Congenital Malformations of the Upper and Lower Airways

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Disclosure

- I have no actual/potential conflict of interests or relevant financial relationships to disclose.

Objectives

- Understand the anatomical separation of the upper and lower airways.
- Review the different types of malformations that affect the airways.
- Discuss the presentation of such malformations and how they cause a disorder is ventilation/oxygenation.

Case Presentation

- 12 yo male with the previous diagnosis of asthma presents to OSH with wheezing and inc wob.
- He is given steroids, albuterol, epi, mag, and placed on HFNC.
- He is intubated for worsening failure and transported to Covenant.
- Ventilator was weaned with reassuring graphics and he was extubated the following morning.

Case Presentation

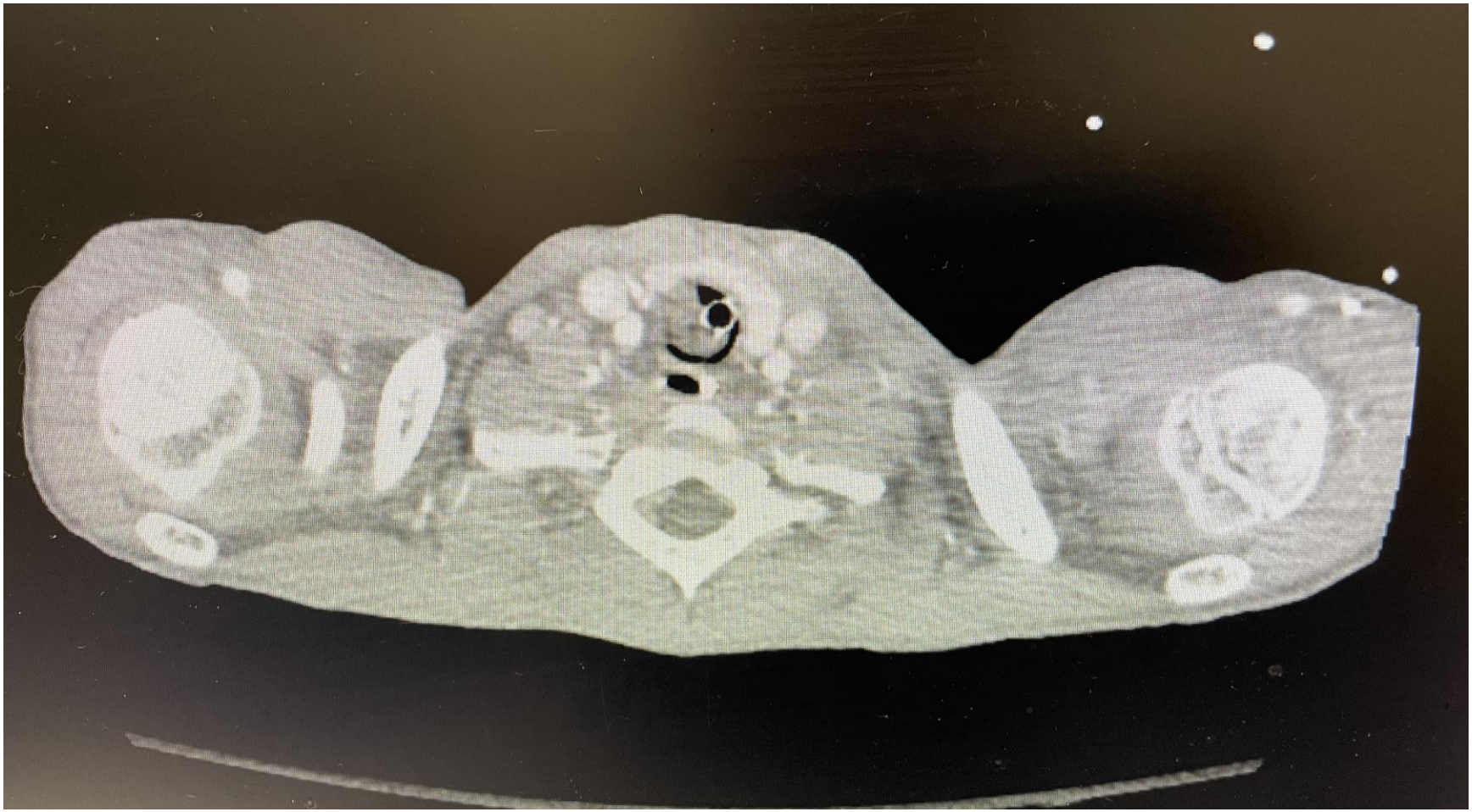
- Shortly after extubation, he again developed severe respiratory failure with retractions and poor air exchange.
- Terbutaline gtt, BiPap, continuous albuterol, mag x 2 were given without improvement.
- He was re-intubated the following day without difficulty. Following intubation, airway obstruction resolved and once again vent was weaned to low settings

At what level is the obstruction occurring???

Case Presentation

- ECHO attempted, but could not rule out a vascular ring. CTA showed a 2 cm subglottic filling defect (mass) in the subglottic trachea above the thoracic inlet adjacent to the ET tube.

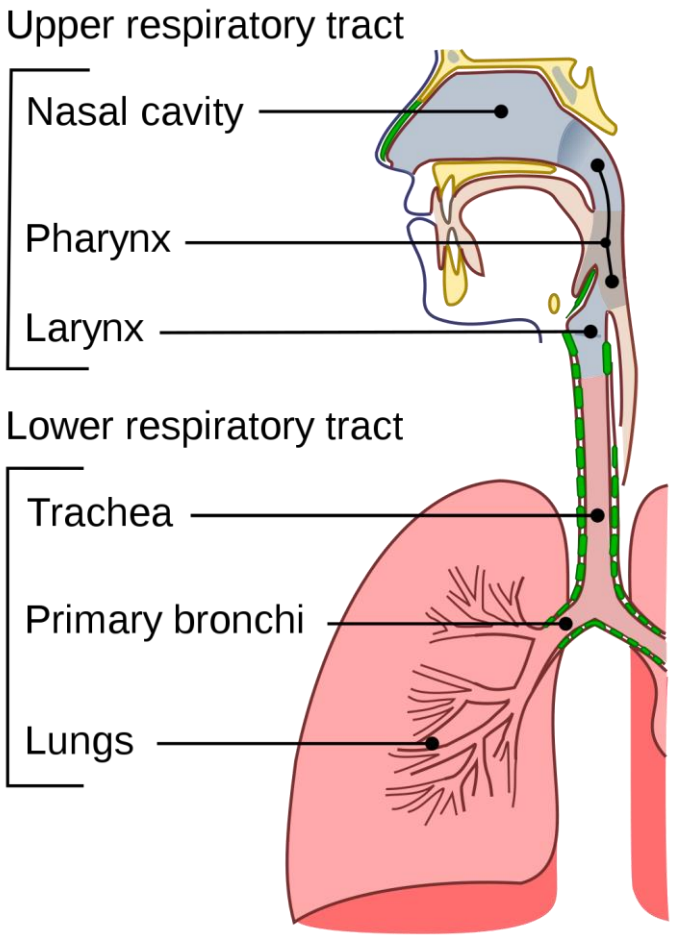
CTA



Case Presentation

- It was decided to transfer the patient to U of M for subspecialty care.
- He underwent placement of tracheostomy prior to the resection of the mass.
- Diagnosis- tracheal chondroid hamartoma
- He then underwent a tracheal reconstruction without issue and extubated

Division of the Respiratory System



Anomalies of the Larynx

- Laryngomalacia= collapse of the supraglottic structures during inspiration
- Most common congenital anomaly
- Inspiratory stridor
- Increase risk of GERD
- Loudest at 4-8 months (resolves by 12-18 months)
- Diagnosed usually on history alone

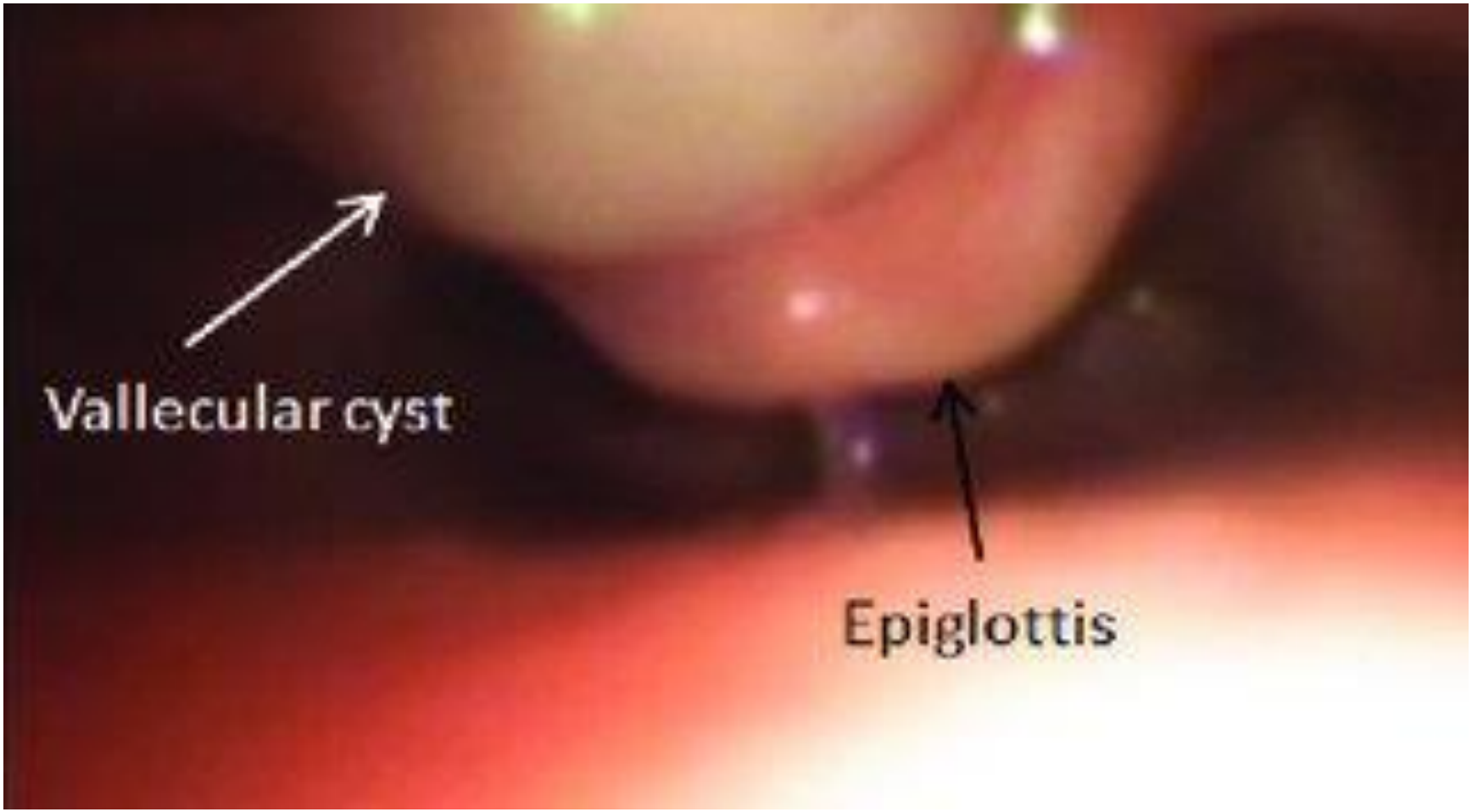
Laryngomalacia cont'd

- Management:
  - Mild, insure weight gain is adequate
  - Moderate, medical management with anti-reflux medications
  - Severe, surgical correction with supragloyyoplasty

Anomalies of the Larynx

- Vallecular cysts= contain respiratory epithelium and mucous glands
- Uncommon cause of respiratory distress in a newborn. Leads to a hoarse cry
- Marsupialization is the treatment

Vallecular Cyst



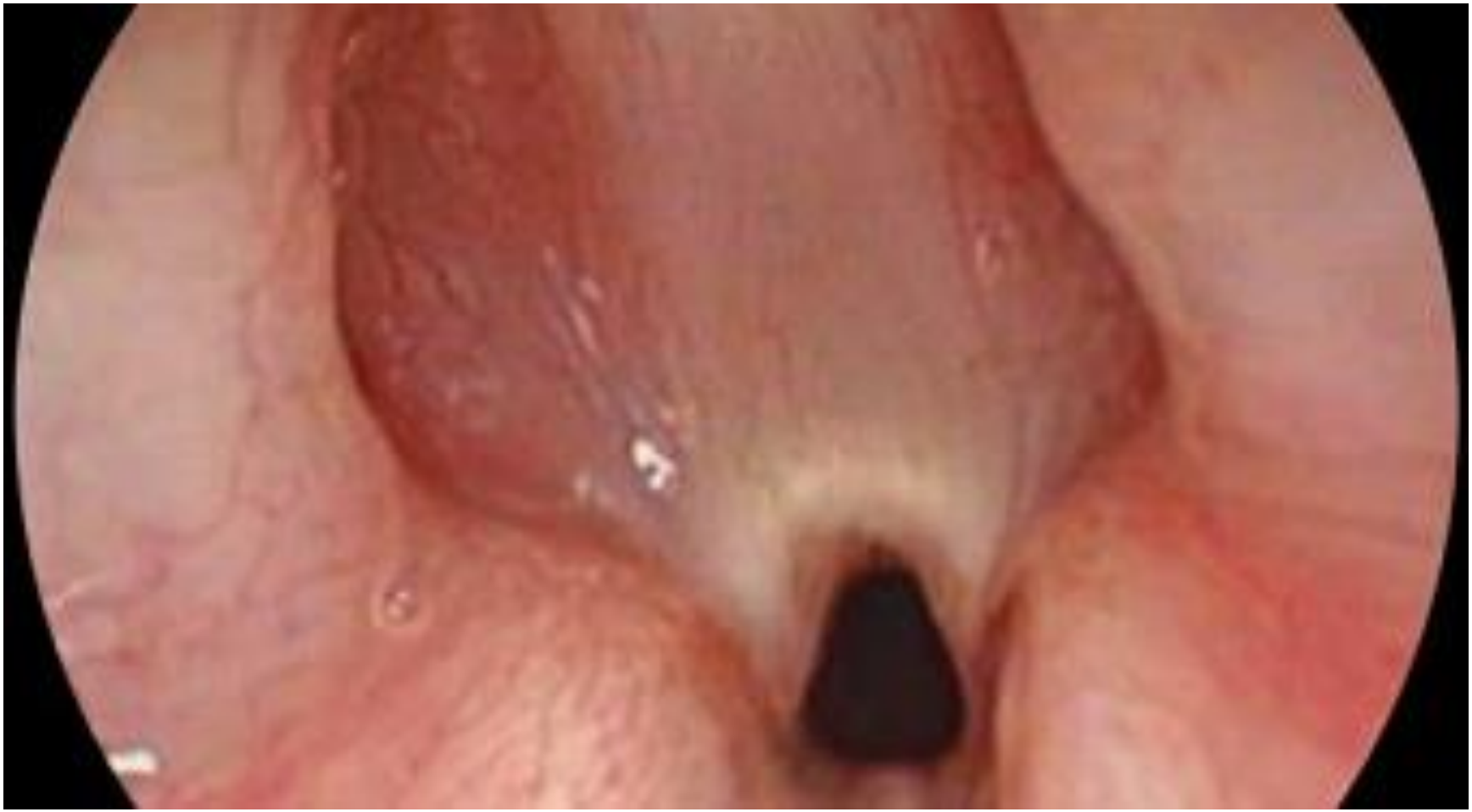
Anomalies of the Larynx

- Laryngeal atresia= failure of epithelial growth and recanalization of the subglottic region
- Asphyxia at birth
- Requires emergent tracheostomy
- Most of the time identified on antenatal ultrasound

Anomalies of the Larynx

- Laryngeal webs= failure of reabsorption of the epithelial layer and leads to incomplete separation of the vocal cords
- Usually presents in infancy with respiratory distress and high pitch cry
- Treatment is with a CO2 laser
- May require reconstruction and stenting

Laryngeal Web



Anomalies of the Larynx

- Congenital Subglottic Stenosis= narrowing of the lumen in the cricoid region
- Most commonly presents as recurrent croup
- Sometimes biphasic stridor in a newborn
- Typically improves as the child gets older, less than 30% require a tracheostomy

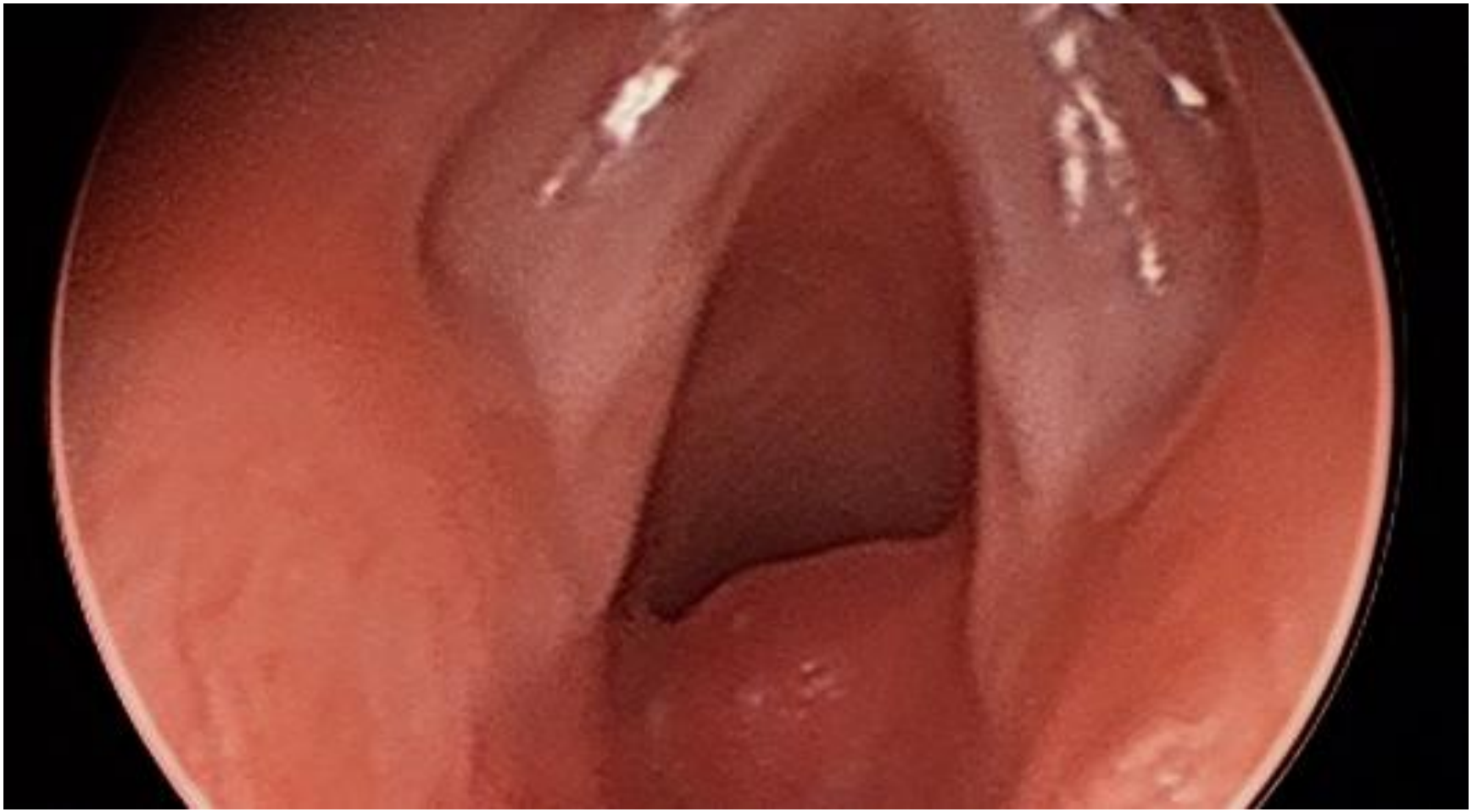
Anomalies of the Larynx

- Laryngeal Cleft= failed fusion of the lateral growth plates of the posterior cartilage (6-7 weeks in the embryonic period)
- More common in boys
- 1/10,000-20,000 live births
- Presents as increased secretions, FTT, wheezing, stridor, aspirations, recurrent infections

Anomalies of the Larynx

- Subglottic hemangiomas= vascular lesions which grow rapidly in the first weeks-months and then stabilizes
- Complete resolution occurs in 50% of patients by age 5
- Propranolol is the first-line therapy. Sometimes require locally injected steroids or surgical intervention

Subglottic Hemangioma



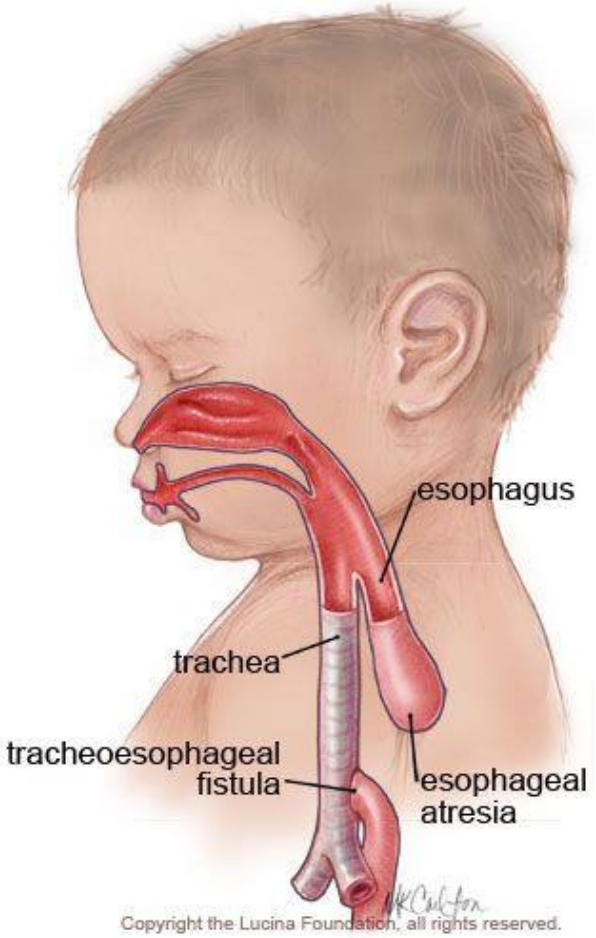
Anomalies of the Trachea

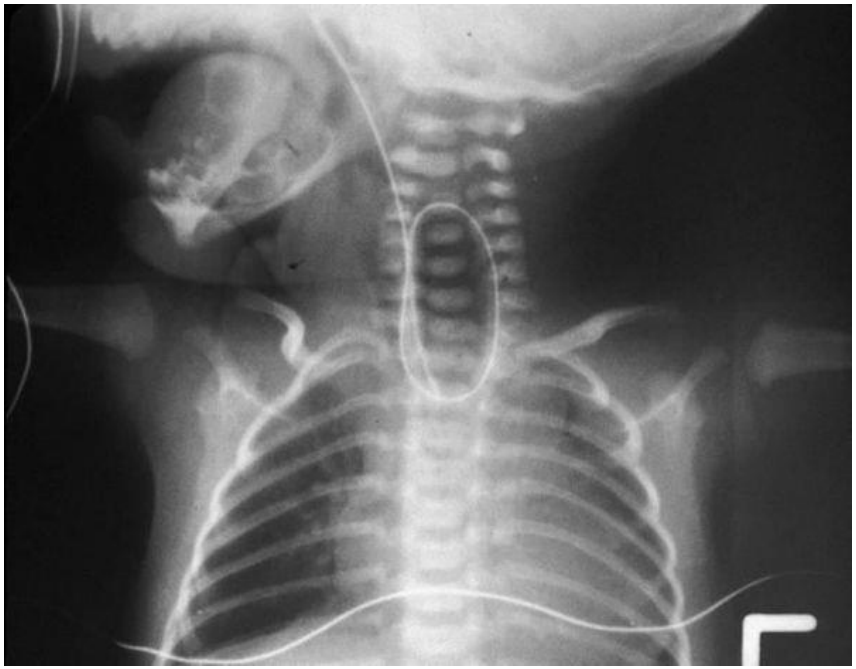
- Tracheoesophageal fistula (TEF)= more common anomaly with ~ 1/3500 births
- Type C accounts for 84% of TEFs
- May have associated syndromes (CHARGE and VACTERL)
- Polyhydramnios occurs in 2/3 of pregnancies
- Symptomatic immediately after birth with excessive secretions and choking

TEF

- Diagnosis by placing an catheter to the stomach. If it is unable to pass 10-15 cm and confirmed coiling in the upper pouch, TEF is confirmed
- Surgical correction is performed as soon as possible
- Prior to surgery, continuous suction to pouch to avoid aspiration

TEF





Anomalies of the Trachea

- Tracheal Atresia= agenesis or aplasia of the trachea which leads to the partial or complete absence of trachea below the larynx
- Severe cyanosis and no audible cry at birth
- Usually lethal and associated with other abnormalities

Anomalies of the Trachea

- Tracheal stenosis= narrowing from complete or near complete cartilage rings
- May be complete stenosis or funnel shaped
- Diagnosed with bronchoscopy
- Treated with balloon dilation, stent placement, laser treatment, or tracheoplasty

Anomalies of the Trachea

- Tracheomalacia= common anomaly characterized by dynamic collapse of the airway during breathing
- Primary or secondary
- Signs and symptoms depend on location and severity. Usually recurrent harsh cough or inspiratory stridor
- Diagnosed with dynamic endoscopy
- Most improve spontaneously by 12 mo

Anomalies of the Bronchus

- Similarly to the trachea the following conditions may occur:
  - Bronchial atresia
  - Bronchial stenosis
  - Bronchomalacia
- Bronchial cysts= anomalous budding of the foregut, may occur anywhere in the tracheobronchial tree
- Usually present in second decade of life with recurrent cough and wheezing

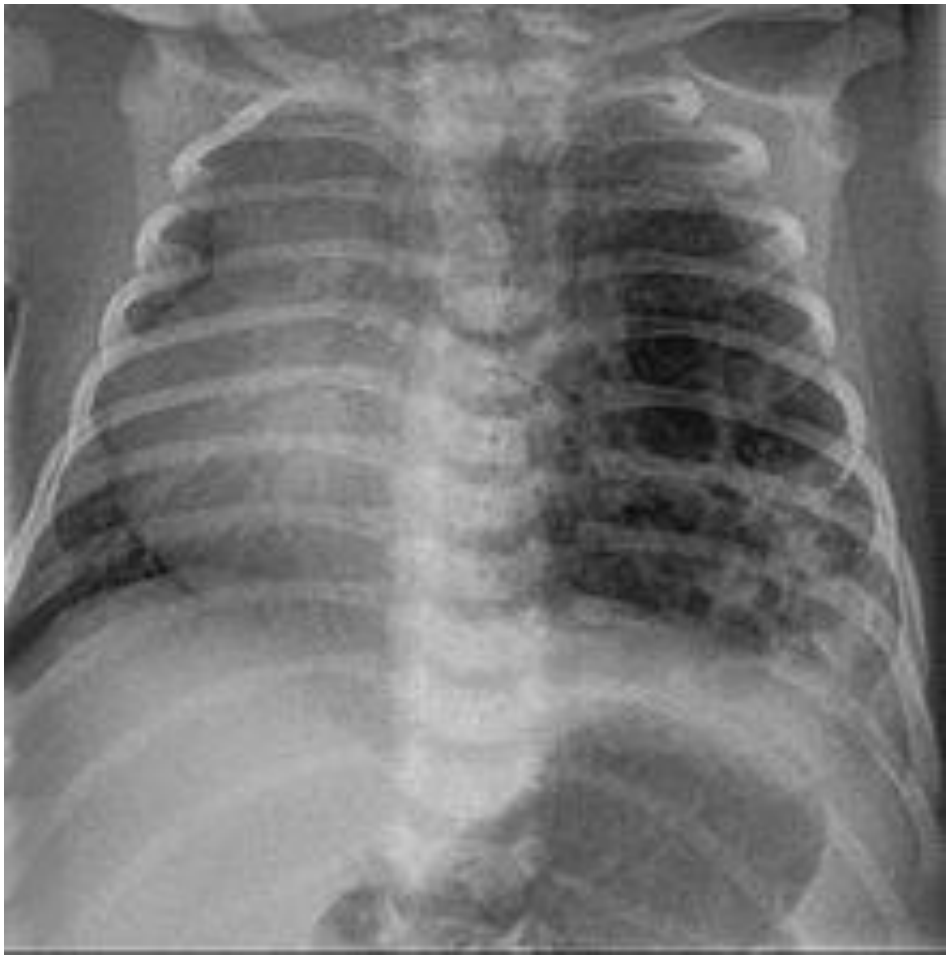
Congenital Pulmonary Airway Malformation (CPAM)

- Mass of abnormal lung tissue, classification is based on the number of cysts present
- Most common CPAM (60-70% of cases) is type 1 and consists of a single, dominant cyst but may include a few cysts 2-10 cm in diameter
- Usually located in one lobe
- Large cysts may cause mediastinal shift and diaphragm compromise

CPAM

- Often diagnosed prenatally on ultrasound
- Chest CT or MRI maybe warranted
- Small CPAMs may be delayed for resection and re-evaluated
- Large CPAMs may be candidates for in utero resection
- May lead to maternal mirror syndrome
- Possible malignancy later in life if not resected

CPAM



Questions?