Masqueraders of Wheezing: Asthma or a Wheeze in Sheep's Clothing

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KEY OBJECTIVES

- Know the differential diagnosis of recurrent wheeze in young children
- Understand the importance of radiographic and potentially bronchoscopic evaluation in children with recurrent wheeze and poor response to inhaled therapies
- Recognize the clinical and spirometry pattern for vocal cord dysfunction in school age children with difficult to control asthma

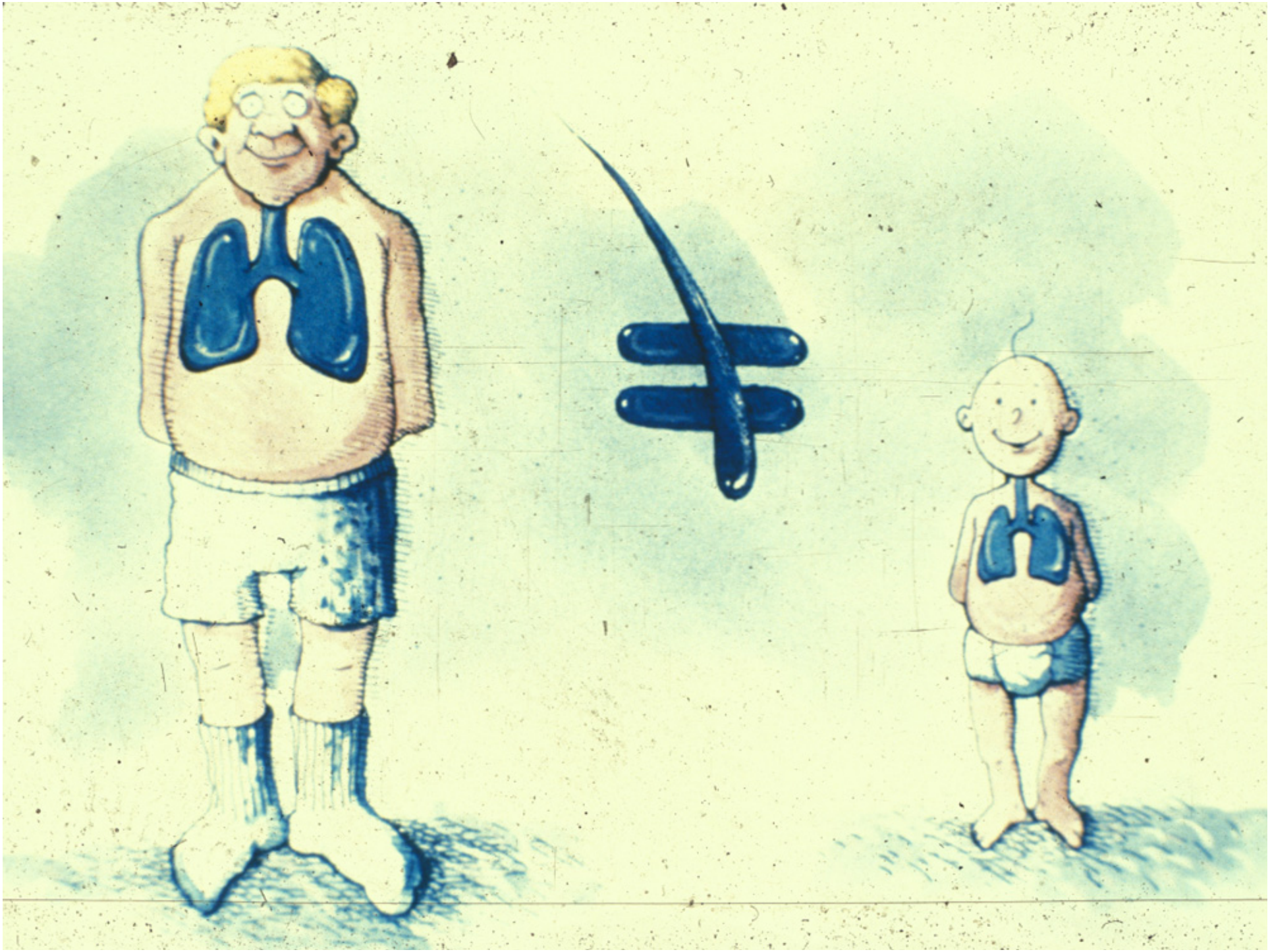
Differential Diagnoses:
Remember all that wheezes is not ASTHMA

PEDIATRICS

- Infection - VIRAL (RSV)
- Asthma
- Anatomic Abnormalities
  - Malacia
  - TEF
  - Vascular ring/sling
  - Mediastinal mass/tumor
  - CAMs, cysts, CLE, sequestration
  - Congenital Heart Disease (L→R shunt)
- Inherited - CF and Immunodeficiency
- BPD
- Aspiration - GERD, FB
- ILD including BO
- VCD

ADULTS

- VCD
- Asthma
- COPD
- Congestive Heart Failure
- Anatomic - Airway tumor, LAD
- Bronchiectasis
- ILD including BO

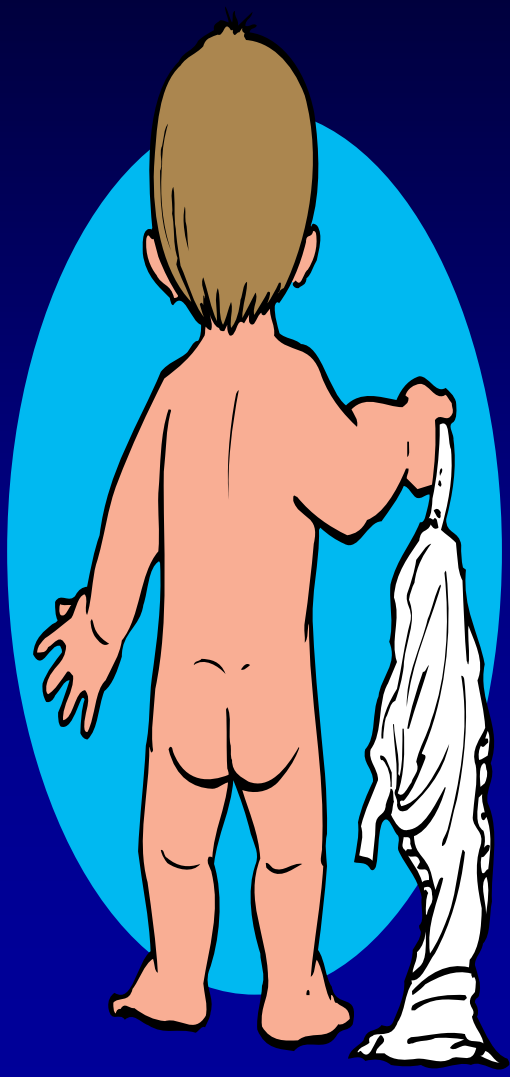


Pathophysiologic Properties Predisposing Infants and Young Children to Wheeze

1. ⇓ Bronchial smooth muscle content
2. Hyperplasia of bronchial mucous glands
3. ⇓ radius of conducting airways
4. ⇑ peripheral airway resistance due to ⇓ size
5. ⇑ Chest wall compliance
6. Diaphragm
   - Horizontal insertion of the diaphragm to the rib cage
   - ⇓ number of fatigue-resistant skeletal muscle fibers
7. Deficient collateral ventilation
8. Females - ⇓ lung size
9. Males - ⇓ Vmax FRC

Asthma in Infants

- Very difficult to diagnose
- Features of disease are less clear
- Clinical diagnosis relies on a history of:
  - **episodes of wheezing**
    - 20% of all children will have wheezed by 1 year, 33% by 3 years, and 50% by 6 years
    - Majority do not progress to asthma
      - 85% are transient wheezers
      - 15% develop persistent wheezing = asthma
  - **Coexistence of atopy or specific trigger**



RS - 8mo male presents with recurrent wheezing and severe cough since birth

- Persistent dry barky cough and wheezing
- Two episodes of respiratory distress requiring hospitalization
- Cough most prominent upon awakening and intermittent throughout the day/night
- Mild to moderate snoring and restless sleep
- “Mouth-breather”
- Recurrent ER and PCP visits due to respiratory symptoms - frequent courses of antibiotics and prednisone

Term infant, Apgars $8^{1}9^{5}$ and Bwt 5#6oz

- Hospitalized 4/20 - 4/25 with +RSV & 10/14 - 10/19 with RSV-
- GERD - based on UGI
- Reactive airways disease

NKDA IZ: UTD

Meds: Ranitidine 2X/day

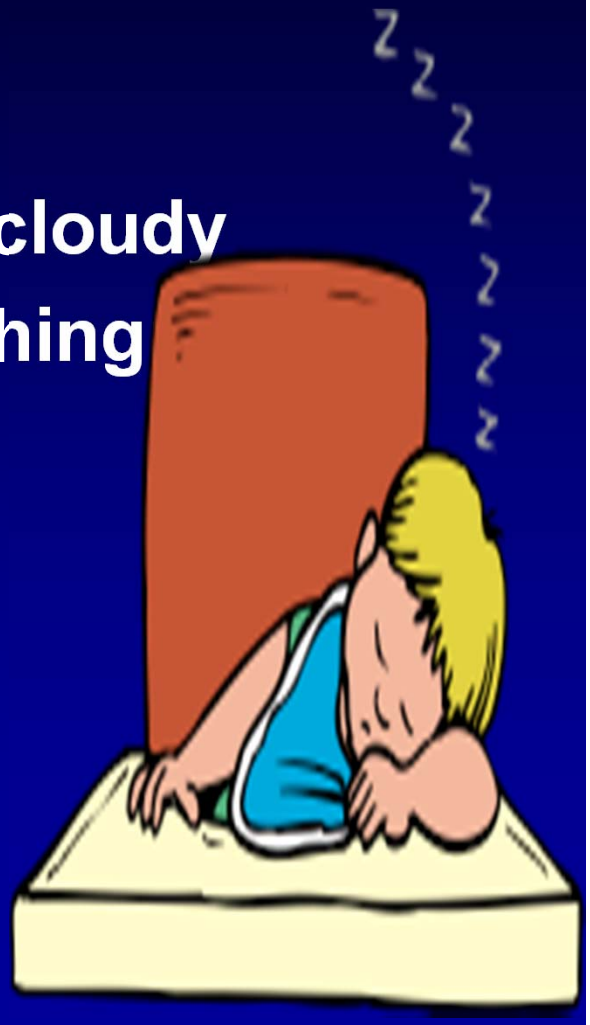
Albuterol nebs q4-6^{o}

Budesonide nebs 2X/day (1 month)

FHx: MOC with asthma and hayfever

SHx: MOC and GPOC smoke outside;
No pets

ROS: Positive for chronic clear → cloudy
nasal d/c; SOB with severe coughing
moderate eczema;
chronic loose stools on Abx



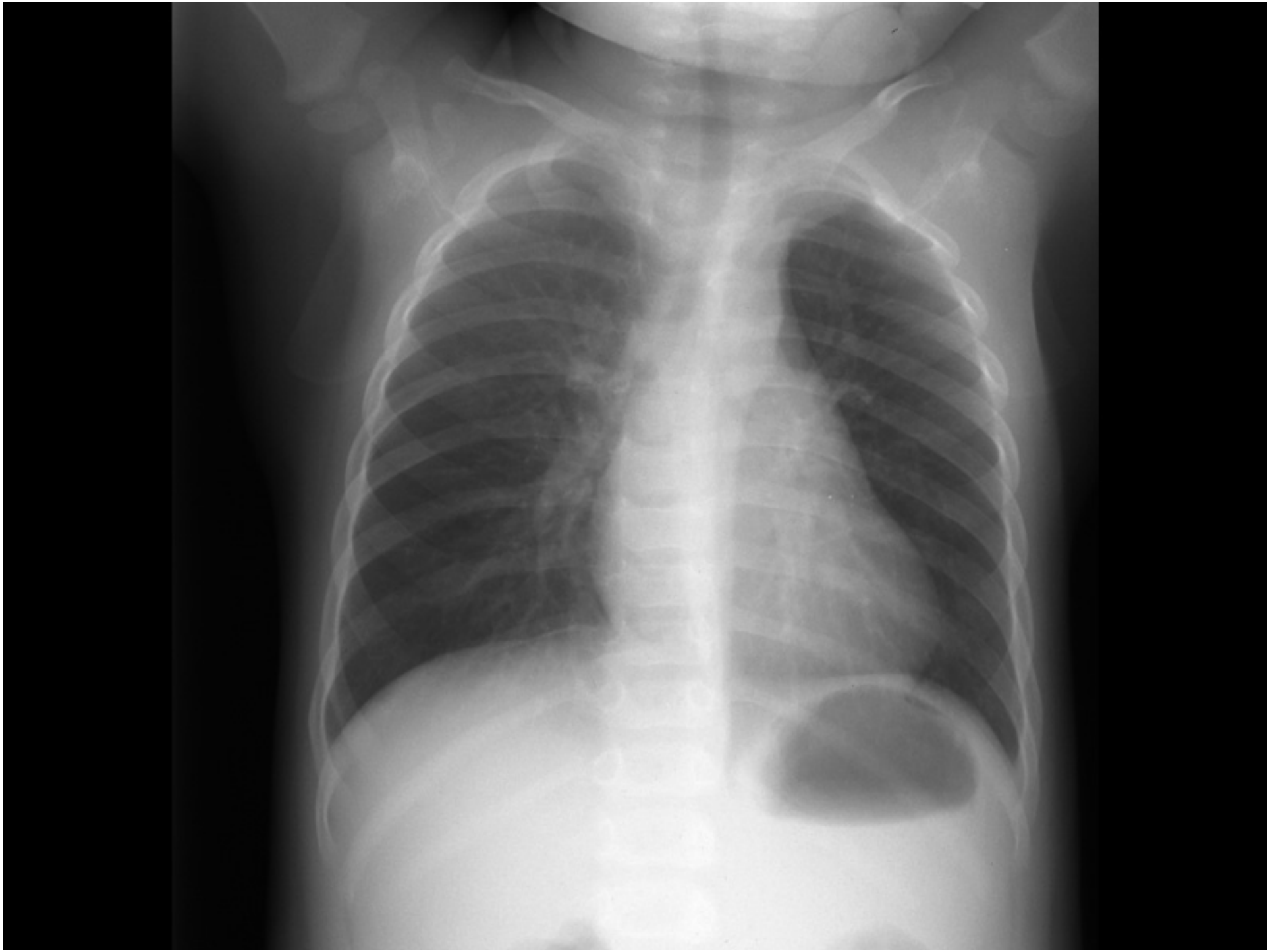
PE: Afebrile HR 156 RR 52 BP 92/60 91% RA
Wt. 6.2 kg (<5%)
Ht. 64 cm (10%)

Gen: alert, small male; severe barky, cough episodes; baseline mild respiratory distress

HEENT: clear, non-injected conjunctiva; boggy, erythematous nasal mucosa with cloudy white drainage R>L; Moderate cobblestoning of the posterior pharynx with mucous noted, tonsils 2+

Lungs: Moderate transmitted UA coarseness. Inspiratory and expiratory stridor, Diffuse end-expiratory wheezes bibasilar

Skin: LE>UE nonexcoriated eczema



Asthma Predictive Index

– H/o ≥ 4 wheezing episodes in the past year (at least one must be MD diagnosed)

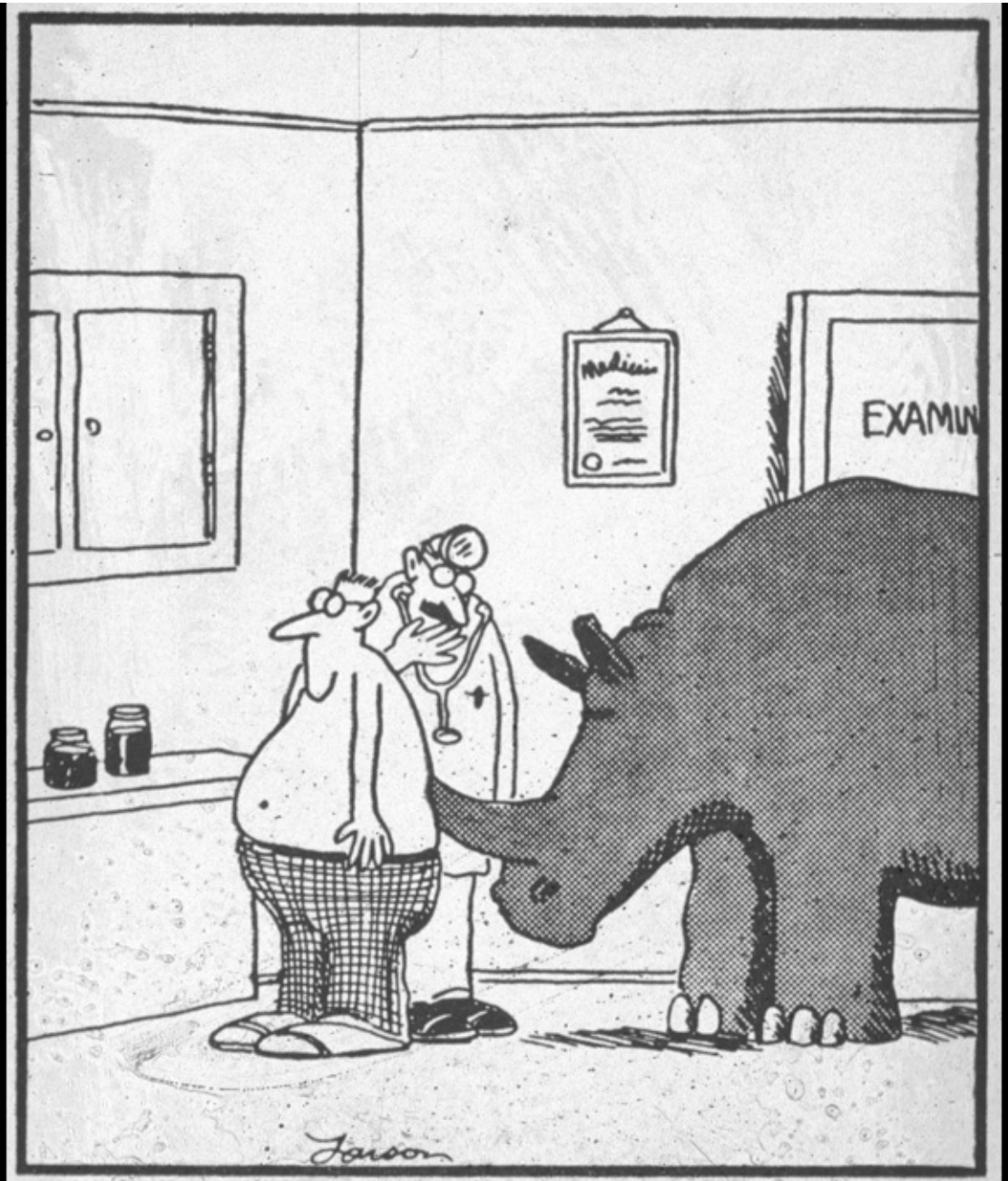
PLUS

<u>One major criteria</u> or <u>Two minor criteria</u>

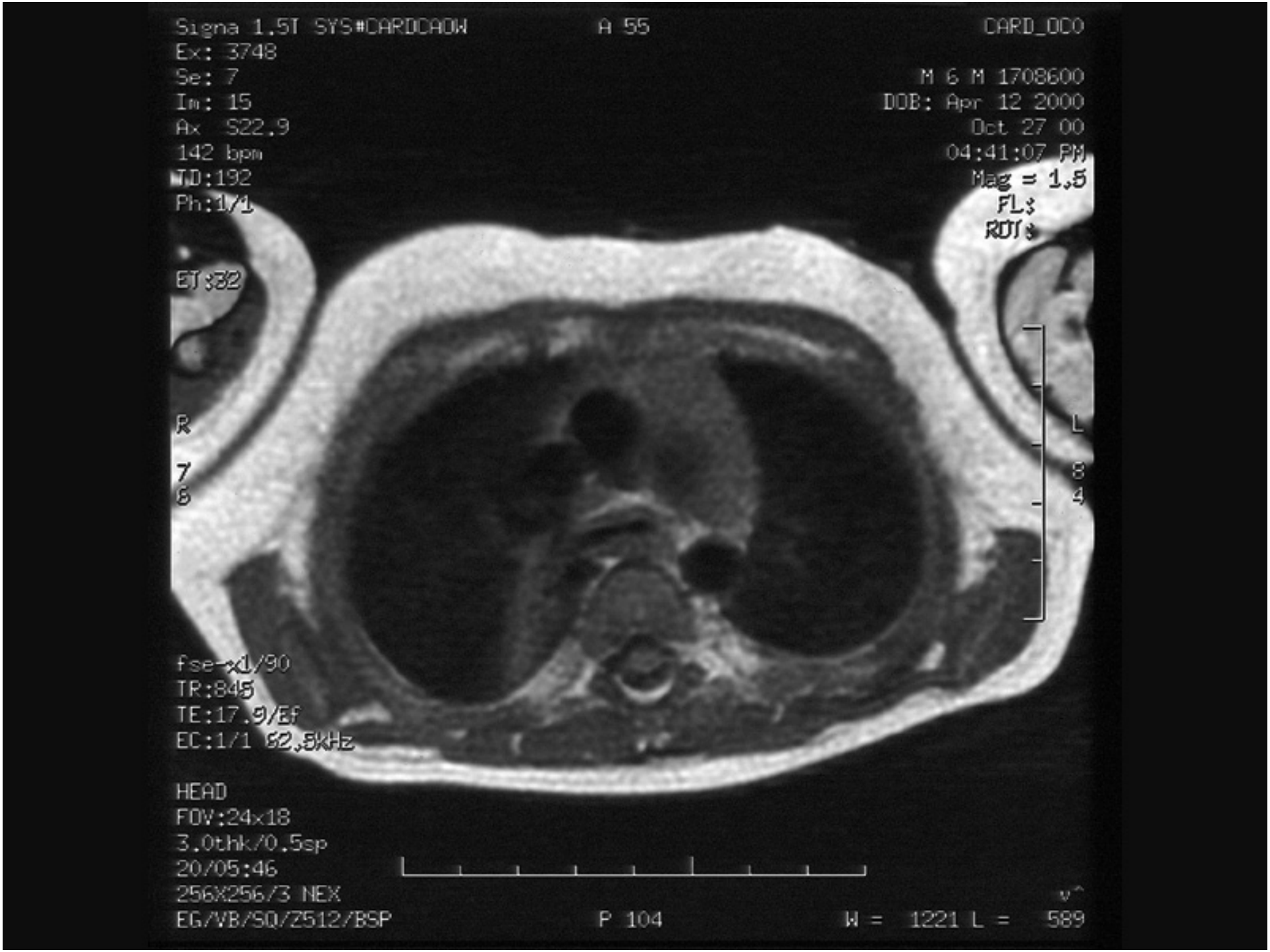
- Parent with asthma
- Atopic dermatitis
- Aeroallergen sensitivity

- Food sensitivity
- Peripheral eosinophilia (≥ 4%)
- Wheezing not related to infection

If +, then 65% likelihood of developing clinical asthma
If -, then 95% likelihood of not developing clinical asthma



"Wait a minute here, Mr. Crumbley. . . . Maybe it isn't kidney stones after all."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Diagnosis Please

- Innominate artery compression
- Moderate tracheomalacia
- Mild to moderate persistent asthma
- Nasal rhinitis and postnasal drip

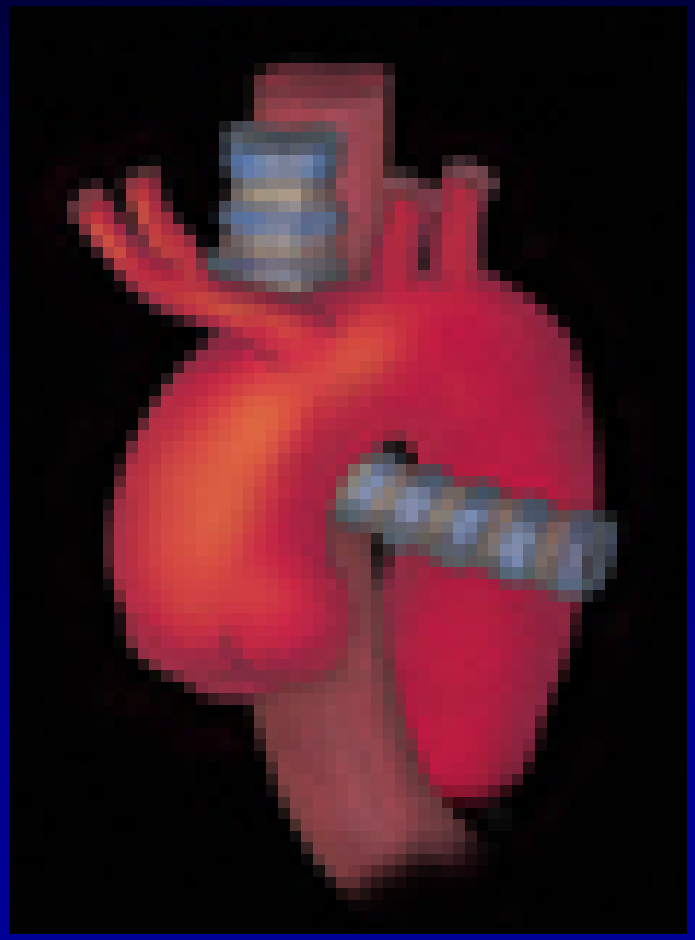
Vascular Anomalies

- First identified in 1750, surgical management was not inaugurated until 1946 by Gross and Ware
- Four basic types resulting in extrinsic tracheal obstruction and often esophageal compression
  - Aberrant right subclavian artery*
  - Anomalous innominate artery* (an incomplete ring)
  - Right aortic arch with left ligamentum arteriosum or PDA*
  - Double aortic arch
  - Pulmonary aa. sling

*Account for the most common vascular anomalies

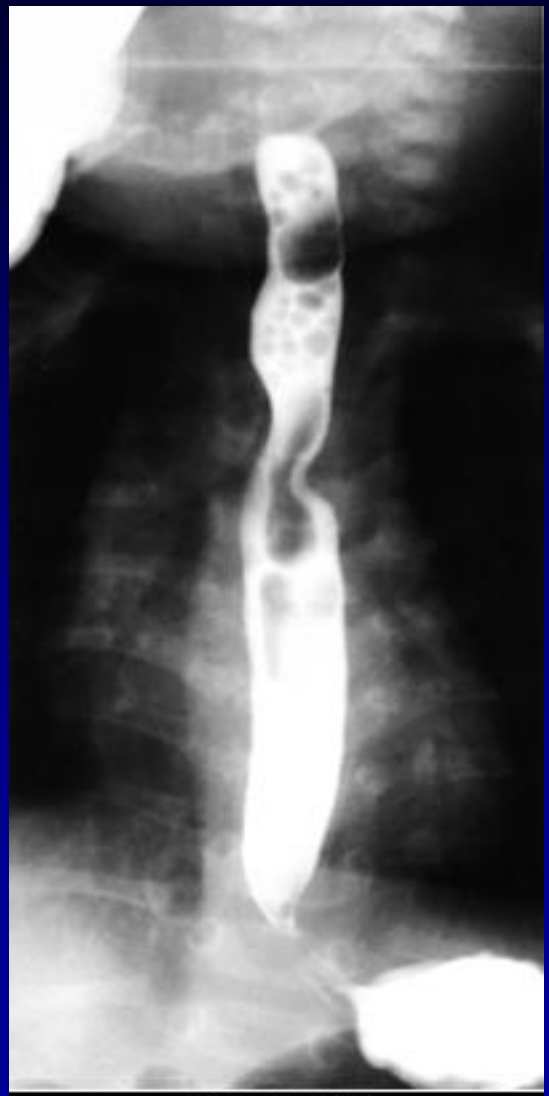
Innominate AA compression

- Common variant of normal
- AKA anomalous innominate or left carotid artery
- An incomplete ring- delayed or premature origin from the arch
- Degree of compression = symptoms
- Symptoms → stridor, brassy cough
- Dysphagia and emesis less likely
- UGI may show anterior compression
- 50% present with apneic spells
- <u>Treatment</u>
  - Reimplant innominate to R and anterior arch
  - Aortopexy (suspend arch to post. Sternum)

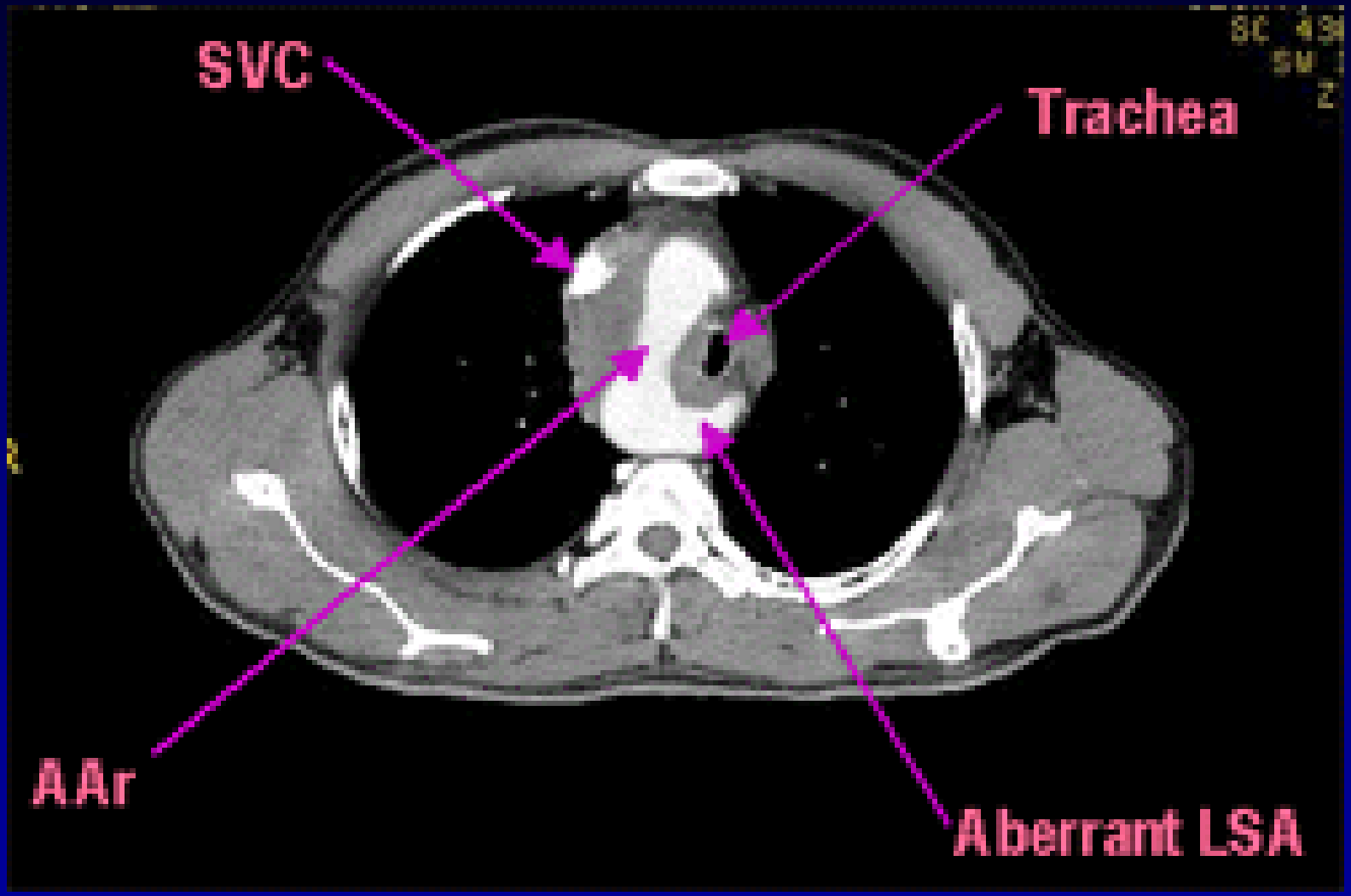


Aberrant Right Subclavian

- Most commonly asx; 1/200
- May constrict the posterior esophageal wall producing dysphagia
  - Courses behind the descending aorta toward the right and behind the esophagus
- The normal aorta produces a lateral indentation on the esophagus and slightly displaces the trachea to the right.

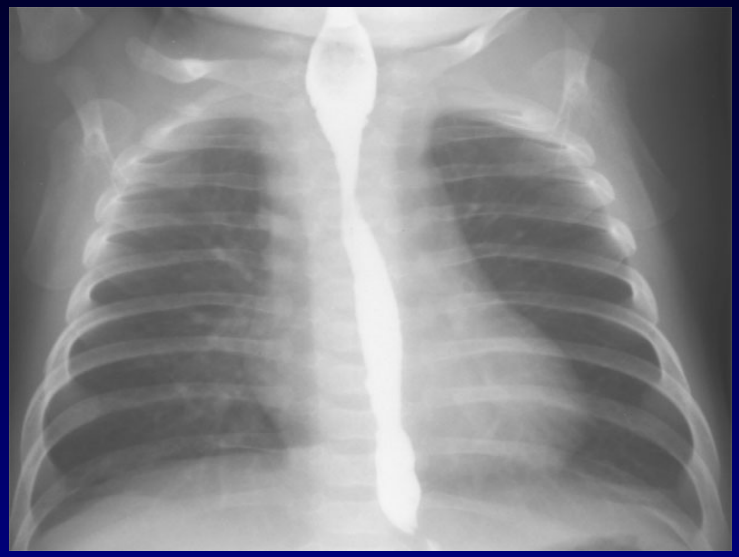


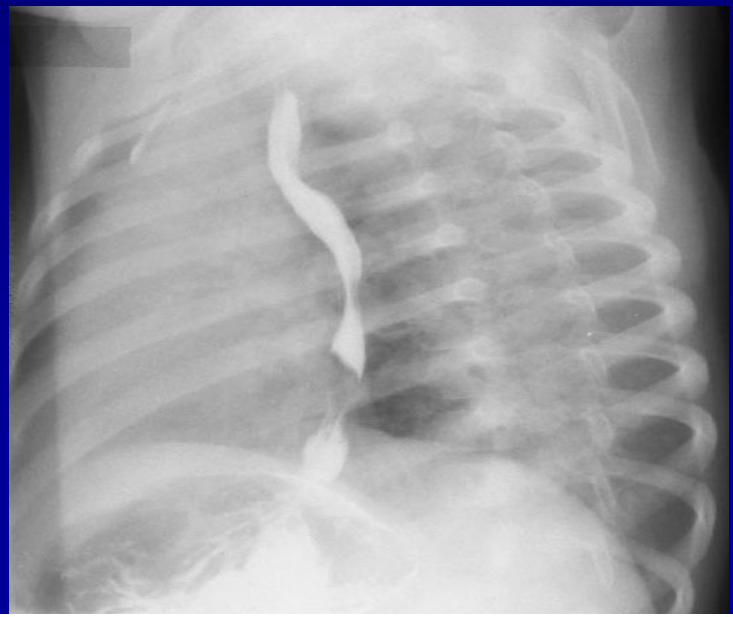
Right Aberrant Subclavian



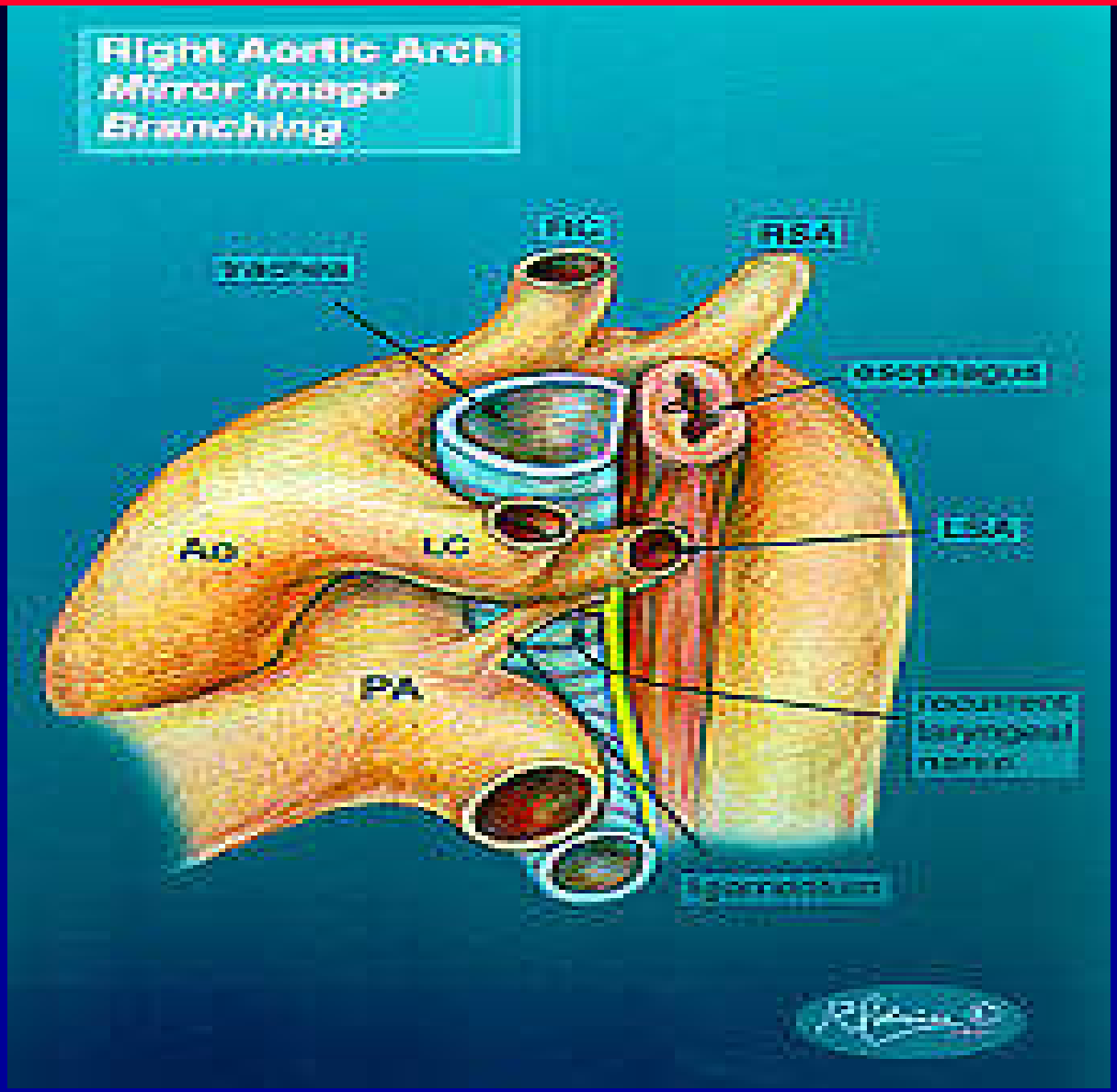
Right Aortic Arch

- Persistent right 4th brachial vessel which typically disappears
- 2nd most common type of ring
  - Most common is associated with an aberrant L. subclavian aa.
  - May or may not be associated with a ductus or ligamentum arteriosum
    - CXR and UGI are very helpful for diagnosis
  - More common in males
- Can be associated with Tetralogy/truncus/transposition





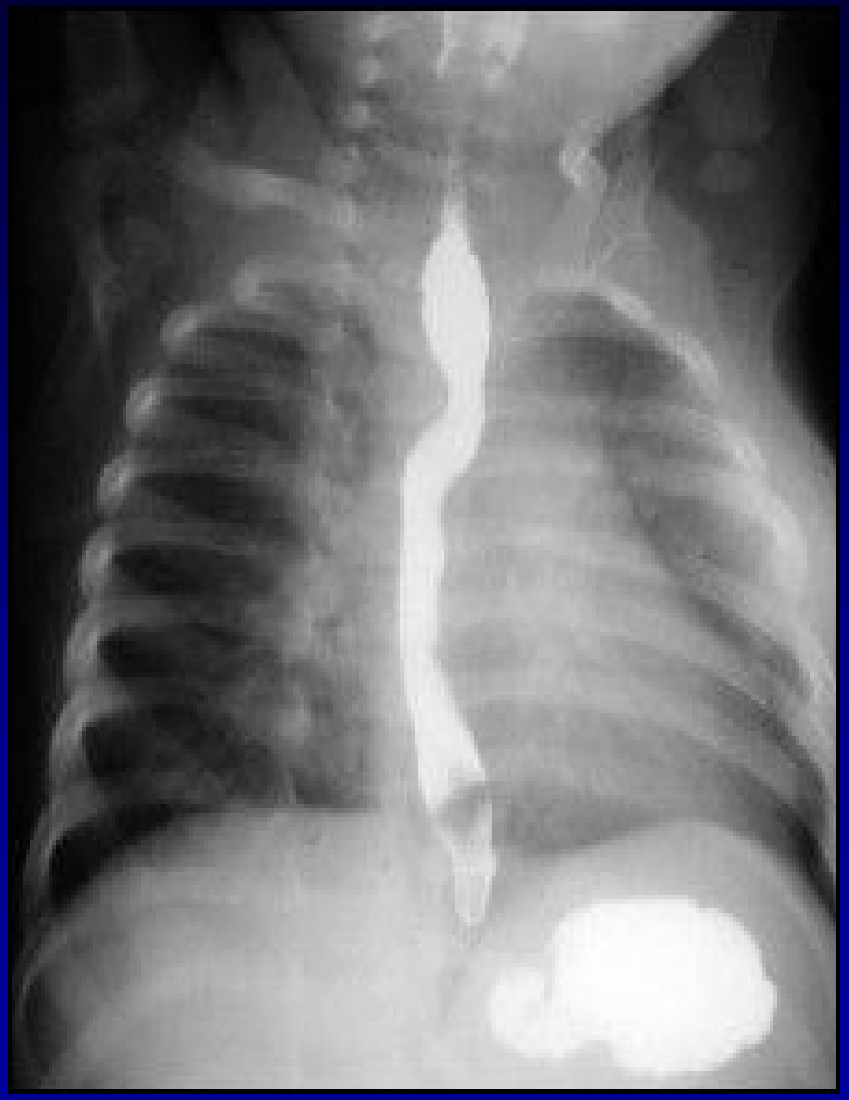
Right Aortic Arch



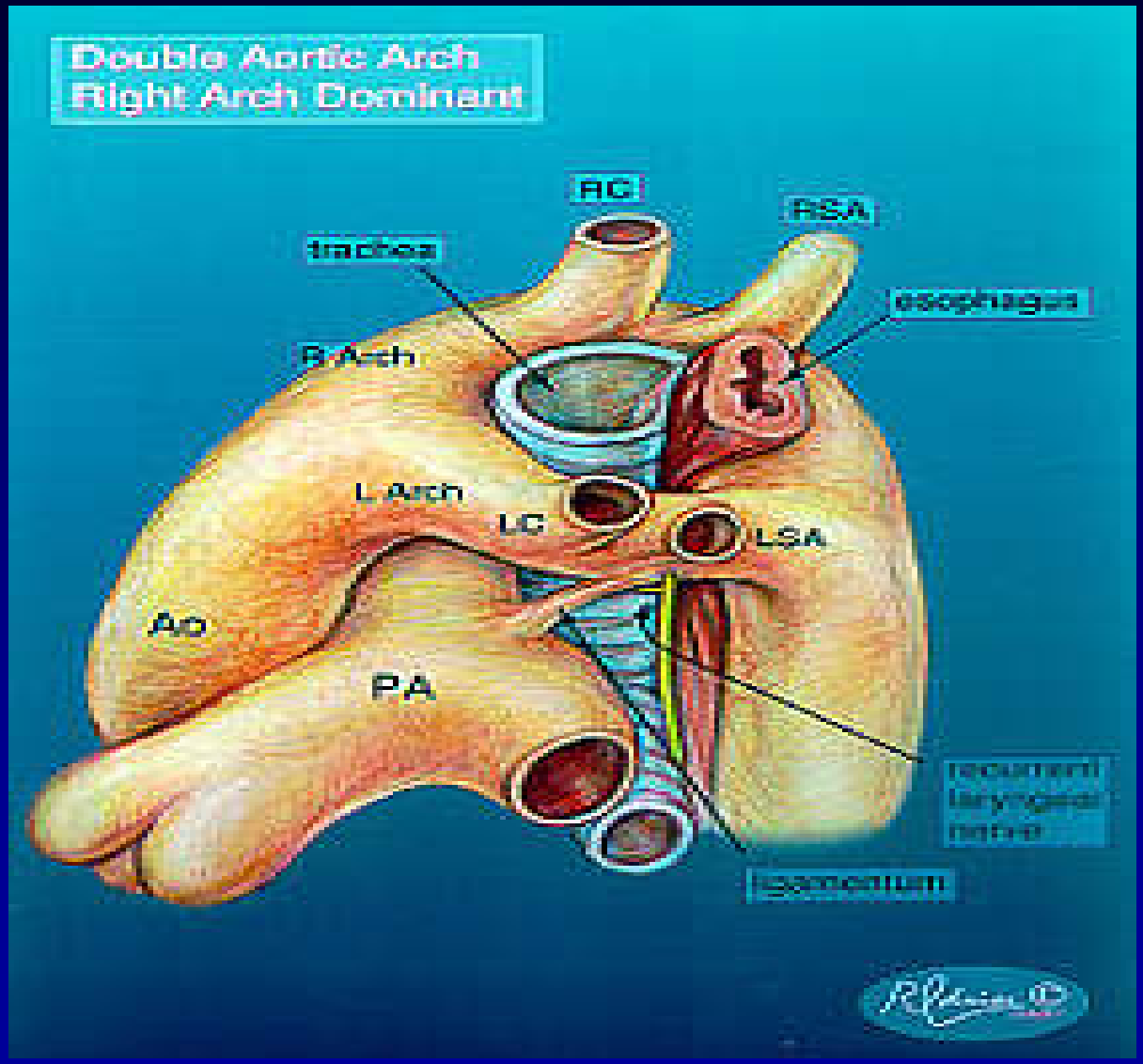
Double Aortic Arch

- The ascending aorta arises anterior to the trachea and divides into 2 arches which pass posterior and to the right and anterior and to the left of the trachea and esophagus.
  - Type 1 has both arches patent and functioning and this type is most common
  - Type 2 has both arches intact but one is atretic, usually the left

- AP views of an esophagram demonstrate bilateral compression of the esophagus forming a reverse “S” sign with the superior curve resulting from the high R arch compression and the inferior curve from the inferior L arch compression.

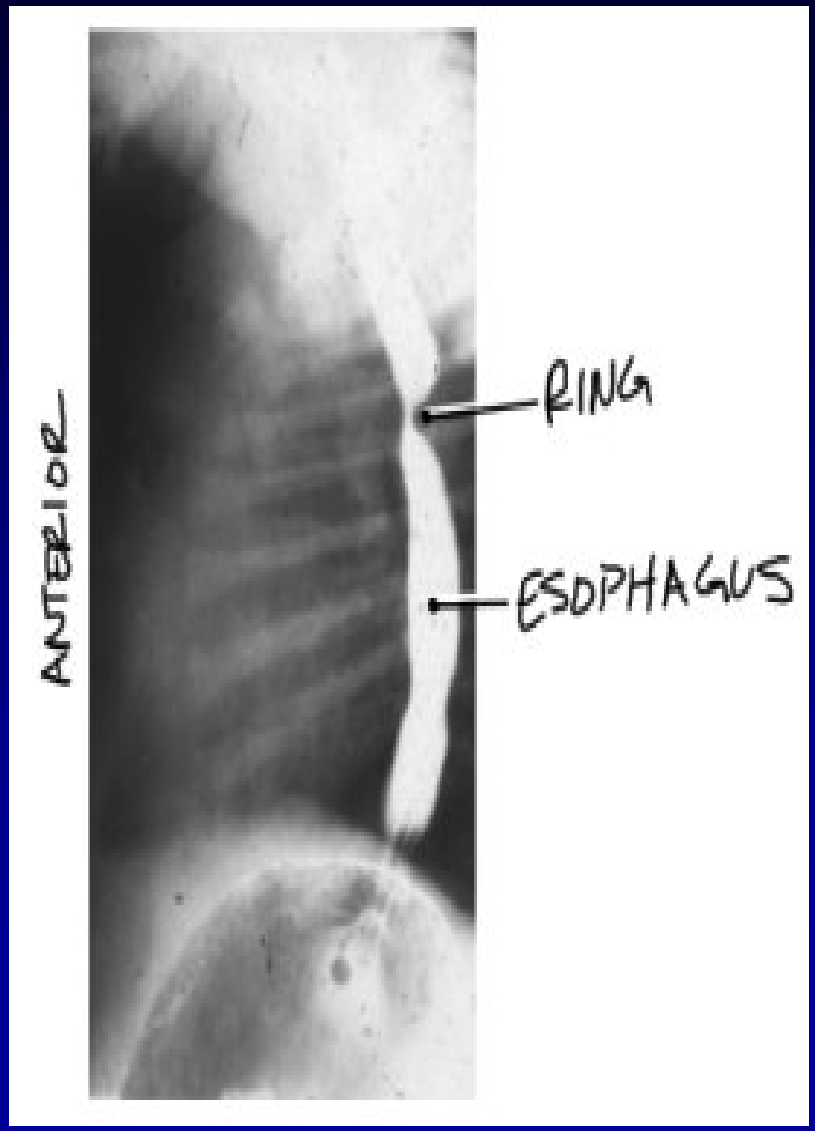


Double Aortic Arch

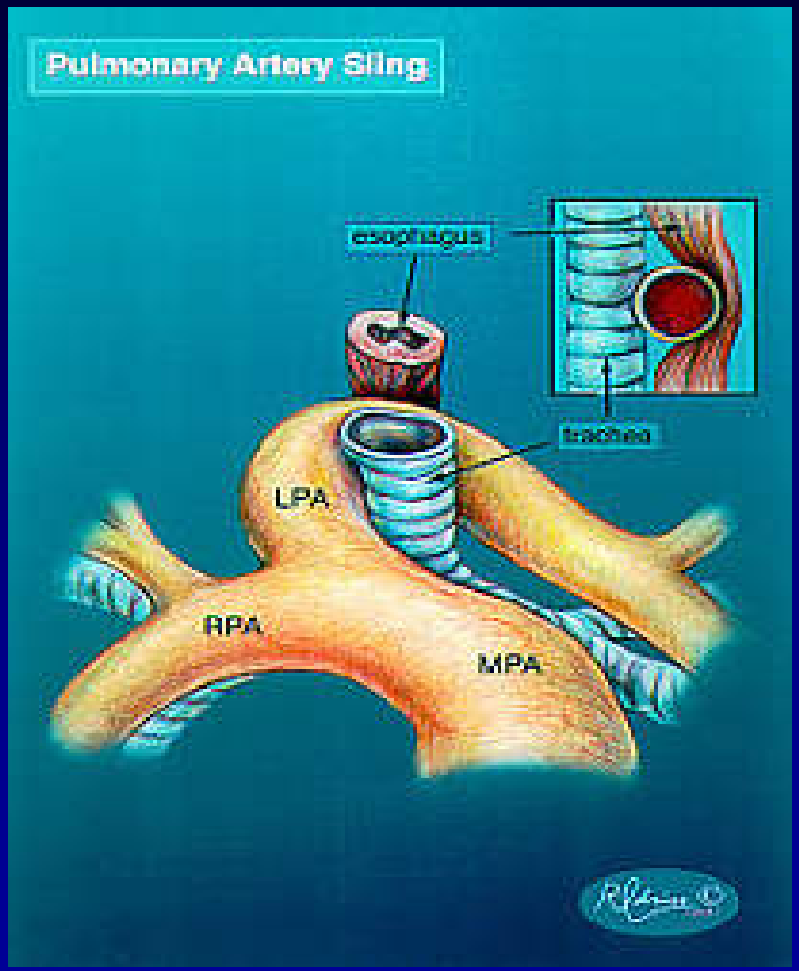


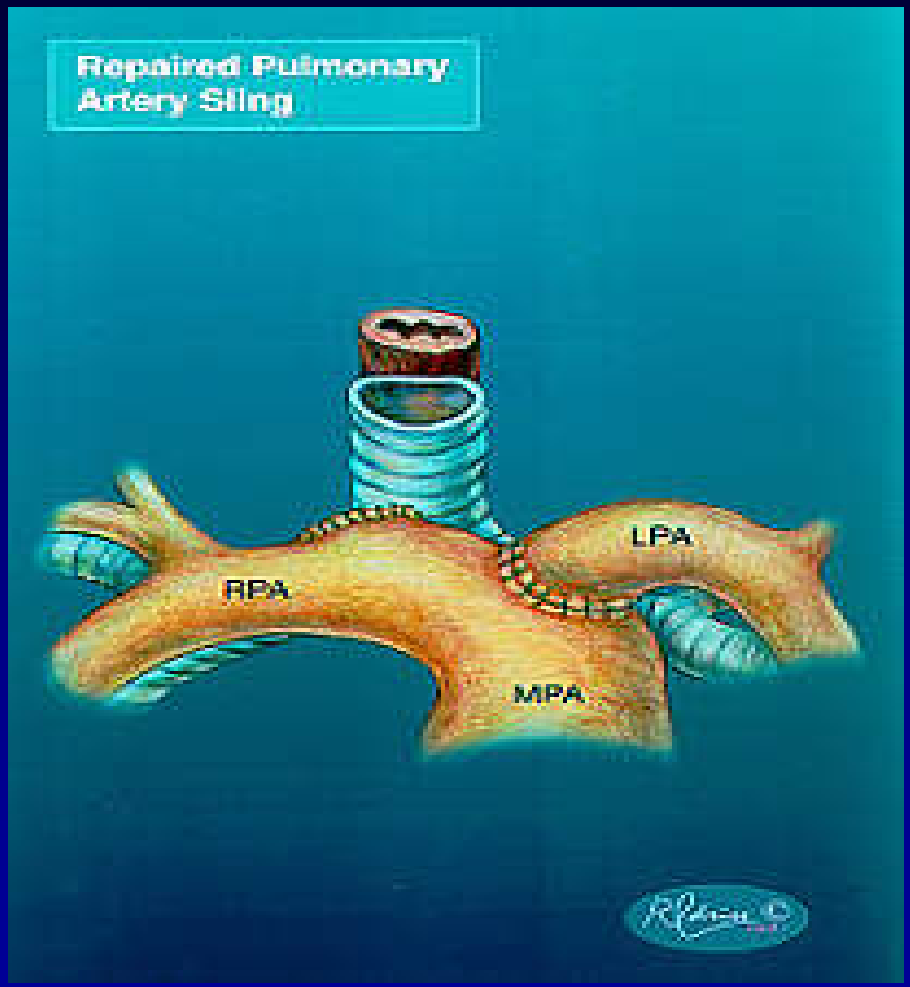
Pulmonary Artery Sling

- Least common vascular anomaly to cause tracheal compression
- Created by anomalous origin of the left pulmonary artery from the posterior aspect of the right pulmonary artery.
- Associated tracheobronchial abnormalities:
  - Complete tracheal rings, tracheomalacia.
  - Hypoplasia and stenosis of tracheal segments
- Congenital heart defects are found in 50% of pulmonary artery sling cases; most commonly ASD, PDA, VSD, and left superior vena cava.
- Other organ system abnormalities may occur. These include imperforate anus, Hirschsprung disease, biliary atresia, and genitourinary defects. Abnormalities of ovaries, vertebrae, thyroid gland, and pulmonary parenchyma have been reported.

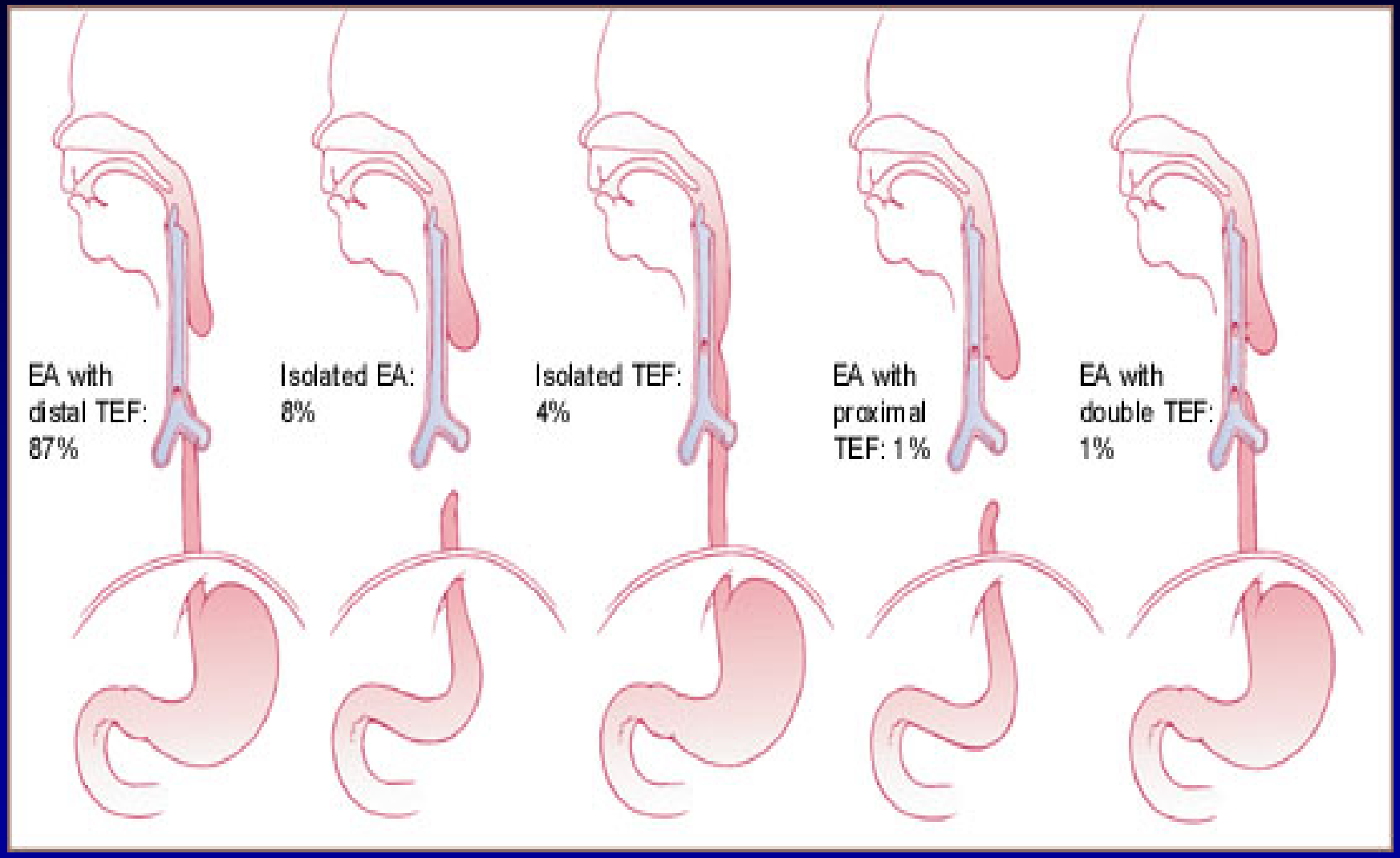


Pulmonary Artery Sling



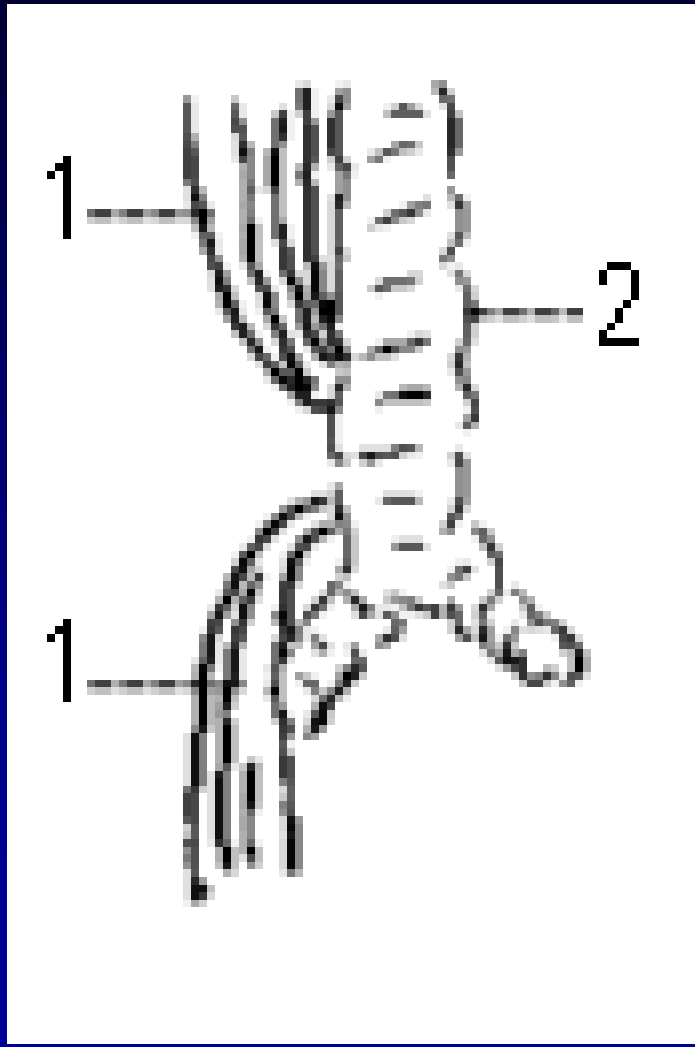


Tracheoesophageal fistula



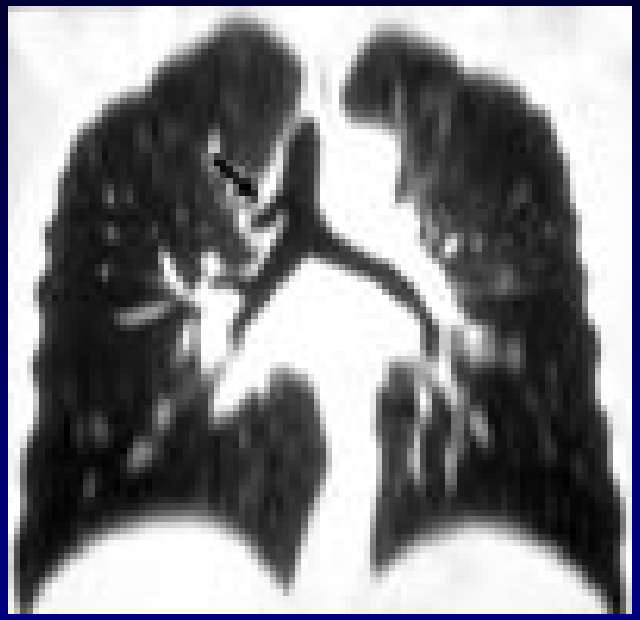
Tracheal esophageal fistulas

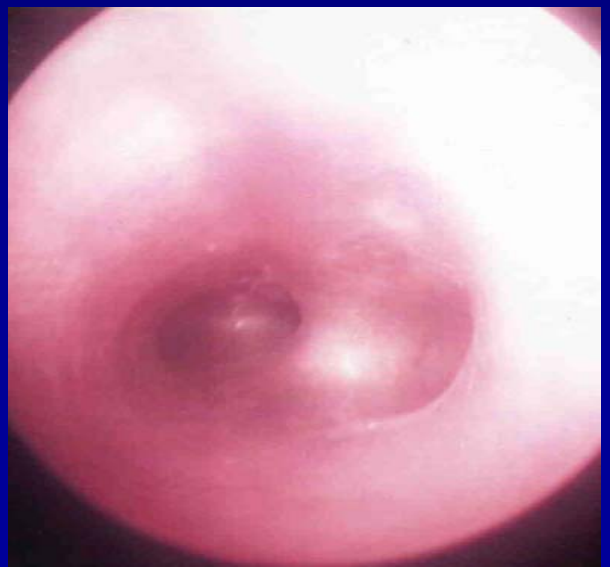
- 1st surgical repair in 1888 with esophageal atresia
- 17-70% have associated anomalies including Down's syndrome
  - CV: VSD, PDA, Tetralogy, ASD, R sided arch
  - GI: dudonal atresia, imperforate anus, malrotation, Meckel's diverticulum, annular pancreas
  - GU: horseshoe kidney, polycystic kidneys, ureteral malformations and/or reflux, hypospadias
  - MS: hemivertebrae, radial anomalies, poly or syndactyly, rib malformations, scoliosis



Tracheal bronchus

- Congenital malformation of the tracheo-bronchial tree.
- Occurs superior to the bifurcation of the trachea - usually on the Right
- Frequently asymptomatic, but can be revealed by recurrent infection, atelectasis or bronchiectasis.





School Age Children

Case #3: AG - 13 y/o Caucasion male

- Presents for evaluation of difficult to control asthma
  - 15-20 exacerbations over the last year
  - Steroid-dependent over the last 2 months
  - Poor exercise tolerance
  - School absences - 2 months in the last year

HPI: Symptom onset at 6 mos with acute croup-like episode

- Recurrent episodes of barky cough and hoarseness dx'd as asthma
- Typical asthma exacerbation starts as a "throat tickle" then proceed to coughing paroxysms
  - Within 24 hours, SOB and chest/throat tightness
  - Requires acceleration of albuterol MDI/ nebulizations
- Prior to last year, 6-8 oral steroid bursts for 5-10 days
- Recurrent episodes of GERD
- Chronic nasal congestion

Steroid Side Effects:

- Cushingoid
- Growth retardation
- Osteoporosis
- Weight gain
- Adrenal insufficiency/ steroid withdrawal
- Acne

Current Medications:

- Medrol 4 mg q.d.
- Fluticasone/salmeterol 500/50 Diskus 1 inhalation twice daily
- Prilosec 40 mg q.d.
- Allegra 60 mg b.i.d.
- Flonase - 2 sprays/nares q.h.s.
- Zithromax 250 M/W/F
- Xopenex and albuterol nebulizers p.r.n.
- Retin-A ointment q.h.s.
- Minocycline 500 mg q.h.s.

Past Medical History:

- **Birth history** - 6# 10 oz term infant
  - C-Section due to placenta previa
  - Fussy in the first 6 months of life; colic/gassy
- **H/o of recurrent OM** - PE tubes at 9 months
- **Sinusitis** - CT scan last year was negative
- **Asthma Hx**
  - Hospitalizations - (1) x 3 days at 14 years of age
  - No ICU admissions or ETT
  - 10 prior ER visits – 4 involving EMS
  - No history of seizures or LOC related to respiratory distress
- **GERD** - clinically diagnosed 2 years ago

- **Family history**
  - FOC with h/o childhood asthma and hayfever
  - MOC with hayfever and mild eczema
- **Social history**
  - Only child living in Kentucky. 10th grade; “A/B” student. Many friends. Active in Boy Scouts.
  - Resides in 25-year-old wood/ brick home. FOC does smoke. + dog in the home.
- **Prior w/u included:**
  - Skin testing approximately 4 years ago
  - CT scan approximately 1 year ago
  - All CXR’s with minimal hyperinflation and no infiltrates
  - Bronchoscopy/BAL approximately 1 year ago
    - Bronchitis
    - Lipid index of 90

- **Allergen history**
  - Perennial nasal stuffiness ± post-nasal drip
  - Seasonal allergic conjunctivitis
  - Skin test (+) - molds, dust mite, feathers, shellfish and peanuts
- **Respiratory triggers**
  - Exercise
  - Viral infections
  - Dust
  - Tobacco smoke
  - Pollution
  - Odors such as perfumes and paint fumes
  - Weather changes specifically cold and damp
  - Feather pillows, mowing the lawn, trees, flowers…….
- **Allergies**: Amoxicillin → hives
- **ROS** - poor growth seen by peds endocrinologist, chronic fatigue and tension HA, no snoring or mouth breathing, no eczema

Physical Examination:

Afebrile **HR** 80 **RR** 16 **Bp** 127/85 **Saturations** - 96%

Wt. 53 kg (25%) **Ht.** 149.8 cm (<5%)

General: small for age, cushingoid male

HEENT: No allergic shiners, Normal fundoscopic, normal nasal mucosa, 1+ tonsils, no cobblestoning of the posterior pharynx

Chest: Truncal obesity

Lungs: Good aeration without wheezing even on forced expiratory maneuver

CV: RRR, nl S1S2 without murmur

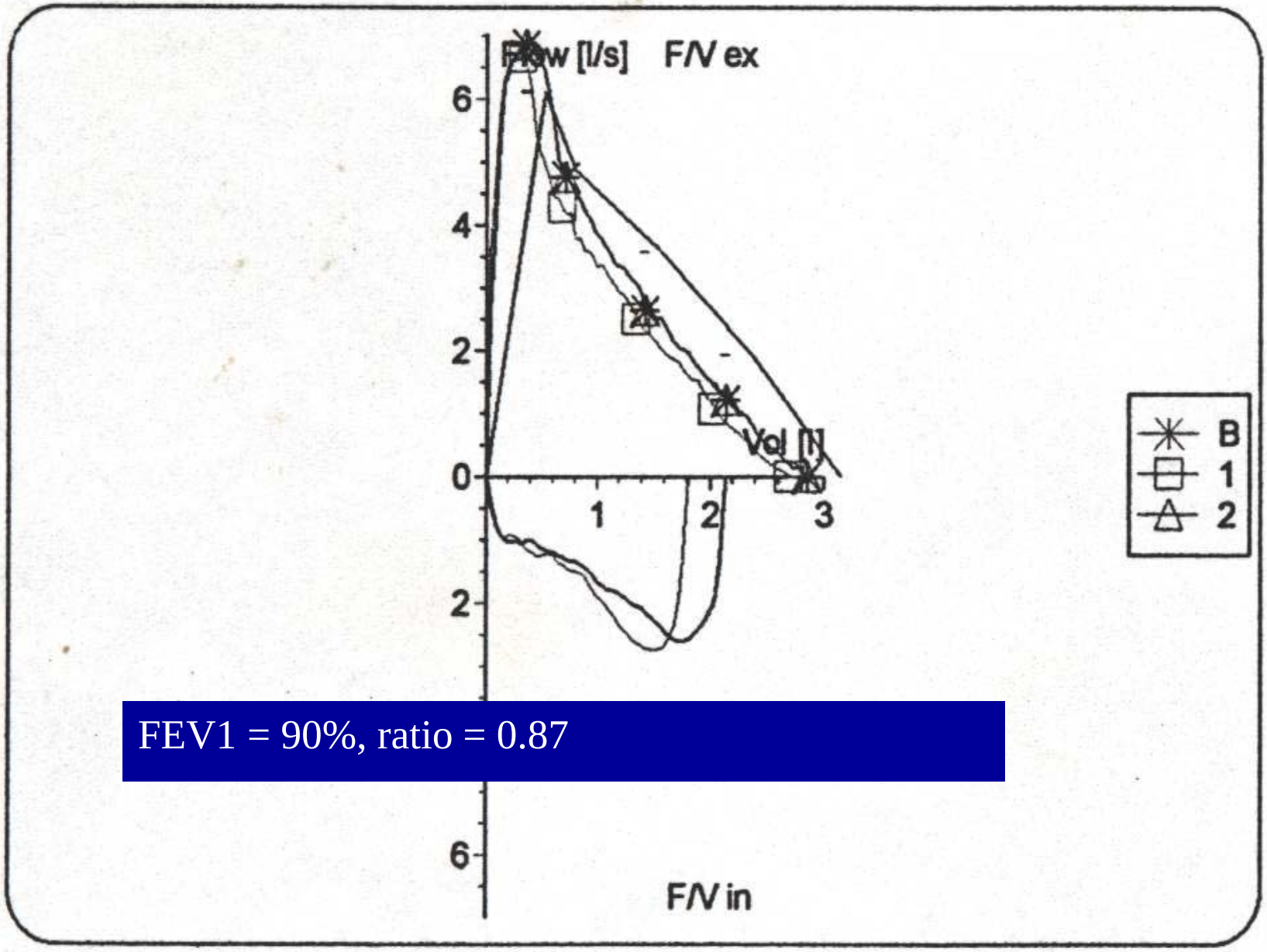
Abdomen: Obese with NABS, soft NTND without HSM/masses

Ext: No clubbing, cyanosis, and/or edema

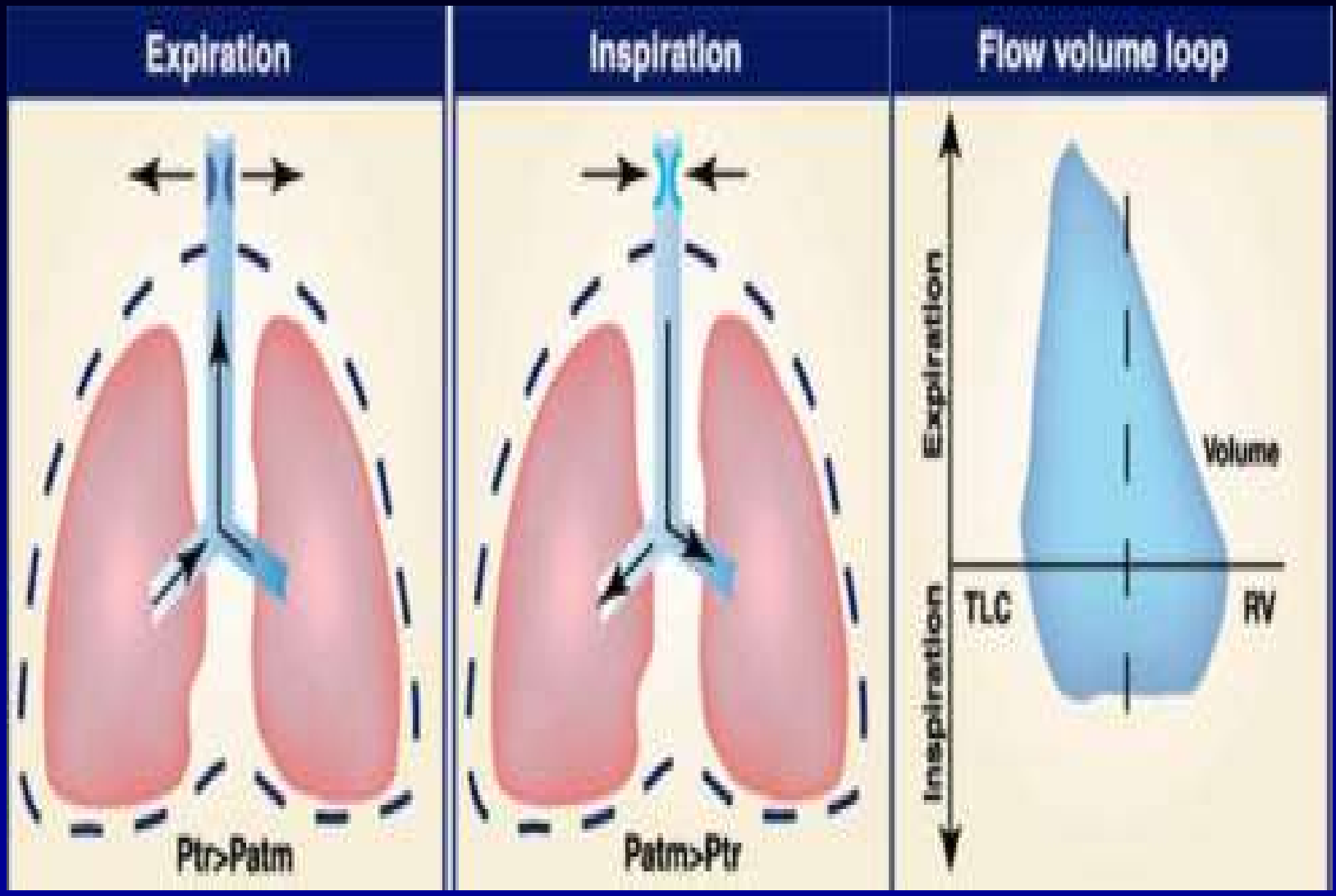
Skin: Acne-form lesions on face, no eczema

Initial Inpatient Evaluations:

- CBC: 3% serum eosinophilia
- IgE 679 IU/ml (0-450)
- Impedance probe positive for multiple episodes of non-acidic reflux
- Mch challenge: negative
- Spirometry pre- and post bronchodilator and volumes
- onsults



FEV1 = 90%, ratio = 0.87


Expiration
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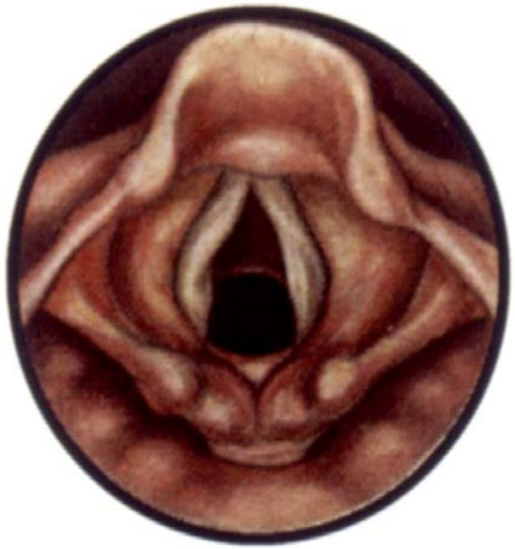
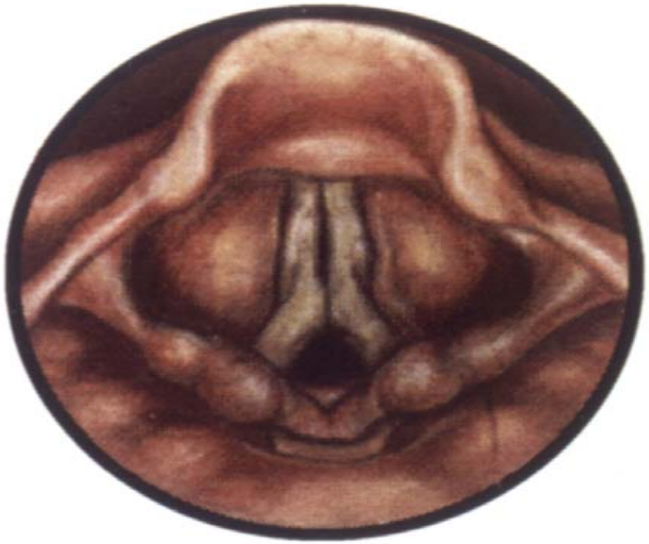
Expiration
Inspiration
Volume
TLC
RV

Signs & Sx of VCD

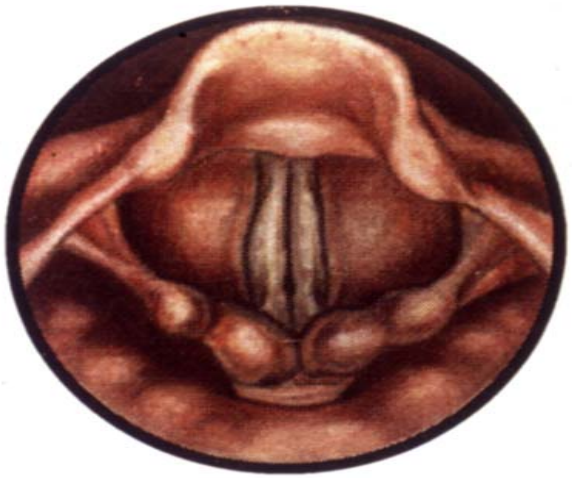
- SOB
- Stridor
- Chest and throat tightness
- Cough
- Wheezing
- Choking
- Dysphonia
- Hyperventilation sx
- “Can’t get enough air”



Normal
Mid-Inspiratory Glottis



Mid-Inspiratory
Vocal Cord Adduction
with Posterior Chinking



Closed
Mid-Inspiratory
Vocal Cord Adduction

Discharge:

Diagnoses:

- VCD
- Asthma
- Anxiety disorder
- GERD
- SSE's

Recommendations:

Drugs

- Omeprazole 20 mg BID
- Flovent 220 w puffs BID
- Paxil 10 mg qd
- NSI/fluticasone nasal qd
- Calcium/multivits qd

Other

- Psychotherapy
- VCD exercises

ACUTE VCD MANAGEMENT

- Calm, reassuring manner
- Slow breathing
- Panting/coughing
- Heliox inhalation (70% He/ 30% O_2)
- Sedation
- CPAP

CHRONIC VCD MANAGEMENT

- Sympathetic approach to the dx disclosure
- Speech therapy
- Treat conflicting diseases -
  Asthma, GERD, Nasal Rhinitis, EIB
- Relaxation therapy/self-hypnosis
- Psychotherapy +/- psychotropic drugs
- Discontinuation of unncessary medications

SPEECH THERAPY

- Place hand on abdomen
- Inhale slowly via nose (need the nose to be clear)
- Exhale slowly out through the mouth making soft “s or sh” sound
- Prevent shoulders from lifting/falling and keep neck relaxed
- Practice 5X daily in sets of 10

6 Month Follow-up Visit

Life would be infinitely happier if we could only be born at the age of eighty and gradually approach eighteen……

Mark Twain