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 children – young to school age
- 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 patterns for vocal cord dysfunction in school age children with difficult to control asthma

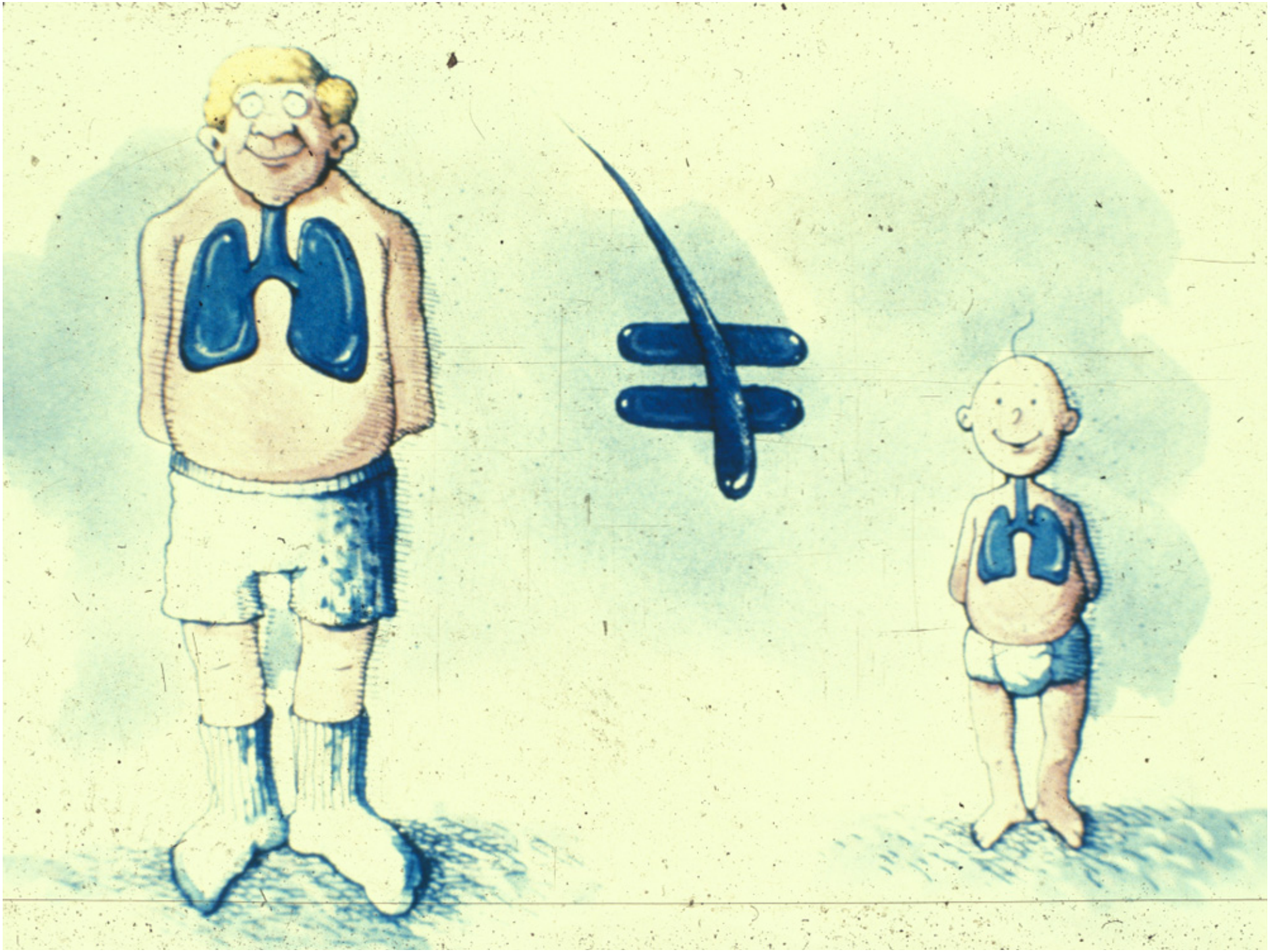
Differential Diagnoses:
Remember all that wheezes is not ASTHMA

PEDIATRICS

- Infection - VIRAL (RV, MPV, 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

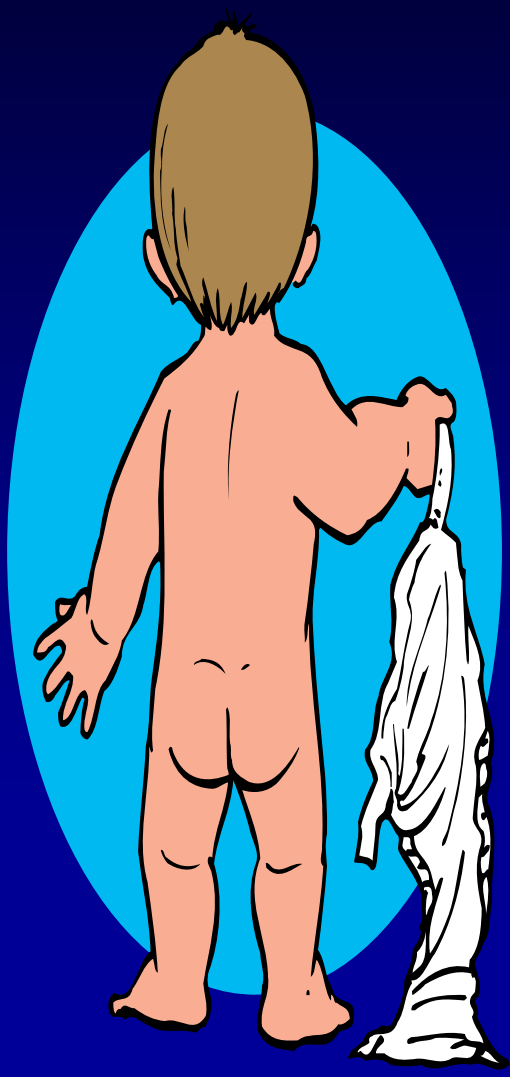


Pathophysiologic Properties Predisposing Infants and Young Children to Wheeze

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

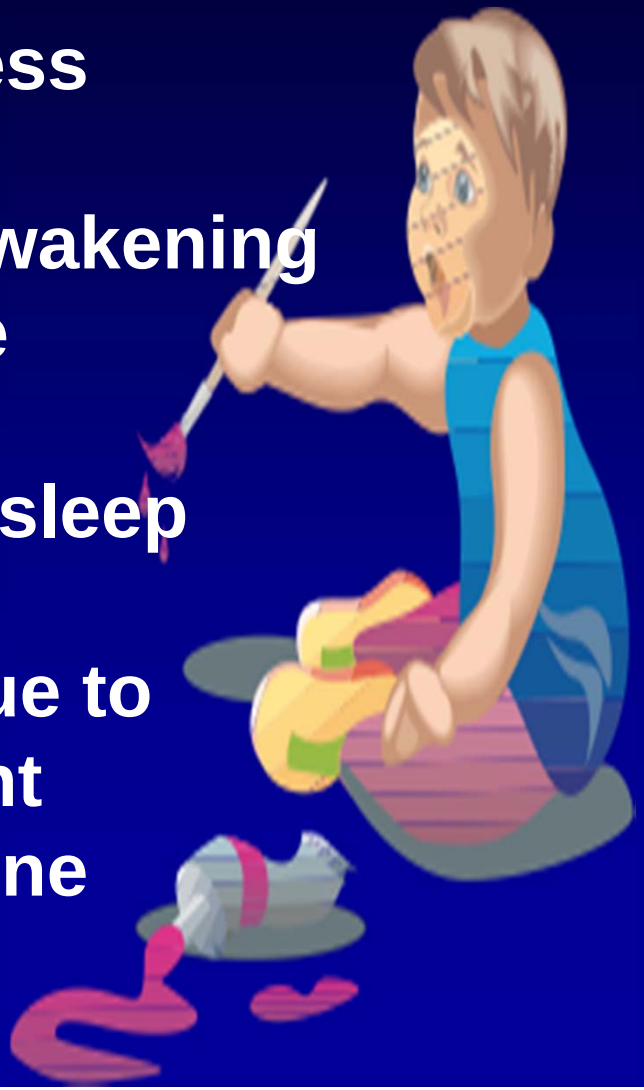
Asthma in Infants

- 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

- 2 episodes of respiratory distress requiring hospitalization
- Cough most prominent upon awakening and intermittent throughout the day/night
- Moderate snoring and restless sleep
- “Mouth-breather”
- Recurrent ER and PCP visits due to respiratory symptoms - frequent courses of antibiotics/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 q4-6º

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 and post-tussive emesis; moderate eczema; chronic loose stools on Abx

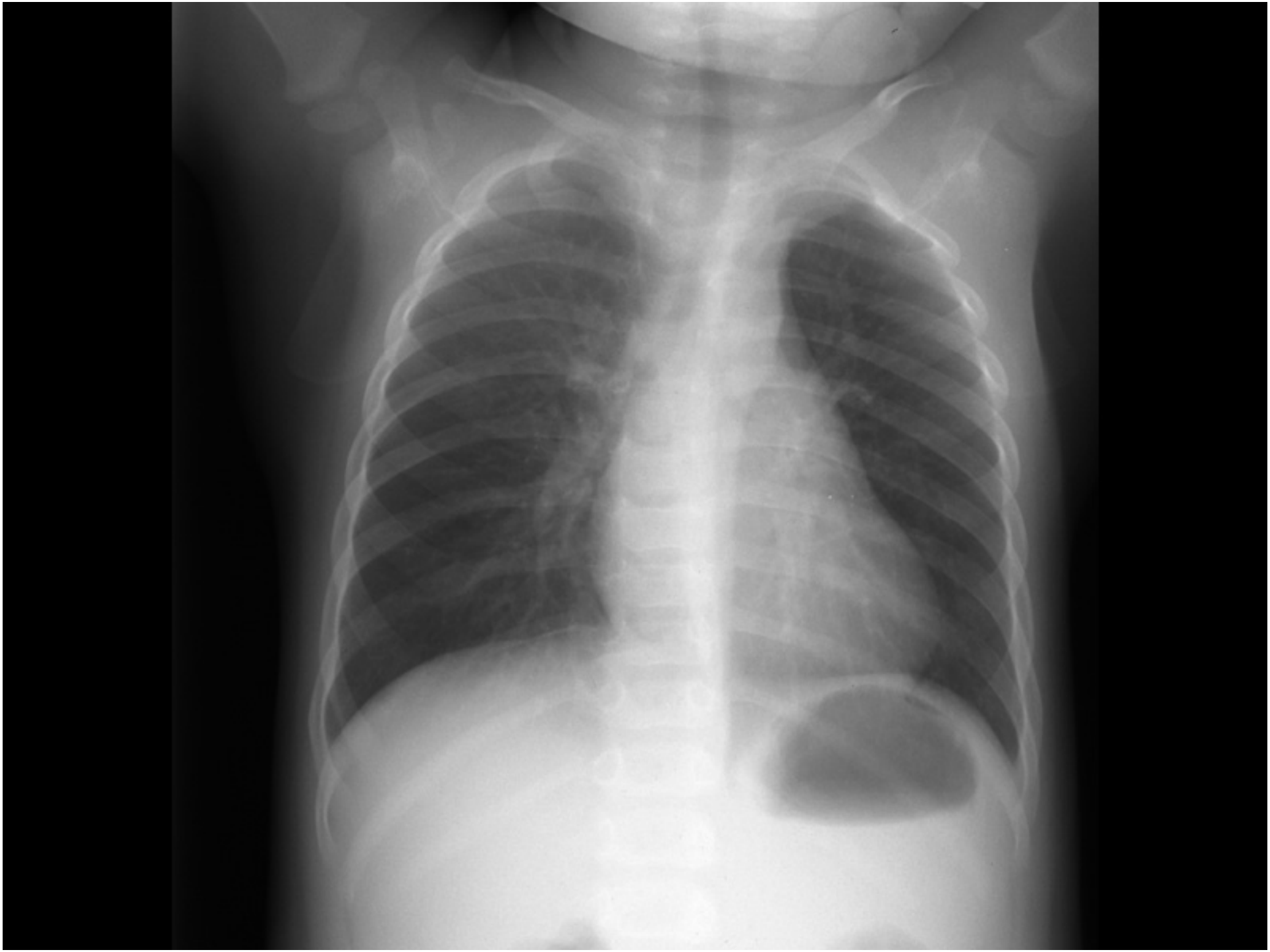
PE: Afebrile HR 156 RR 52 BP 92/60 91% RA
Wt. 6.2 kg (<5%)
Ht. 64 cm (10%)

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)

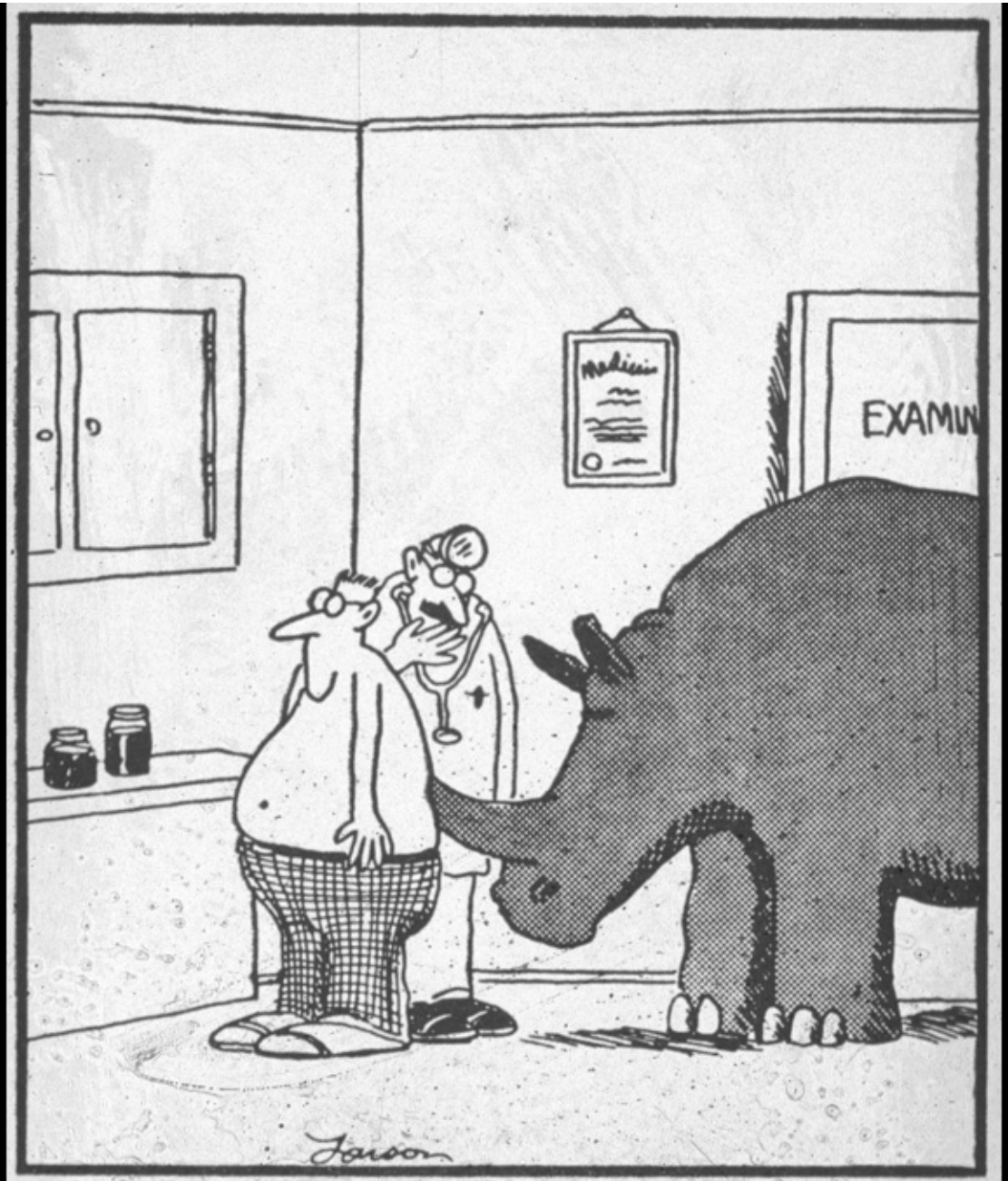
PLUS

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

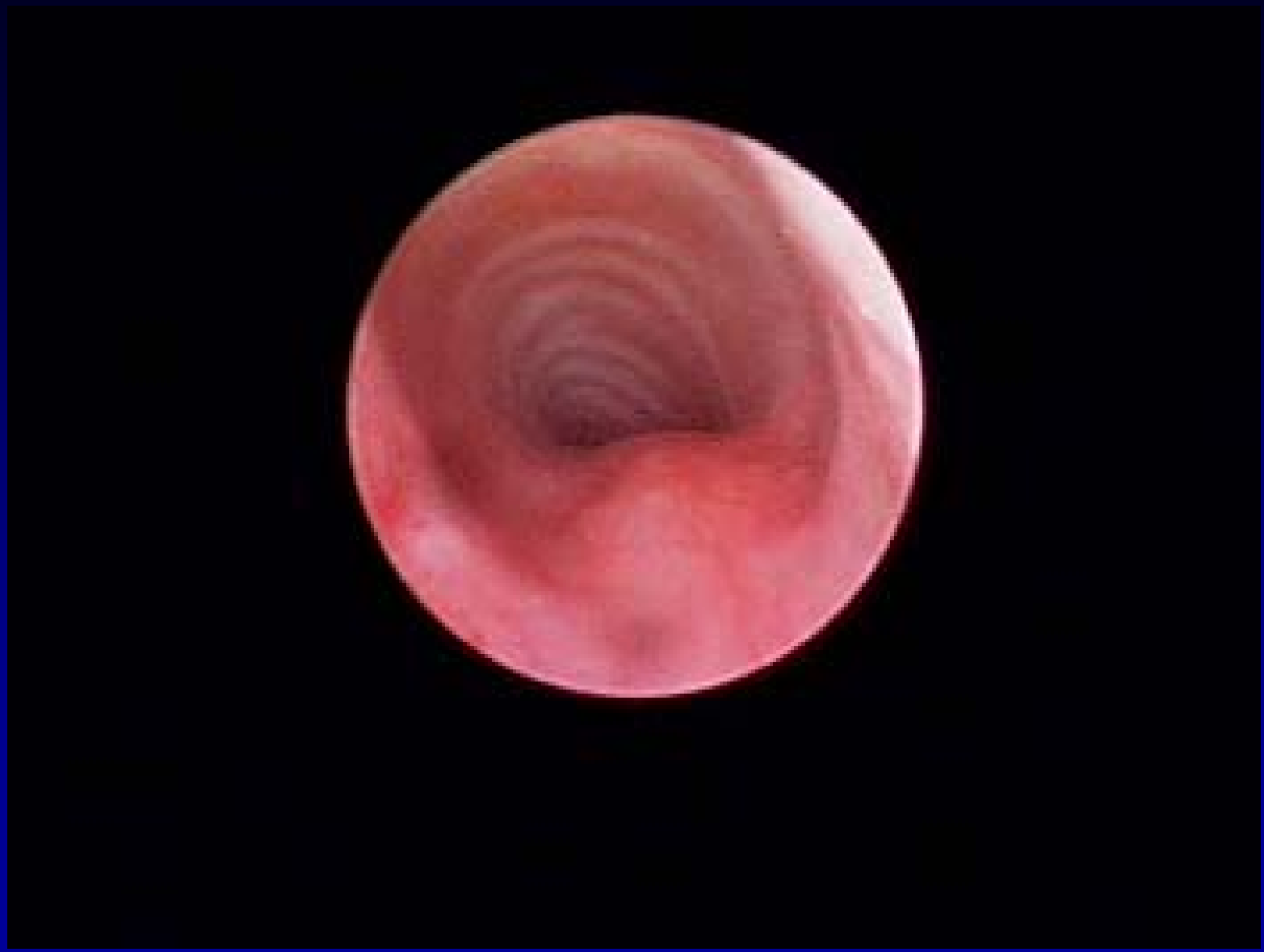
- Parent with asthma
- Atopic dermatitis
- Aeroallergen sensitivity

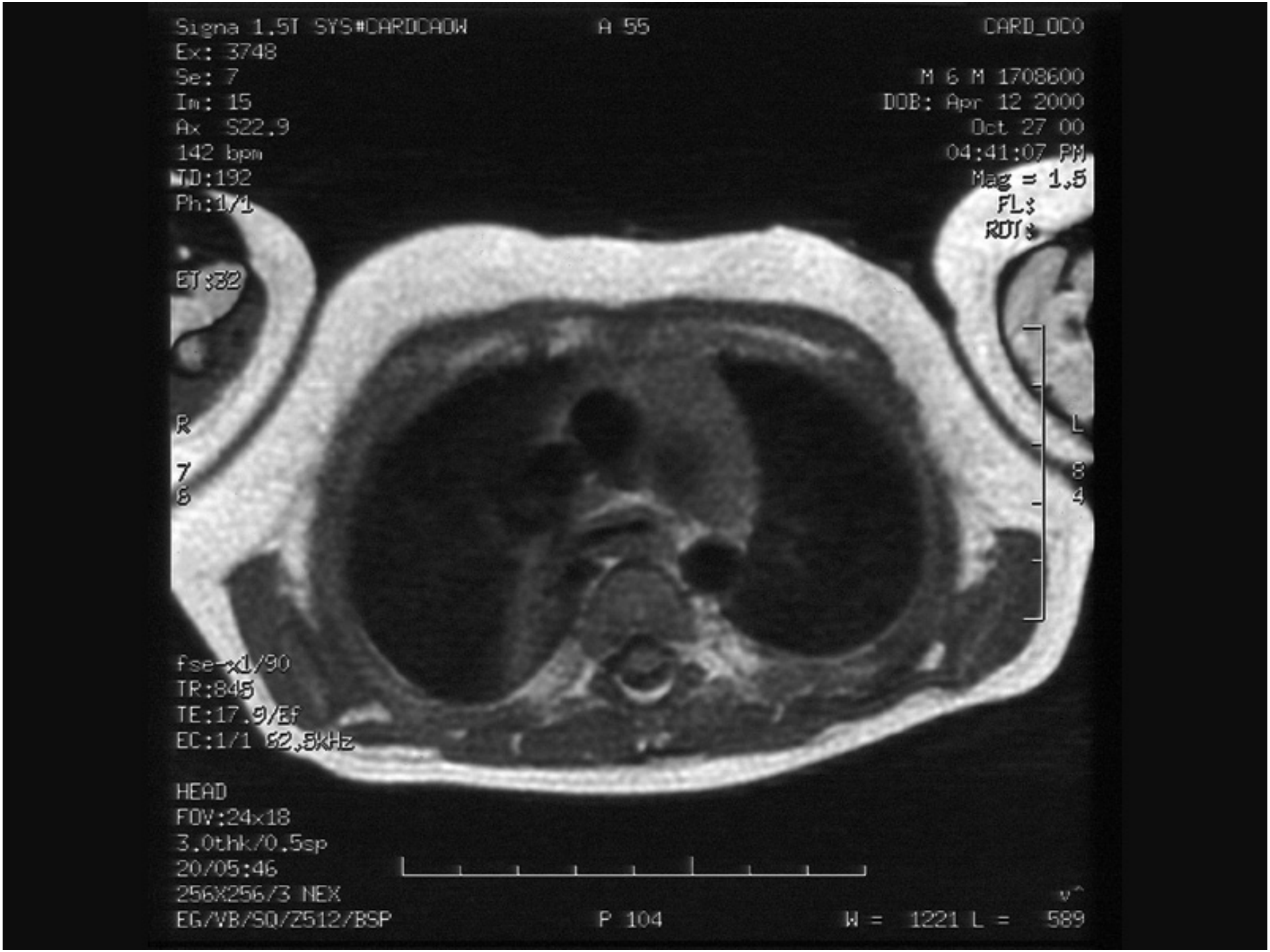
- Food sensitivity
- Peripheral eosinophilia (≥ 4%)
- Wheezing not related to infection

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
- Moderate persistent asthma
  - How do I want to deliver my meds
    - Maximize delivery
    - Maximize compliance
- <u>Nasal rhinitis and postnasal drip</u>

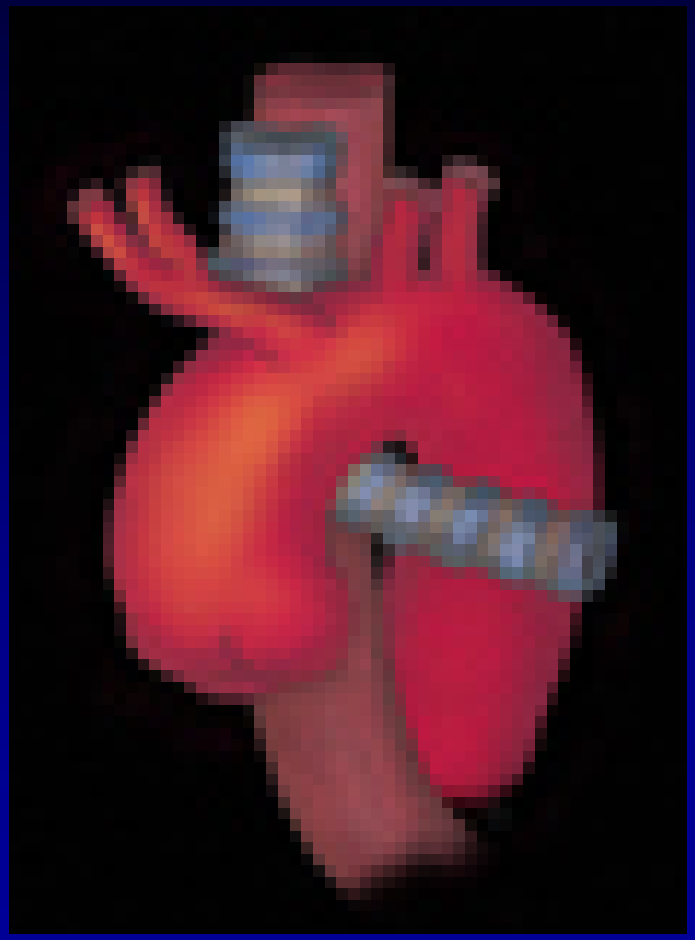
Vascular Anomalies

- 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
  - Anomalous innominate artery* (an incomplete ring)
  - Aberrant R subclavian artery*
  - Right aortic arch
  - Double aortic arch
  - Pulmonary artery sling

*Account for the most common vascular anomalies

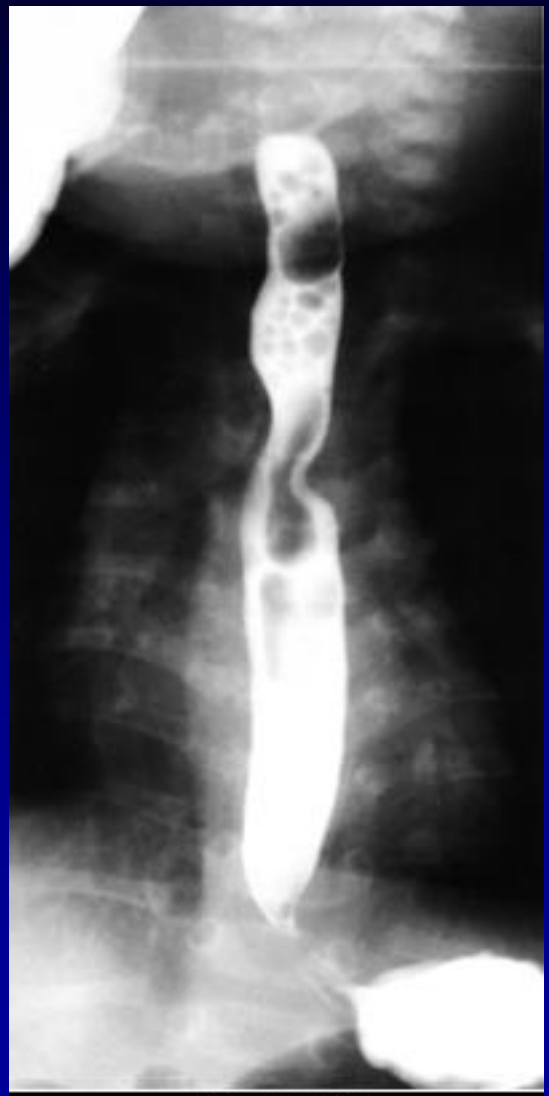
Innominate AA compression

- Common variant of normal – 5%
- 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
- Treatment
  - Reimplant innominate to R and anterior arch
  - Aortopexy (suspend arch to post. Sternum)

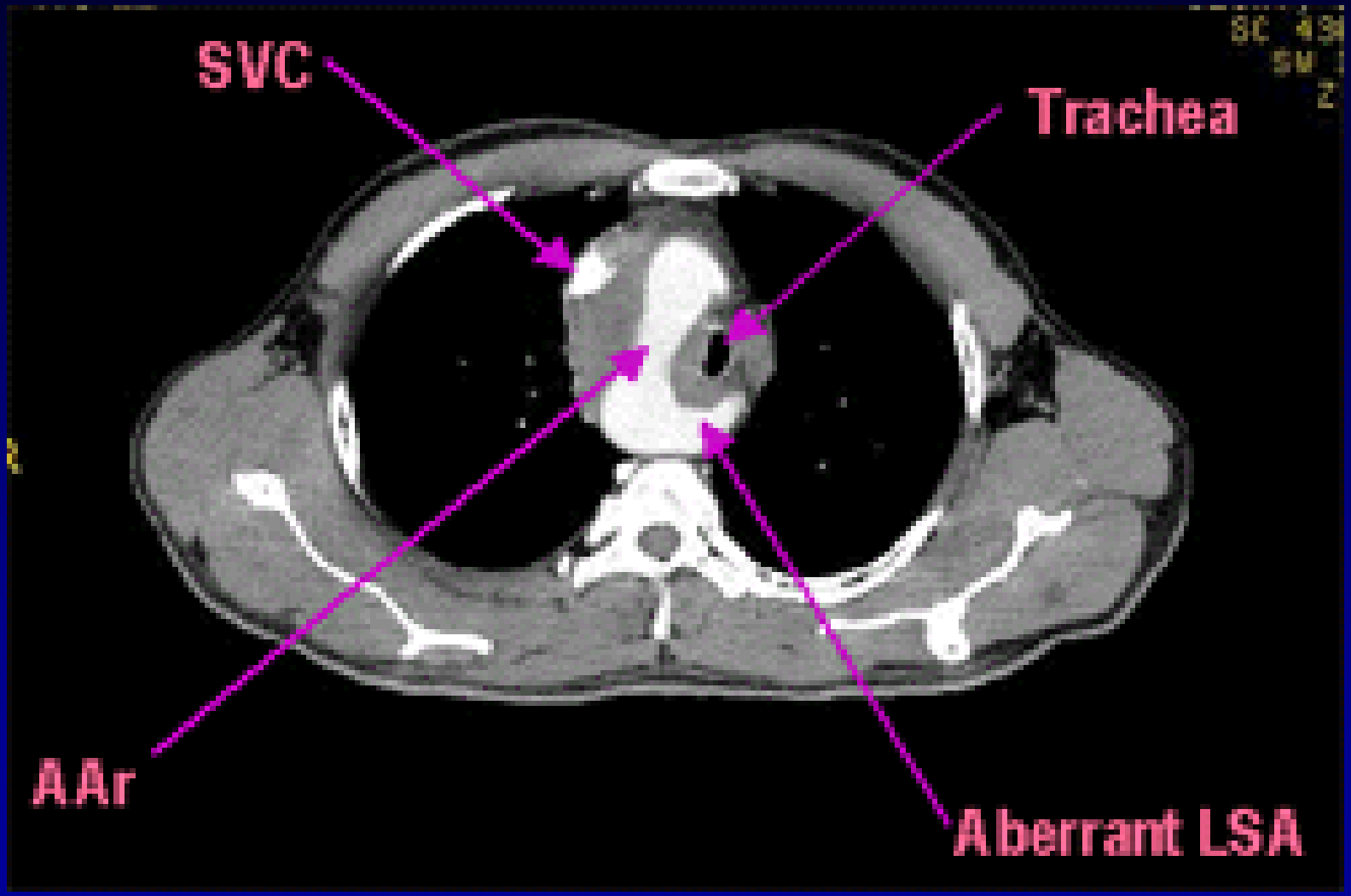


Aberrant Right Subclavian

- 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.

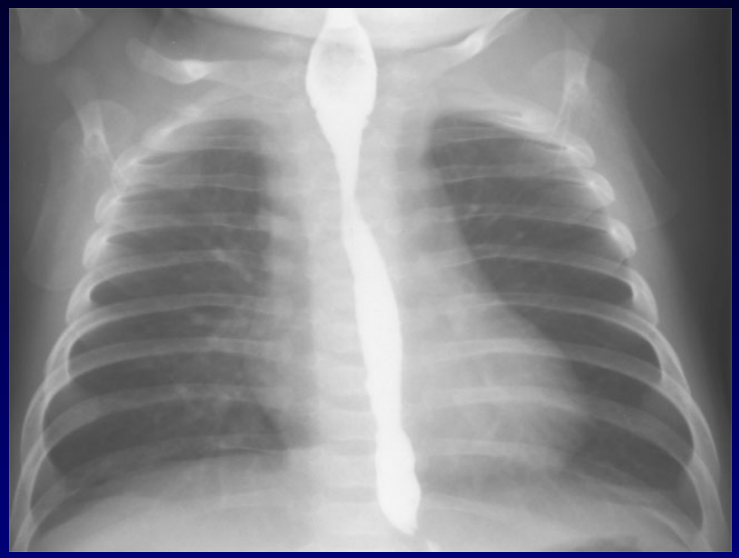


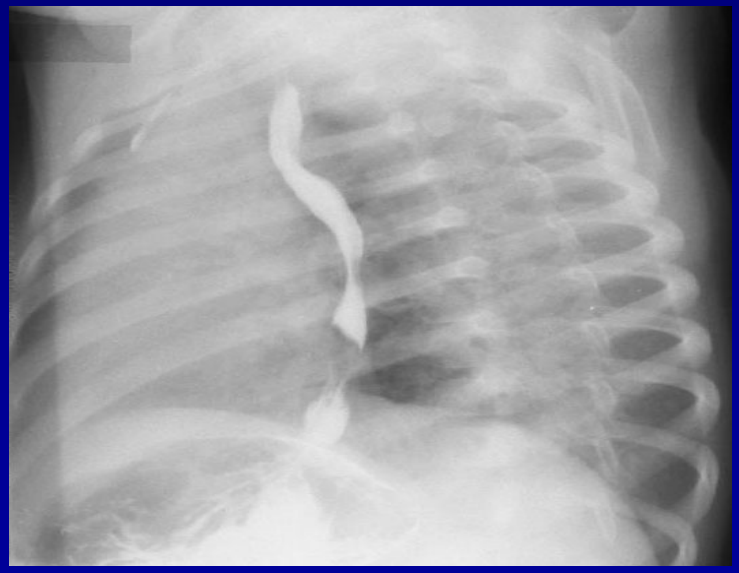
Right Aberrant Subclavian



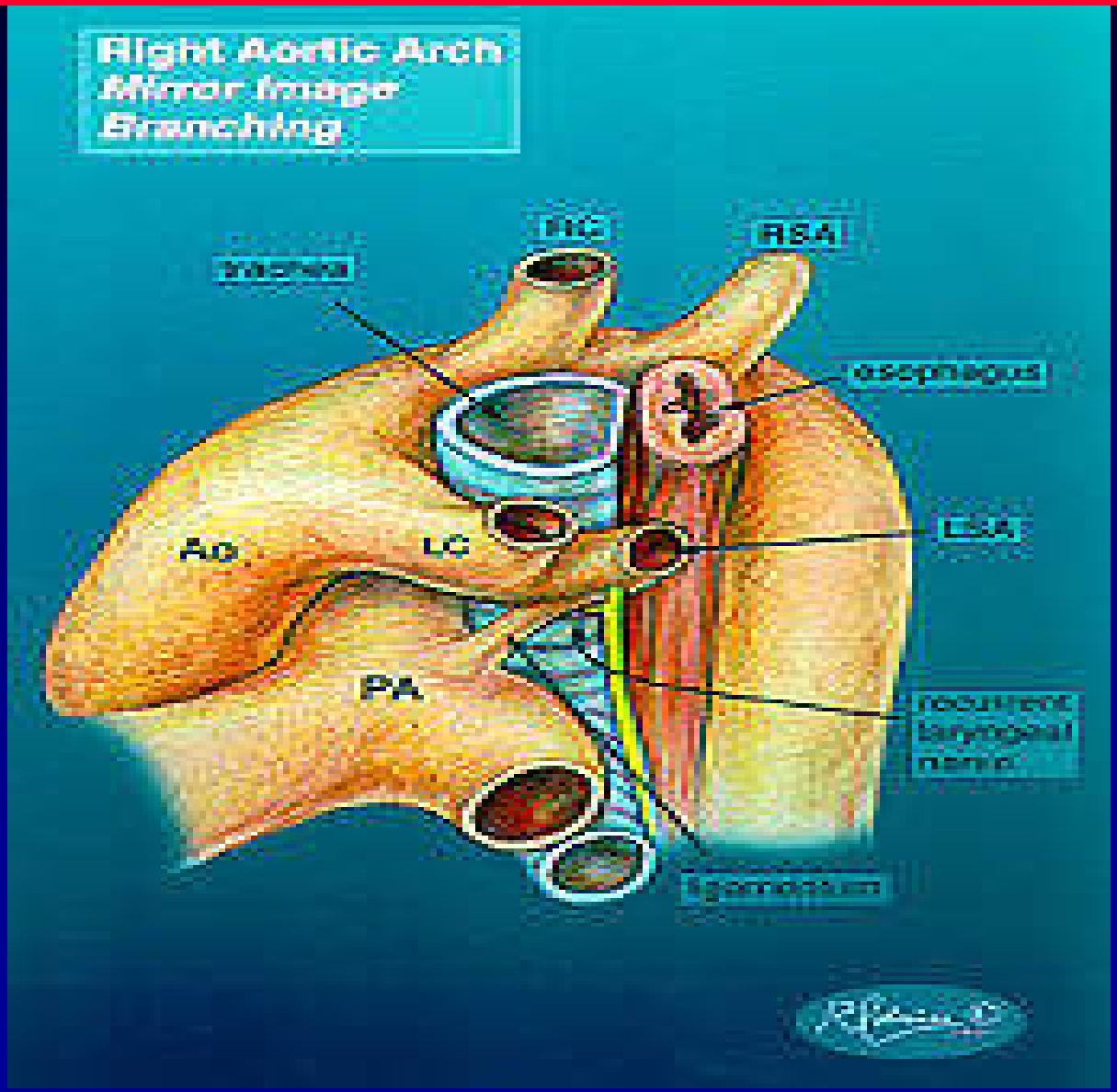
Right Aortic Arch

- 2nd most common type of ring
  - Most common is associated with an aberrant L. subclavian aa.
  - May or may not be associated with a PDA or ligamentum arteriosum
    - CXR and UGI are very helpful for diagnosis
  - More common in males
- Can be associated with Tetralogy/truncus/ transposition





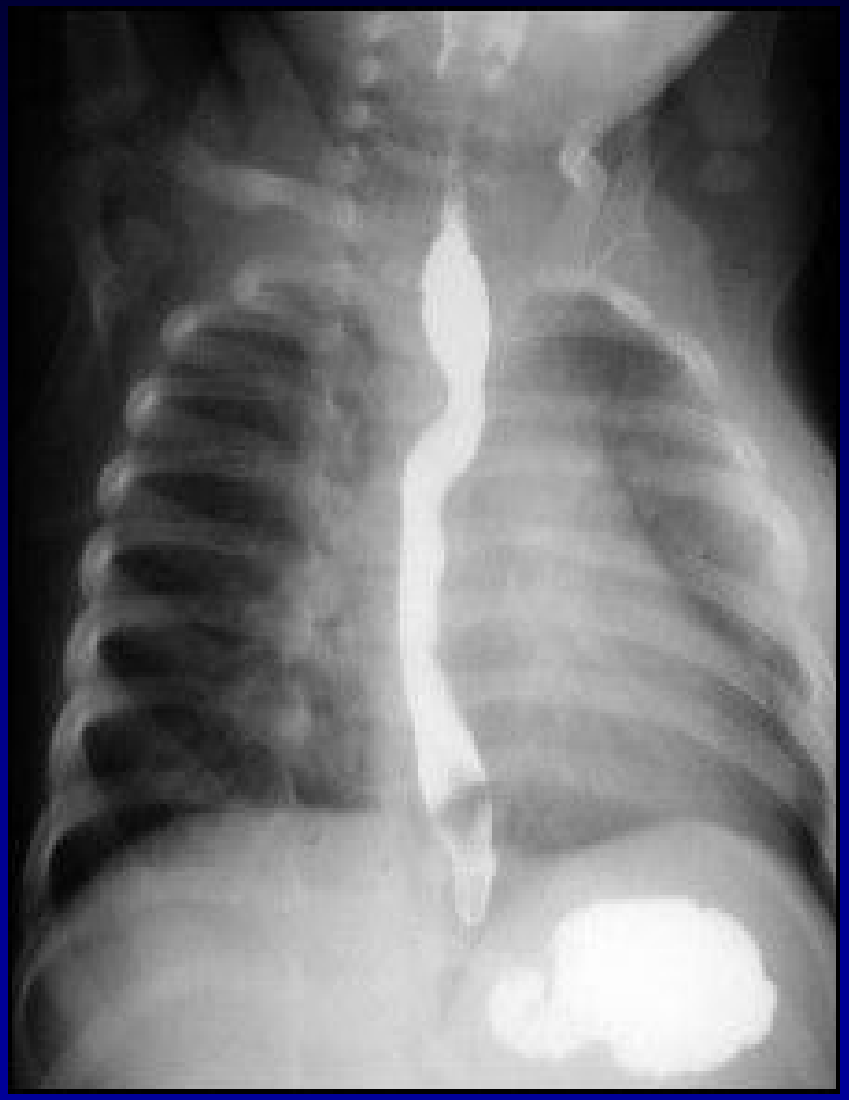
Right Aortic Arch



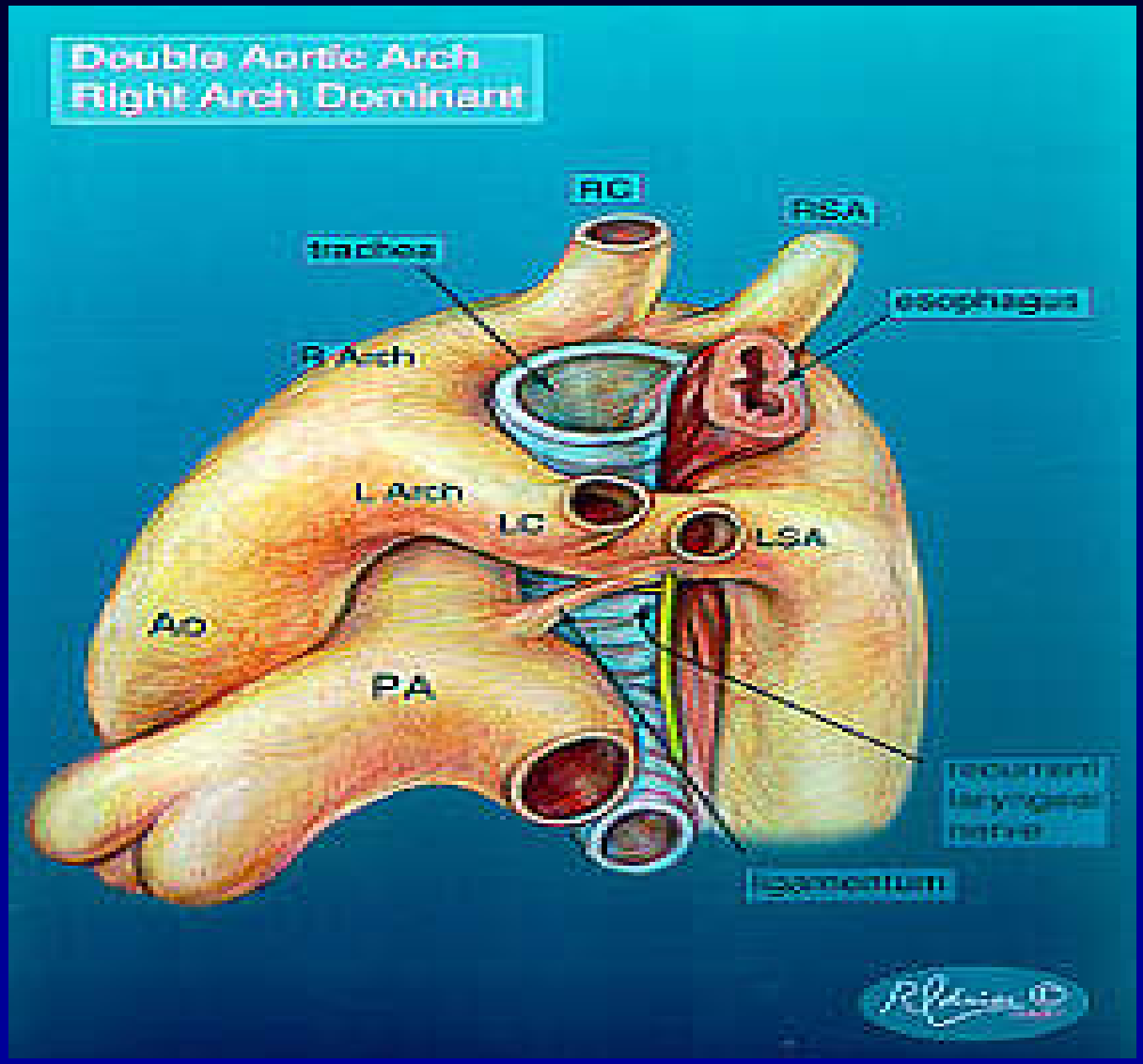
Double Aortic Arch

- 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.

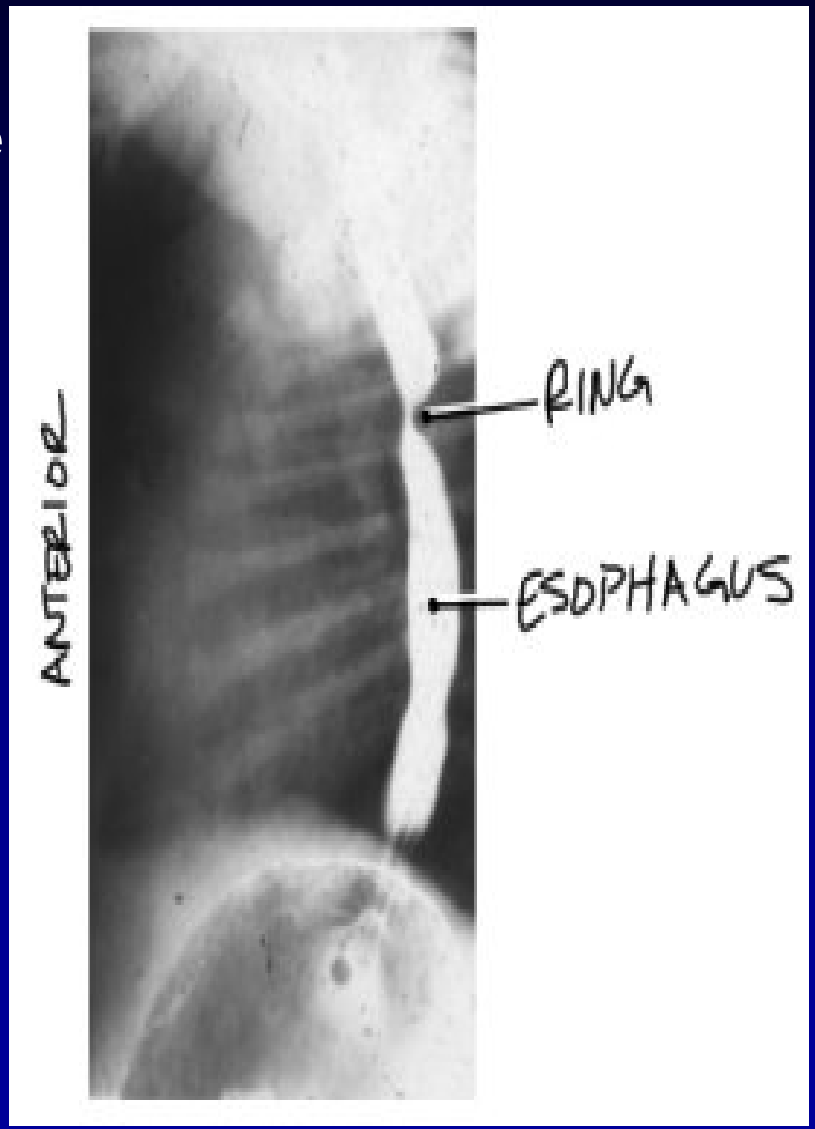


Double Aortic Arch

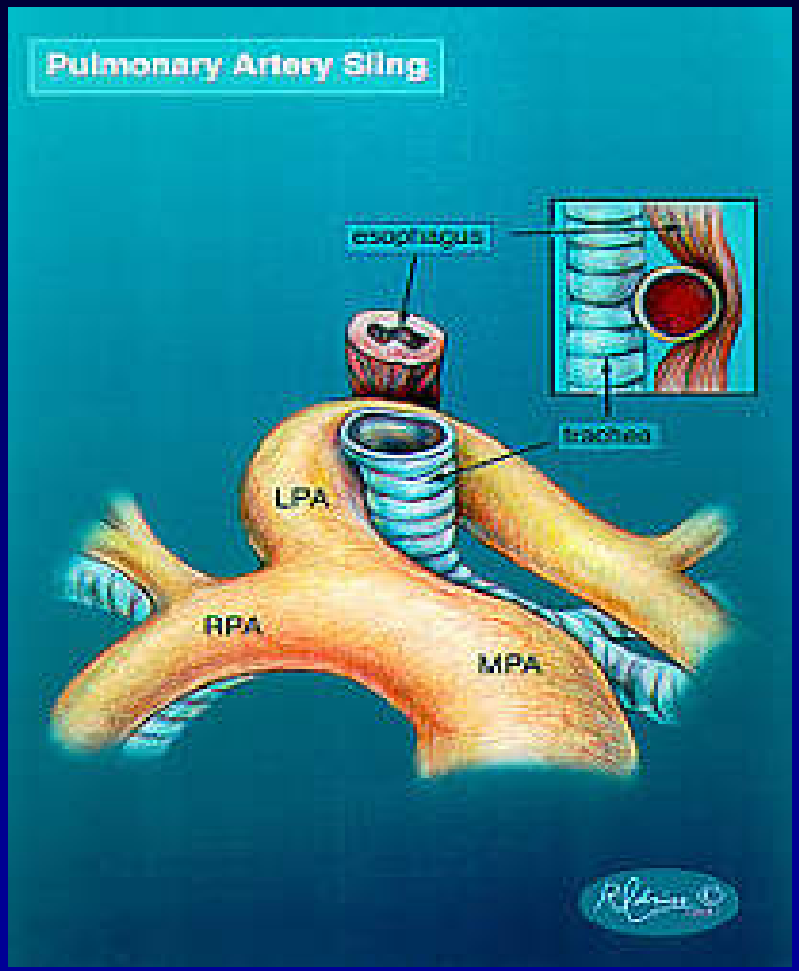


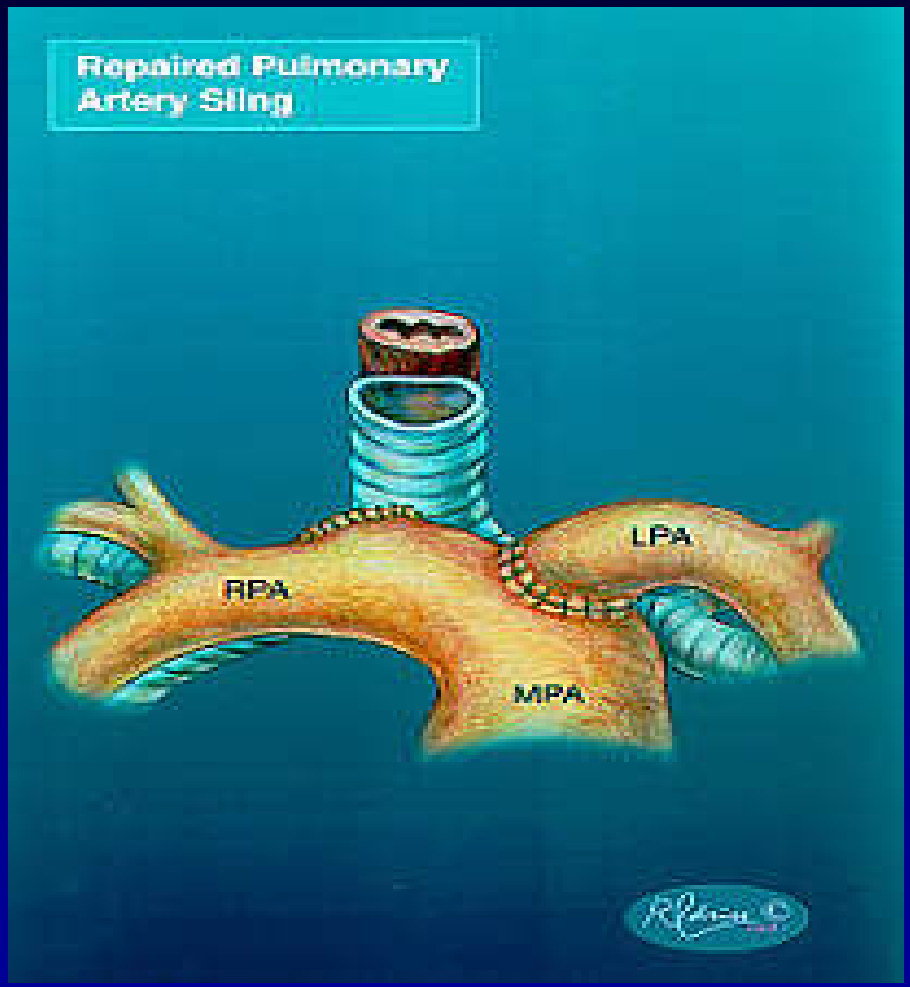
Pulmonary Artery Sling

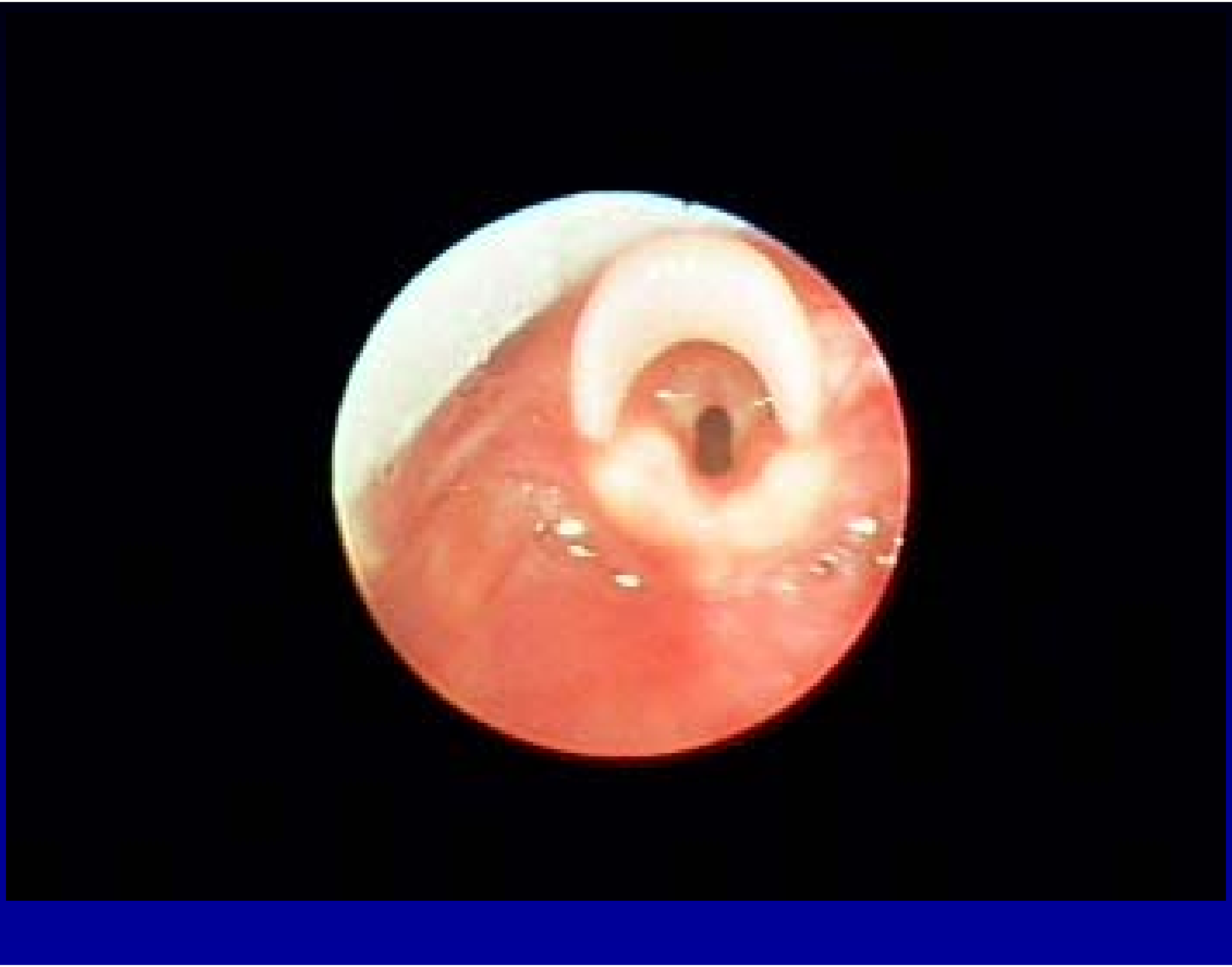
- Least common to cause tracheal compression
- Left pulmonary artery arises from the posterior aspect of right pulmonary artery.
- Associated tracheobronchial abnormalities:
  - Complete tracheal rings, tracheomalacia.
  - Hypoplasia and stenosis of segments
- Congenital heart defects in 50%
  - ASD, PDA, VSD, and left SVC
- Other organ system abnormalities: imperforate anus, Hirschsprung's, biliary atresia, GU defects (ovaries), vertebrae, and thyroid.



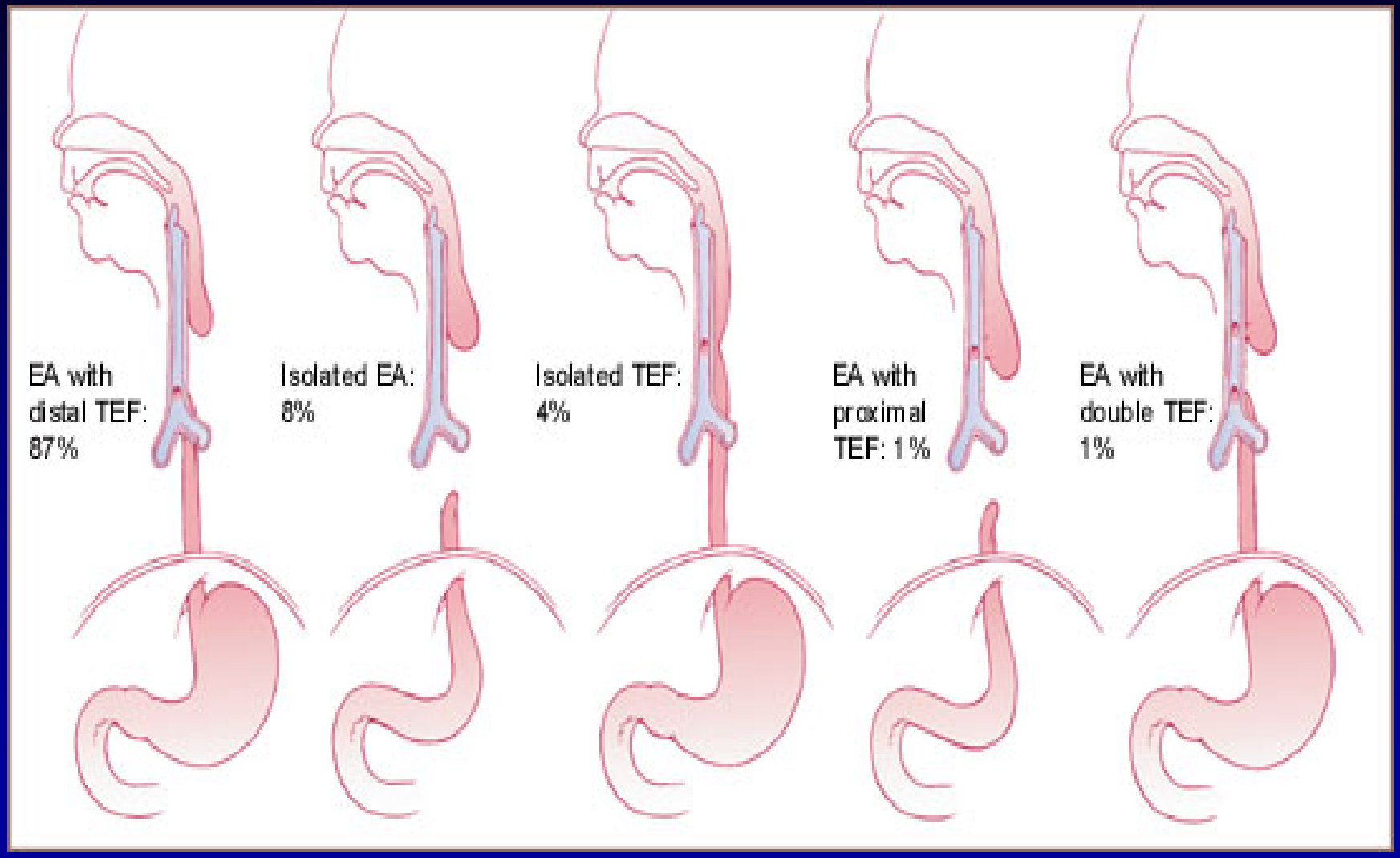
Pulmonary Artery Sling





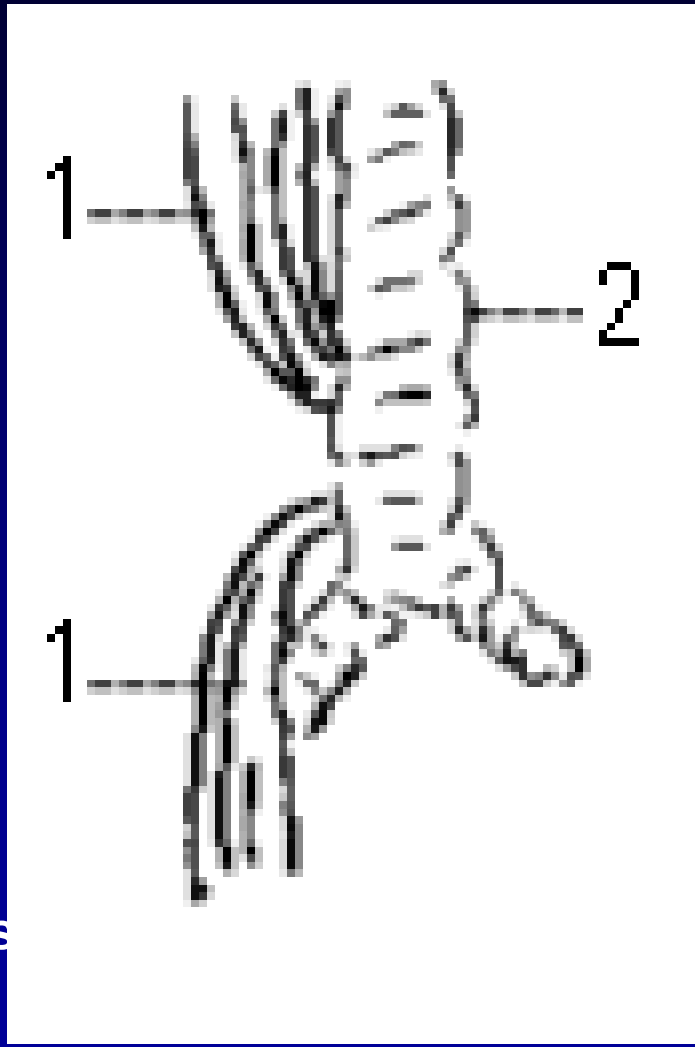


Tracheoesophageal fistula



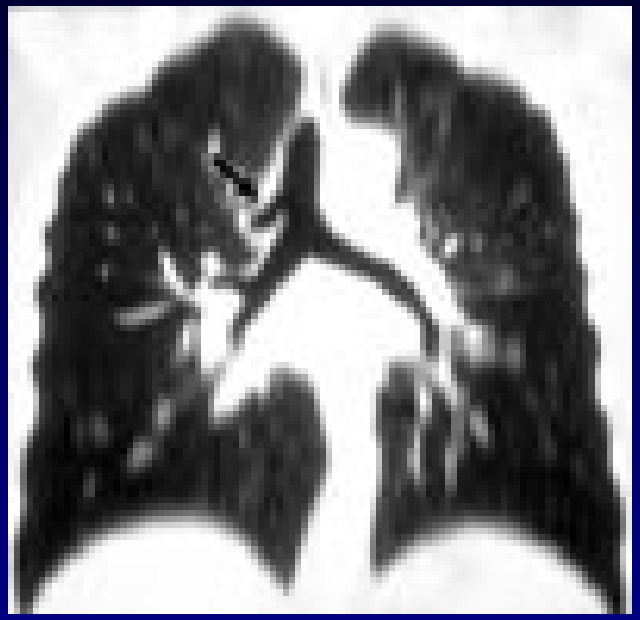
Tracheal esophageal fistulas

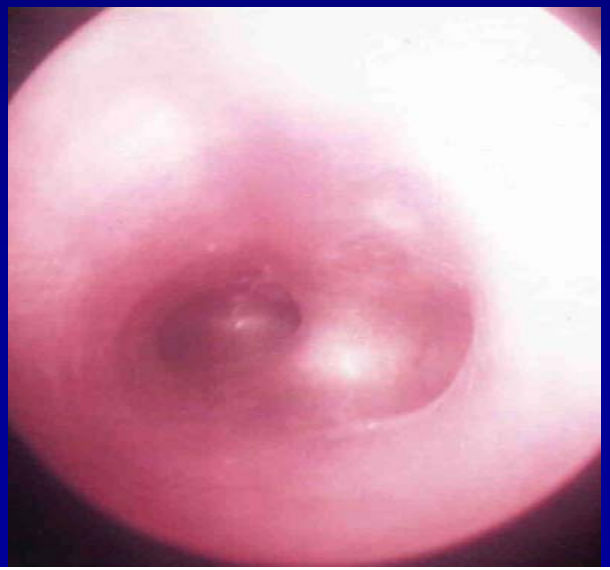
- 1st surgical repair in 1888 with esophageal atresia
- Up to 70% with associated anomalies
  - Down's syndrome
  - CV: VSD, PDA, Tetralogy, ASD, R sided arch
  - GI: dudonal atresia, imperforate anus, malrotation, Meckel's diverticulum, annular pancreas
  - GU: horseshoe kidney, polycystic kidneys,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



Tracheal bronchus (Pig bronchus)

- Congenital malformation of the tracheo-bronchial tree.
- Occurs superior to the bifurcation of the trachea - usually on the Right
- Frequently asymptomatic, but can present with recurrent infection (RUL), atelectasis or bronchiectasis.





School Age Children

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 with asthma at 4 years, increased difficulty over the last 2 years
- 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 5-7 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 12 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

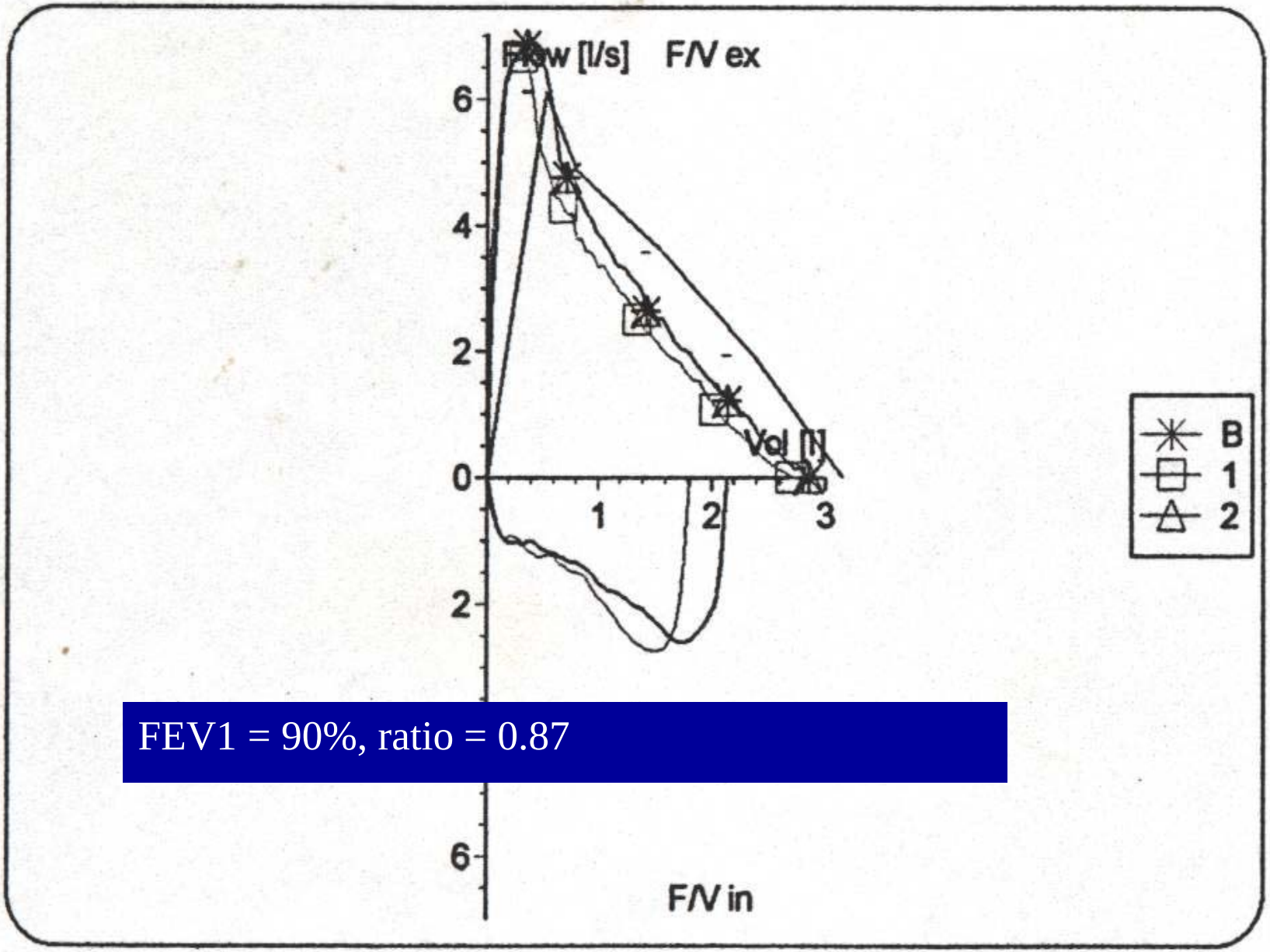
Abdomen: Obese with NABS, soft NTND without HSM/masses

Ext: No clubbing, cyanosis, and/or edema

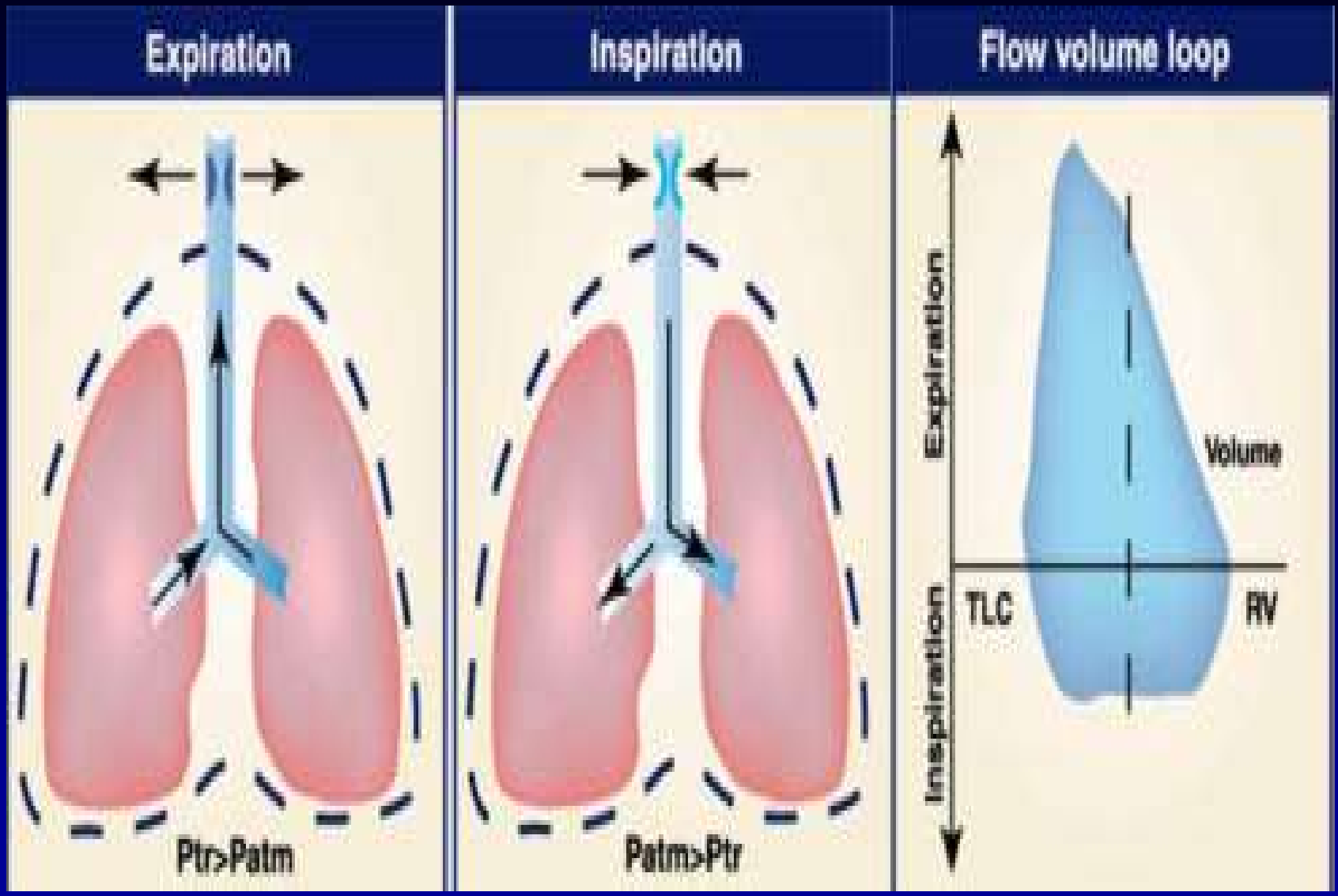
Skin: Acne-form lesions on face, no eczema

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



FEV1 = 90%, ratio = 0.87


Expiration
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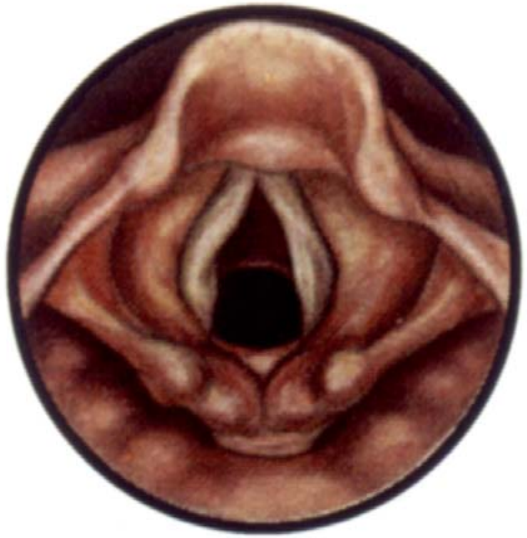
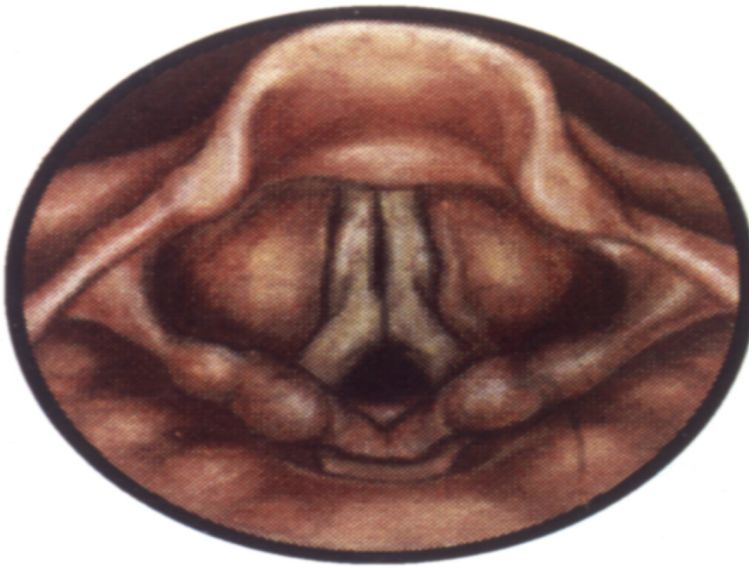
Inspiration
Volume
TLC
RV

Signs & Sx of VCD

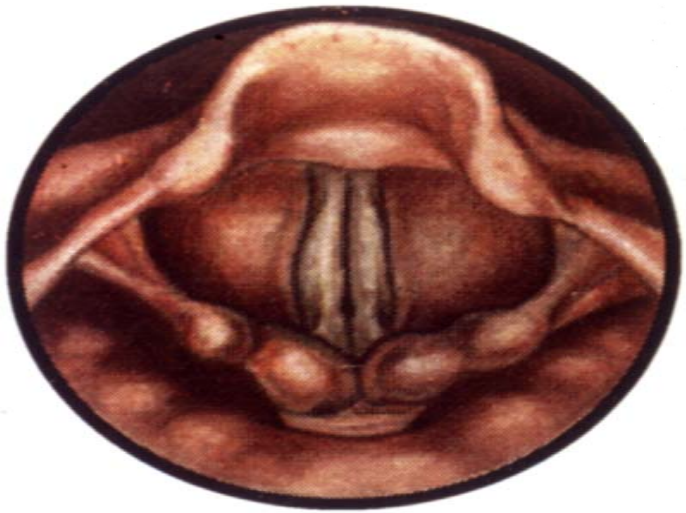
- SOB
- Stridor
- Chest and throat tightness
- Cough
- Wheezing
- Choking
- Dysphonia
- Hyperventilation sx
- “Can’t get enough air in”



Normal
Mid-Inspiratory (



Mid-Inspiratory
Vocal Cord Adduction
with Posterior Chinking



Closed
Mid-Inspiratory
Vocal Cord Adduction

Discharge:

Diagnoses:

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

Recommendations:

Omeprazole 20 mg BID
Fluticasone/Salmeterol 230/21 2 puffs BID
NSI/fluticasone nasal BIDCalcium/MVI qd

Other

VCD exercises

ACUTE VCD MANAGEMENT

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

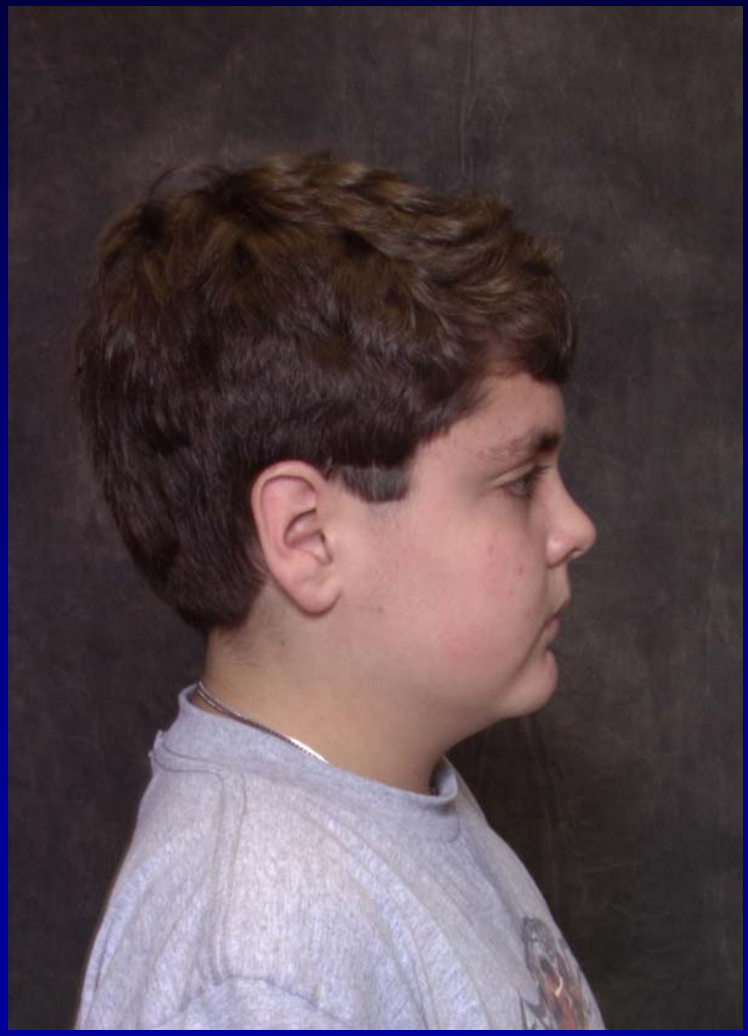
CHRONIC VCD MANAGEMENT

- 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 a clear nose)
- 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