Pulmonary Issues in Neuromuscular Diseases

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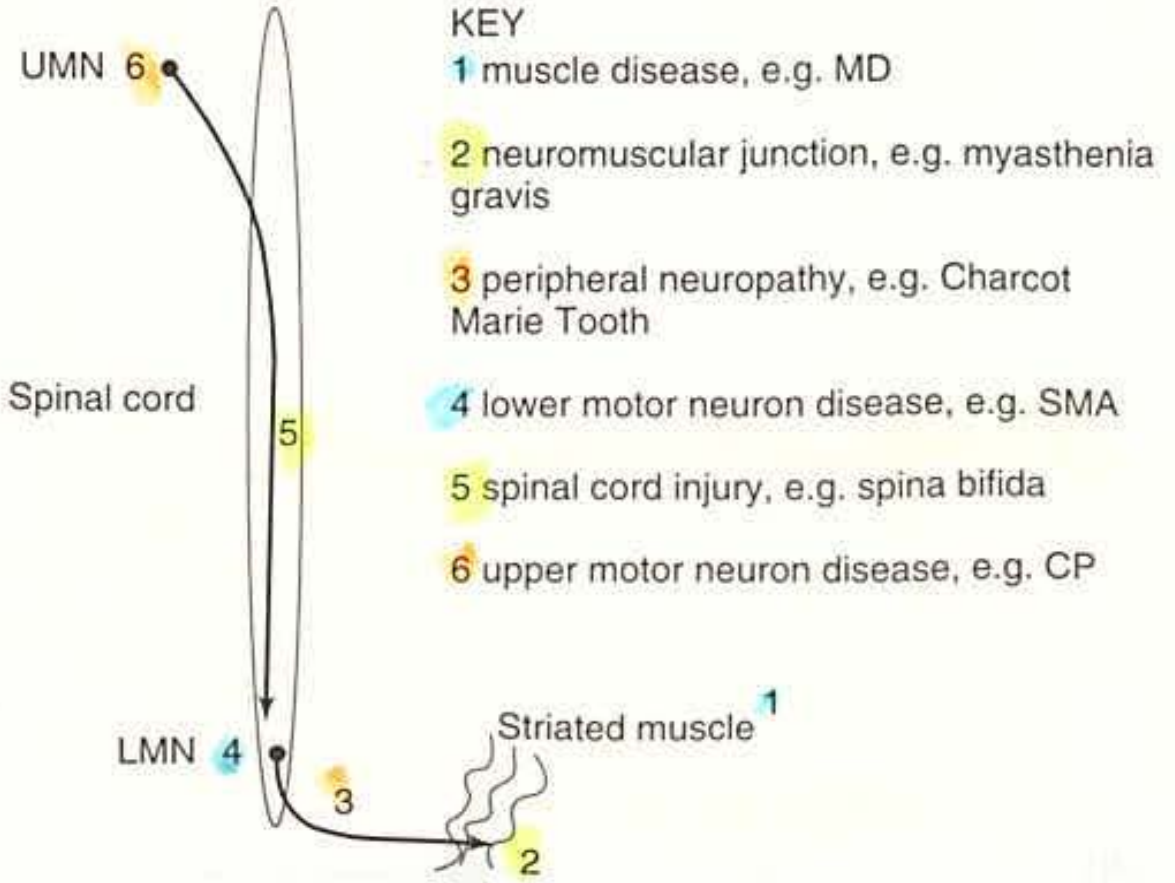
- Neuromuscular Disorder does ***NOT*** mean mental retardation .
- It may ***OR*** may ***NOT*** be associated with MR.
- It could be congenital or acquired.

Conflict of Interest?

- My life is full of conflicts
- None of which is interesting


UMN 6
Spinal cord
5
LMN 4
3
Striated muscle 1
2
KEY
1 muscle disease, e.g. MD
2 neuromuscular junction, e.g. myasthenia gravis
3 peripheral neuropathy, e.g. Charcot Marie Tooth
4 lower motor neuron disease, e.g. SMA
5 spinal cord injury, e.g. spina bifida
6 upper motor neuron disease, e.g. CP

Upper Motor Neuron

- Cerebral Palsy

Lower Motor Neuron

1. Spinal Motor Atrophy (SMA).
2. Peripheral Neuropathy (GBS, CMT).
3. NM Junction (Myasthenia Gravis).
4. Muscular Dystrophies.

Combined Upper and Lower

1. Spinal Cord Injuries.

2. Spina Bifida

Normal Respiratory Muscles Function

Diaphragm

- Acts as a piston that decreases intrapleural pressure.
- Normal motion: BOTH chest wall and abdominal expand “outside”.
- Patients with NMD do NOT report dyspnea until the diaphragm is involved.

Intercostal Muscles

- External: primarily inspiratory.
- Internal: Primarily expiratory.
- Normally: Inspiration is active and expiration is passive.

Abdominal muscles

- Contraction of RA and AO increases intra abdominal pressure and helps active expiration (exercise, Asthma--)

Upper Airway Muscles

- Contract with inspiration, preventing airway collapse.
- Healthy infants: “glottic breaking” is partial ADDUCTION of VC to maintain end exp. Volume .
- “Glottic breaking” is impaired with NMD, resulting in atelectasis

Swallow Function

- Oral phase: voluntary.
- Pharyngeal and esophageal: involuntary.
- Weakness: results in food going thru nose and trachea.

Cough

A. Starts with irritation of receptors of vagus nerve. Inspiration to near TLC.

B. Closure of glottis followed by contraction of abdominal muscle.

C. Glottis suddenly opens resulting in an upward movement in Diaphragm and expulsion of air @ 300 mi/hr.

Cough

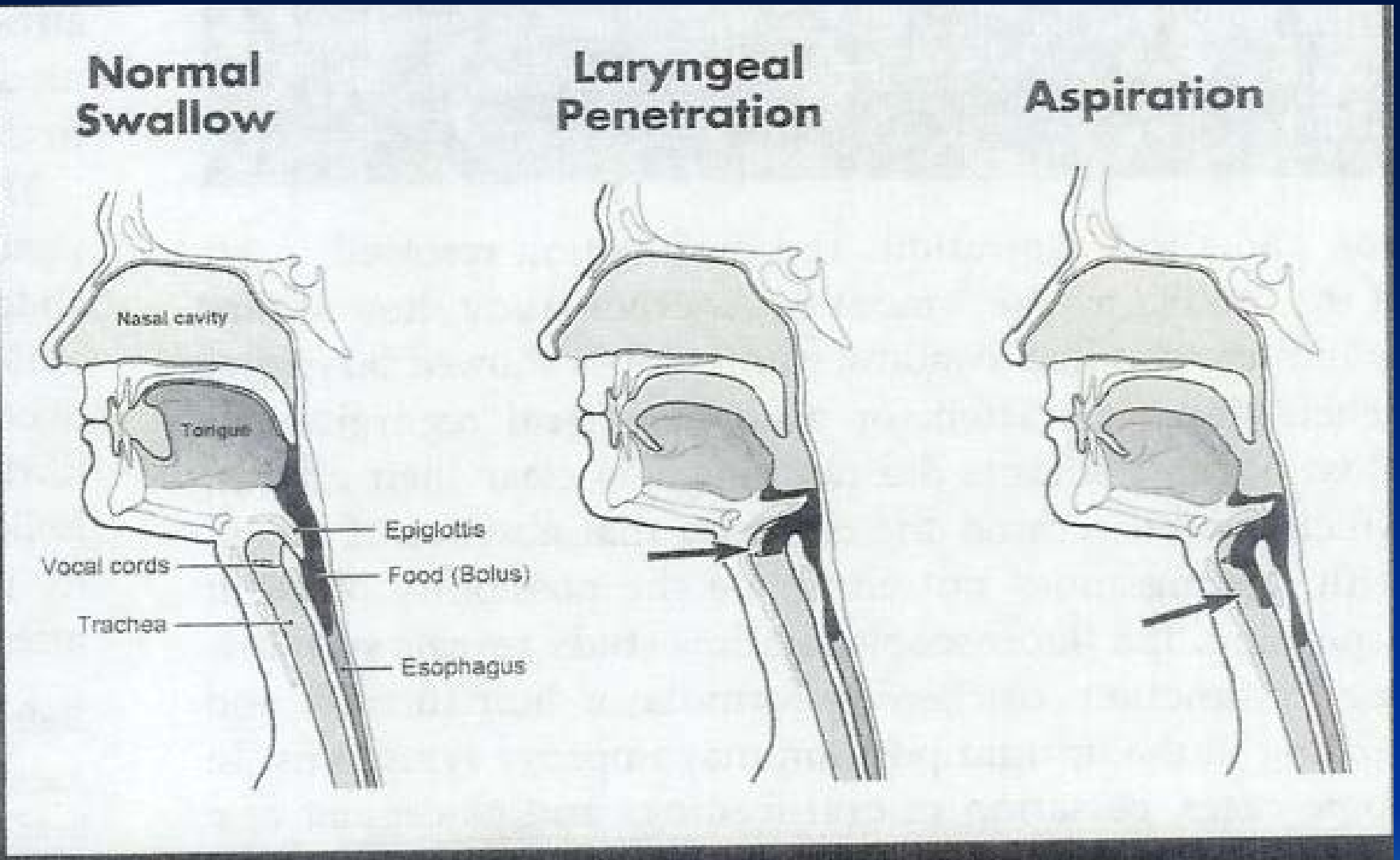
- Weakness of the abdominal muscles reduces the effectiveness of cough

SLOW Changes in NMD

- In the early stage little if any paranchymal lung disease is present.

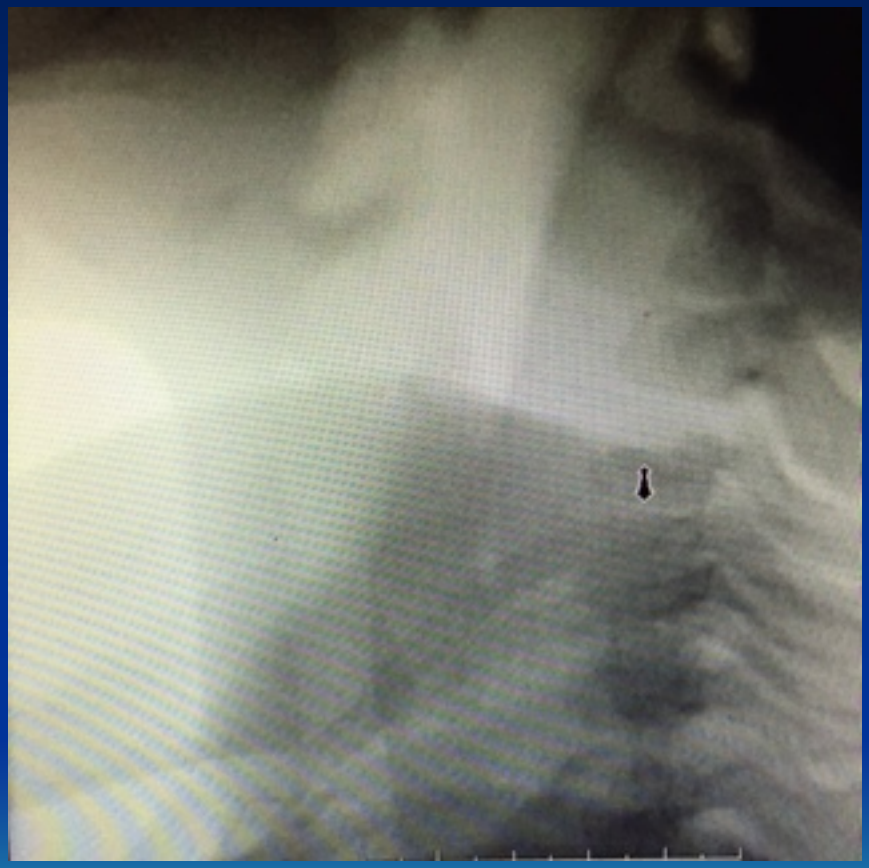
With **TIME**

- Weak muscles and often chest wall deformity lead to poor lung expansion and ultimately reduced lung growth.
- Weak VC and oropharyngeal muscles lead to recurrent aspiration pneumonias.
- Poor expansion and cough leads to recurrent atelectasis



Courtesy of A. Johnson

ITEM 164A: Normal and abnormal swallowing: In normal swallowing (left), the food bolus enters the esophagus. Laryngeal penetration occurs when food enters the airway above the vocal cords (center). In aspiration (right), food travels below the vocal cords into the trachea.



- Poor swallowing impairs nutrition and growth which worsen the muscle weakness and weakens the immunity.

- Progressive muscle weakness lead to worsen chest deformity and further decrease in lung volumes.

Pulmonary Consequences of NMD

1. Recurrent Pneumonia and aspirations.
2. Recurrent Bronchospasm.
3. Recurrent Atelectasis.
4. OSA.
5. Daytime sleepiness and fatigue.
6. Drooling, difficulty swallowing.
7. Nasal regurgitation.
8. Decreased lung volume.
9. Death from aspiration pneumonia is the most common cause of death.

- All leads to **Restrictive Lung Disease**.
- Often with **Obstructive** component as well

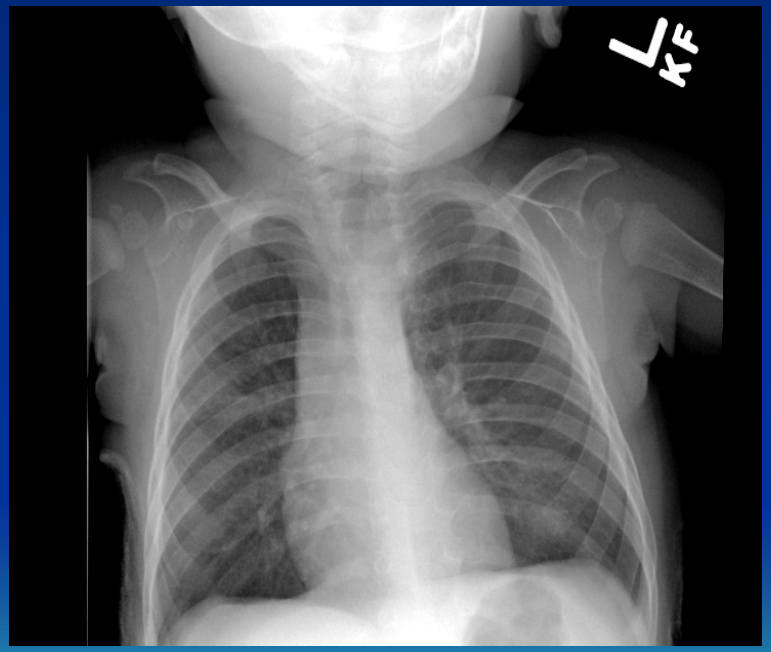
The GOAL

- Delay/minimize the progression of lung disease.
- Prevent/Treat complications such as Asthma, pneumonia, OSA, --
- Improve the quality of life.

- Lung disease is a “secondary complication/natural progression” of NMD.
- Little if any lung disease is present in the early stages.

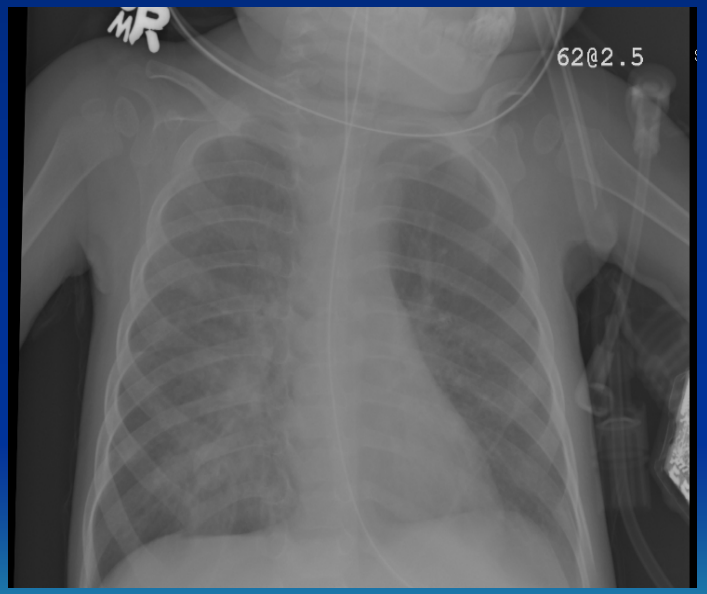
Sept/2009

- 7 mos. with SMA-I
- IMPRESSION:
  Negative chest x-ray.

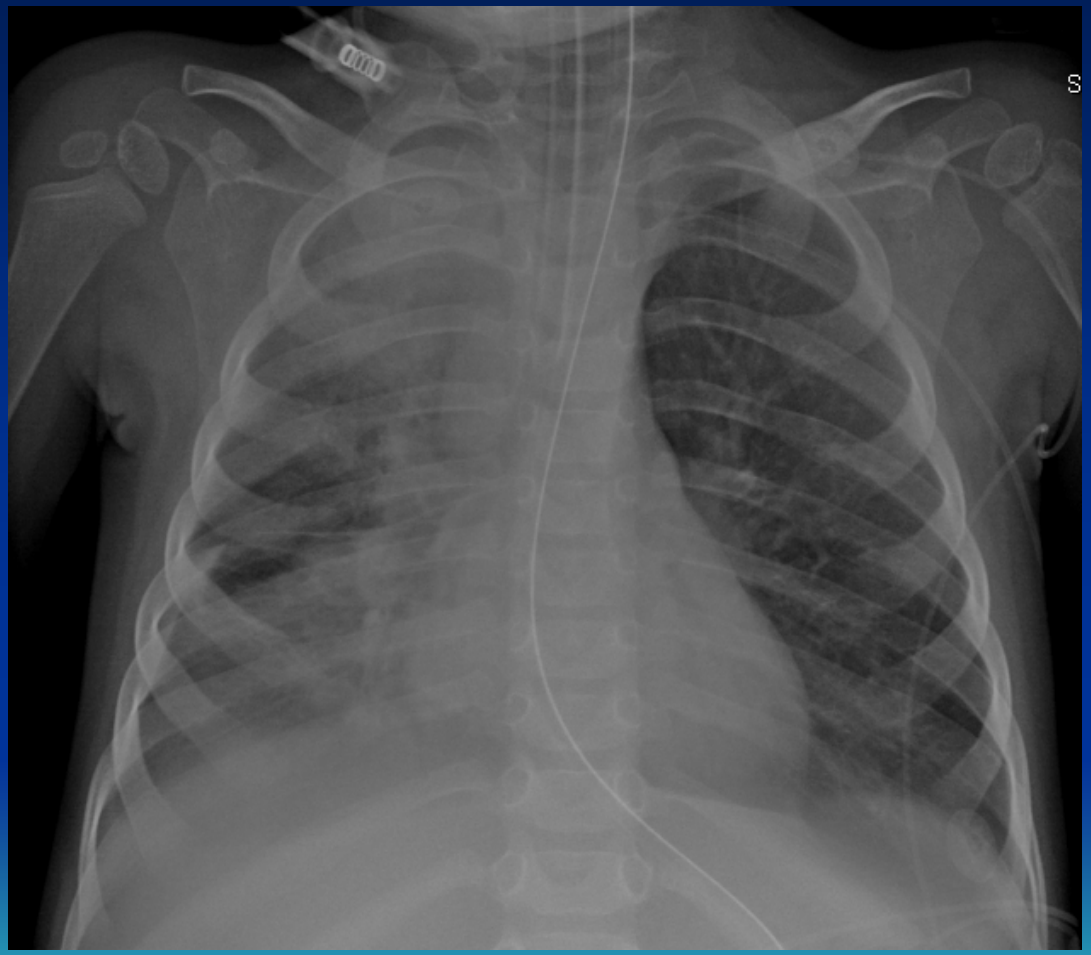


Jan/2010

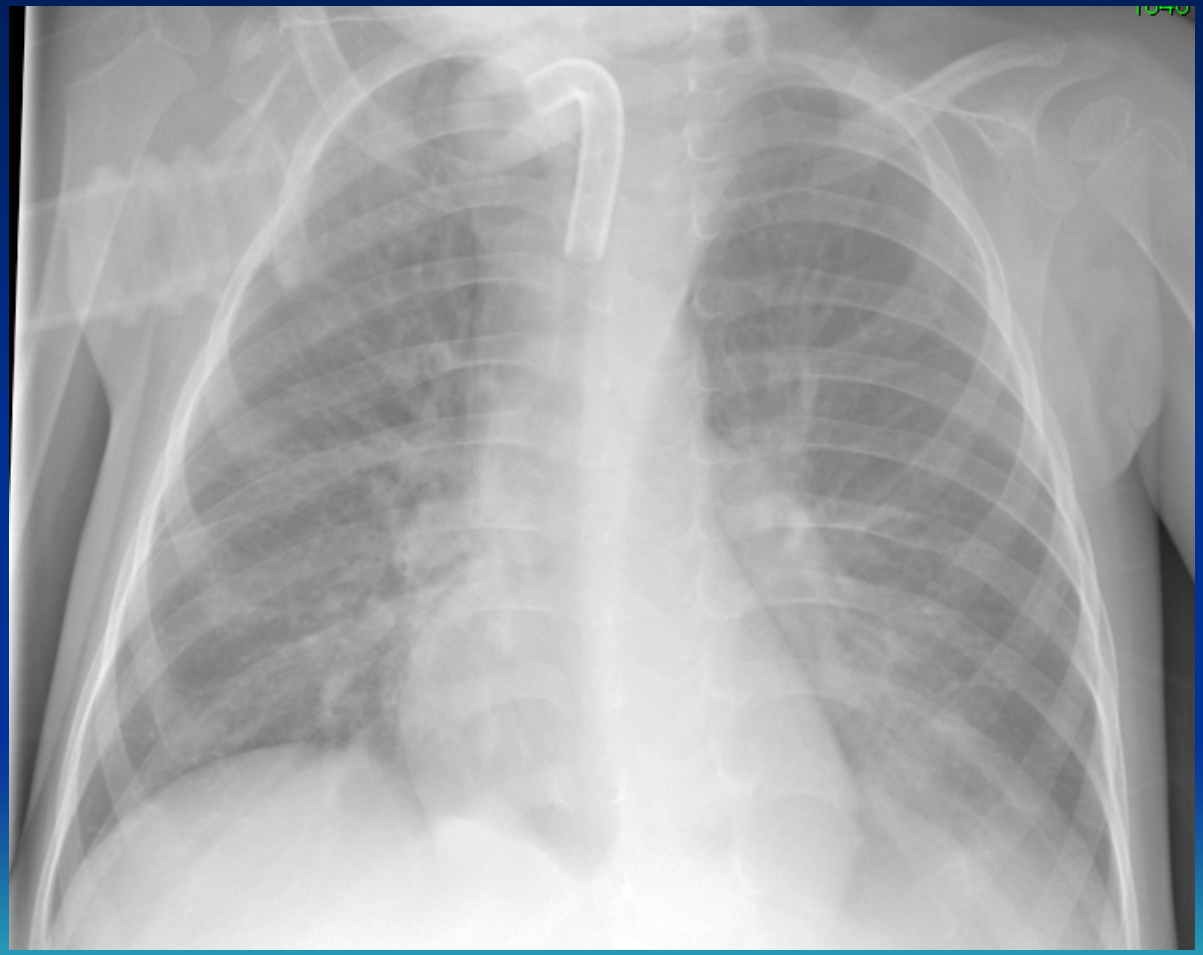
- Same pt with fever
  DIB



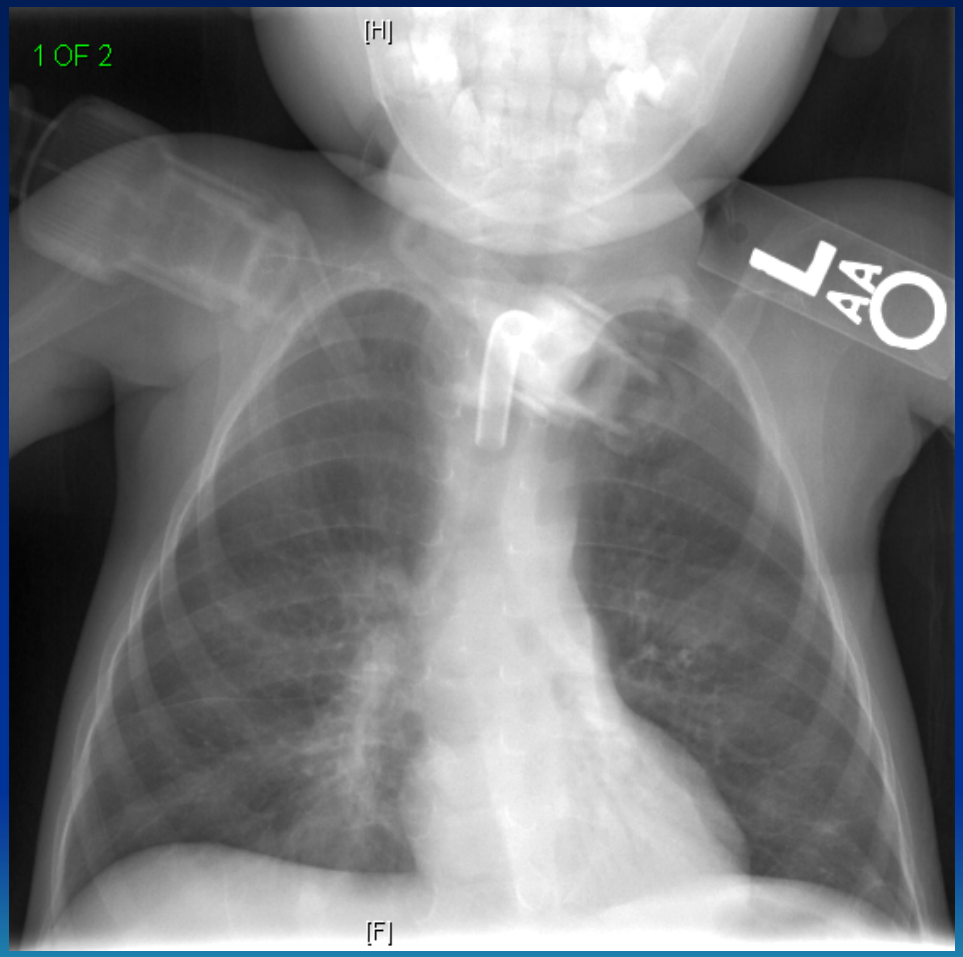
March/2010



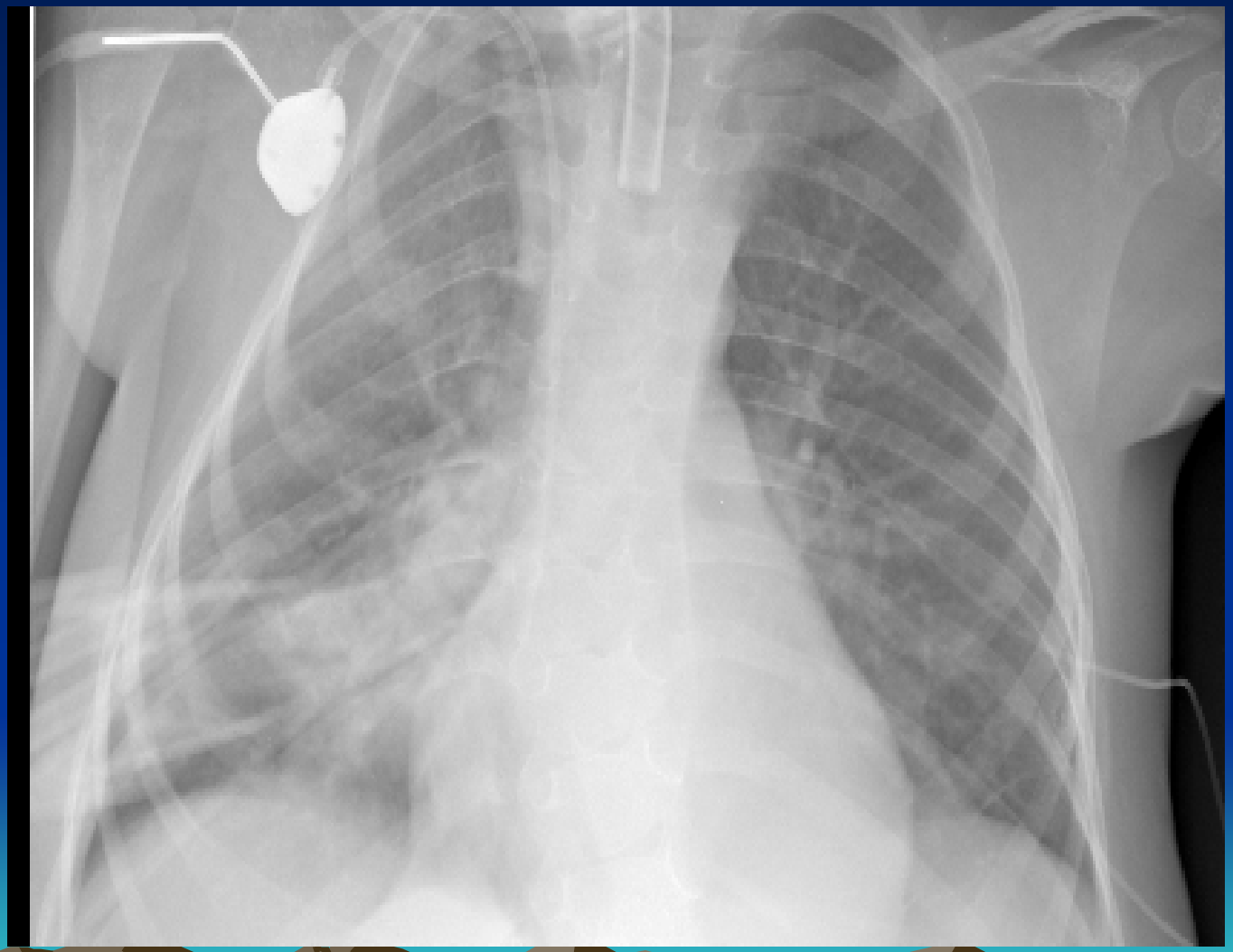
Aug/2010



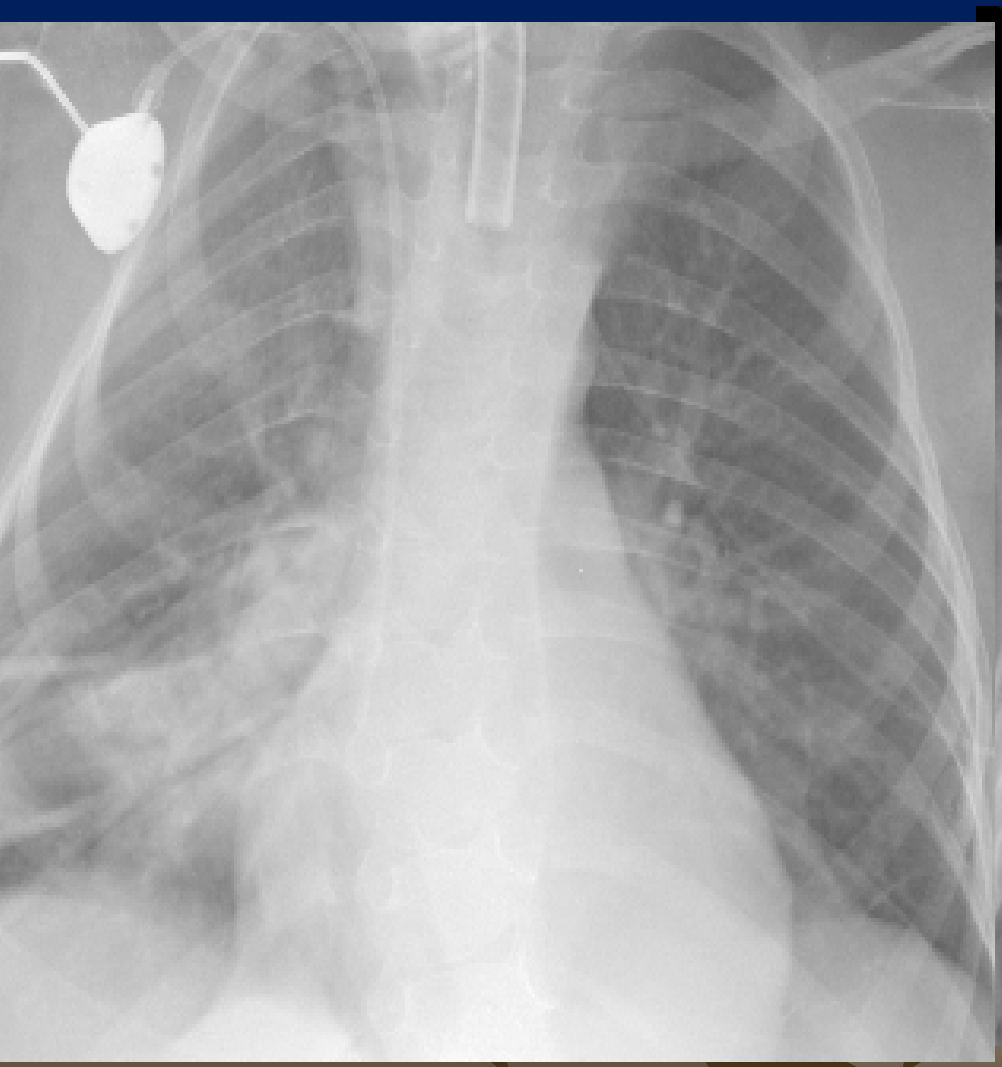
Dec/2010

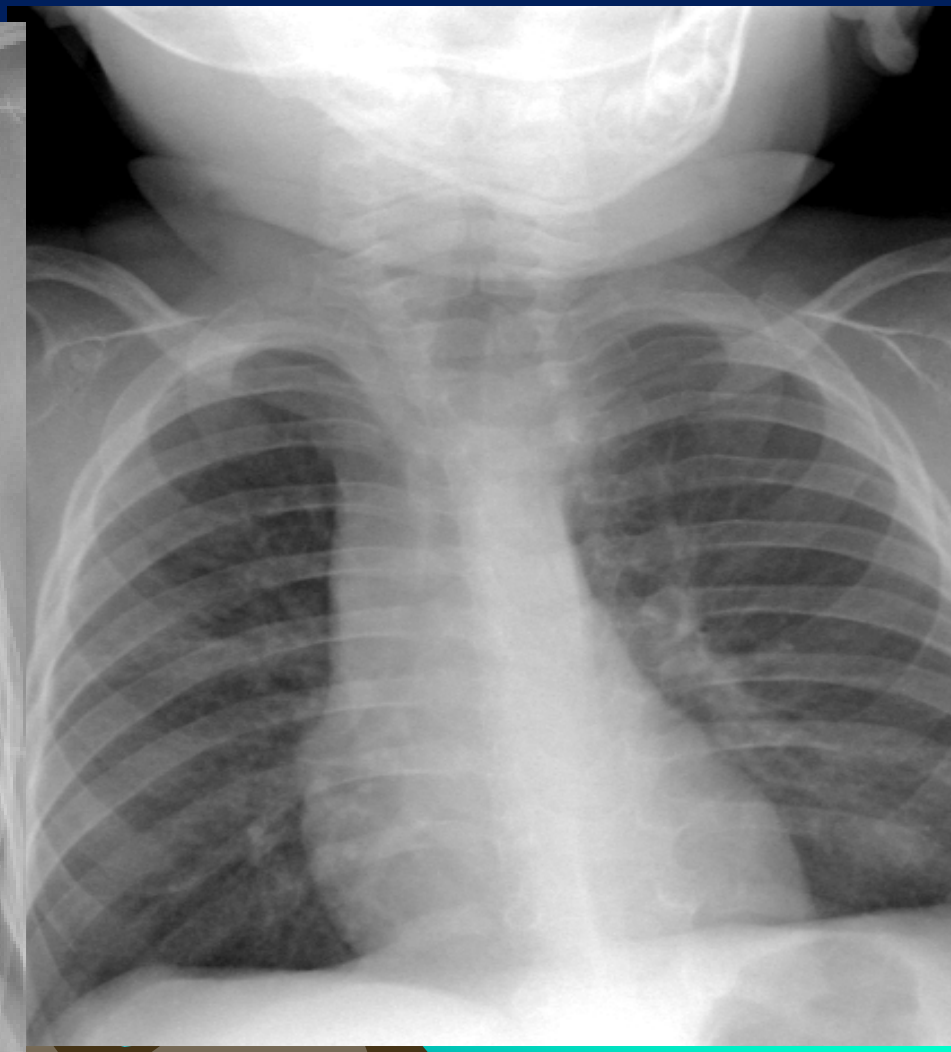


Dec/2011



Goal





Multi Specialty Approach Neurology and

- GI: Swallowing and feeding evaluation, nutrition
- Orthopedics: Scoliosis/Kyphoscoliosis evaluation and repair.
- ENT: OSA, Tracheotomy care.
- PM&R: Exercise, Botox,--
- Pulmonary: ACT, Trach. and Ventilator care.
- Palliative care: **SHOULD BE DISCUSSED EARLY AND BEFORE COMPLICATIONS OCCUR**.

Approach to NMD History

- Multiple pneumonia?
- Drooling, aspiration, difficulty in swallowing, time needed to feed?
- Weak cry?
- Vomiting, GERD?
- Sleep history; snoring, day time sleep--?

Physical Examination

- Scoliosis?
- Digital clubbing?
- Cyanosis?
- Gag reflux?

Digital Clubbing

Diagnostic work Up

1/ Sat O2.

2/ CO2 via Capnography or CBGs.

3/ FVC, MIP, MEP (normally about -100 and +200)

4/ Sleep assessment, PSG.

5/ Annual CXR .

Primary Care Visits

Same as per AAP plus

Flu vaccine

Pneumococcal Vaccine

Pulmonary F/U

Routine visit is recommended every 3-9 months depends on severity

Pulmonary Management

- Airway Clearance.
- OSA, Assisted Ventilation.
- Treatment of Acute exacerbations.
- Pre-operative evaluation.

Airway Clearance:

1. Essential to prevent PNA, atelectasis, and progressive lung disease

2. Drooling and ineffective cough is a major cause of morbidity and mortality.

3. In cooperative patient, this could be assessed by measuring cough peak flow.

Airway Clearance:

1. **CPF< 160** L/min is associated with poor airway clearance.

2. **Normal CPF : 147 to 488 L/min in females and from 162 to 728** L/min in males, age range of 4-18 yrs.

3. **MEP < 45 cm H2O** is also associated with poor airway clearance (normal value is 150-230 cm H2O)

Airway Clearance

1. Manual: CPT with position changes.

2. Vest: uncomfortable due to chest wall deformity and poor cough.

3. Cough assist devices.

4. Medications: DNAse, 3% saline, Robinol

Mucomyst

NAC is ***NO LONGER*** used for ACT

CAD

- Provides a (+30) positive pressure breath.
- Followed by (– 30) negative pressure, air is “sucked out” of the chest.

CAD

VEST

Vest

Is more of “ secretions mobiliser’ and best use for CF

Assisted Ventilation

- **Night time:** if a PSG shows significant OSA, Hypoxemia, increased A/HA index
- **Continuous:** if Sat O2 <92%, PaCO2 is > 55. while awake.
- **BiPAP** for advanced disease or if CPAP fails, newer recommendations of trying BiPAP 1st

CPAP/BiPAP

- Applied thru a face mask or nasal prongs.
- "**Effectiveness**" is assessed by number of OSA, DeSats, A/HA index on PSG, and by changes in morning PCO2.
- In severe cases, Tracheostomy and home ventilator is needed

Acute Exacerbations

- Cough.
- Increased secretions.
- Fever.
- Increased WOB.

????

- Is it a PNA/ Trachiatis??

Work Up

- CXR.
- CBGs.
- CRP.
- Respiratory C/S

CBCs

- WBC increased?
- Bands?
- Anemia ?

CBGs

- Always compare with a previous values.
- Many patients with NMD may have a CO2 retention (PCO2 46-55).
- PH, Bicarb normal or abnormal

Respiratory Culture

Are there any WBCs ?

Most Trached patient are colonized with Staph or Ps.A.

The presence of WBCs on gram stain suggest acute infection

CRP

- Early, non-specific acute phase reactant.
- CRP> 20 “suggests” bacterial pneumonia.

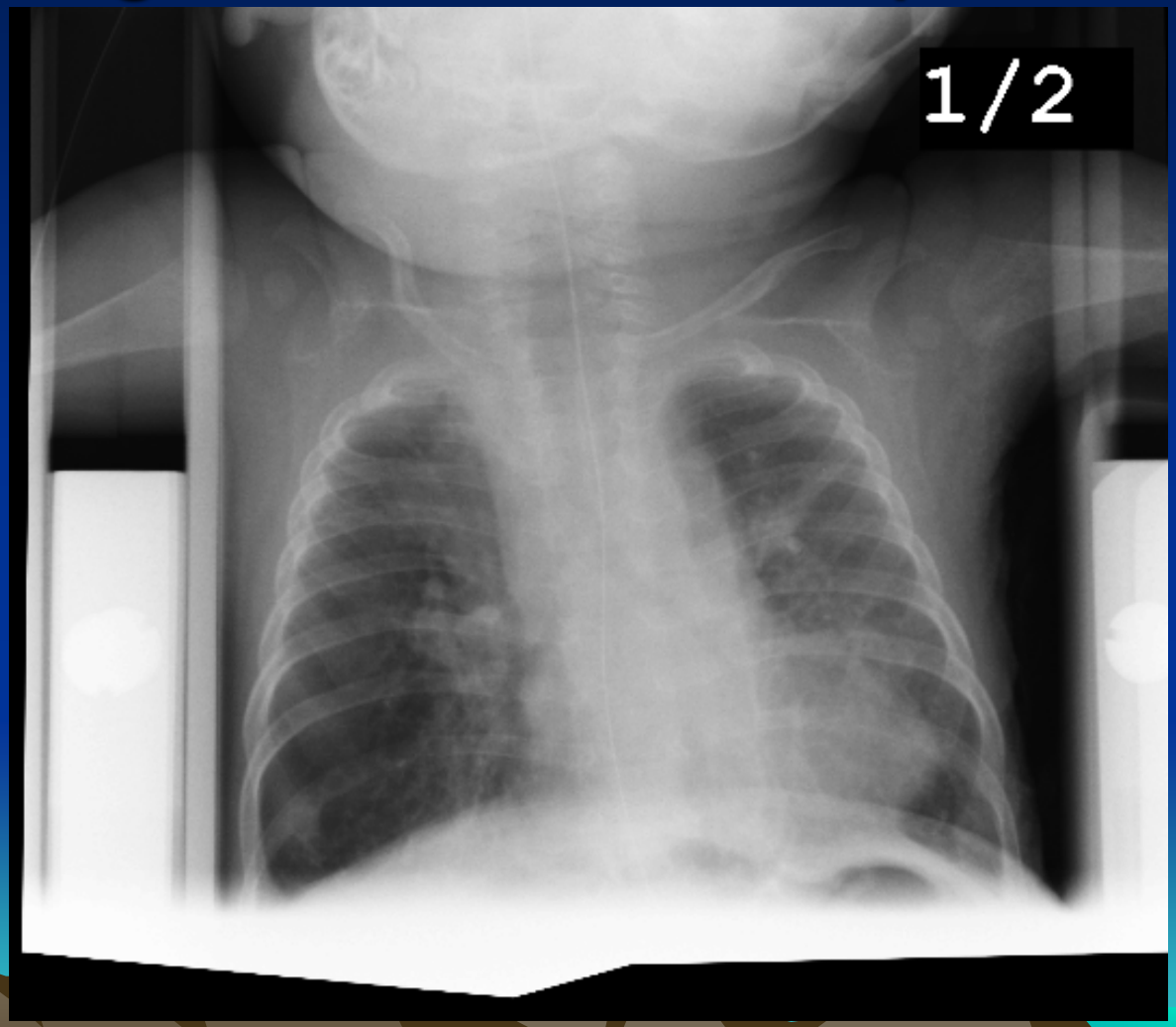
CXR

MUST be compared with previous ones

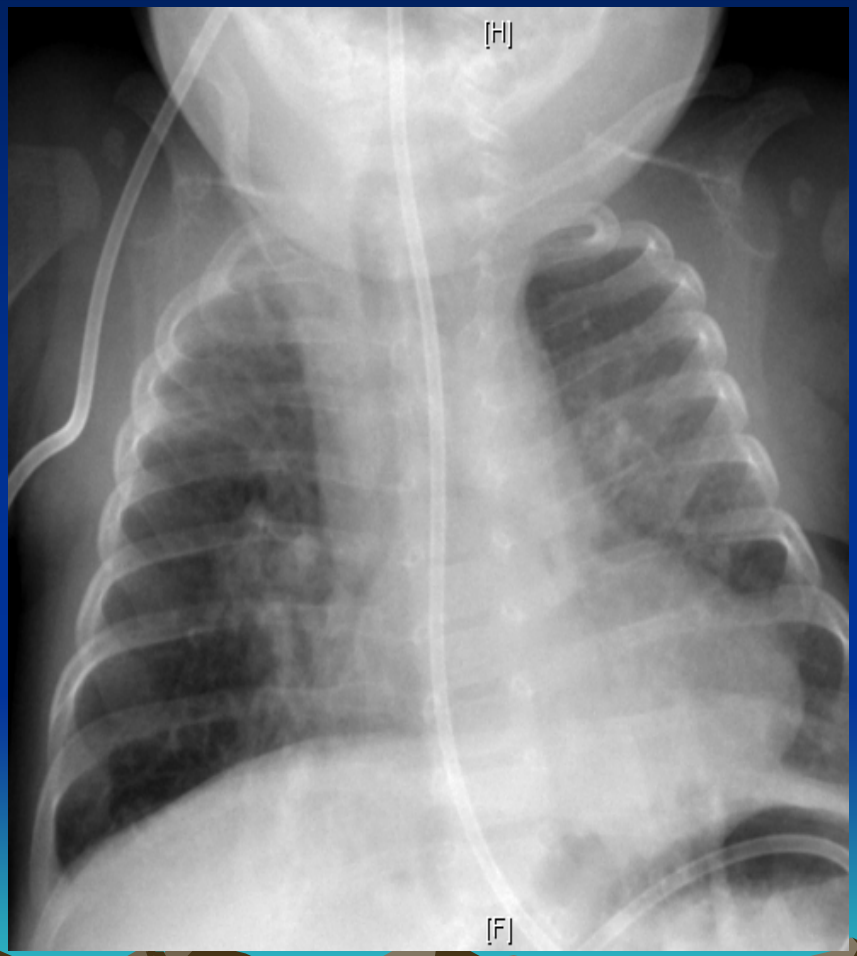
Antibiotics pending C/S

- Cover MRSA, Anaerobes, Pseudomonas
- Clindamycin, Cefepime,Tobramycine, Cipro
- If resp/tracheal c/s shows little or no PMNs, and CRP is < 15 likely viral and may not need abx

2 yrs. With MD comes with 2 days cough, ? Tactile temp.11/01/11



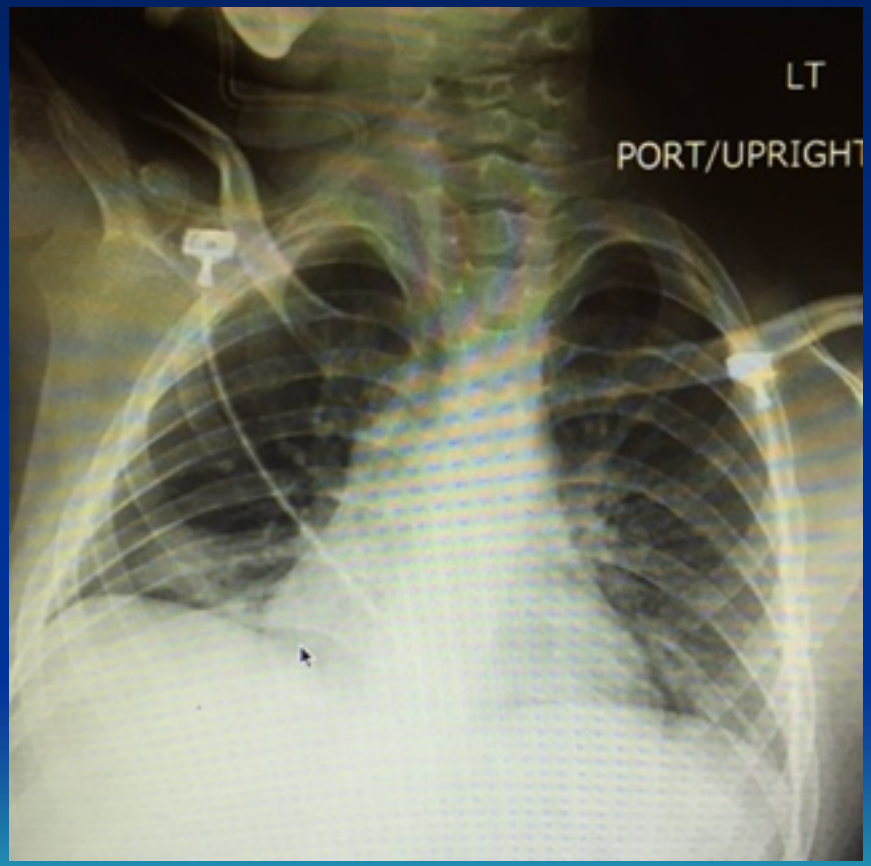
CXR 9/26/2011



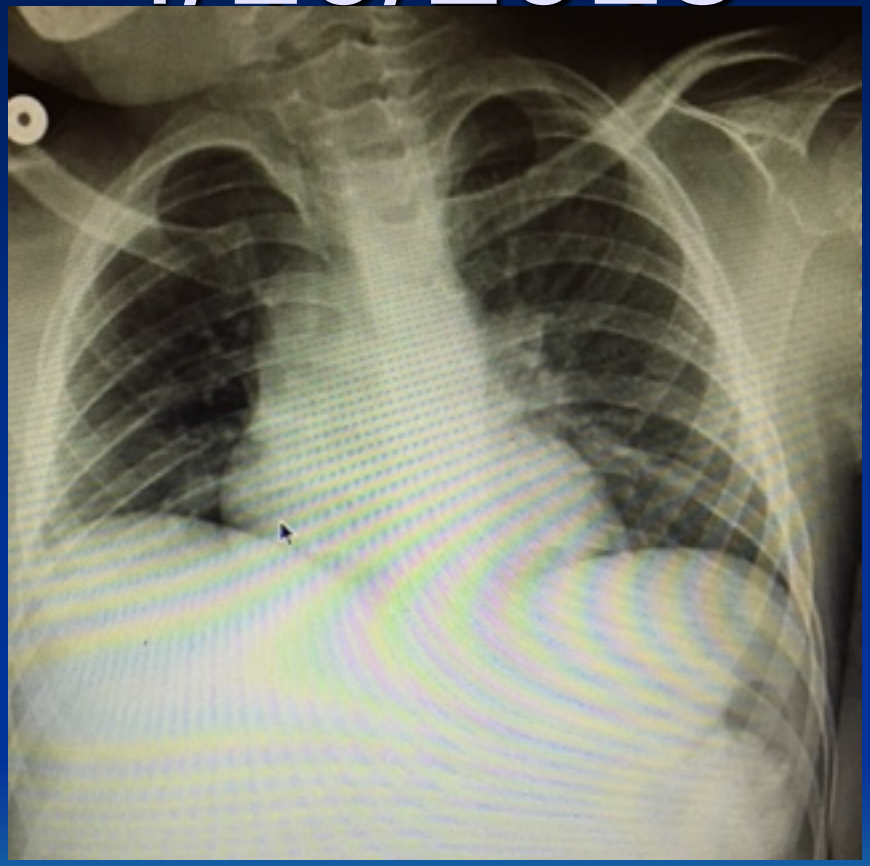
- NO changes, Normal CBC, CRP< 4.

- Viral illness. No Abx given

9 yrs with CP cough, DIB 4/09/15



4/10/2015



Note the resolution of RLL density in 24 hrs

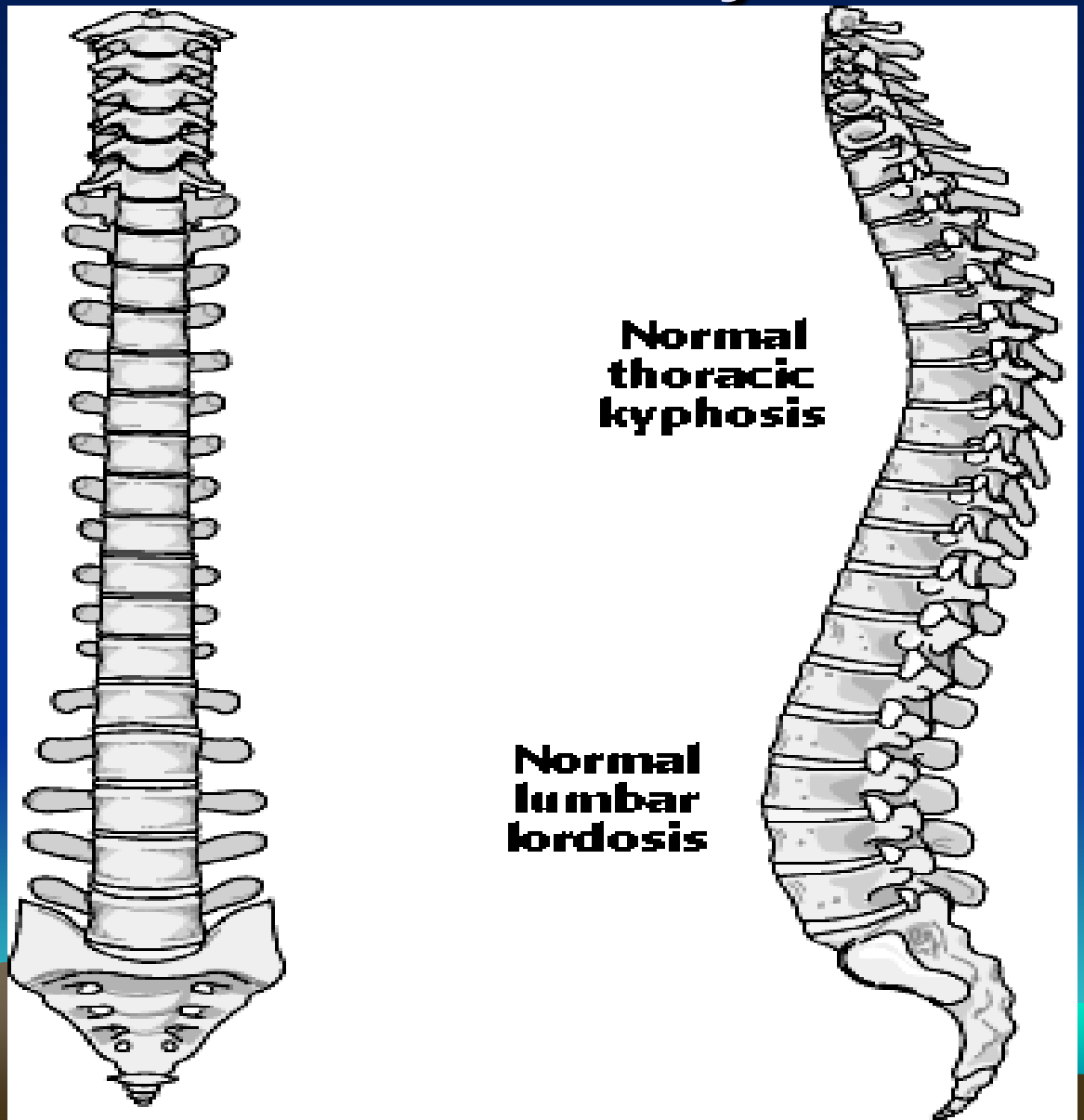
When to discharge ?

- Better clinically
- FiO_2 back to home dose (usually < 0.4)
- $PaCO_2 < 55$

Scoliosis

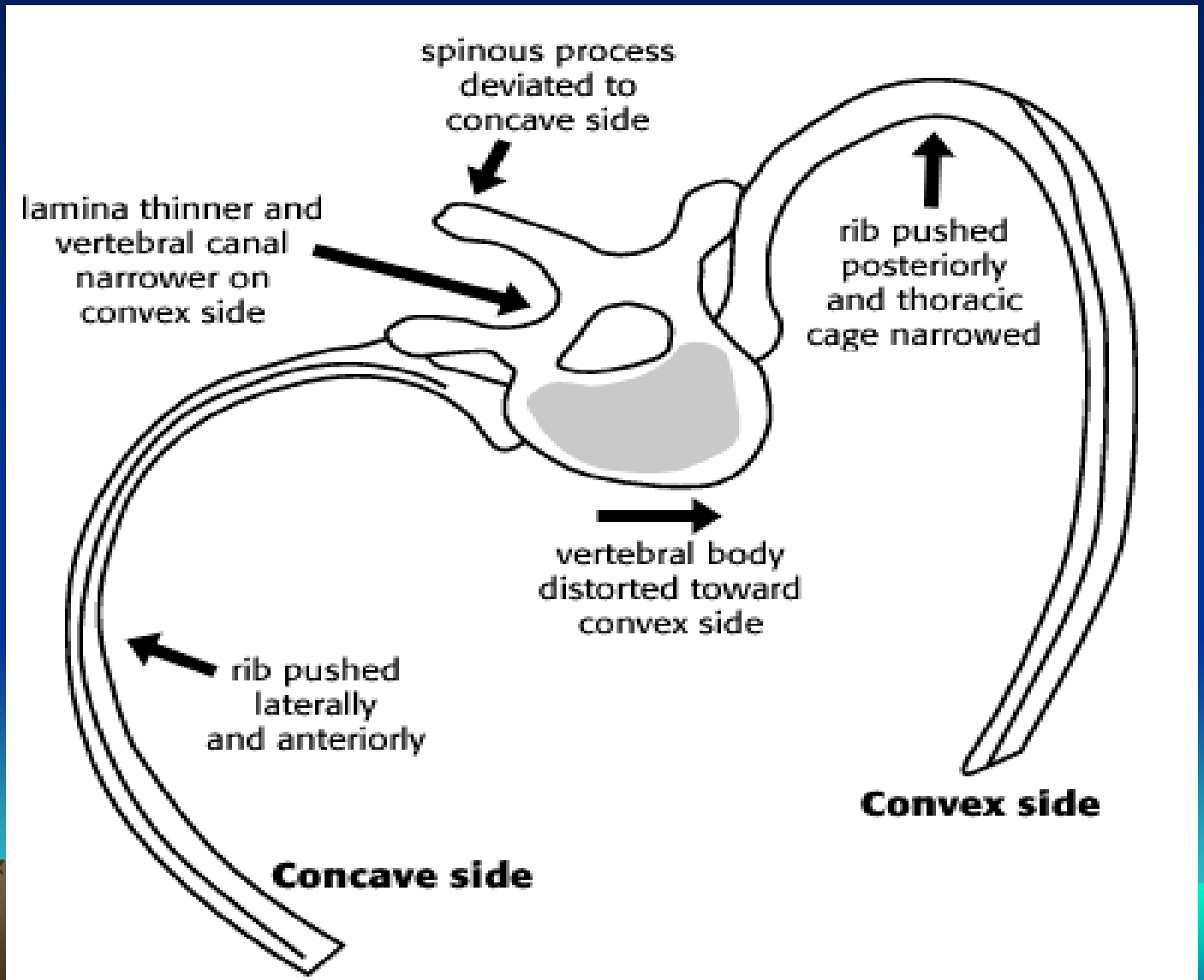
- Almost **ALL** patients with have chest wall deformity of some degree.
- Serial exams (including CXR, PFT)is recommended.
- Orthopedic evaluation is recommended.

Normally

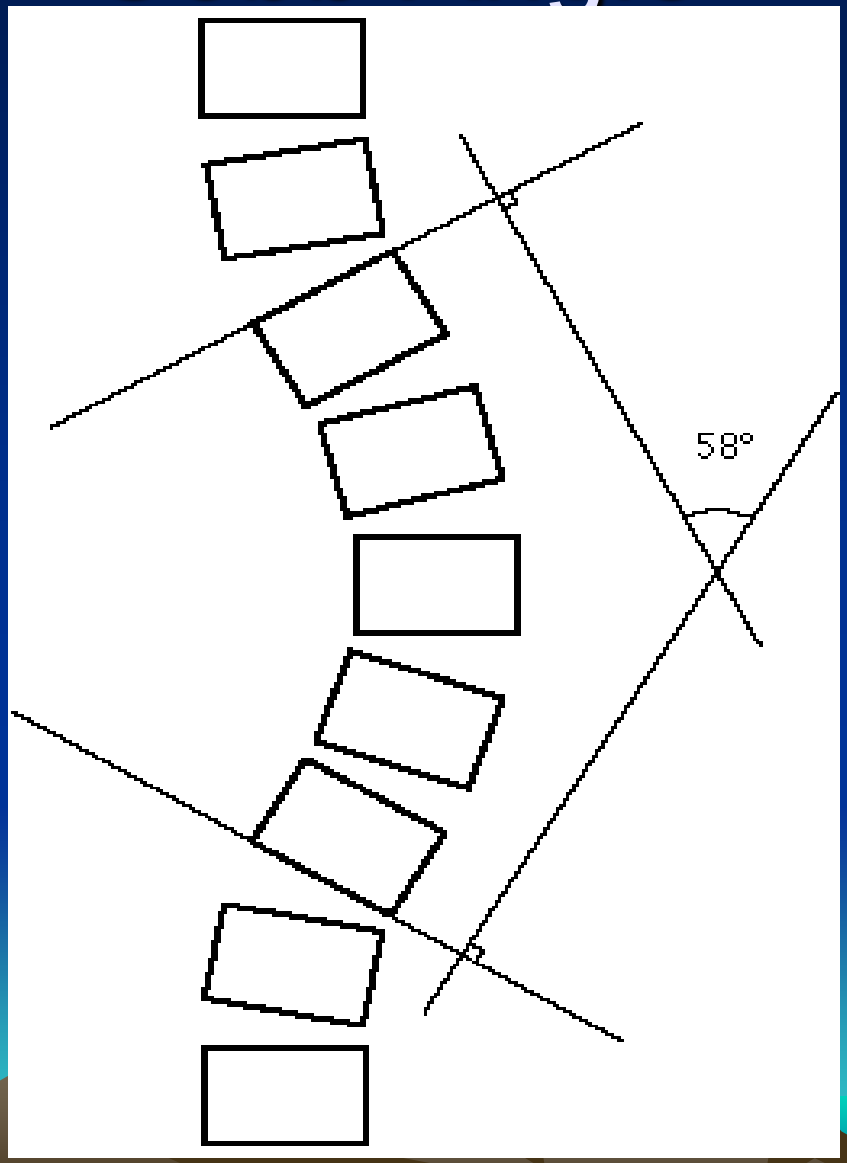


- Normally there should be no lateral curvature of the spine.
- The normal thoracolumbar spine is relatively straight in the sagittal plane and has a double curve in the coronal plane.
- The thoracic spine in convex posteriorly (kyphosis).
- The lumbar spine is convex anteriorly (lordosis).

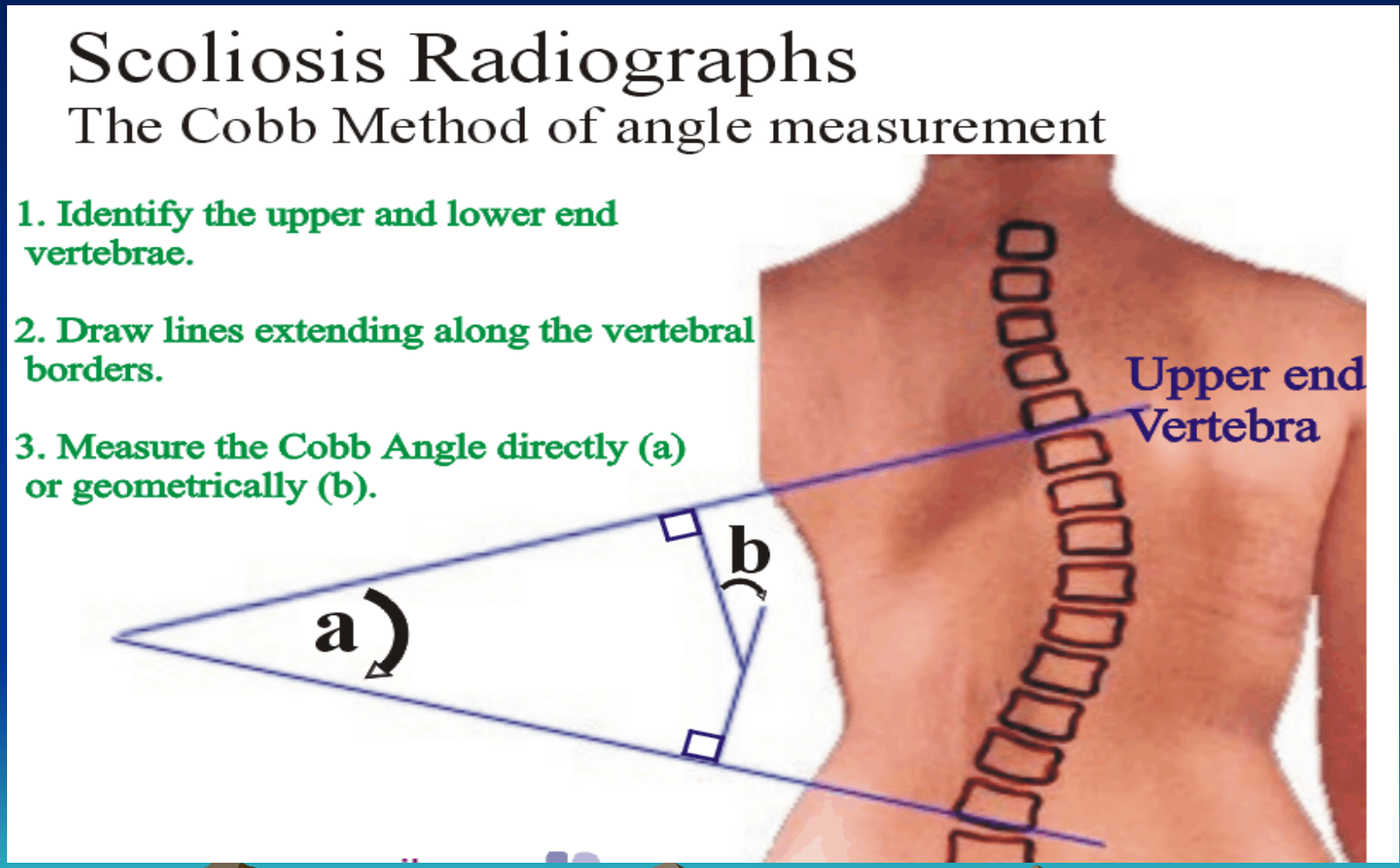
Changes in Scoliosis

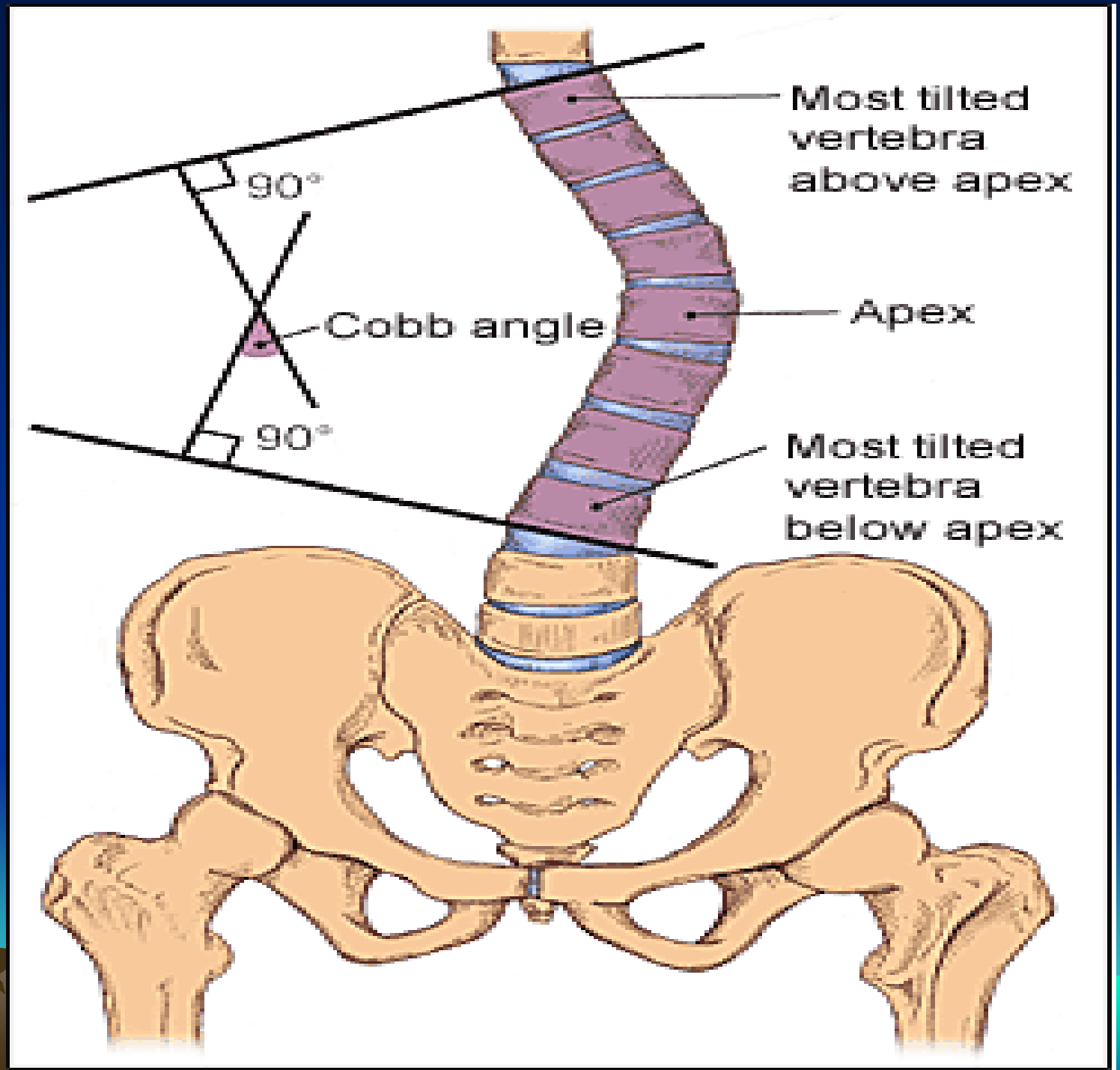


Cobb Angle



Cobb Angle


Scoliosis Radiographs
The Cobb Method of angle measurement
1. Identify the upper and lower end vertebrae.
2. Draw lines extending along the vertebral borders.
3. Measure the Cobb Angle directly (a) or geometrically (b).
a
b
Upper end Vertebra


Most tilted vertebra above apex
90°
Cobb angle
Apex
90°
Most tilted vertebra below apex

Cobb angle

When to repair scoliosis

- Cobb's angle 30-50.
- FVC >40 %.
- Cardiac evaluation (DMD, Pulmonary HTN)
- Sleep evaluation.

Evaluation for Sedation and GA

- Airway: Hypotonia, drooling, gag, loose teeth, small jaw, tongue??
- Lungs: SatO2, CO2, scoliosis, pneumonia.
- Heart: PHT, arrhythmia, CHF??

Evaluation for Sedation and GA

- PFTs: MIP, MEP, FVC (a value < 50% is associated with increased risk, and <30% is suggest the need for preoperative preparation for BiPAP /CPAP

- Sat.O2, CO2, CXR.

Evaluation for Sedation and GA

- MUST explain to the parents that in severe cases pts. may be unable to get off the ventilator in the presence of severe lung disease.

- This should be discussed **BEFORE** sedation/GA.

GI and Nutrition

- Most have poor swallowing.
- GERD is often present.
- Above problems often results in a poor nutrition.
- CLD and chronic increased WOB leads to increased caloric requirments.

GI and Nutrition

- G-tube insertion is the end result to ensure sufficient nutrition.
- High caloric formulas are usually used.

Thank you

References

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