications for this include but is not limited to: 1) patients who do not respond to medical therapy; 2) patients who relapse after the discontinuation of medical therapy; 3) evaluation prior to antireflux surgery; and 4) when atypical sx are present. A pH probe with two sensors are used to determine the upward extent of reflux (located 5 and 20 cm above the LES), and tracing is analyzed for a temporal correlation between sx and reflux episodes.
  - **Demeester Scoring** - The DeMeester score uses 6 parameters (supine reflux, upright reflux, total reflux, number of episodes, number of episodes longer than 5 min, and the longest episode) to calculate a score that indicates the severity of reflux. It is calculated from the equation (patients value - mean)/standard deviation +1 for each parameter, the final score being the sum of the scores for the 6 different parameters.

**Treatment:**

- **Lifestyle Modifications** - Avoid fatty, spicy foods, alcohol, smoking, chocolate, and coffee. Eat frequent small meals and no less than 2 hours prior to bed.

- **Medications** - PPIs, H2 blockers, antacids.

- **Laparoscopic Fundoplication** - This procedure increased the resting pressure and length of the LES and decreases the number of transient LES relaxations. Ideal for the patient who has a good response to PPIs, is young and chooses operation early in the course of disease to avoid a lifelong commitment to lifestyle change and medications, and in whom complications such as strictures are rarely seen.

5. (Josh) Surgical and nonsurgical treatment for esophageal hypertonia and dysphagia after laryngectomy.

If dysphagia prevents adequate caloric or fluid intake, pharyngoesophageal stenosis and stricture should be investigated as late complication of total laryngectomy. It is usually secondary to radiation fibrosis of the neck some months or years after completion of the treatment. A MBS can help diagnose which component or components of the swallowing tract or mechanism are faulty. Tumor recurrence should be suspected but once excluded by endoscopy and biopsy, repeated outpatient dilatation is usually effective. If dilatation is unsuccessful, flap augmentation, such as jejunal free flap may be necessary for successful rehabilitation. Neuromuscular paralysis or weakness may be either preexisting or iatrogenic from radiotherapy or from surgical damage to the hypoglossal nerve. In these patients, time and speech therapy exercises assist recovery. Occasionally, circumferential spasm of the pharyngoesophageal segment may be sufficient to cause dysphagia, in which case Botox A injections or myotomy may be helpful.

6. (Jeff) Discuss the treatment of cervical esophageal cancer and reconstructive options.

Partial Pharyngectomies: primary closure, local rotation flaps, tongue flap.
Larger defects: Pectoralis or radial forearm free flap.
For circumferential defects: radial forearm, jejunal interposition, gastric pull-up
7. (Caroline) What are the 4 types of esophageal diverticula? Discuss the location and anatomy of each.

<table>
<thead>
<tr>
<th>Diverticula</th>
<th>Location</th>
<th>Associated conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Zenker's diverticula</td>
<td>Proximal to the cricopharyngeus at the level of C5-C6, formed by the herniation of mucosa through an area of weakness in the posterior wall of the hypopharynx (the Killian triangle)</td>
<td>Reduced upper esophageal sphincter compliance</td>
</tr>
<tr>
<td>Midthoracic</td>
<td>Mid-esophagus, near tracheal bifurcation</td>
<td>Mediastinal inflammation: fungal infections, tuberculosis Motility disorders: achalasia, diffuse esophageal spasm</td>
</tr>
<tr>
<td>Epiphrenic</td>
<td>Distal 6-10 cm of the esophagus</td>
<td>Motility disorders: achalasia, diffuse esophageal spasm Esophageal obstruction</td>
</tr>
<tr>
<td>Diffuse intramural diverticulosis</td>
<td>Segmental or diffuse involvement of the body of esophagus</td>
<td>Chronic inflammation, stasis, or distal obstruction</td>
</tr>
</tbody>
</table>

8. (Tali) What are the treatment options for a Zenker’s diverticulum?

The etiology of Zenker’s is incompletely understood. It is thought that patients have improperly timed relaxation of the cricopharyngeus muscle during swallowing. Over time, the increased pressure causes herniation of the esophageal mucosa posteriorly (the weakest area), between the inferior pharyngeal constrictor and the cricopharyngeus muscle. Zenker is a false diverticulum. Often food particles retained in the pouch leading to halitosis, regurgitation, aspiration, and dysphagia.

Surgical approaches include (1) stapled or hand-sewn diverticulectomy with cricopharyngeal myotomy, (2) stapled or hand-sewn diverticulopexy with cricopharyngeal myotomy, and (3) endoscopic division of the diverticular wall with an endoscopic stapler. Historically, myotomy alone was performed, with a lower rate of relief of symptoms and more frequent complications. Myotomy alone is associated with persistent symptoms in up to 30% of patients. Recurrence requiring repeat surgery is necessary more frequently than with other procedures.

1)Diverticulectomy with cricopharyngeal myotomy: With a stapled or hand-sewn diverticulectomy and cricopharyngeal myotomy, the pouch neck is either oversewn or stapled, and the pouch is excised. The cricopharyngeus muscle is divided longitudinally no less than 5 cm. This is typically performed through a left neck incision and is primarily closed with a closed suction drain in place.

Complications:
- Mortality (0-9.5%) Morbidity (4-47%)
- RLN paralysis Esophageal stenosis
- Mediastinitis Pharyngocutaneous fistula
- Hematoma Esophageal perforation

Advantages
- Removes the diverticulum Provides tissue for pathologic review
- Highly effective

Disadvantages
- Longer surgical intervention Longer hospitalization
- Delayed alimentation Immediate symptom relief (90-100%)

Long-term symptom recurrence (2-33%). Gutschow compared the success rates with "open" versus endoscopic techniques. He found that in sacs <3cm the open technique achieved 98% long-term success while the endoscopic approach achieved only a 57% rate. With >3cm sacs the success rate was 97% and 88%, respectively.

2)Diverticulopexy with cricopharyngeal myotomy: In the diverticulopexy with cricopharyngeal myotomy, the diverticulum is inverted and sutured to the prevertebral fascia, and the cricopharyngeus muscle is divided as above. The difference in this procedure is that the pouch is not excised. This procedure is more commonly advocated in the severely debilitated patient because there is no division of the esophagus, pharynx, or diverticulum, and there is no suture line.

3)Endoscopic myotomy/diverticulectomy: In the endoscopic myotomy, a double-bladed rigid endoscope is placed into the pharynx with one blade positioned in the esophagus and the other in the diverticulum. A reticulating endoscopic linear stapler is introduced into the pharynx with one jaw of the stapler in the pouch and one jaw in the esophagus. The stapler is locked across the common septum of the two and is fired. If necessary, this is repeated until the bottom of the pouch is reached. This results in an opening of the pouch and a division of the cricopharyngeus muscle. The pouch wall becomes incorporated as a wall of the esophagus.

Complications:
Dec 20: Esophageal Disorders/Dysphasia (updated 08/06)

- Mortality (0-1%)
- Morbidity (10-31%)
- RLN paresis/paralysis
- Bleeding/Cervical emphysema
- Mediastinitis
- Dental injury
- Esophageal perforation
- Diverticulum perforation

**Advantages:**
- Short surgical procedure
- Easily repeated
- Quick return to oral alimentation
- Short hospitalization
- Less tissue trauma
- Highly effective

**Disadvantages:**
- Unable to perform in some patients
- Does not remove the diverticulum
- No tissue for path
- Immediate symptom relief (94-100%)
- Long-term symptom recurrence with stapling technique (0-47%): According to Gutschow, the recurrence rate with any endoscopic approach is greater if the sac is smaller. Recurrences may be successfully managed with a repeat endoscopic or an "open" approach; when performing the open approach, the surgeon needs to recognize the altered anatomy present as a result of the prior endoscopic procedure.

**Cautery or laser technique (1-79%; varies widely by reporting site)**

9. (Scott) Discuss the phases of swallowing; what diseases can cause problems at each phase?

Swallowing has four phases: oral preparatory, oral propulsive, pharyngeal and esophageal. The oral preparatory phase processes the bolus and the oral propulsive phase propels food from the oral cavity into the oropharynx. These are voluntary actions.

The pharyngeal phase involves a rapid sequence of overlapping events. 1) The soft palate elevates; 2) The hyoid bone and larynx move upward and forward; 3) The vocal folds move to the midline, and the epiglottis folds backward to protect the airway; 4) The tongue pushes backward and downward into the pharynx to propel the bolus down. It is assisted by the pharyngeal walls, which move inward with a progressive wave of contraction from top to bottom; 5) The upper esophageal sphincter relaxes during the pharyngeal phase of swallowing and is pulled open by the forward movement of the hyoid bone and larynx. This sphincter closes after passage of the food, and the pharyngeal structures then return to reference position.

In the esophageal phase, the bolus is moved downward by a peristaltic wave. The lower esophageal sphincter relaxes and allows propulsion of the bolus into the stomach. Unlike the upper esophageal sphincter, the lower sphincter is not pulled open by extrinsic musculature. Rather, it closes after the bolus enters the stomach, thereby preventing gastroesophageal reflux. The swallow reflex is a complex neurologic event involving participation of high cortical centers, brain stem centers such as the tract of the nucleus solitarius and nucleus ambiguous, and cranial nerves V, VII, IX, X, and XII. Neurologic deficits in any of these areas can result in dysphagia.

**Disorders affecting swallowing:**

**Local disorders:**
- Foreign Bodies: foreign body sensation, +/- fever, dysphagia, and odynophagia, excessive salivation, vomiting, chest pain, +/- wheezing
- Structural disorders: Cricopharyngeal Hypertonicity, Zenker's Diverticulum, Lateral Pharyngeal Pouches and Diverticula, Esophageal Webs, Esophageal Ring
  - Achalasia: Dysphagia for solids greater than liquids. Radiology reveals a bird's beak deformity.
  - Cervical Spine Disease
- Diffuse Esophageal Spasm, Gastro-esophageal Reflux Disease (GERD)
- Tracheostomy: Prevents proper laryngeal elevation during the pharyngeal phase of swallowing. Increases extrinsic esophageal pressure and leads to regurgitation and even aspiration.
- Cancer of the Larynx or Pharynx: Odynophagia
- Cancer of the Esophagus: Dysphagia to solids more than liquids
- Surgical resection
- Radiation fibrosis

**Systemic Disorders:**
- Stroke: Swallowing difficulty after stroke in 47% of cases. Most patients recover within one week. Causes: delayed triggering of the swallow reflex, cricopharyngeal dysfunction, reduced lingual and pharyngeal control, and weak cough. With brain stem strokes, direct cranial nerve deficits can lead to further impairment of glottis closure and coordination of phases of swallowing.
- Neurological disorders: Amyotrophic Lateral Sclerosis Parkinson's Disease, Multiple Sclerosis, Muscular Dystrophy
Dec 20: Esophageal Disorders/Dysphasia (updated 08/06)

- Myasthenia Gravis: Disorder of acetylcholine receptors. Dysphagia and fatigue are typically worse later in the day.
- Laryngeal Nerve Injury: Decreased glottic closure pressures and neurogenic dysfunction of the inferior constrictor and cricopharyngeus muscles.
- Autoimmune Disorders: Systemic sclerosis, system lupus erythematosis, dermatomyositis, mixed connective tissue disease, mucosal pemphigoid, epidermolysis bullosa, Sjogren's syndrome (xerostomia) and Rheumatoid arthritis (cricoarytenoid joint fixation).
- Other causes: Psychiatric disorder, Psychogenic dysphagia, Iatrogenic causes, Medications

Elderly: Oral preparatory phase problems result from poor dentition, and oral phase problems result from loss of tongue connective tissue. Pharyngeal phase changes include increased pharyngeal transit time and prolonged upper esophageal sphincter relaxation time.

Children: nasal obstruction (nasal masses, choanal atresia, and choanal stenosis) may present with feeding difficulties. Cleft lip or palate, mucoceles, ranulas, and Warthin's duct stenosis may cause dysphagia. Laryngomalacia, vocal cord paralysis, laryngeal clefts, tracheo-esophageal fistula, foregut malformations, or vascular rings of the aorta or pulmonary arteries that compress the esophagus or trachea may all contribute to feeding problems and dysphagia. Tumors of the aerodigestive tract which can certainly cause dysphagia include hemangiomas, lymphangiomas, papillomas, leiomyomas, and neurofibromas.