HRCT IN PEDIATRIC DIFFUSE LUNG DISEASE

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OBJECTIVES

- How I deal with a HRCT request in a child?
- Technical challenges- getting better images
- How to formulate a useful report
- Terminology
- Latest classification of ILD in children & infants

HRCT

- Check Chest radiograph(s) and prior CTs
- Low dose technique
- Sampling diffuse or widespread abnormalities
- Concentrates on pulmonary parenchyma
- Does NOT evaluate large airways or mediastinum
- Does NOT help if conventional CT is normal
- Most cases- thin sections from multislice low dose acquisition suffice

Motion Artifact on High-Resolution CT Images of Pediatric Patients: Comparison of Volumetric and Axial CT Methods- Bastos M, Lee EY et al – AJR November 2009, Vol 193, Number 5

Technical aspects

- Weight/size/age-based dosing
- 80-120 kVp
- 1 mm slices
- 7 mm to 20 mm intervals
- Fast acquisition
- 6-8 slices in inspiration
- Minimum of 4 expiratory slices

Dose

- Compare CT scanners for relative dose
- Use breast shields
- Scan limited area if possible
- Increased sharpness (but increased noise)

Prepare the patient

- Patient cooperation or control is key
- Inspiration in 4 to 6 year olds
- Expiration 6 to 8 year olds
- Child-friendly scanner
- Technologist/Radiologist/play therapist in room to coach until 10-12 years old

Patient Preparation

- Explain before entering the room
- Repeat and practice again on the table
- Speak to patient during the procedure
- Helps to have parent in room during scan

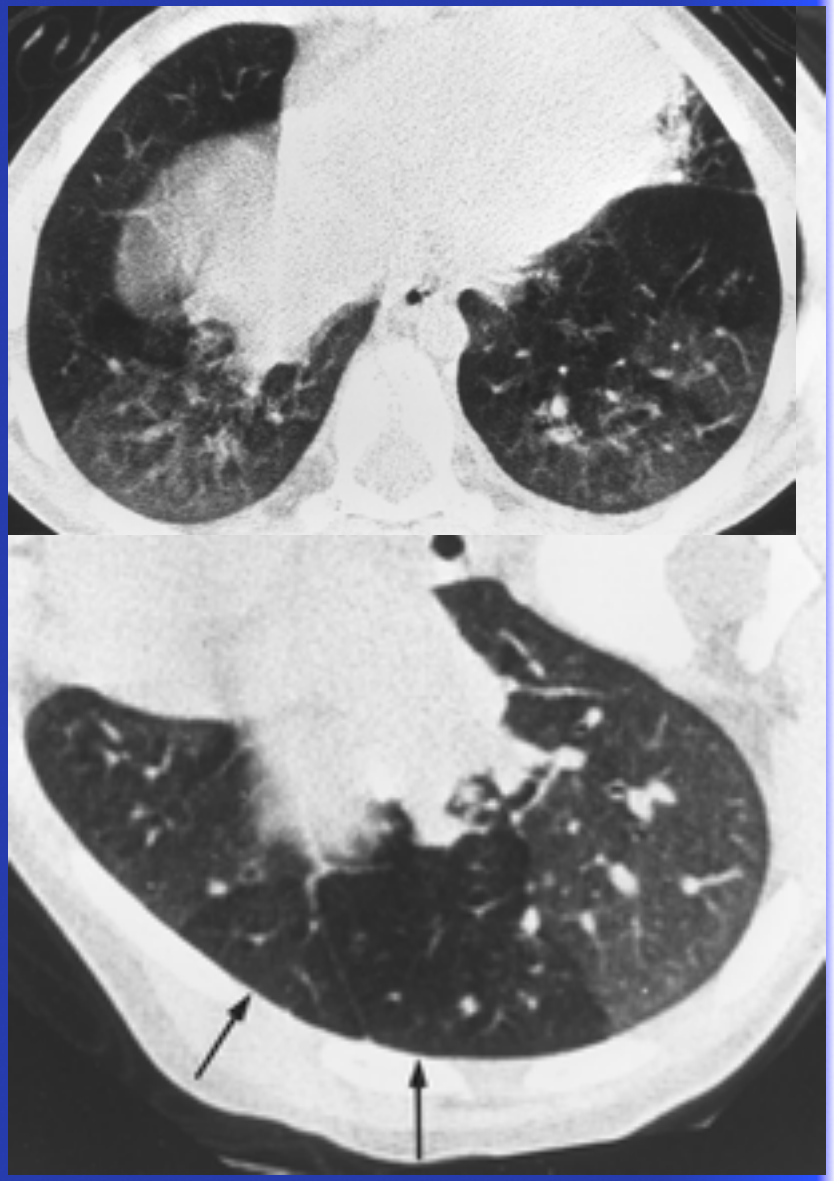
Younger children

- Quiet breathing is not adequate for HRCT
- Motion degrades images
- Consider decubitus imaging
- Controlled ventilation CT
- Intubation cannot be avoided in the young child

Decubitus imaging

- Perform initial HRCT
- Lateral decubitus position
- Up side is inspiratory
- Down side is expiratory

Lucaya et al AJR 2000 174:235-41

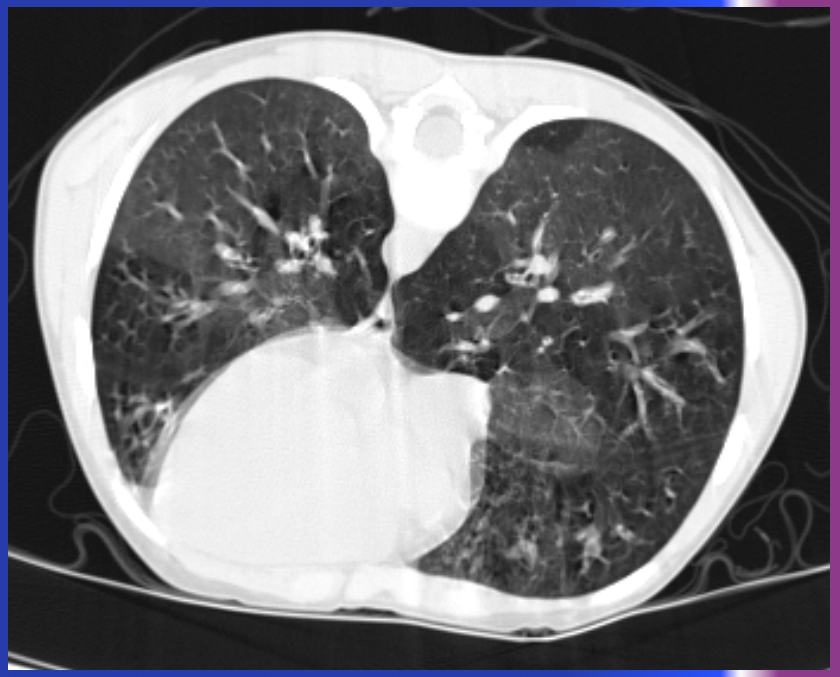


Controlled ventilation CT

- Ventilation with face mask
- CO_2 and chest stretch receptors produce 10-15 seconds of apnea
- Inspiration and expiration images during apneic period

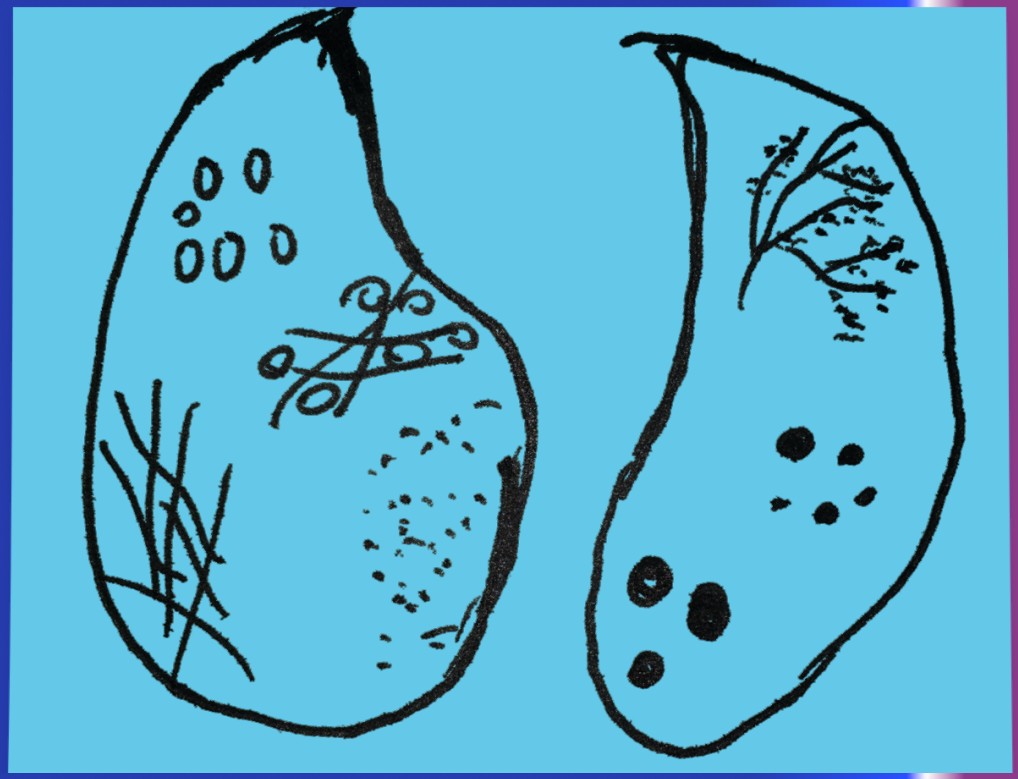
General anesthesia

- Main challenge is atelectasis
- Scan as soon as possible after intubation
- Maintain relatively high inspiratory pressure (30 cm water) with frequent sighs
- Prone scanning helps reduce the atelectasis

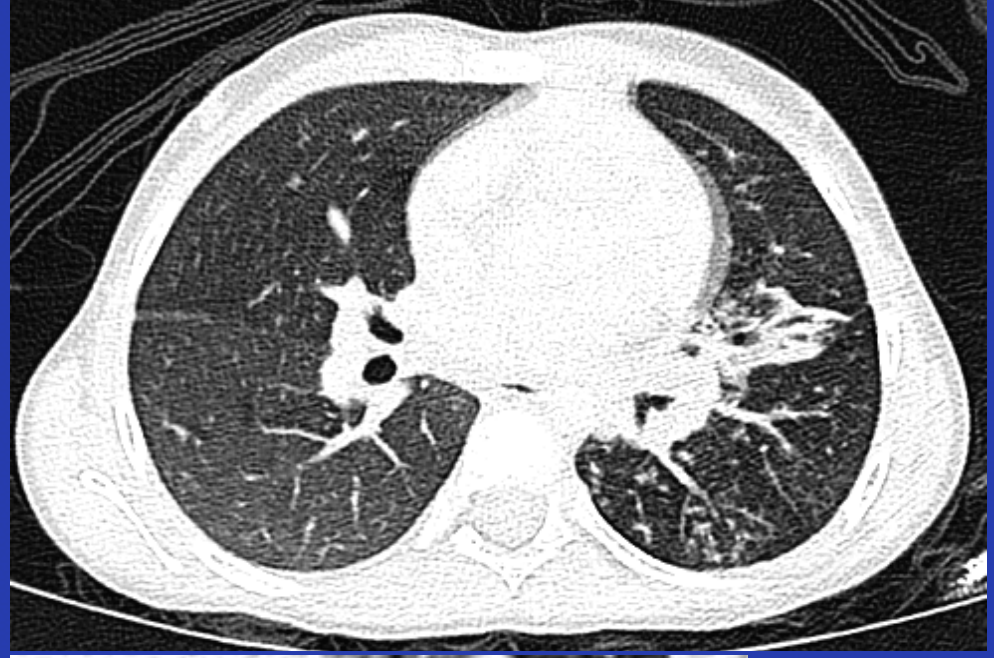


Formulating the report

- ✧ Start with large & small airways
- ✧ Define parenchymal abnormality
- ✧ Ground glass/nodules/ cysts/Emphysema/ Linear/reticular
- ✧ Terminology similar to adult HRCT (use Glossary*)
- ✧ Diagnoses different

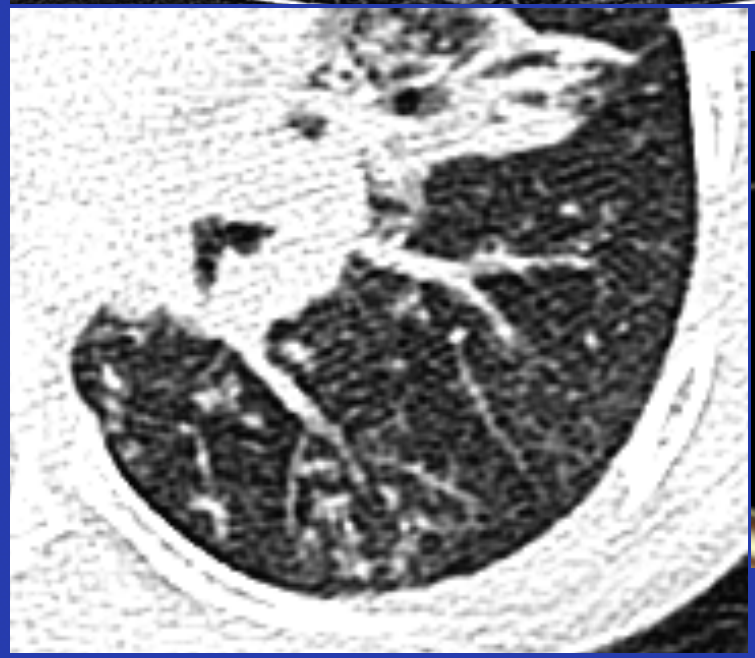


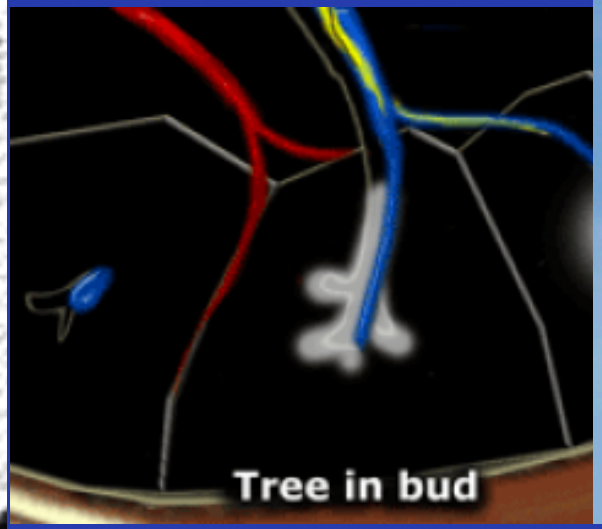
Hansell DM et al Fleischner Society: glossary of terms for thoracic imaging. Radiology. 2008;246:697–722.



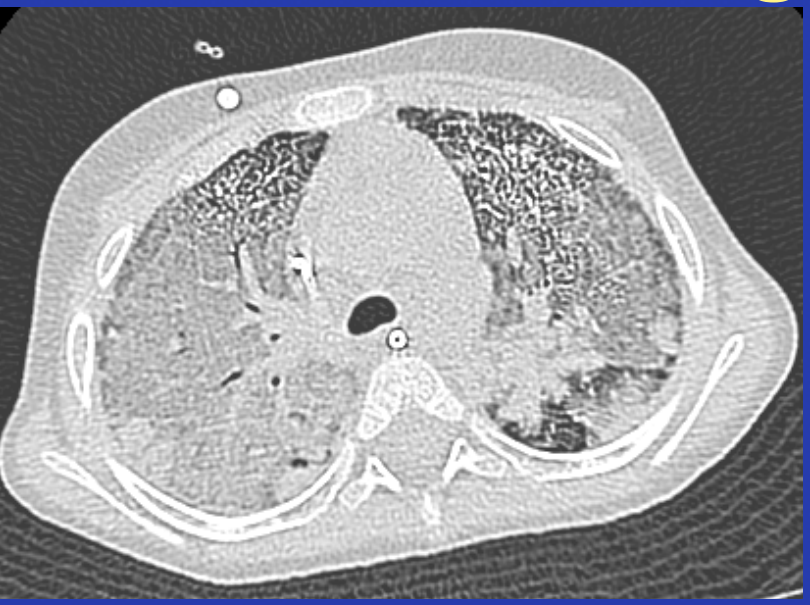
Tree in bud

- Inspissated *material* in distal bronchioles
- MAI and other infections
- Chronic aspiration





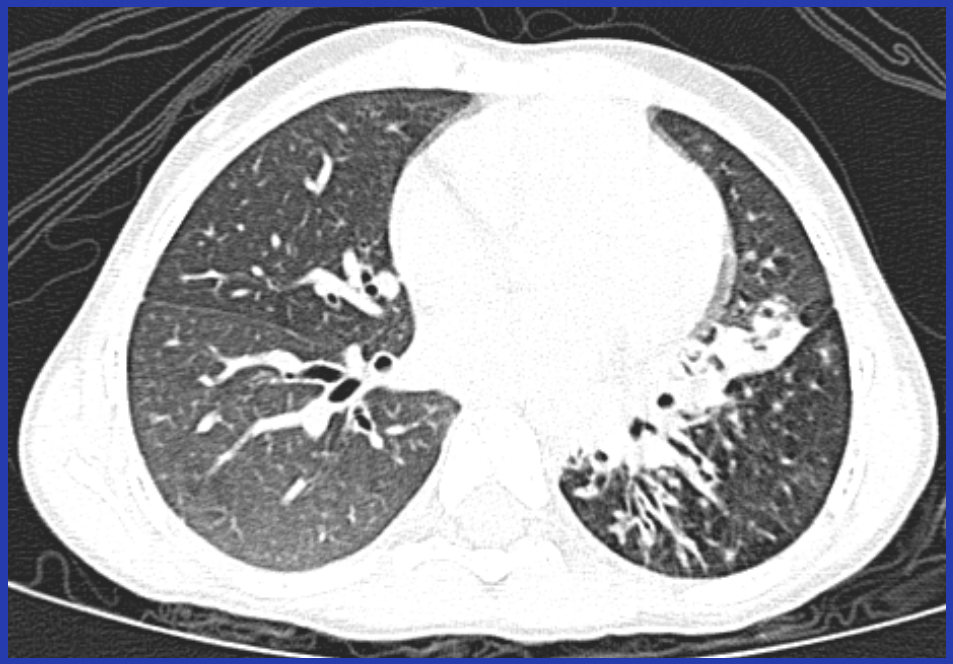
Ground glass attenuation



Niemann-Pick type C disease

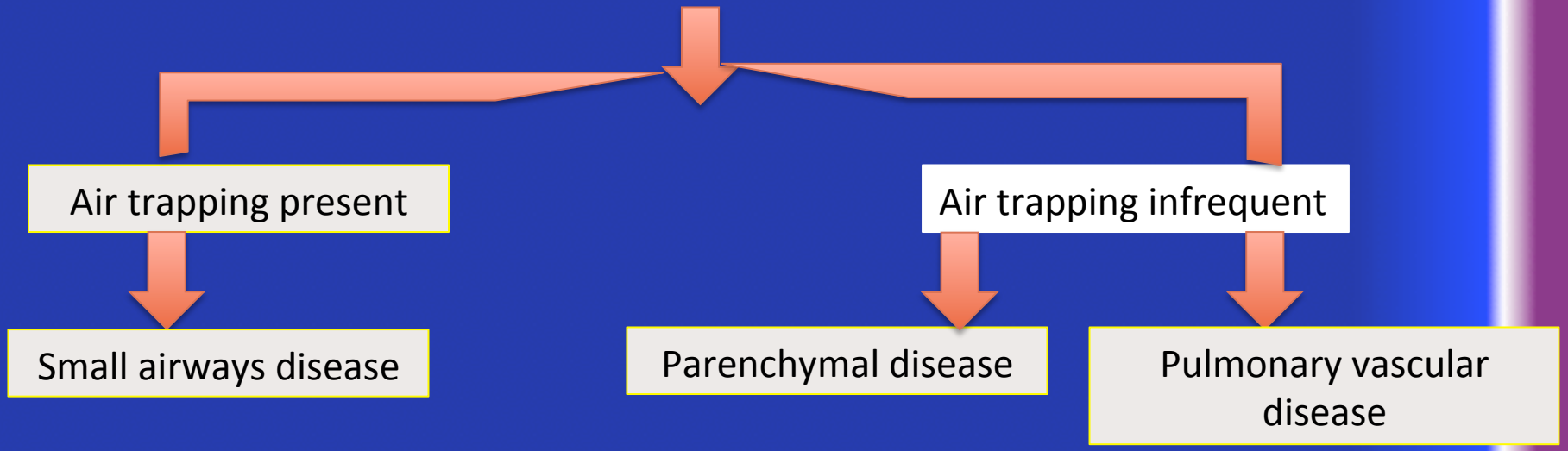
- Hazy increased attenuation of lung with preserved visibility of bronchovascular structures
- Very non-specific
- Increased capillary blood volume (shunting) OR
- Interstitial thickening OR
- Alveolar filling with cells, fluids or other material

Mosaic attenuation



- Attributable to patchy interstitial disease or small airway disease with air trapping
- **BUT** can also be seen with pulmonary hypertension, thromboembolism, or other occlusive vascular disease that results in mixed oligemic and plethoric lung.

Mosaic attenuation



Diagnoses and classification

- Adult classification DOES NOT work in childhood ILD or diffuse lung disease
- Childhood diagnoses are different
- ChILD characterized by-
  - Combination of hyperinflation, mosaic attenuation, air trapping, ground-glass opacities, consolidation, linear/reticular opacities, nodules, or cysts

Clinicopathologic classification of childhood ILD

I. Disorders of Infancy
II. Disorders of the normal host
III. Disorders related to systemic diseases
IV. Disorders of the immunocompromised host
V. Disorders masquerading as ILD

DISORDERS OF INFANCY

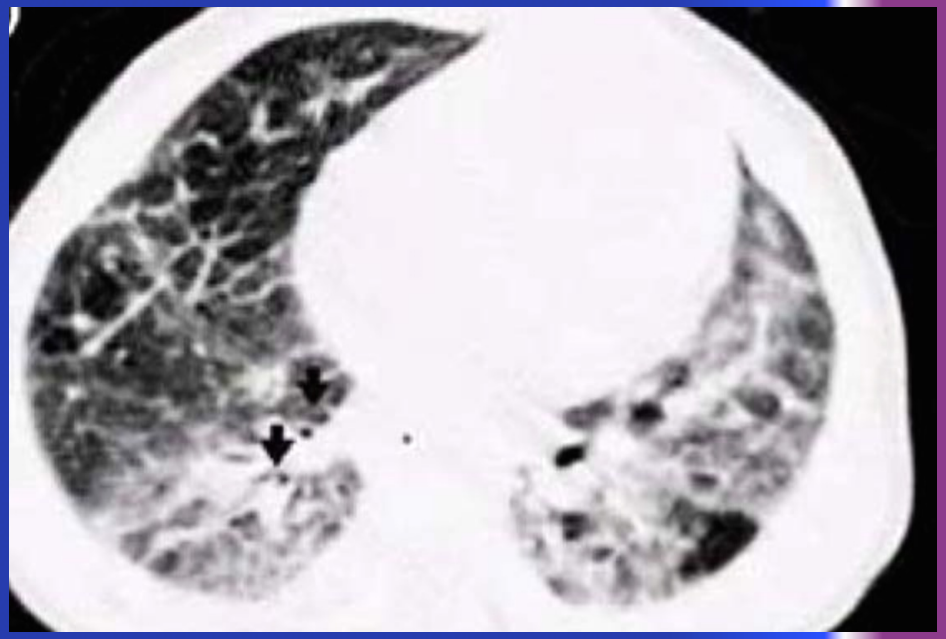
Disorders of infancy- 1

- **Diffuse developmental disorders**
  - Acinar dysplasia
  - Congenital alveolar dysplasia (CAD)
  - Alveolar capillary dysplasia with misaligned pulmonary veins (ACDMPV)

- Term infants, rapid and progressively worsening hypoxia often with severe PHT after birth or early neonatal period
- **Early death, so HRCT findings are not readily available**
- Radiographs- Normal to decreased lung volumes with diffuse opacities resembling hyaline membrane disease

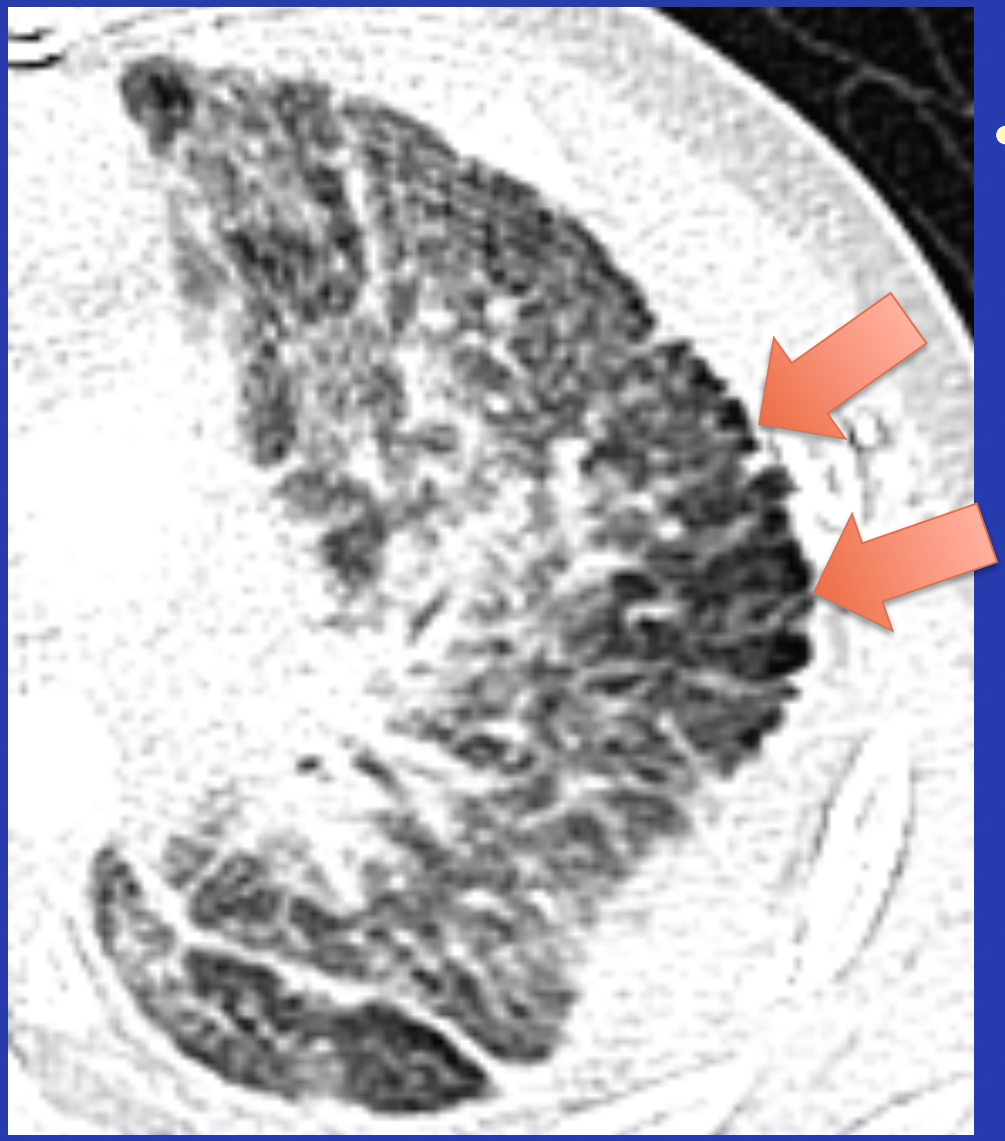
Disorders of infancy- 2

- **Growth disorders**
  - Pulmonary hypoplasia assoc with conditions like oligohydramnios or neuromuscular disease
  - Prematurity associated BPD
  - Structural abn associated with conditions like Trisomy 21 and Down syndrome



- **43% diffuse lung disease in infants**
- Look for underlying conditions

Growth disorders- Imaging features



- Small peripheral cysts as in Trisomy 21 or Turner syndrome

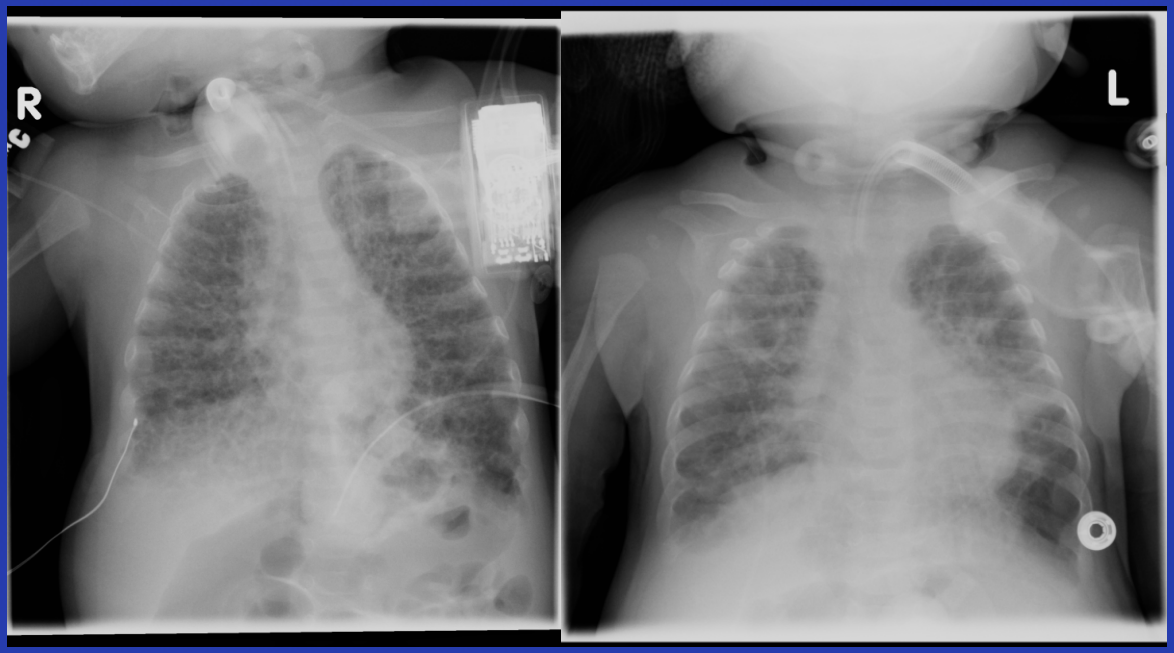
Disorders of infancy - 3

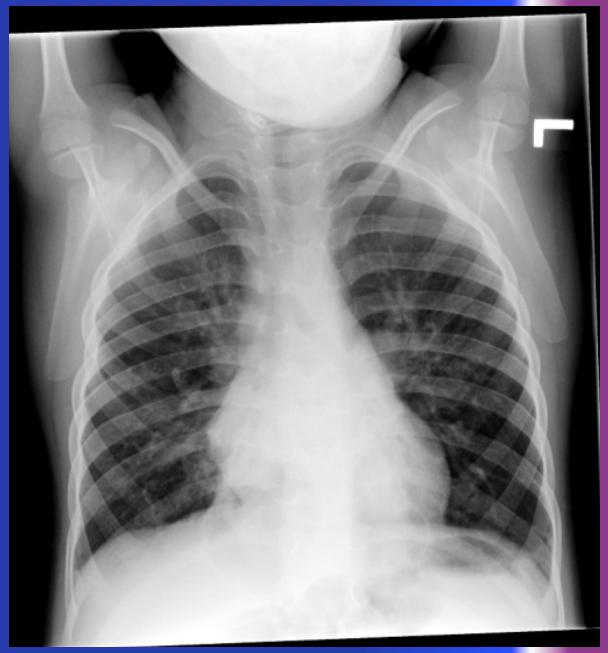
- **Surfactant dysfunction disorders**
  - Surfactant protein B & C, ATP-binding cassette transporter protein A3 (ABCA3) deficiencies
  - Rarer disorders TTF1 and lysinuric protein intolerance

  - Respiratory failure at birth (SpB & ABCA3) or later postnatally with persistent tachypnea and hypoxemia (SpC & ABCA3)
  - Family history of lung disease

Surfactant dysfunction – imaging features

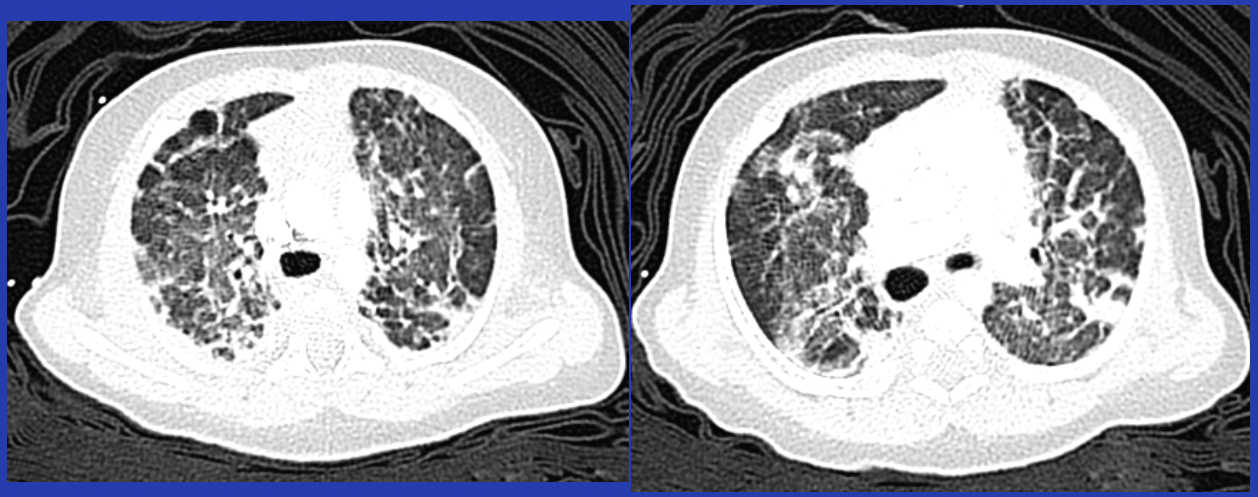
- Diffuse hazy or granular parenchymal opacities (ground glass opacities) on CXR
- GGO & variable interlobular septal thickening on HRCT

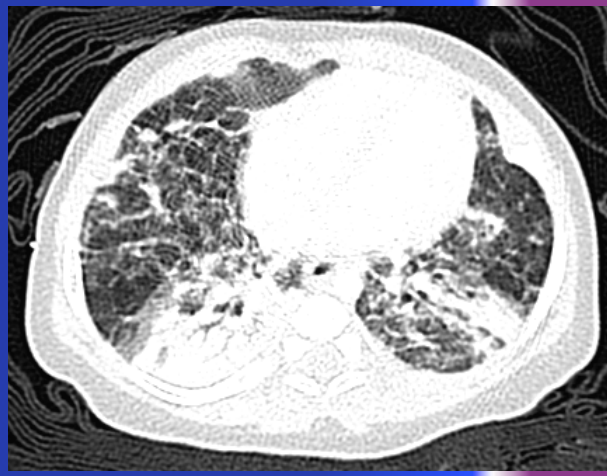




3 months

5 months





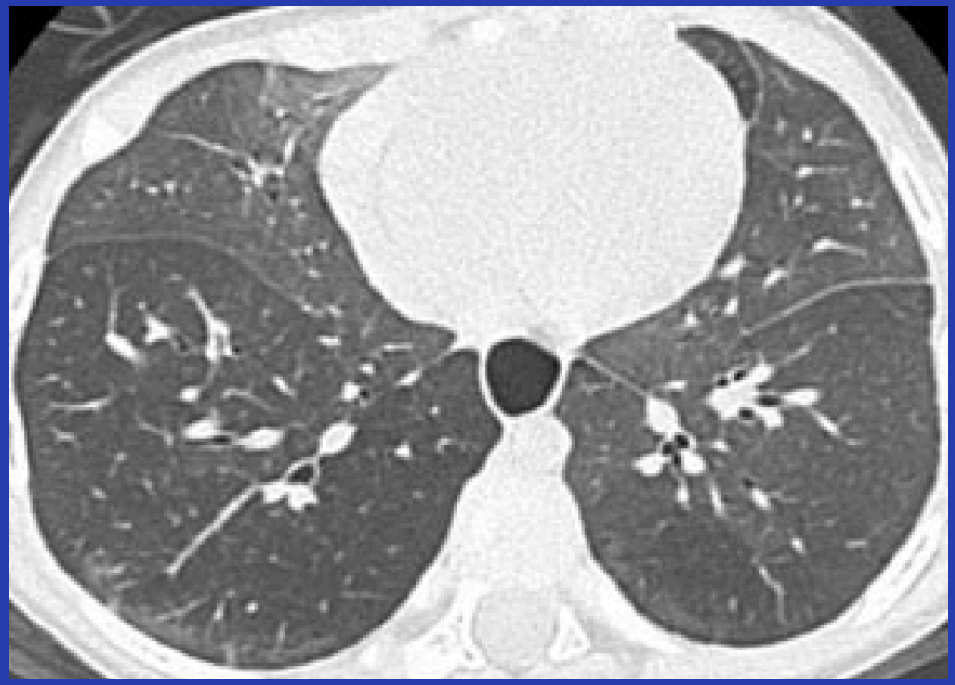
Disorders of infancy -4

Specific conditions of undefined etiology

- NEHI- neuroendocrine hyperplasia of infancy
- PIG- pulmonary interstitial glycogenosis

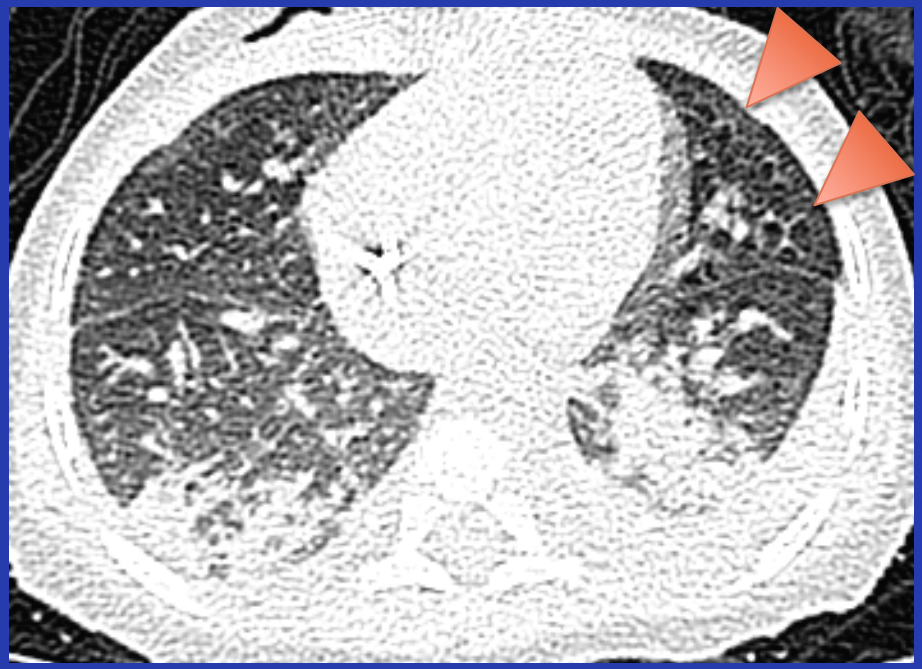
- NEHI- Persistent tachypnea of infancy or follicular bronchitis- Term infants- initially well and then persistent tachypnea, retractions, hypoxemia and crackles without cough or wheezing by 3 months age
- PIG- Preterm & term infants with tachypnea soon after birth

Imaging features



- NEHI
  - hyperinflation, perihilar opacities on CXR
  - GGO with central predominance in lingula and RML
  - Marked hyperinflation on expiration
  - **HRCT is 78% sensitive and 100% specific for NEHI**
  - **Central and anterior distribution**
  - **Path: routine staining may be non-specific or show minor change**
  - **Bombesin staining required**

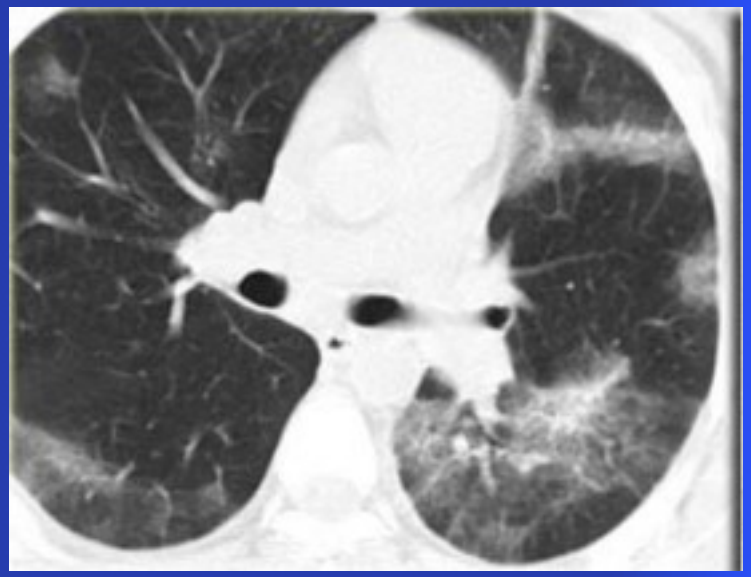
PIG



- Pulmonary interstitial glycogenesis
- Bilateral hyperinflation
- Diffuse interstitial markings on CXR
- GGO
- Interlobular septal thickening in mainly subpleural distribution
- Air-filled cysts have been also described

Disorders of the normal host

- Acute infection
- Post infectious airway injury
- Non-infectious disorders
- Aspiration syndromes
- Eosinophilic pneumonias
- Acute Interstitial pneumonia
- Idiopathic pulmonary hemosiderosis

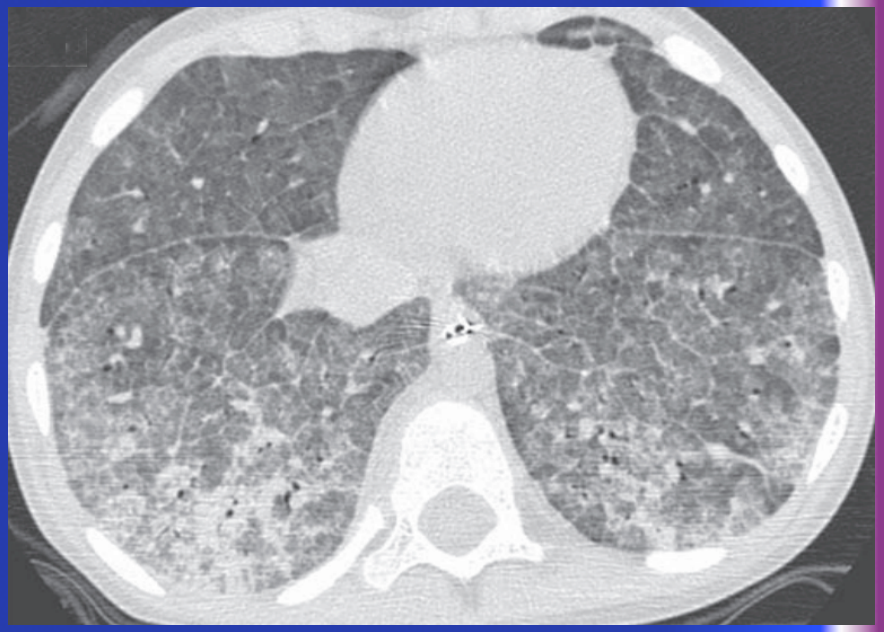


Chronic eosinophilic pneumonia

DISORDERS OF SYSTEMIC DISEASE PROCESSES

Immune related disorders

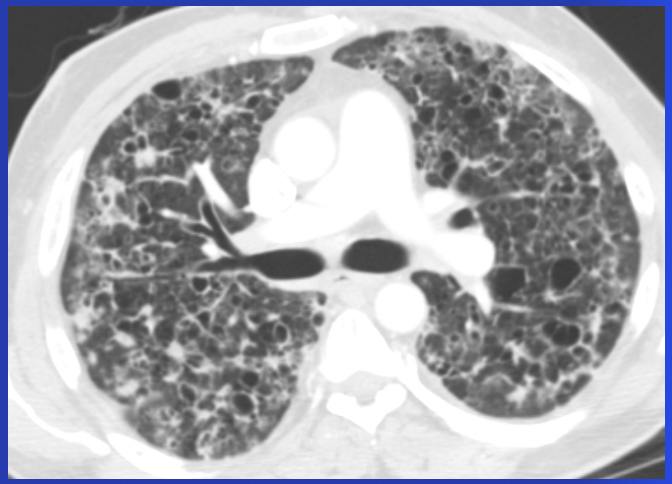
- Acquired pulmonary alveolar proteinosis
- Immune mediated pulmonary hemorrhage syndromes
- Nonhemorrhagic parenchymal disease (collagen vascular disease)
- Wegener's granulomatosis



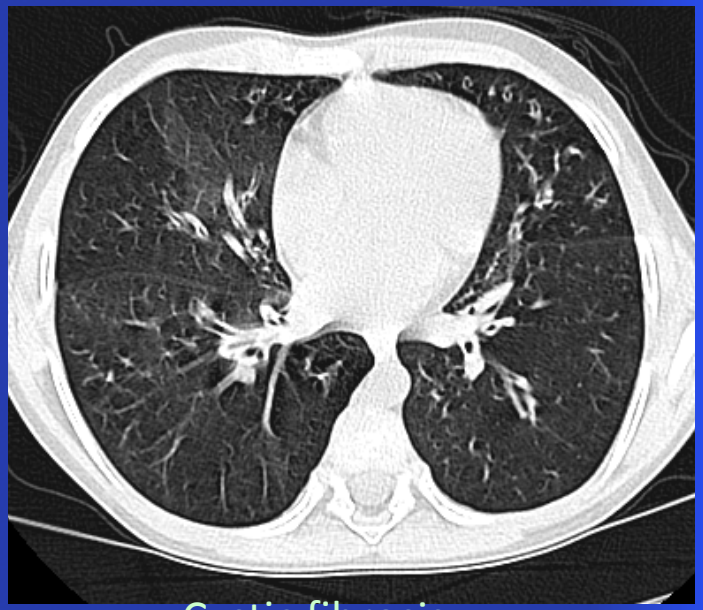
Pulmonary hemorrhage in 4 year-old with anemia and hemoptysis

Non immune related disorders

- Sarcoidosis
- Langerhans cell histiocytosis
- Cystic fibrosis
- Marfan associated pulmonary disorders
- Malignant infiltrates



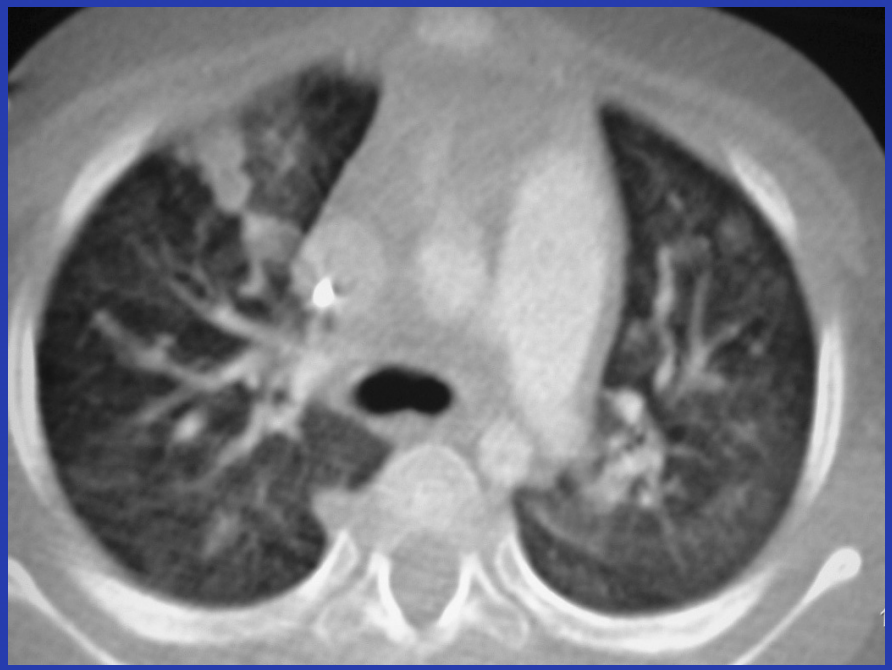
LCH lung



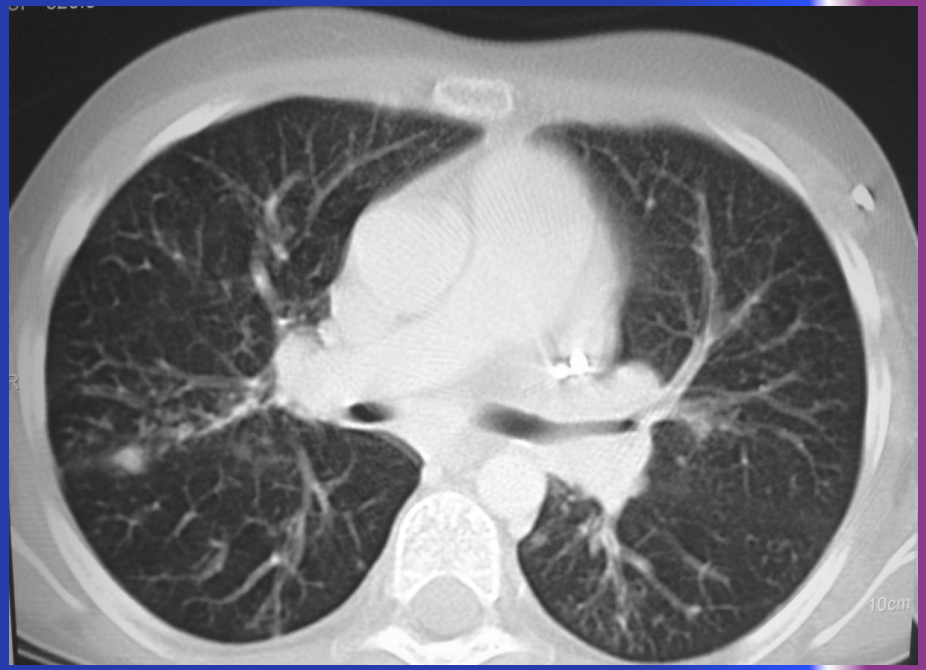
Cystic fibrosis

IV. DISORDERS IN THE IMMUNOCOMPROMISED HOST

Opportunistic infections



CMV infection in post BMT patient



Candida infection in post BMT patient

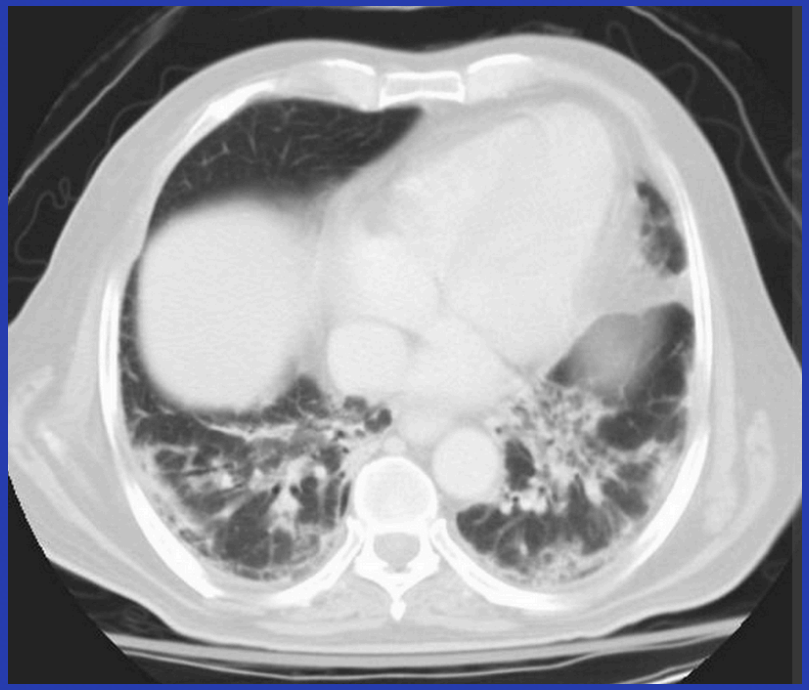
Congenital immunodeficiency

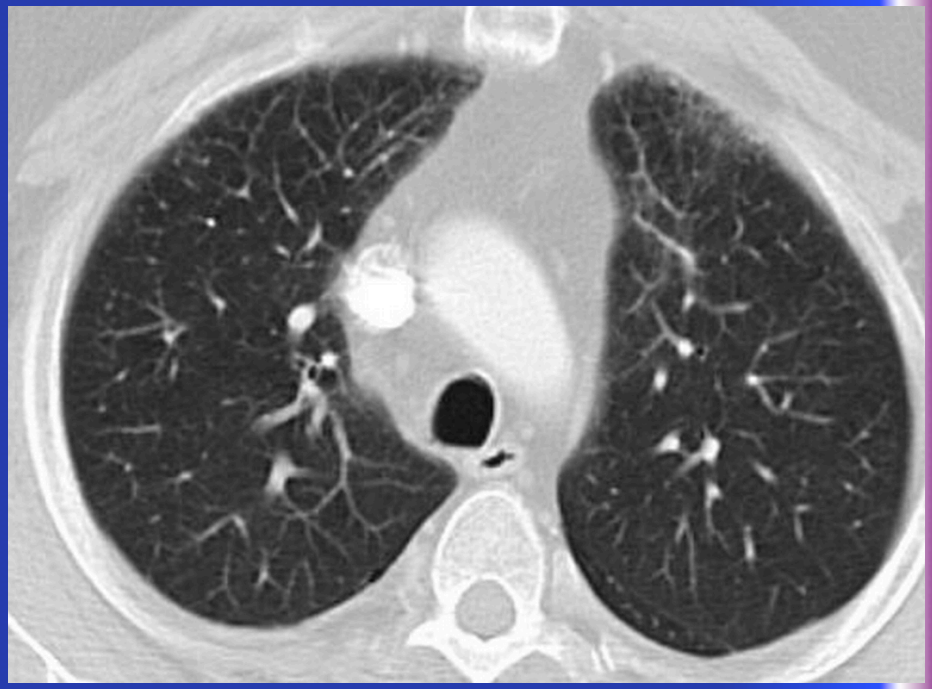
- Chronic Granulomatous Disease
- Common variable immunodeficiency (CVID)

Acquired immunodeficiency

- Non-infectious chemotherapeutic drug related lung injury
- Radiation related lung disease

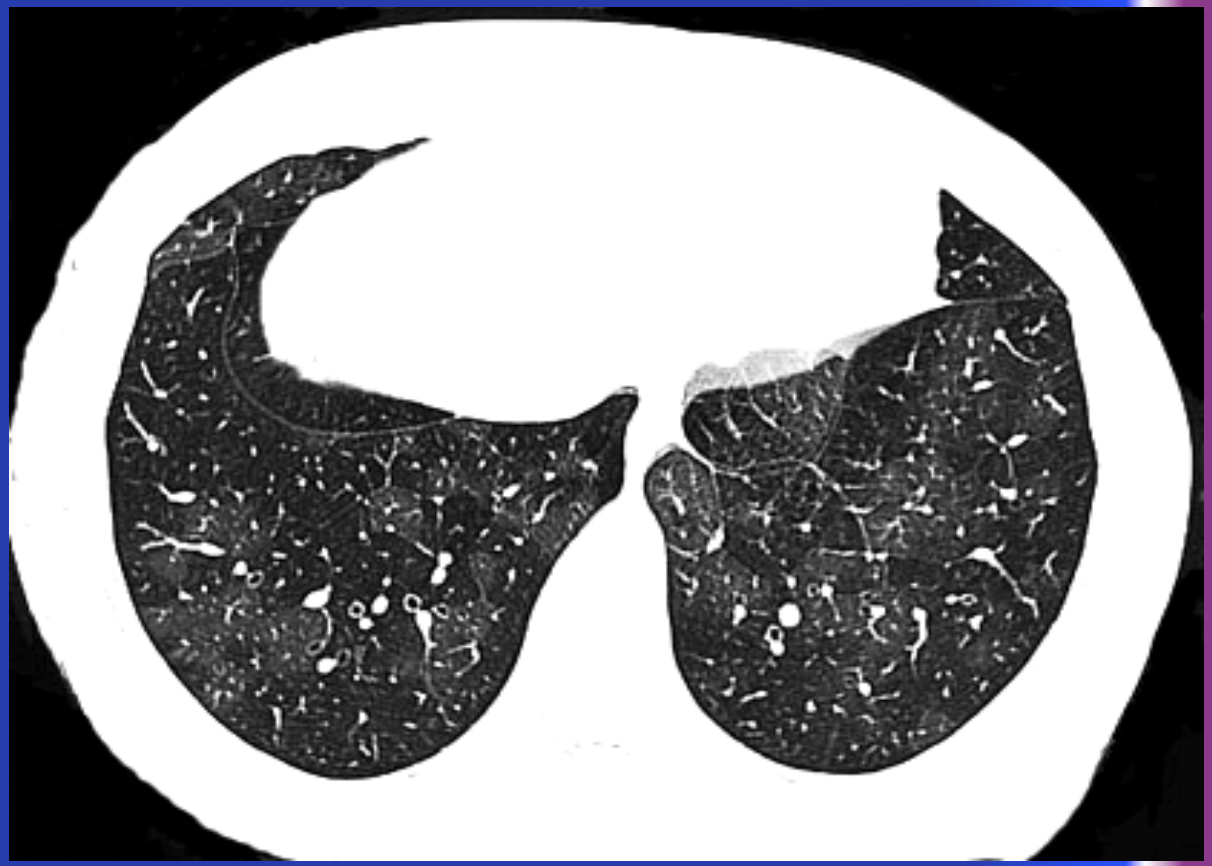
- Look for interstitial prominence and alveolar opacities
- Eventually fibrosis results





Disorders related to lung, BMT & solid organ transplantation

- Rejection
- GVHD
- PTLD



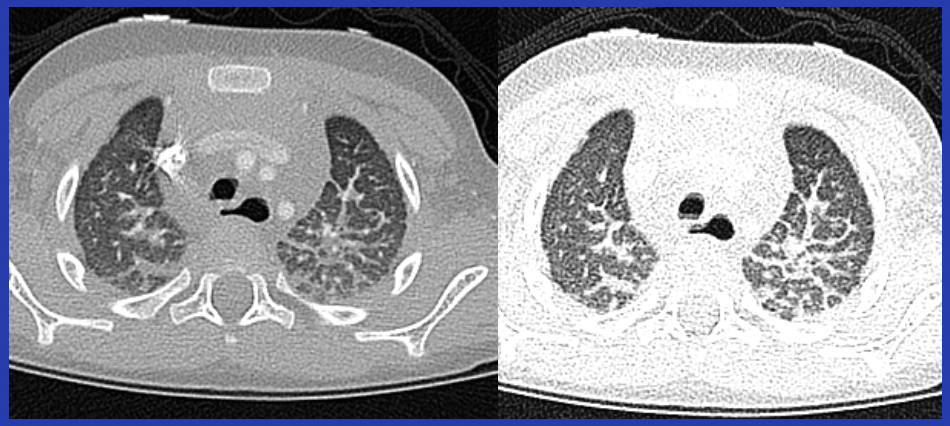
Obilterative bronchiolitis due to graft versus host disease

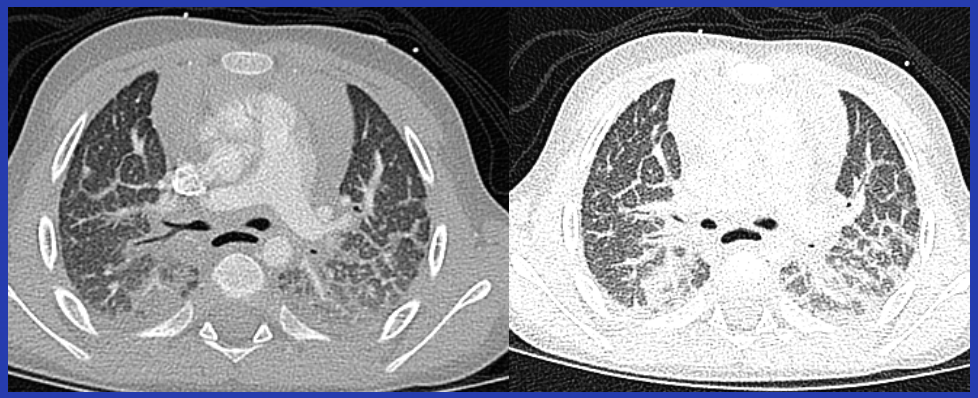
V. DISORDERS MASQUERADING AS ILD

Disorders masquerading as ILD

- Arterial hypertensive vasculopathy
- Congestive vasculopathy
- Lymphatic disorders
- Pulmonary alveolar proteinosis
- Pulmonary edema

Lymphatic disorders

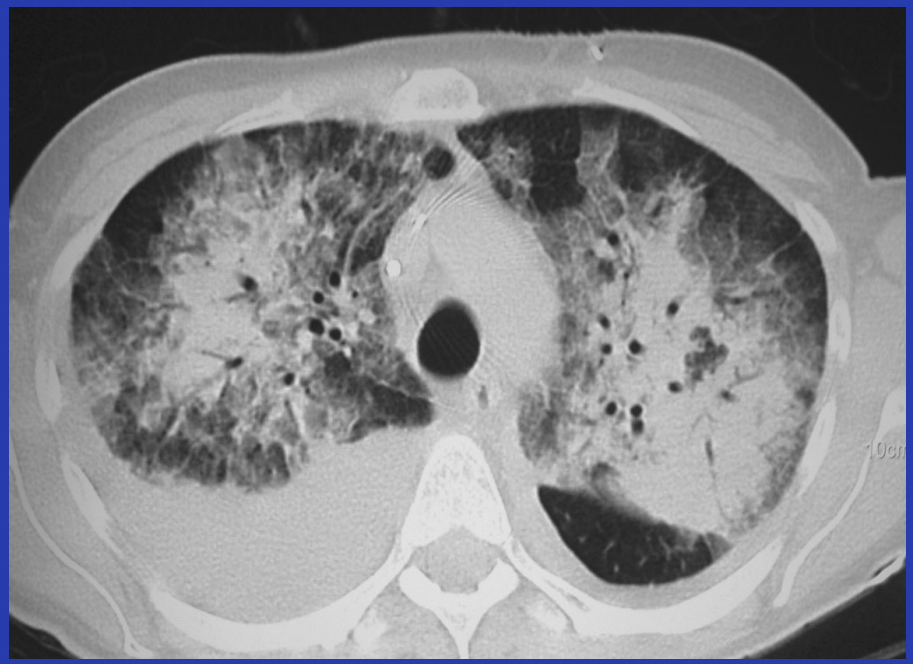




Infant with large cervicothoracic LM

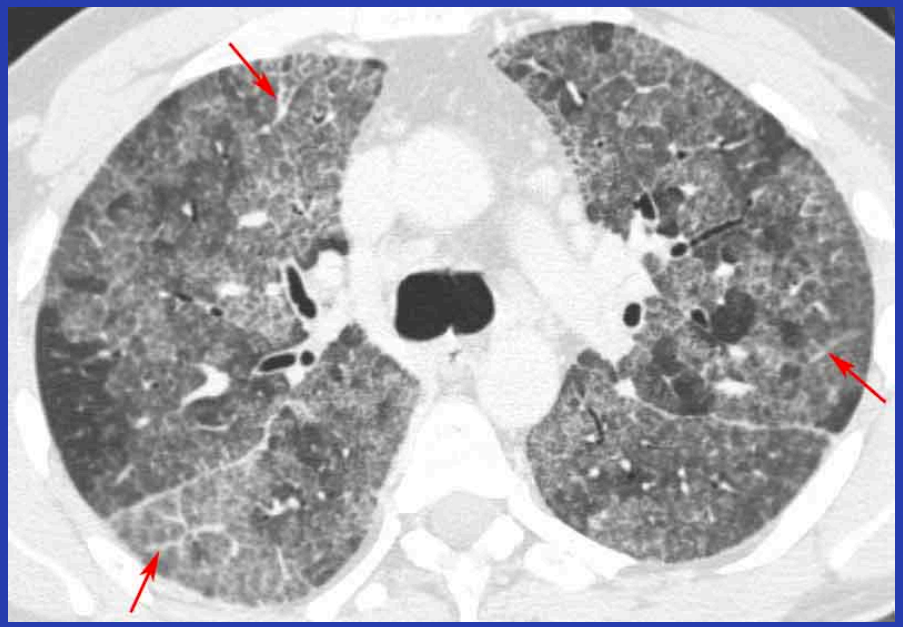
- Diffuse peribronchovascular interstitial thickening
- Interlobular septal thickening
- Chylous pleural effusion common

Pulmonary edema



- Imaging features follows stages of increasing severity
- Ground glass opacity
- Smooth intralobular septal thickening
- Fissural thickening
- Pleural effusions
- Look for a left atrial & ventricular enlargement in cardiogenic edema

Pulmonary alveolar proteinosis



- Pulmonary alveolar proteinosis
- “Crazy-paving”
- **Specific response to an insult, but not etiology**
- Look for surfactant protein deficiency , inhalation exposure, lipoid aspiration and immune compromise

Summary

- Technique of HRCT
- Role of expiratory, prone and decubitus imaging
- Terminology
- Updated classification of childhood ILD with examples
- Clinical-radiological correlation