

Diffuse Lung Lesions (including Interstitial Lung Disease)

Hao-Chien Wang

Department of Internal Medicine
National Taiwan University Hospital

Diffuse Lung Disease

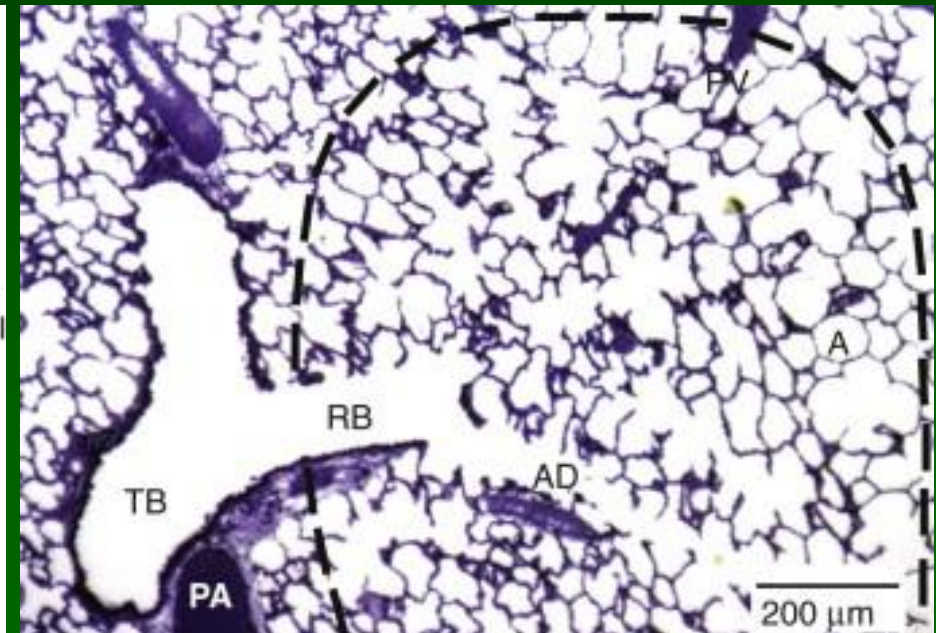
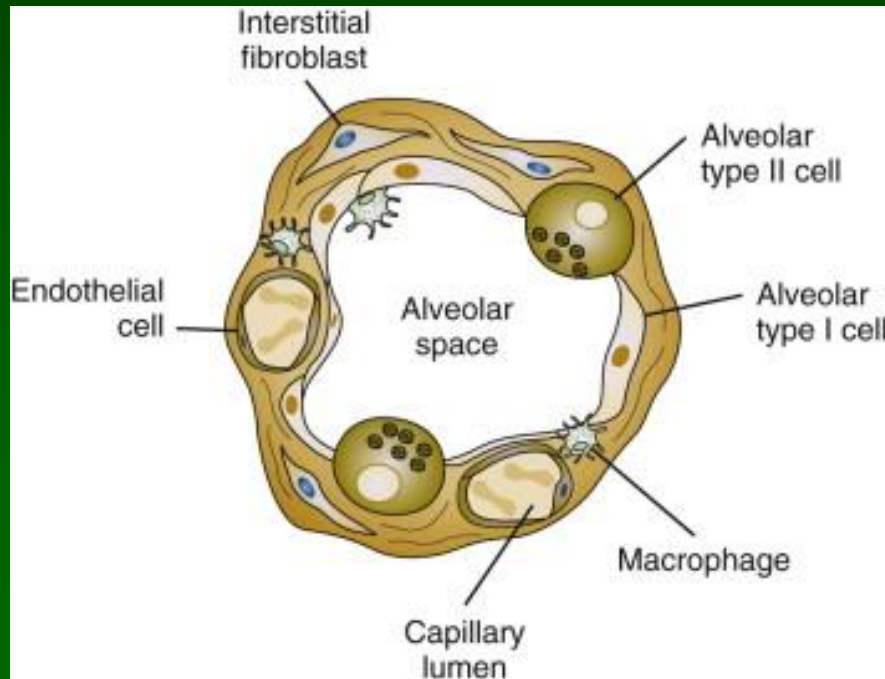
- Alveolar vs. interstitial process
- Over 400 causes
- Known causes:
 - Infection
 - Neoplasm
 - Drug/Radiation/Oxygen
 - Pulmonary emboli
 - Organic and inorganic dust/gases
 - Some causes of ARDS
- Unknown causes: connective tissue disease, UIP/IPF, etc.

Pathogenesis of DLD

- Hematogenous
 - Miliary tbc., hematogenous metastasis
- Bronchocentric, bronchiolocentric
 - RB, RB-ILD, BOOP, HP, Tb
- Lymphatic
 - Sarcoidosis, lymphangitis carcinomatosa
- Mixed

Pulmonary Interstitium

- The *interstitium* includes the space between the **epithelial** and **endothelial** basement membranes.



Secondary Pulmonary Lobule

Secondary lobule

- Polygonal
- 1-2.5 cm
- Smallest unit demarcated by connective tissue septa
- Readily identified on HRCT

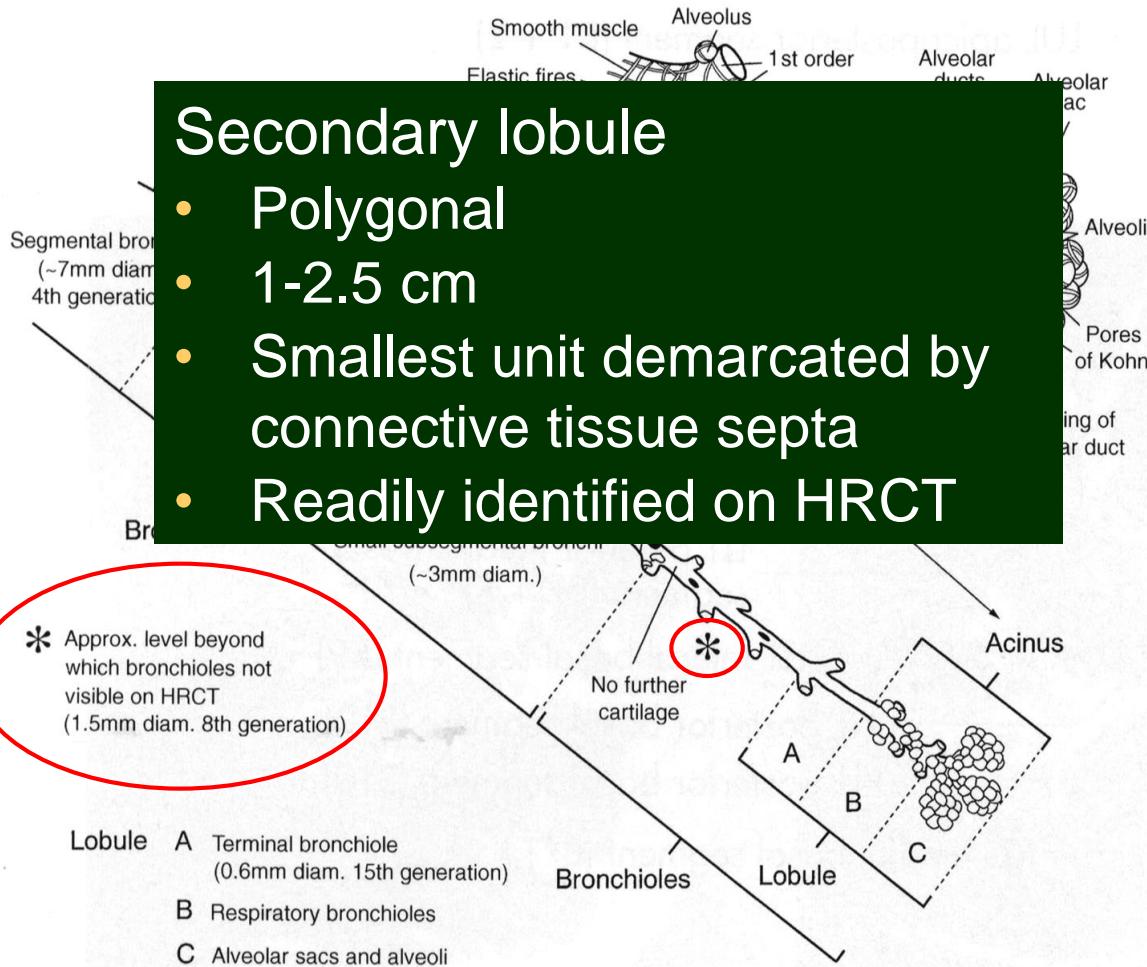
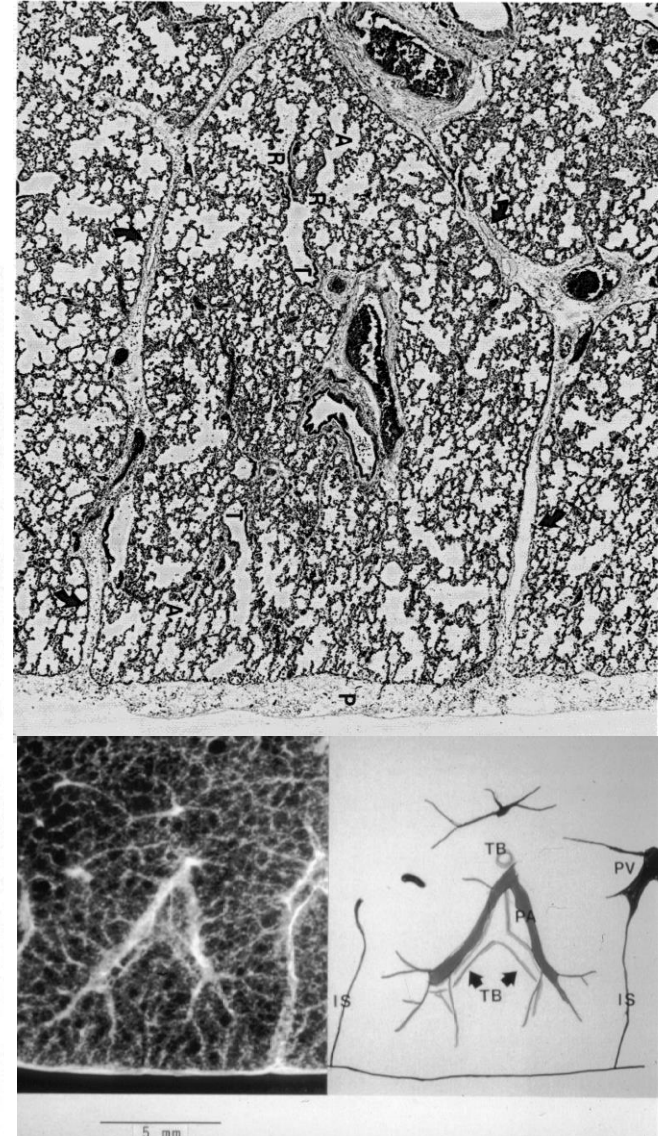
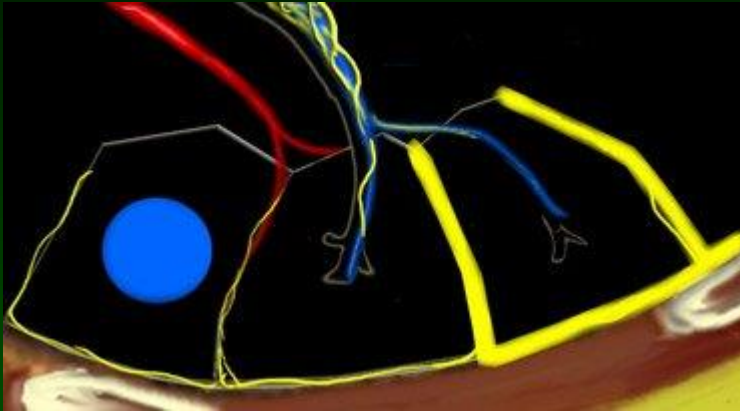


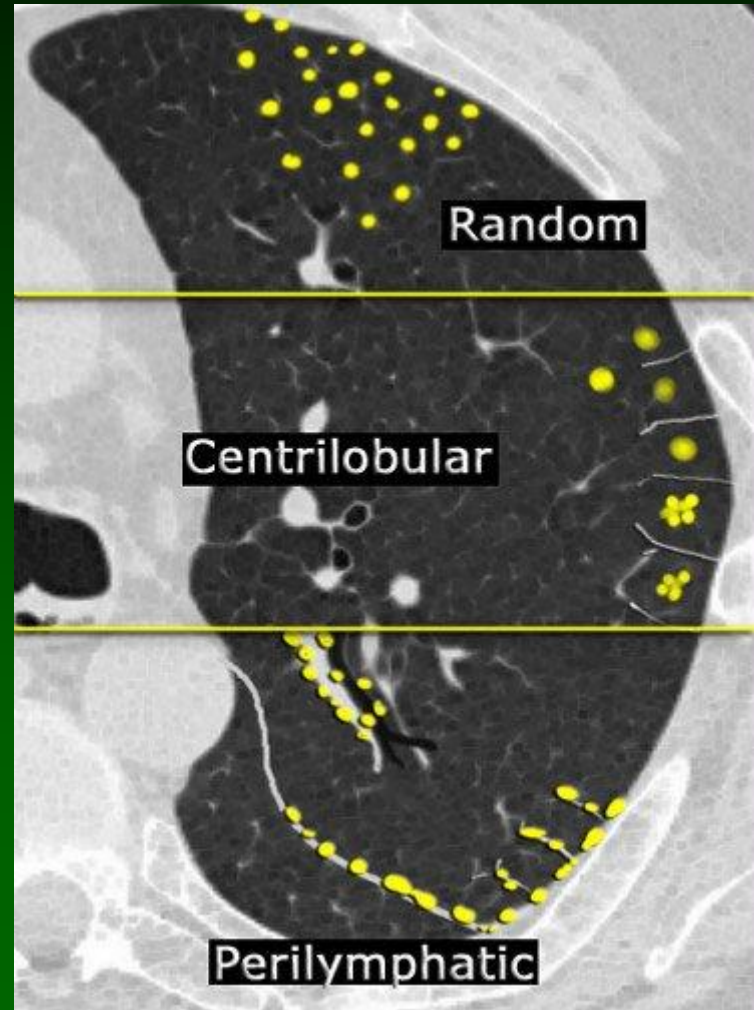
Fig. 2.4 Diagram of branches of airways beyond segmental bronchi.



Secondary Lobule



- Centrilobular
- Perilymphatic



(<http://www.radiologyassistant.nl/en/42d94cd0c326b>)

Approach to Diffuse Lung Disease

- Tempo of disease
- Radiographic pattern
 - Lung volume
 - Pattern of opacities
 - Distribution
 - Ancillary findings
- Clinical context

Approach to Diffuse Lung Disease – Tempo of disease

- Acute
 - Progress within days
- Subacute
 - Change in 2 to 8 weeks
- Chronic
 - No significant change over 8 weeks

Clinical context

- History (occupations, drugs, smoking, collagen disease, toxins, geography, symptoms)
 - Occupational history: asbestos (railway, building, carpenters, joiners); coal mining, metal polishing, fettling or sandblasting of cast iron; organic dusts or allergens (moldy hay, mushroom, birds, pets)
 - Drug histories: amiodarone, chemotherapy, radiotherapy, paraquat ingestion, crack cocaine or heroin inhalation, IV drug abuse
 - Smoking related: IPF, RB-ILD, DIP, PLCH

Clinical context

- Physical examination (clubbing, basal crackles, no crackles, skin lesions, and musculoskeletal)
 - Collagen vascular disease: joints, skin, eye and mouth, muscular weakness; vasculitides (Churg-Strauss syndrome, Wegener's granulomatosis, Behcet's disease and others)
 - Finger clubbing: IPF or fibrosis associated with collagen vascular disease; rare in sarcoidosis and HP
 - Basal inspiratory crackles: common in IPF, asbestosis; rare in sarcoidosis, coal worker and silicosis

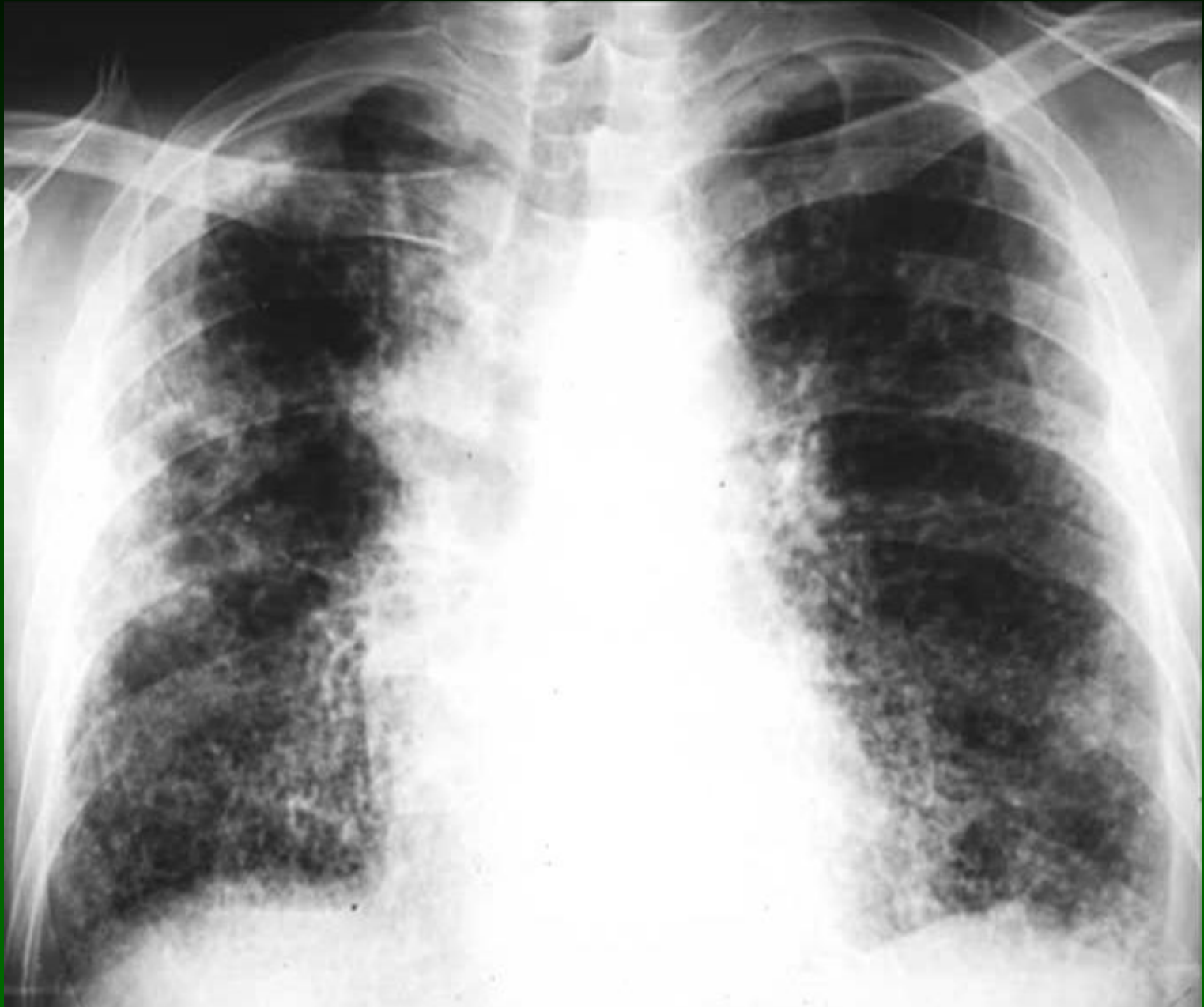
Chest Radiography for DLD

- The first method of detection
- In about 10% of cases, the CXR may look normal
- The pattern of opacities on CXR may be viewed differently when compared with the pattern on HRCT
 - e.g. LAM, PAP

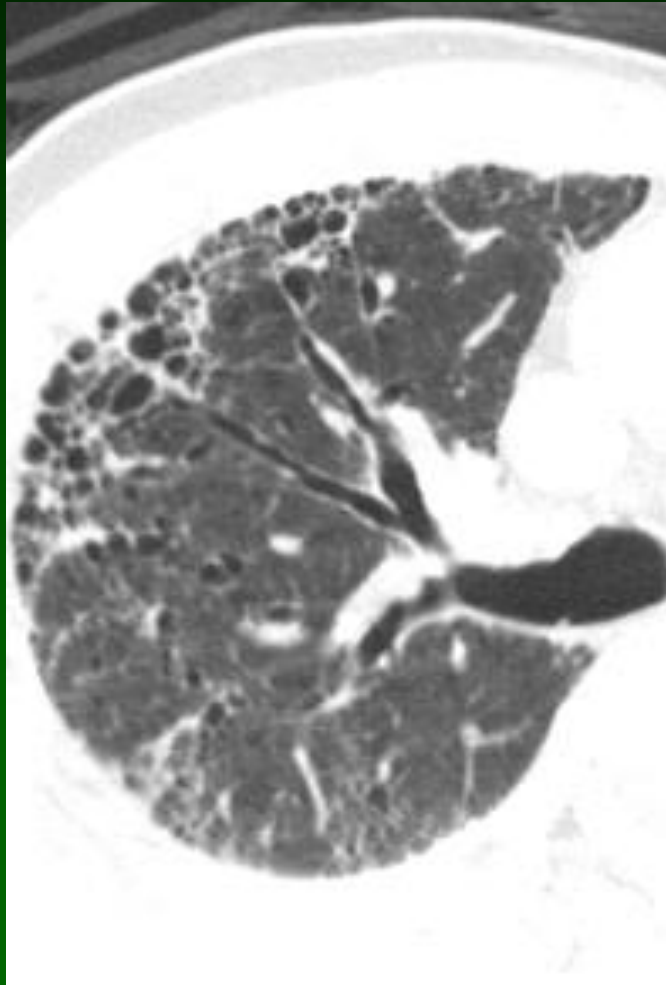
Computed tomography for DLD

- Conventional CT: 7~10-mm slices at 10-mm intervals
- HRCT: 1-1.5 mm slices at 10-mm intervals
 - For patients with suspected diffuse parenchymal lung disease (early detection, pattern identification, extent of opacities, diagnosing bronchiectasis, identifying associated features)
 - Prone position
 - Expiratory images

Idiopathic Pulmonary Fibrosis



Idiopathic Pulmonary Fibrosis



Hypersensitivity Pneumonitis



Approach to Diffuse Lung Disease - Radiography

- CXR
 - Lung volume
 - Opacity
 - Distribution
 - Ancillary findings
- Computed tomography
 - Opacity
 - Distribution

Lung Volume

- Reduced
 - Pathology distal to the airway
 - Fibrosis
 - IPF, asbestosis, sarcoidosis, chronic hypersensitive pneumonitis
- Increased
 - Pathology of the airway
 - emphysema, asthma, bronchitis, obliterative bronchiolitis, PLCH, LAM, neurofibromatosis

Approach to Diffuse Lung Disease – Distribution

	Infiltrative disease	Airway disease
Upper lung predominance	Sarcoidosis, Coal worker's pneumoconiosis, Silicosis, AS, Eosinophilic pneumonia, PLCH, chronic HP, granulomatous infection	Cystic fibrosis, ABPA
Lower lung predominance	IPF, asbestosis, collagen vascular disease	Chronic aspiration, bronchiectasis
Peripheral predominance	IPF, asbestosis, collagen vascular disease, EG, BOOP	Panbronchiolitis
Central predominance	Sarcoidosis, lymphangitis carcinomatosa	ABPA

Approach to Diffuse Lung Disease – Ancillary findings

- Pleural effusion – lung edema, cancer, pneumonia, rheumatoid, LAM
- Pleural plaque: asbestosis
- Pneumothorax: EG/PLCH, LAM
- Cardiomegaly: CHF
- Pericardial effusion: CHF, CVD, metastasis, TB
- Lymphadenopathy: granuloma, neoplasm

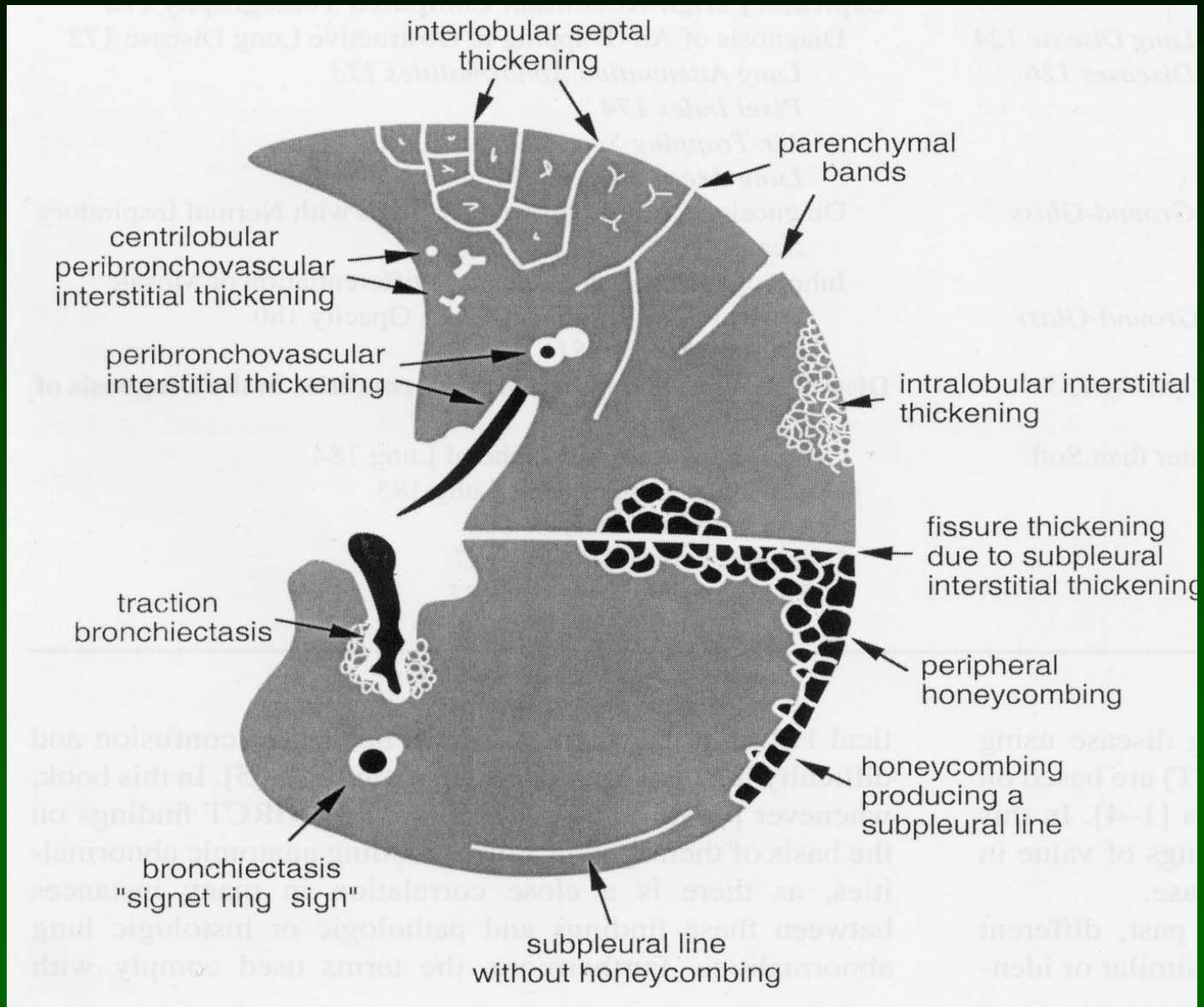
Approach to Diffuse Lung Disease – Ancillary findings

- Esophageal air-fluid level: aspiration, scleroderma
- Spinal fusion: ankylosing spondylitis
- Soft tissue calcification: scleroderma (CREST syndrome), dermatomyositis
- Subcutaneous nodules: neurofibromatosis
- Hepatosplenomegaly: lymphoma, metastatic disease
- Joint abnormalities (humerus, clavicle): rheumatoid disease

Radiological Opacities

- Nodules
 - Sarcoid, silicosis, HP, metastasis
- Reticulation and lines
 - IPF, asbestosis, sarcoidosis, chronic HP
- Ground-glass opacities
 - UIP, DIP, NSIP, AIP, DAD, infection, drug, hemorrhage
- Consolidation
 - BOOP, chronic EP, lymphoma, BAC, infection, hemorrhage
- Cystic airspaces
 - IPF, LAM, LCH

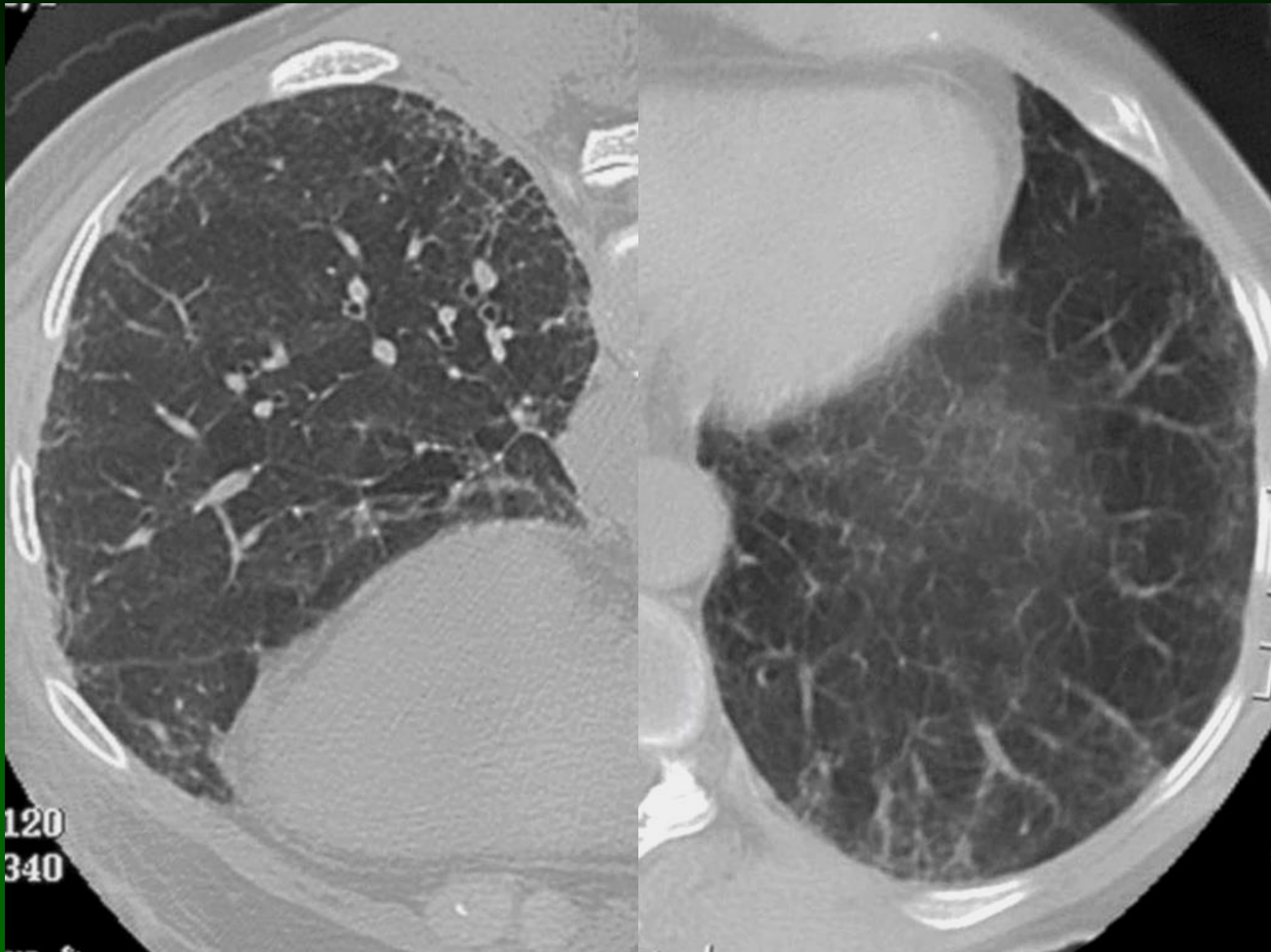
Standardized Glossary in HRCT



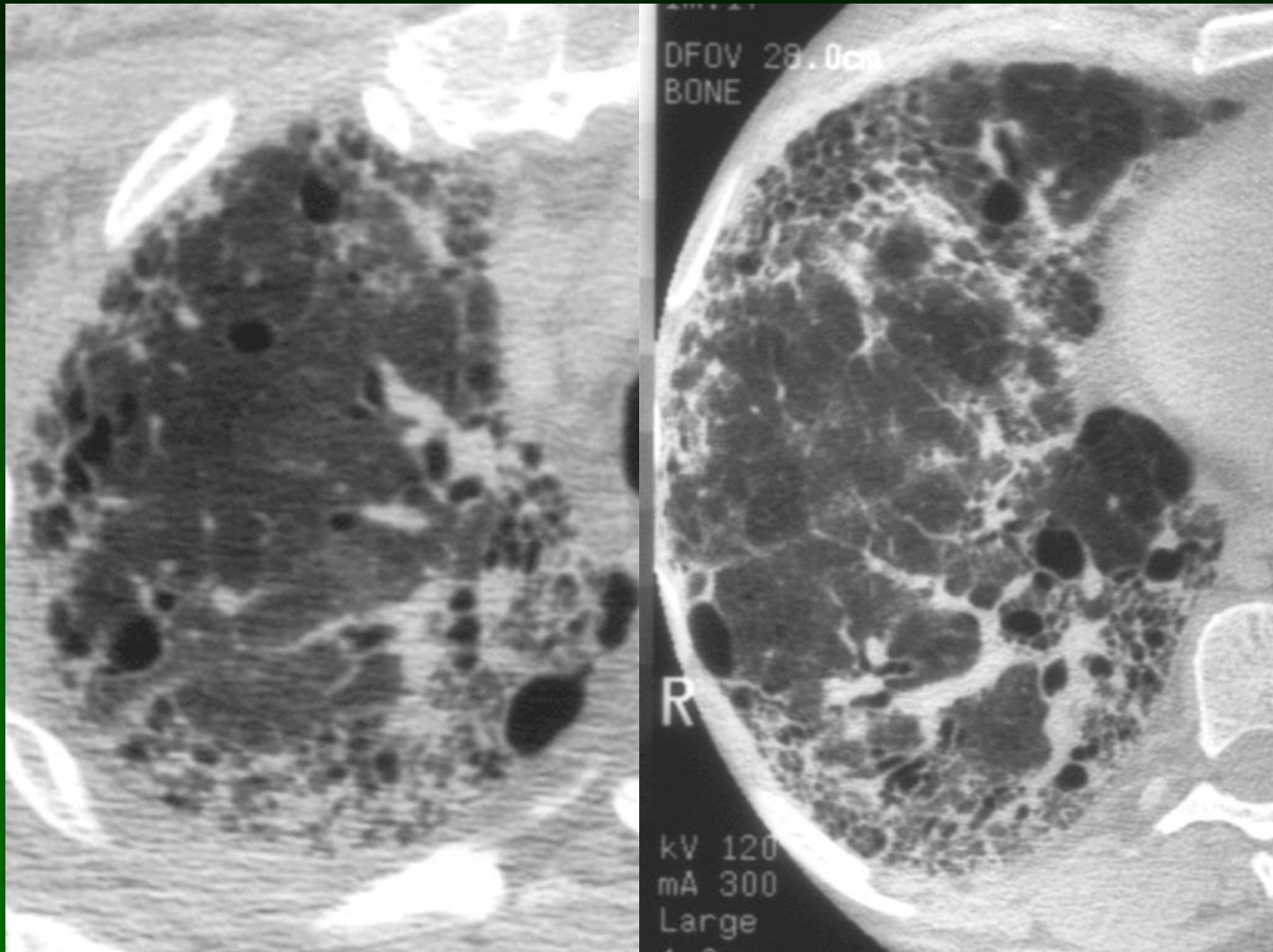
Reticular Pattern

- Reticular pattern
linear (+cystic) opacities
 - Fine
 - Medium (3-10mm)
 - Coarse (>10mm)
- Reticulonodular
 - Reticular, or
 - Reticular + nodular
- Septal thickening
 - Kerley's lines

Reticular Pattern in UIP



Honeycomb Pattern in UIP



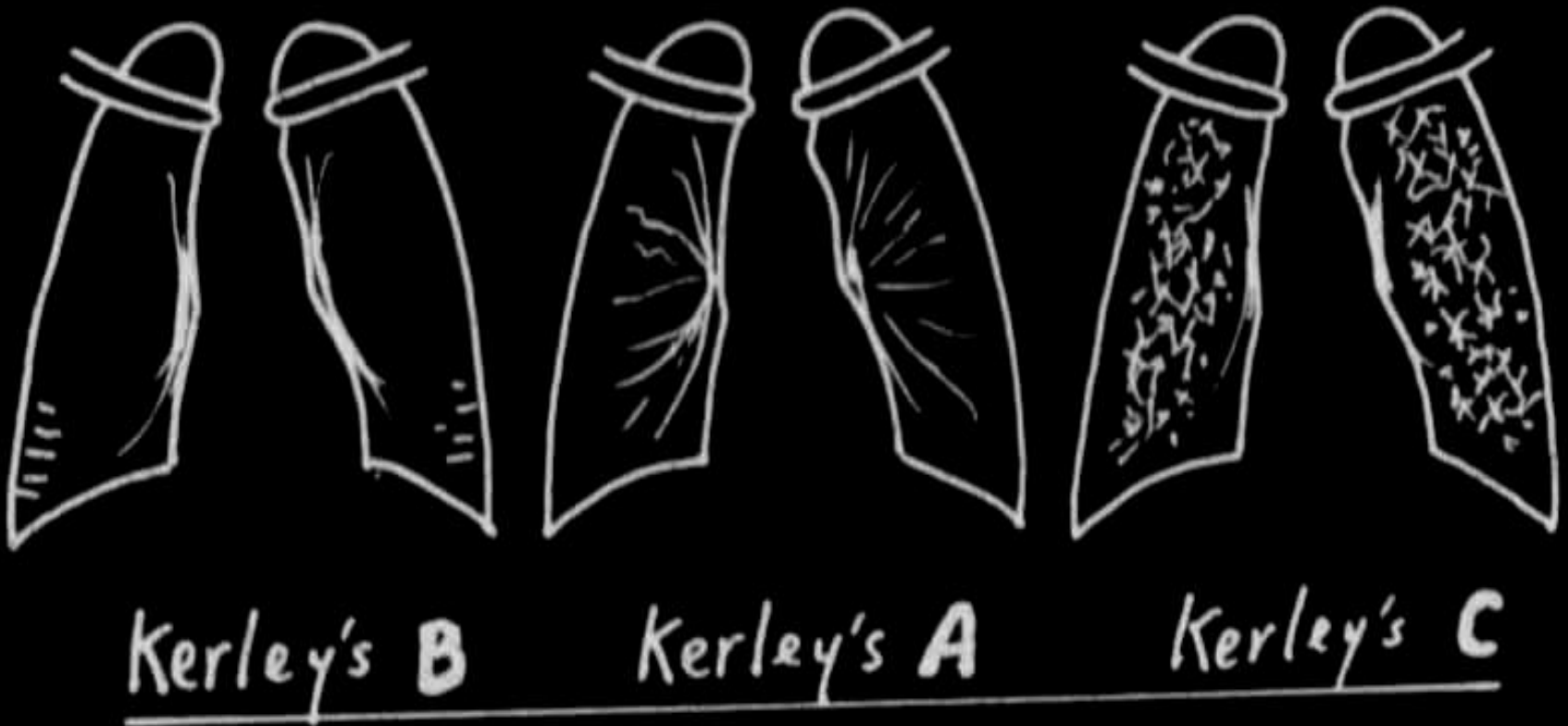
Fine reticular pattern and pleural effusion

- Acute
 - Edema
 - Infection
 - Malaria
- Chronic
 - CHF
 - Rheumatoid disease
 - Lymphangitic spread of tumor
 - Lymphoma and leukemia
 - Lymphangiectasis
 - LAM

Fine reticular pattern and Hilar lymphadenopathy

- Viral pneumonia (rare)
- Sarcoidosis
- Lymphoma and leukemia
- Primary lung cancer
- Metastases
- Silicosis

Septal lines

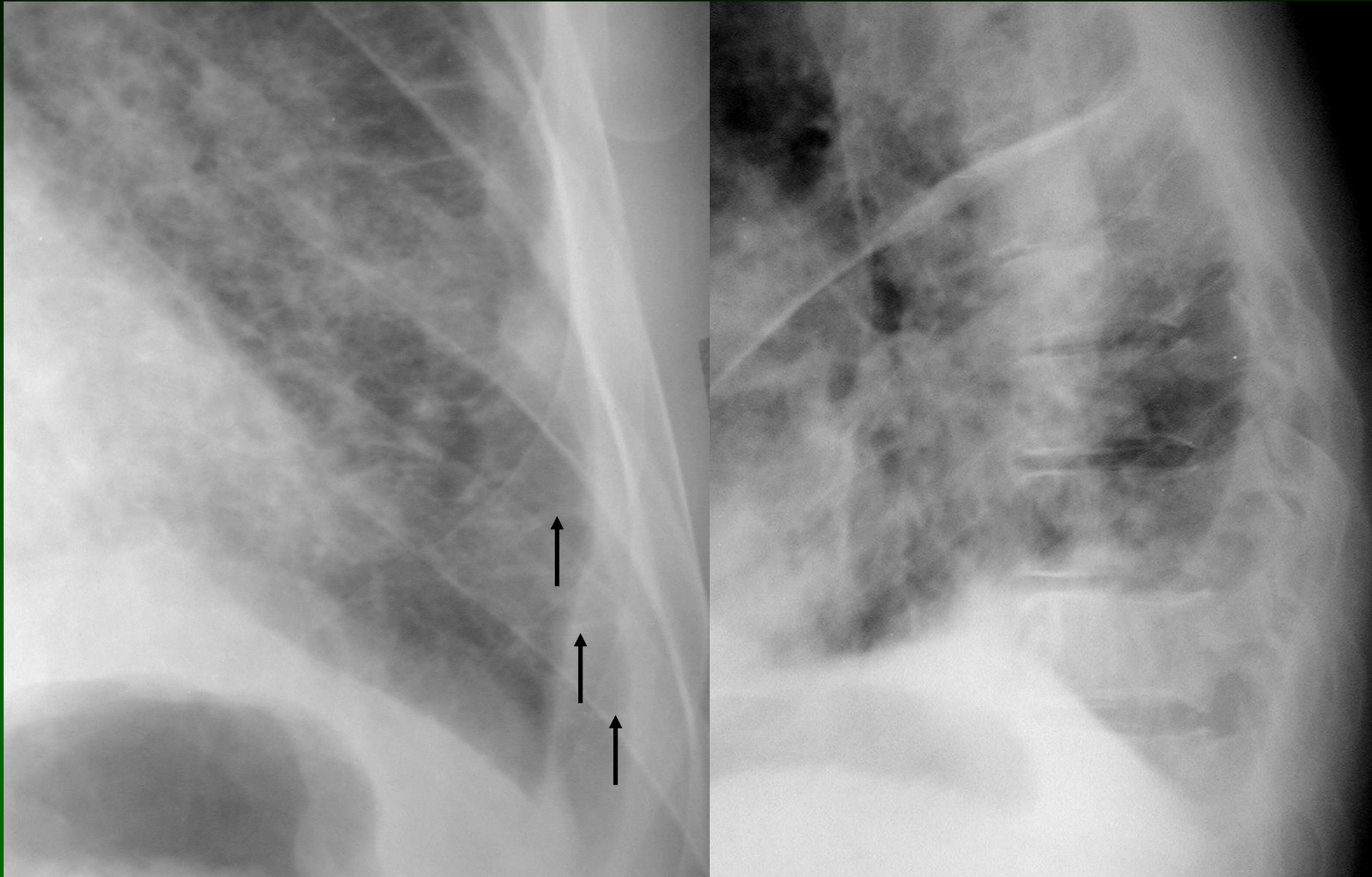


B line: Parallel thin lines at right angle to the pleura, usually at lung base, less than 1 cm in length (interlobular septal line)

A lines: radiate from hila to central portion of the lung, do not reach the pleura, at middle and upper zones, up to 4 cm in length

C line: crisscrossing lines, from superimposition of many B lines

Septal line - Cardiogenic pulmonary edema



Septal thickening

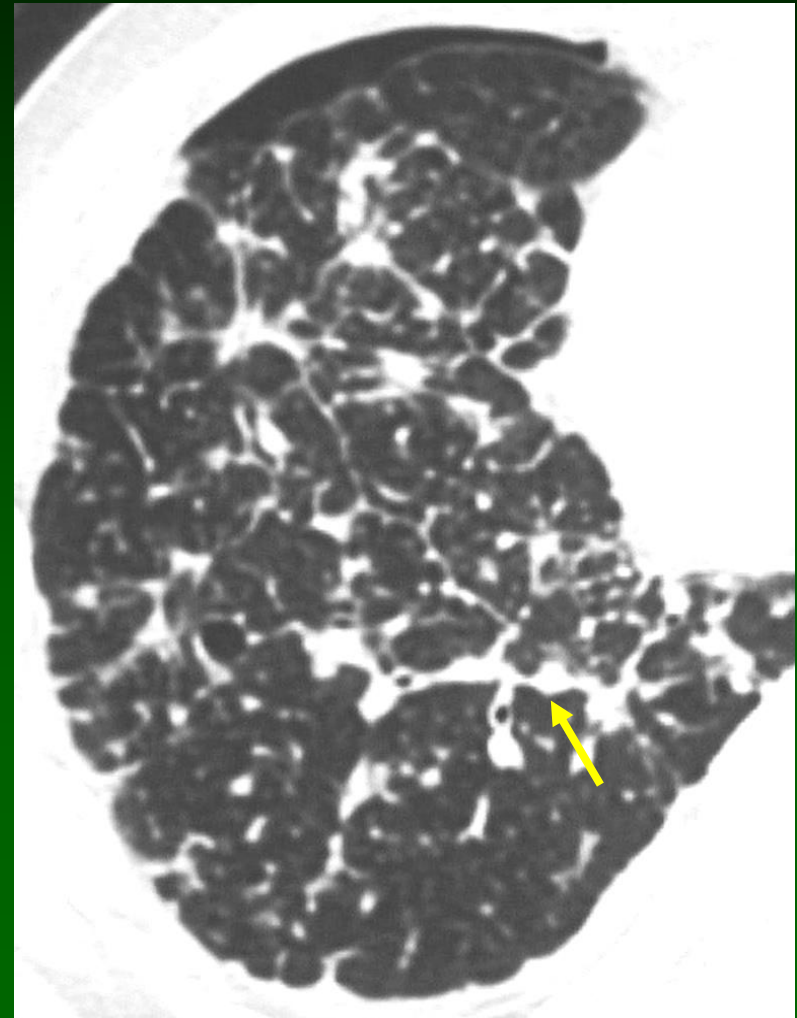
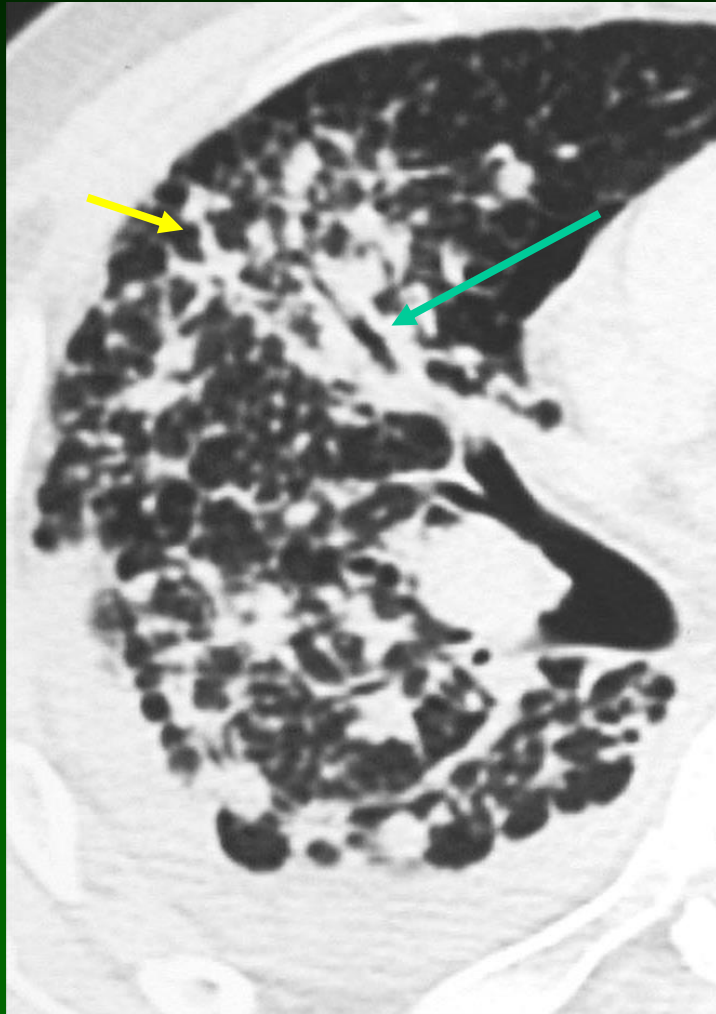
Acute

- Pulmonary edema

Chronic

- Lymphangitis
carcinomatosis
- Pulmonary alveolar
proteinosis
- Sarcoidosis
- Pulmonary VOD

Lymphangitis Carcinomatosa : Beaded thickening of interlobular septa and broncho-vascular bundle (Gastric Cancer)



Lymphangitis Carcinomatosa

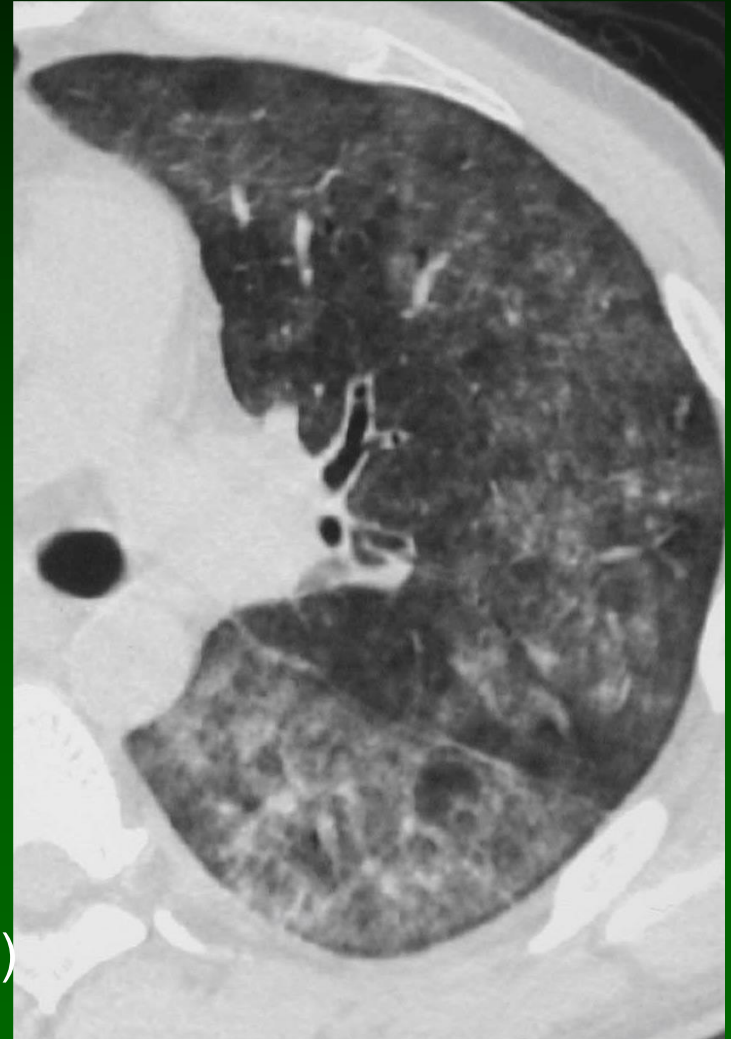


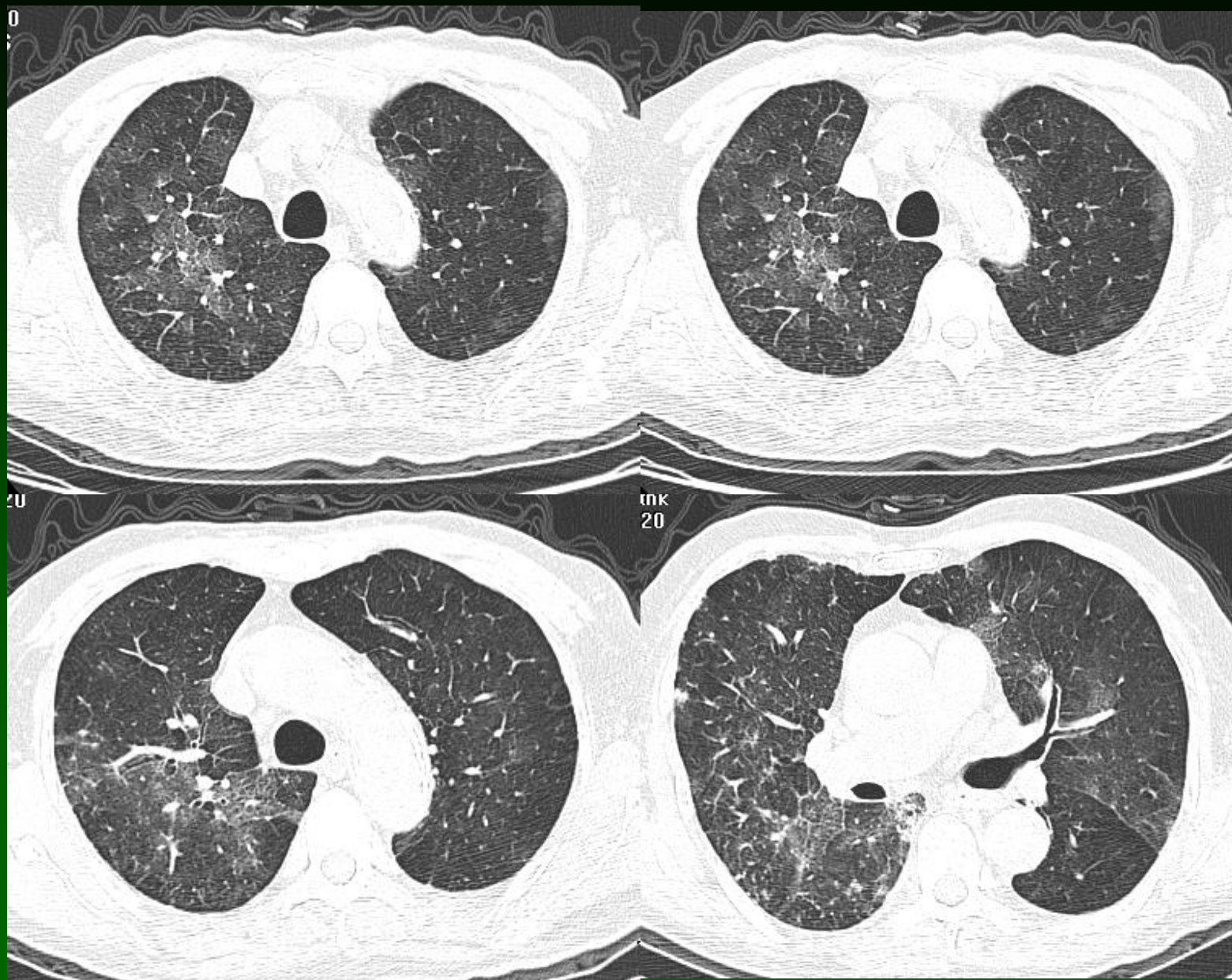
Ground-Glass Pattern

- Hazy increased attenuation of lung, with preservation of bronchial and vascular margins
- Caused by partial filling of air spaces, interstitial thickening, partial collapse of alveoli, normal expiration, or increased capillary blood volume
- Differentiated with Consolidation
- Common but nonspecific
- Clinical history, distribution, associated HRCT findings
- 80% reversible, inflammation/ 20% irreversible, fibrosis

Pneumocystis Jiroveci Pneumonia

- Exudative alveolitis: accumulation of fluid, organisms, fibrin and debris within the alveolar spaces (GGO)
- Mosaic distribution with normal lung
- Intralobular reticulation within GGO: interstitial and interlobular septal infiltration by mononuclear cells and edema
- Subacute stage: interlobular septal lymphatics clearing the alveolitis, with plasma cell and macrophages
- Chronic stage: interstitial fibrosis
- HIV (+) with GGO, highly sensitive and suggestive (94%) (Hartman et al. AJR 1994)





ABCs of Ground Glass Opacification

A	Alveolar proteinosis	C	Cytomegalovirus and other pneumonias (PJP)
	Acute chest syndrome		Cancer and lymphoproliferative disorders
	Acute rejection of lung transplantation		Collagen vascular disease
	ARDS/ AIP		Contusion
	Acute radiation pneumonitis	D	Drug toxicity
			DIP
B	Blood	E	Extrinsic allergic alveolitis
	BOOP		Eosinophilic pneumonia
	BAL		Edema
	Bronchiolitis (respiratory)-associated ILD	F	fibrosis
	BAC carcinoma	G	Granulomatous disease (e.g. sarcoidosis)

Consolidation on CT vs. GGO

- Infiltration of the lung parenchyma completely obliterate the air spaces
 - Filling of the airspace
 - Filling of the interstitium
- More patchy than GGO
- Inspected on mediastinal windows
 - Vessels completely obscured or not
 - CT-angiogram sign

COP/BOOP

- Patchy subpleural and/or peribronchovascular air-space **consolidation** (open arrows)
- often associated with areas of ground-glass attenuation (curved arrows);
- may show **centrilobular nodules** (solid straight arrow) or bronchial wall thickening



(Radiology.
1999;211:555-560.)

Crazy-Paving Pattern

- GGO is superimposed on a background of polygonal reticular lines and septal thickening, without architectural destruction, resembling irregular patio stones.
- Pulmonary alveolar proteinosis: partial filling of alveolar space and lymphatics by proteinaceous material
- Lipoid pneumonia, mucinous BAC

Pulmonary Alveolar Proteinosis

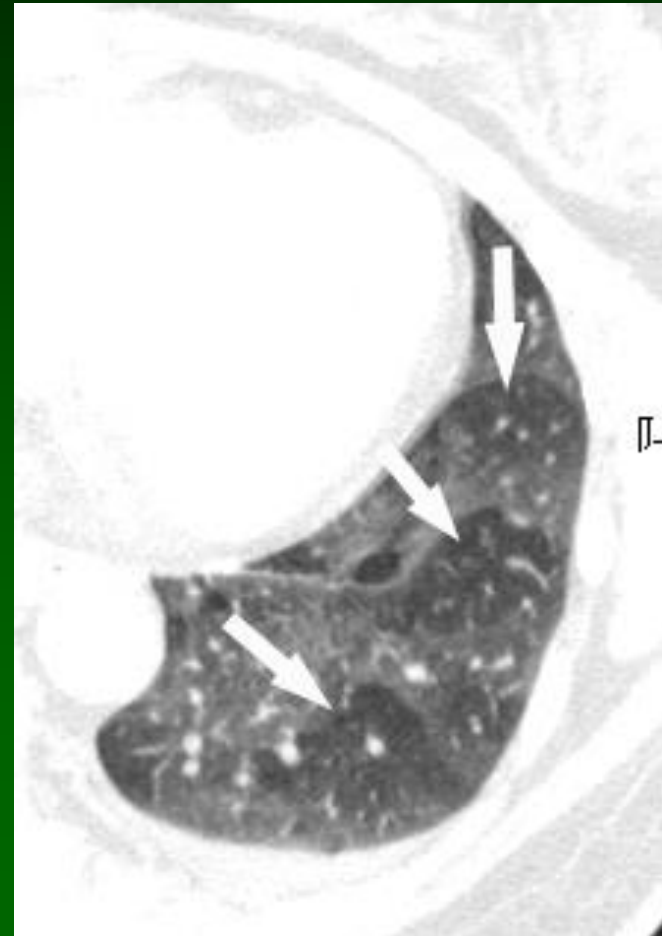
- Filling of alveolar spaces with periodic acid-Schiff-positive proteinaceous material, rich in lipid
- Unknown pathogenesis; silicosis, immunodeficiency
- GGO, geographic distribution
- Smooth thickening of the interlobular septa,
- Crazy-paving appearance (not pathognomonic, but suggestive) of PAP



Mosaic Pattern

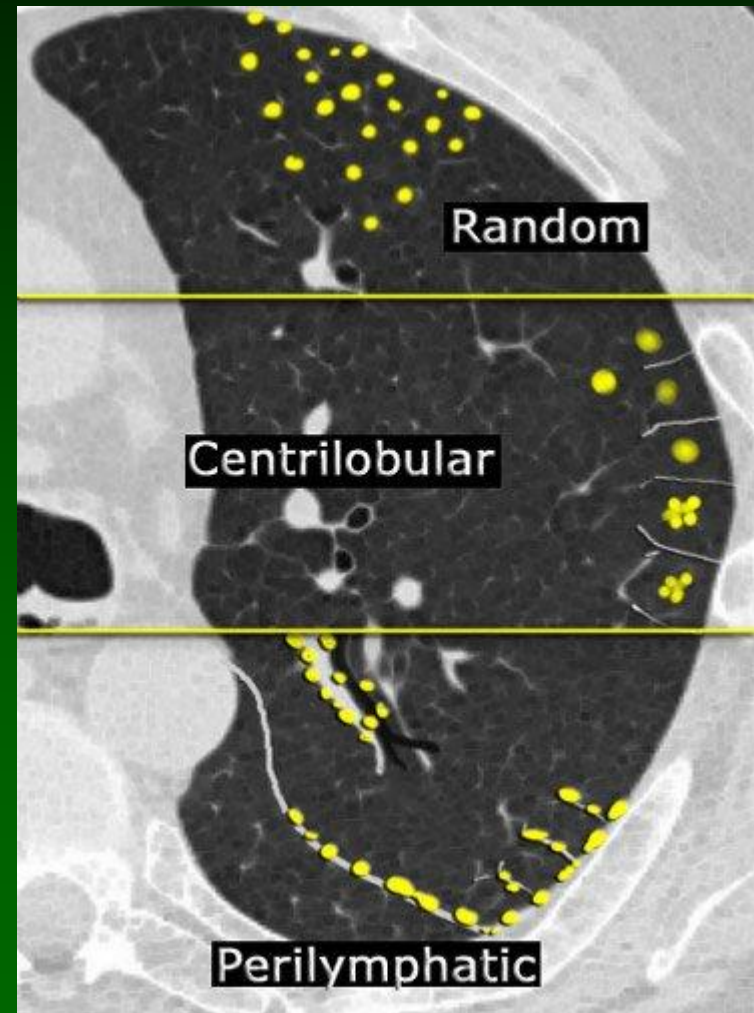
- Decreased lung attenuation
 - Lung destruction: panlobular emphysema
 - Decreased blood flow: pulmonary thromboembolism
 - Decreased ventilation: obliterative bronchiolitis
- Using expiratory CT to differentiate airway disease from vascular disease

Mosaic pattern



Nodular pattern

- Centrilobular
 - HP, RB, BAC, infection
- Perilymphatic
 - Sarcoid, silicosis, lymphangitic ca
- Random
 - Miliary infections, metastasis



Miliary nodules

- Nodules < 5 mm in diameter

Acute - - - - - Viral pneumonia

Miliary tuberculosis

Chronic

Upper lung zone - - Sarcoidosis

Coal workers pneumoconiosis

Silicosis

Eosinophilic granuloma

Hypersensitivity pneumonia

Diffuse - - - - - Any of the above

Miliary carcinomatosis

Miliary Nodules in Miliary Pulmonary TB



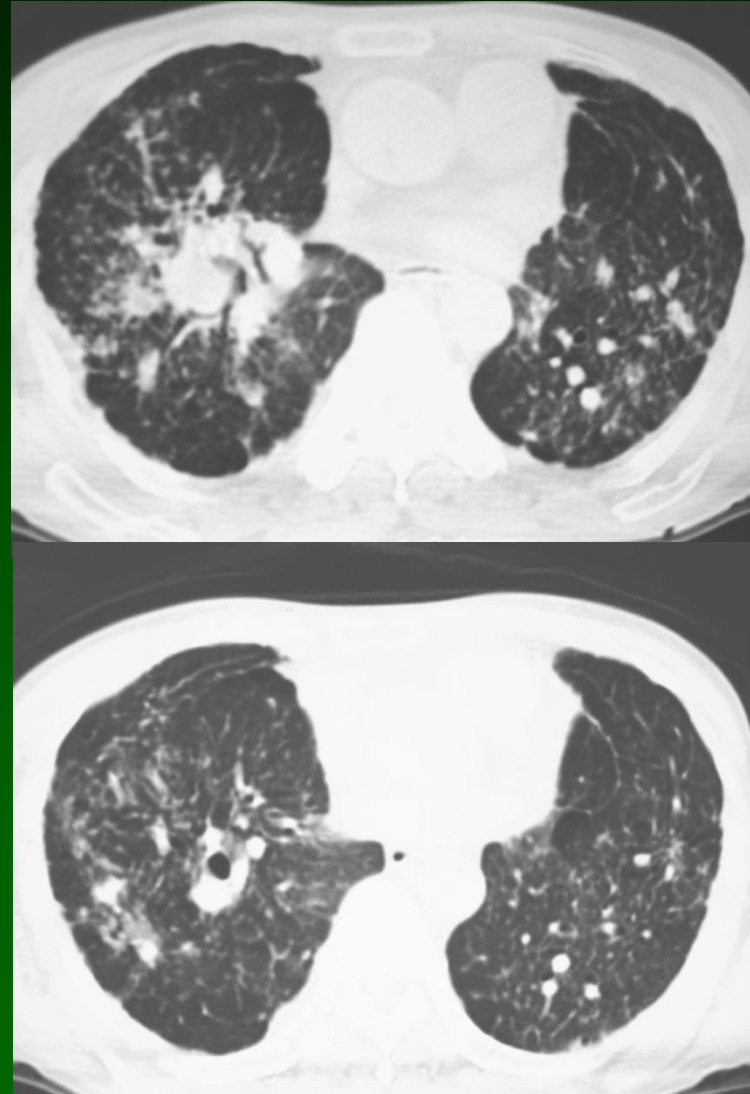
Sarcoidosis

- Bilateral hilar and mediastinal lymphadenopathy with or without parenchymal abnormalities
- Sarcoid granuloma distributed along the lymphatics in the bronchovascular sheath and in the interlobular septa and subpleural regions.
- Nodules, consolidation, fibrosis



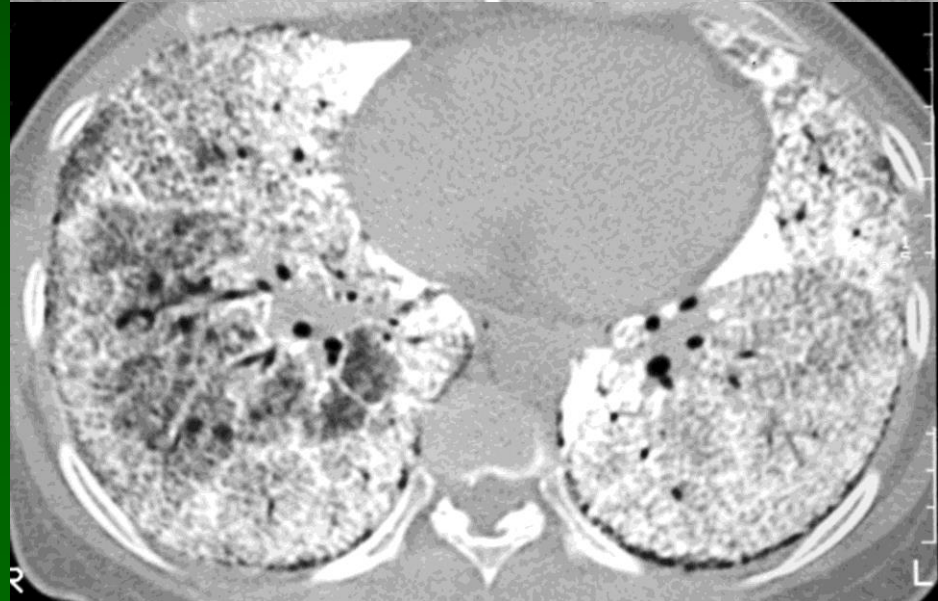
Silicosis/Pneumoconiosis

- Small, well-circumscribed nodules, usually 2-5 mm in diameter, involving mainly the upper and posterior lung zones
- Large opacities: progressive massive fibrosis
- Egg-shell calcification of hilar lymphadenopathy: pathognomonic of silicosis



Pulmonary Alveolar Microlithiasis

- widespread localization of calcispherites or microliths in the alveolar space
- most in adults in Turkey, rare in Taiwan
- calcified miliary or reticular pattern, so called **“sandstorm appearance”**
- subpleural cysts or paraseptal emphysema
- diagnostic images on HRCT



Diffuse fine nodular disease

- Inhalational disease (dust)
 - Silicosis and coal-worker's pneumoconiosis
 - Berylliosis
 - Siderosis
 - Allergic alveolitis
- Eosinophilic granuloma
- Sarcoidosis
- Tuberculosis
- Fungus infections
 - Histoplasmosis, blastomycosis, coccidioidomycosis, aspergillosis, cryptococcosis
- Bacterial infections
 - Nocardiosis, bronchopneumonia (unusual)
- Viral pneumonia
 - varicella

Diffuse fine nodular disease

- Tumor
 - Primary lung cancer
 - Metastasis
 - Thyroid carcinoma, melanoma, other adenocarcinomas (e.g. pancreas)
- Others
 - Bronchiolitis obliterans
 - Alveolar microlithiasis
 - Gaucher's disease
 - Wegener's granulomatosis (rare)

Small nodules

- Nodules > 5 mm in diameter
 - Smooth margins - - - Metastatic disease
 - Rheumatoid nodules
 - Wegener's granulomatosis
 - Lymphoma
 - Irregular margins - - Septic emboli
 - Bronchioloalveolar carcinoma
 - Tuberculosis
 - Lymphoma

Tuberculosis (DM)



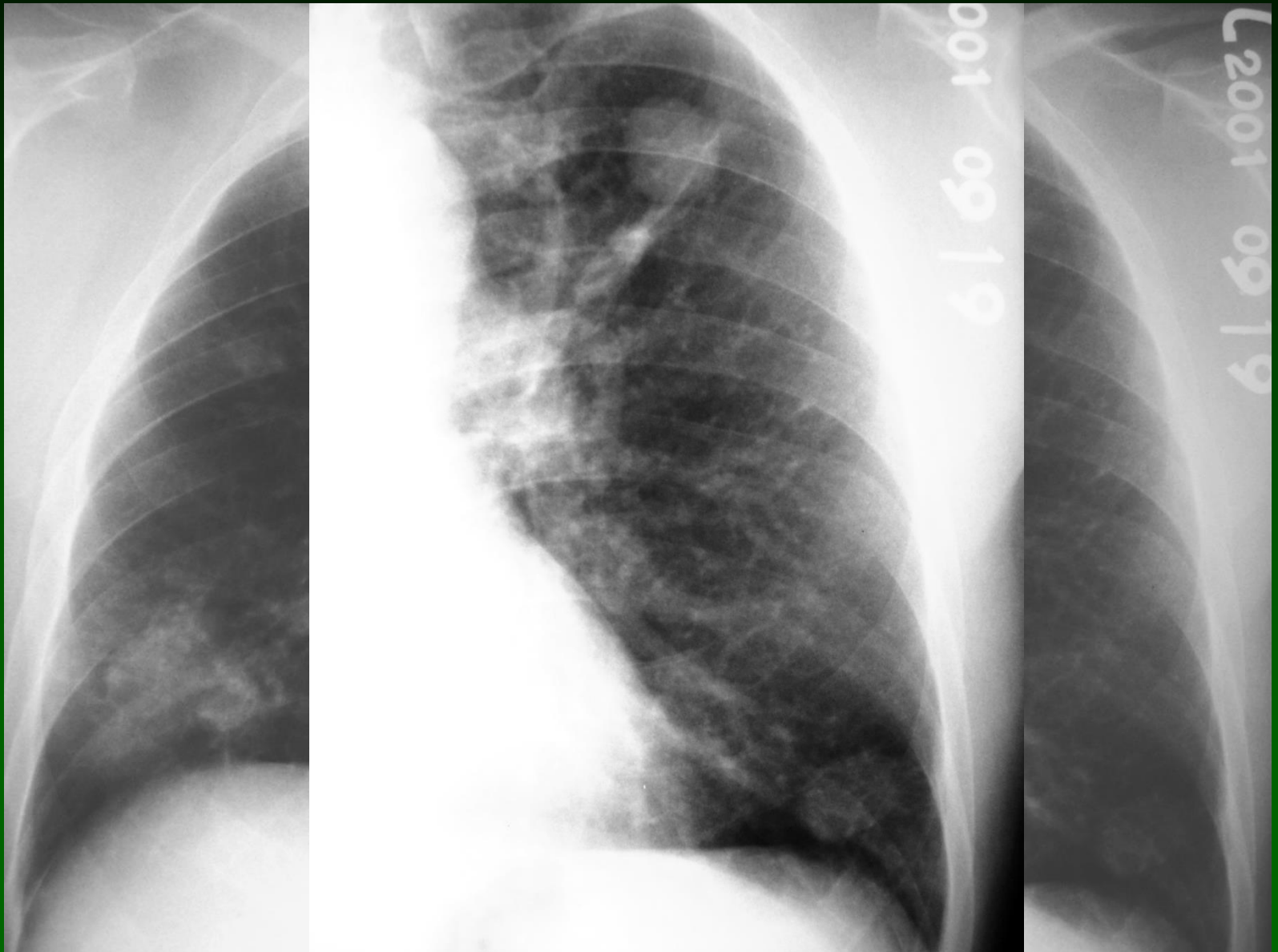
Clues to Interpretation of Multiple Nodular Lesions

- Localization and distribution
 - Metastasis vs. tuberculoma
- Margin
- Calcification
 - Tuberculosis, osteogenic sarcoma
- Cavitation
 - Septic lung, WG
- Changes in size and number
- Hilar shadow
- Pleural effusion
- Same or different etiology

Metastatic malignancy



Tuberculosis



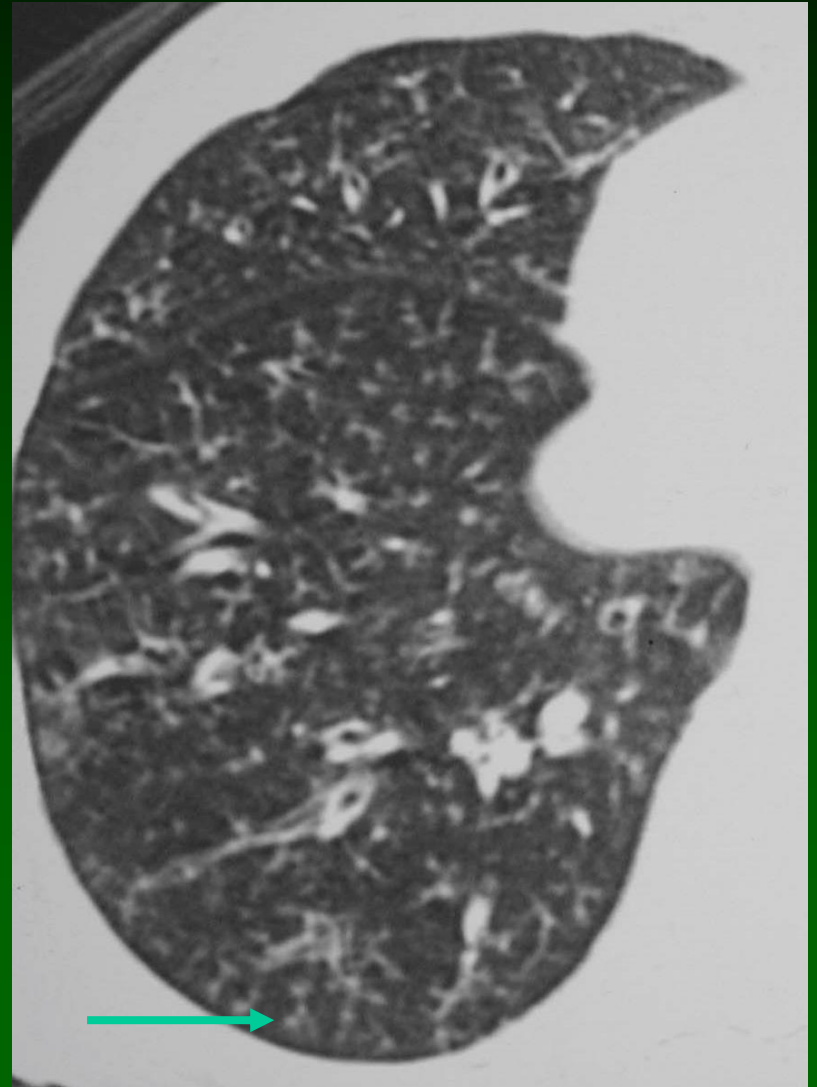
Tree-in-bud (TIB) Pattern

- Centrilobular nodular and branching linear opacities, representing dilated and impacted (mucus or pus) centrilobular bronchioles
 - Infection
 - Allergic bronchopulmonary aspergillosis
 - Cystic fibrosis
 - Aspiration
 - Diffuse panbronchiolitis
 - Obliterative bronchiolitis
 - Asthma

Bronchiolar diseases with TIB pattern

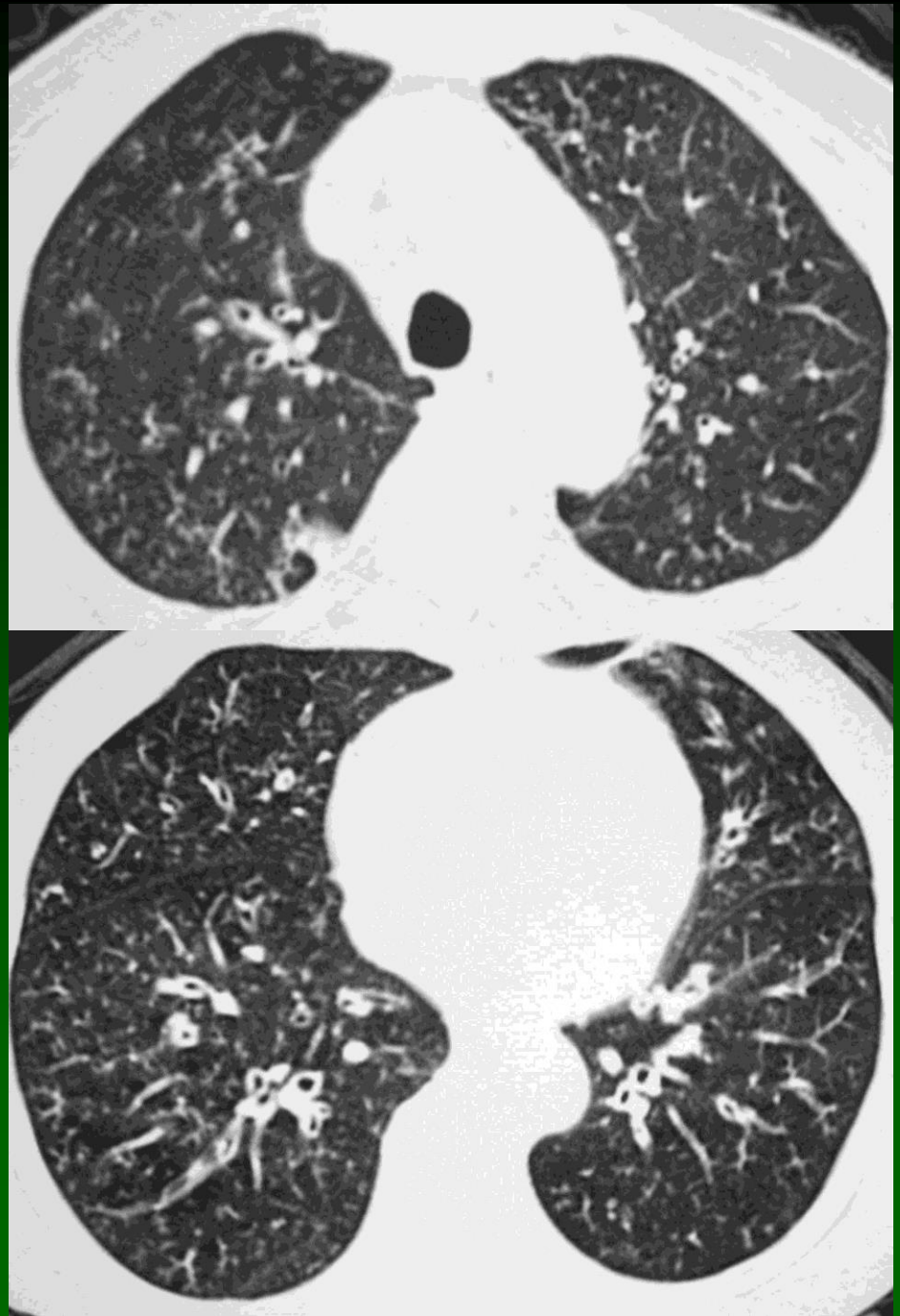
Diffuse panbronchiolitis

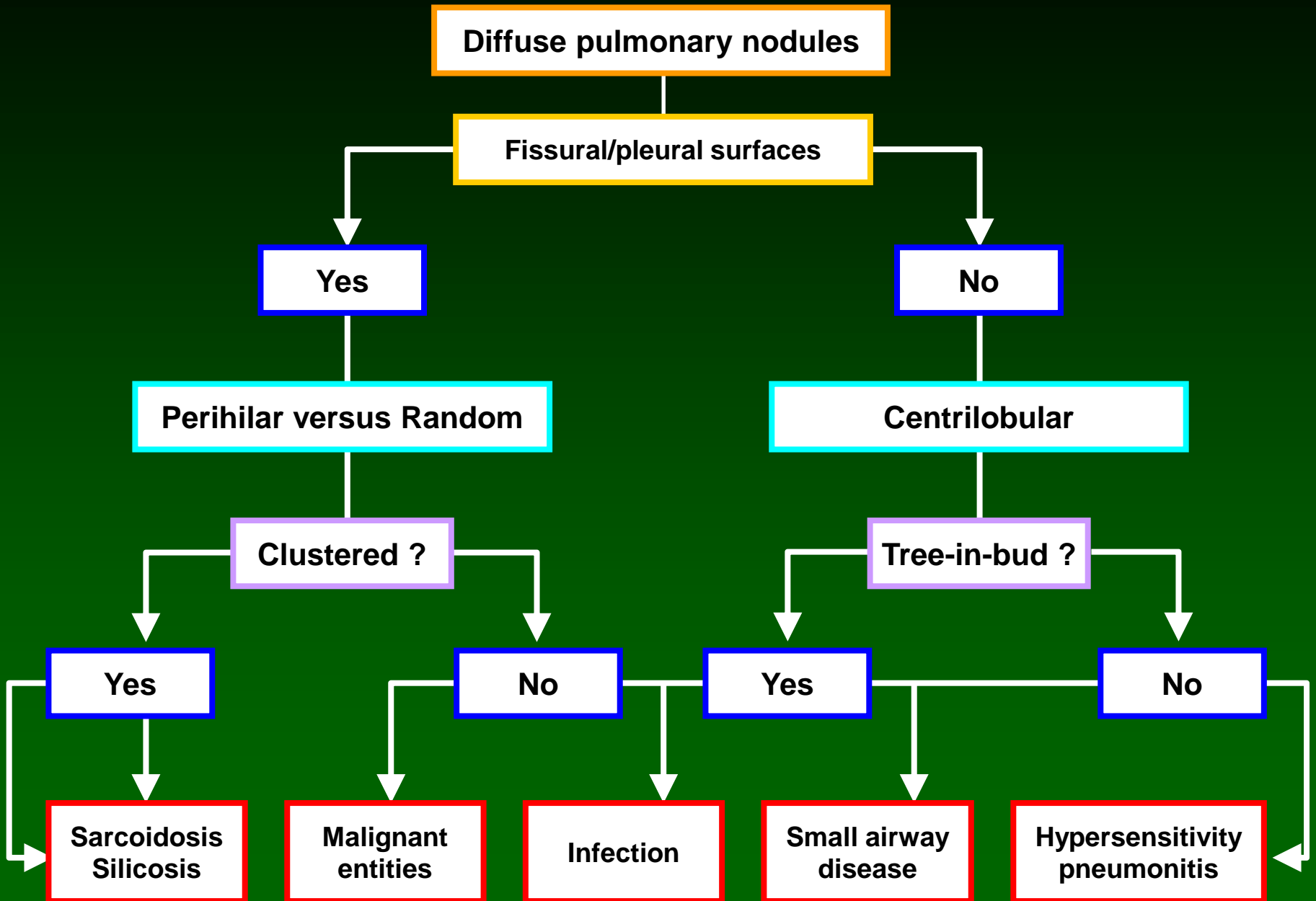
- Subacute development of airway obstruction
- Centrilobular peribronchiolar infiltrates of acute and chronic inflammatory cells
- May associated with bronchiectasis
- Increased lung volume
- Air trapping on expiration



DPB

- Chronic cough, dyspnea, sputum
- PFT: mild-moderate airway obstruction
- CXR: ill-defined nodular infiltration, usually basilar
- Chronic inflammation at respiratory bronchioles, alveolar ducts and alveoli
- Diffuse “TIB” pattern, predominantly lung bases
- Responsive to Erythromycin
- Usually progressive, 5 and 10-year survival rate- 60%, 30%



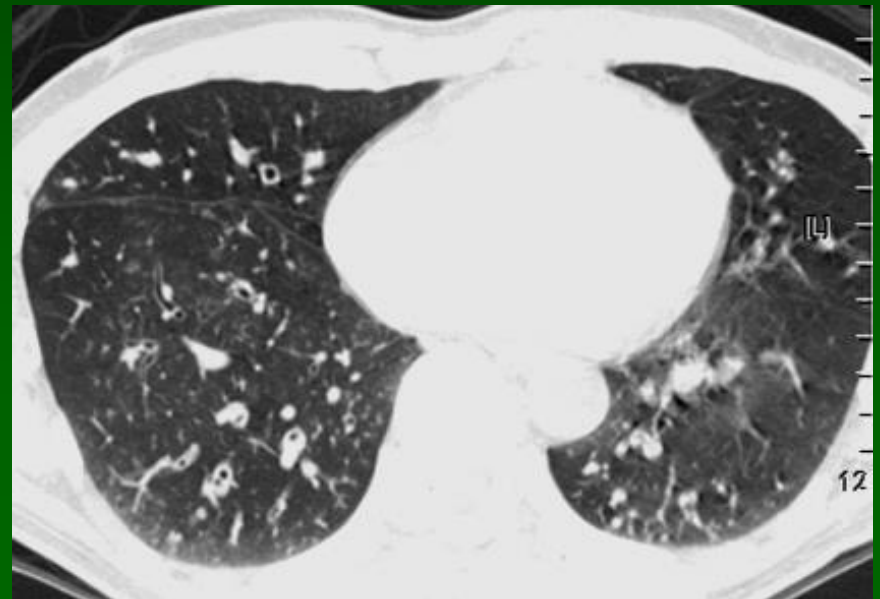
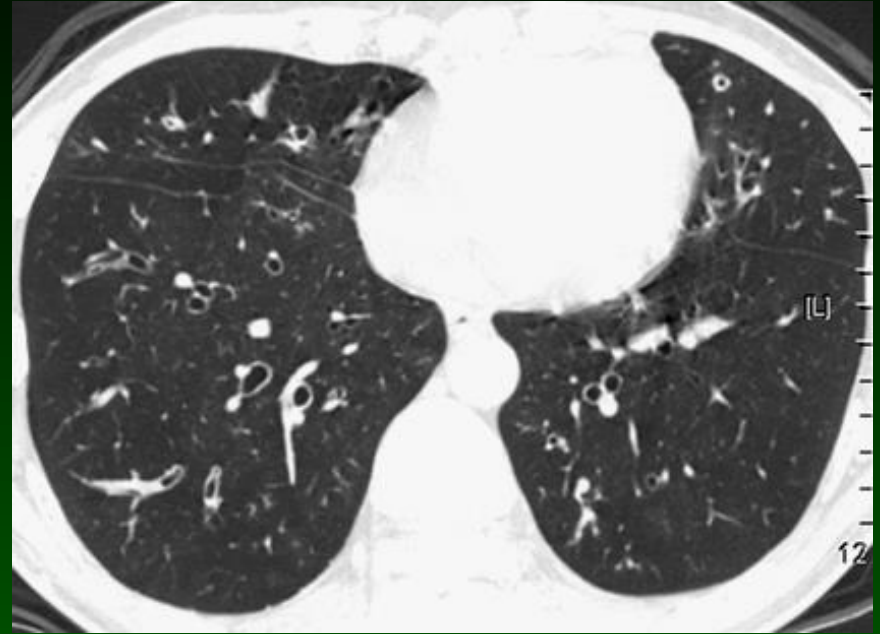


Cystic lung disease

Diagnosis	Comments
Langerhans histiocytosis	Cysts with unusual shapes, larger and more in apices, spared CP angles, small nodules (±)
LAM	Cysts usually round and relatively uniform in size and shape; diffuse lung involvement, exclusively in women
LIP	Less numerous than in LAM and EG; nodules or GGO may be associated
Honeycombing	Subpleural predominance; multiple layers at the pleural surface; cysts care walls; findings of fibrosis
Centrilobular emphysema	Lucencies small, centrilobular, visible walls absent or inconspicuous; upper lobe distribution
Bullae	Subpleural distribution in most cases; single layer at the pleural surface; centrilobular emphysema
Pneumatocele	Scattered; patchy distribution; limited in number; findings of pneumonia
Cystic bronchiectasis	Clustered or parahilar distribution; air-fluid levels; branching lucencies, signet ring sign
Cavitary nodules	Wall usually thick and irregular

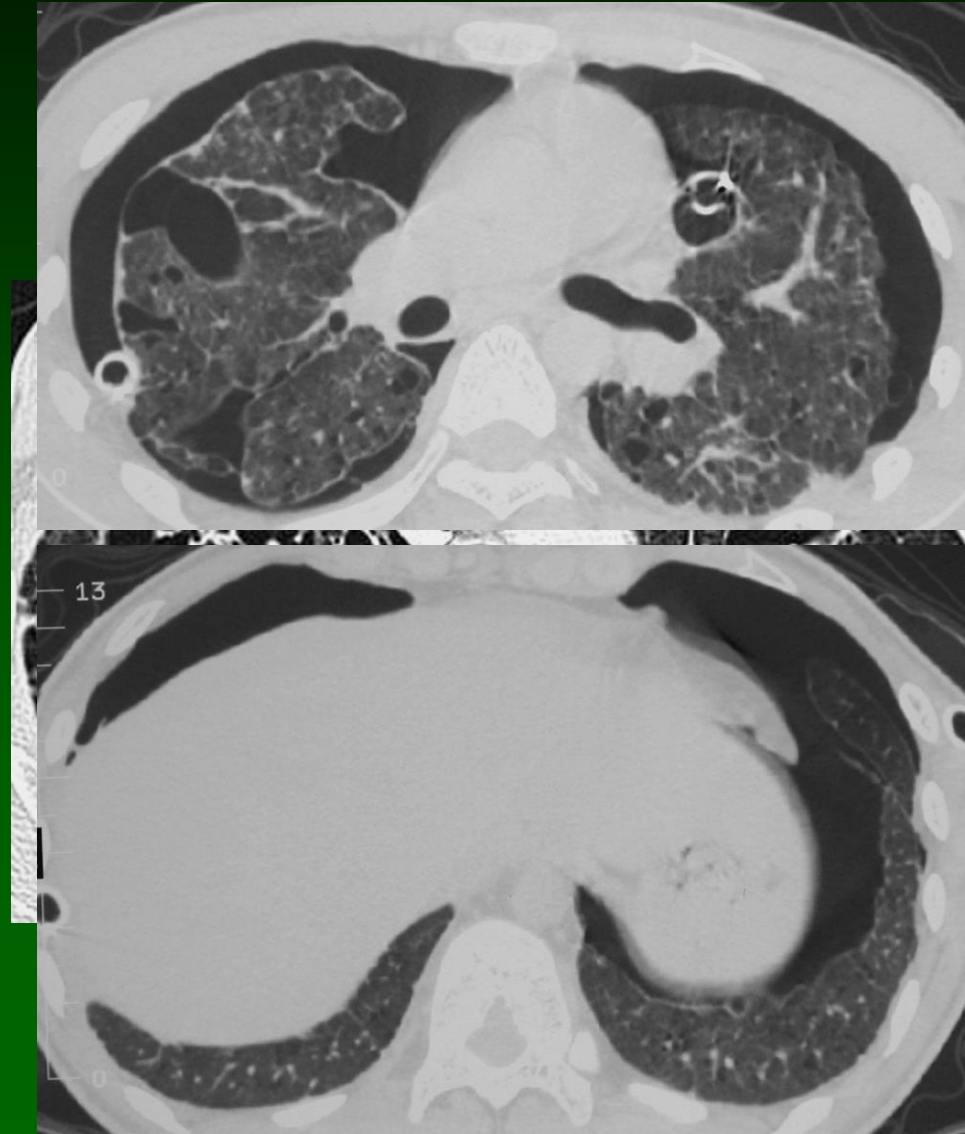
Bronchiectasis

- Signet ring sign
- Kang's study shows 85% pathologic evidence of bronchiolitis in 47 resected lobes with documented bronchiectasis
- HRCT findings: mosaic perfusion, centrilobular nodular or branching opacities, or both (TIB)
- Bronchiolar disease may precede or even lead to the development of bronchiectasis



Pulmonary Langerhans Cell Histiocytosis

- Thin-wall cysts, some confluent or with bizarre shapes, usually smaller than 1 cm
- Thick wall cysts
- Nodules, usually smaller than 1-5 mm
- Upper lobe predominance and CP angle sparing



Lymphangioleiomyomatosis

- Numerous thin-walled cysts, surrounded by normal lung parenchyma
- 2 mm-5 cm in size
- No lung zone is spared
- Cysts may enlarge and wall may thicken



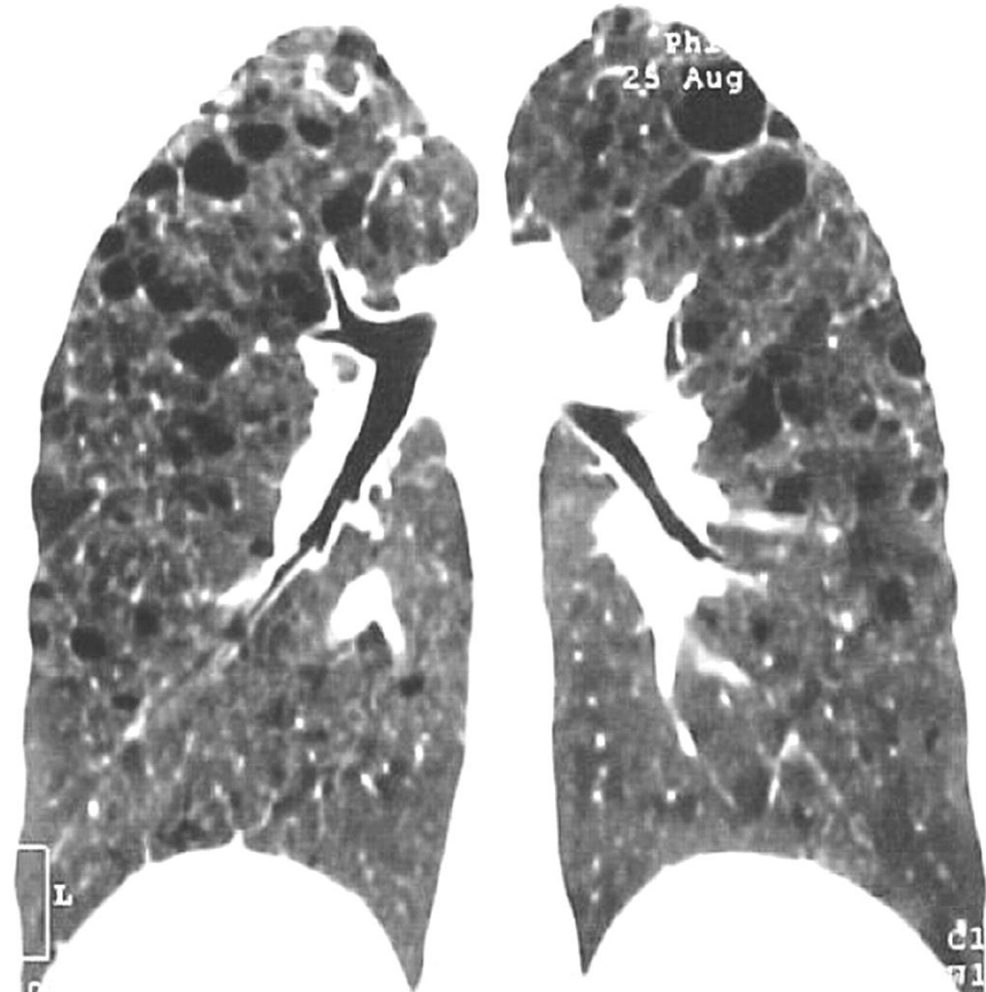
PLCH vs. LAM

- Upper lung predominance
- Irregular and complex cyst
- Nodules
- Lung base (CP angle) spare
- Pleural effusion (LAM)

LAM



PLCH



Cystic Metastases

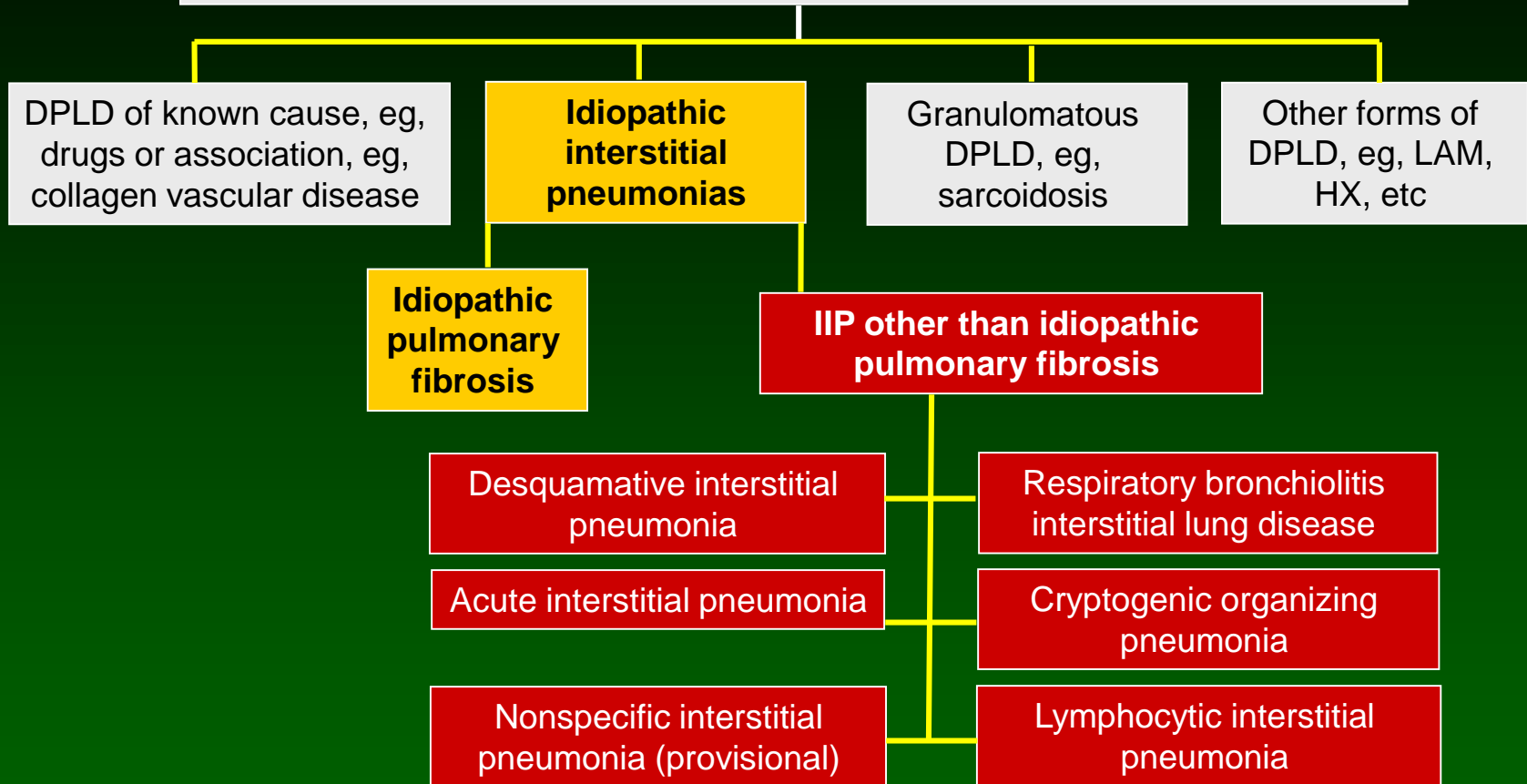


Centrilobular Pulmonary Emphysema

- Most common form
- Cigarette smoking
- Upper lobe predominance
- Central portion of 2nd pulmonary lobule
- Confluent when disease progressive
- Vessel attenuation when disease progresses

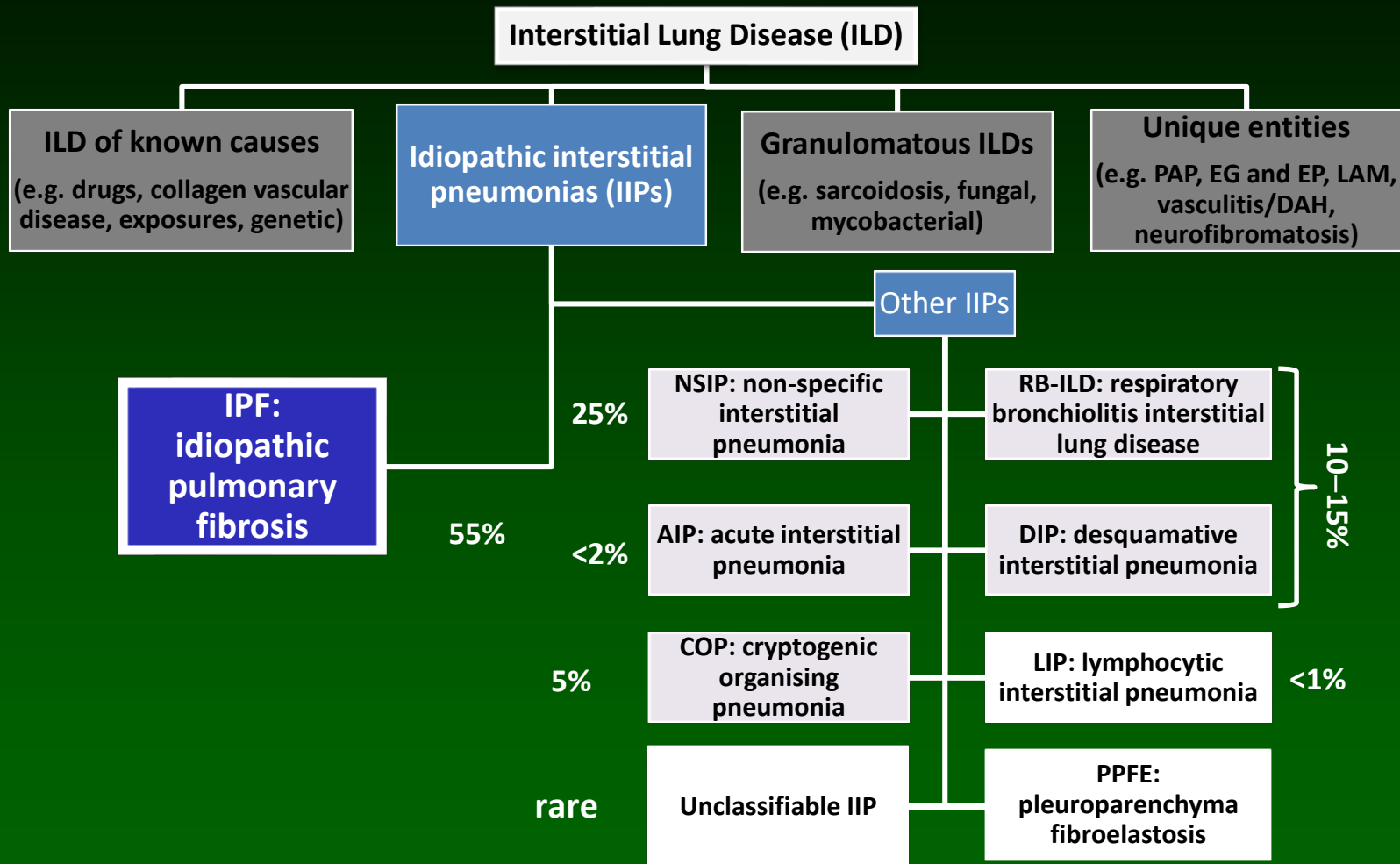


Diffuse Parenchymal Lung Disease (DPLD)



ATS/ERS IIP statement
Am J Respir Crit Care Med 2002

Classification of Interstitial Lung Diseases (ILDs)



2013 ATS/ERS update of international multidisciplinary classification of the IIP

PPFE = pleuroparenchymal fibroelastosis, EG = eosinophilic granuloma, EP = eosinophilic pneumonia,
LAM = lymphangioleiomyomatosis, PAP = pulmonary alveolar proteinosis,

Travis WD. *Am J Respir Crit Care Med* 2013;188:733, Ryerson CJ and Collard HR. *Curr Opin Pulm* 2013;19:453-459.

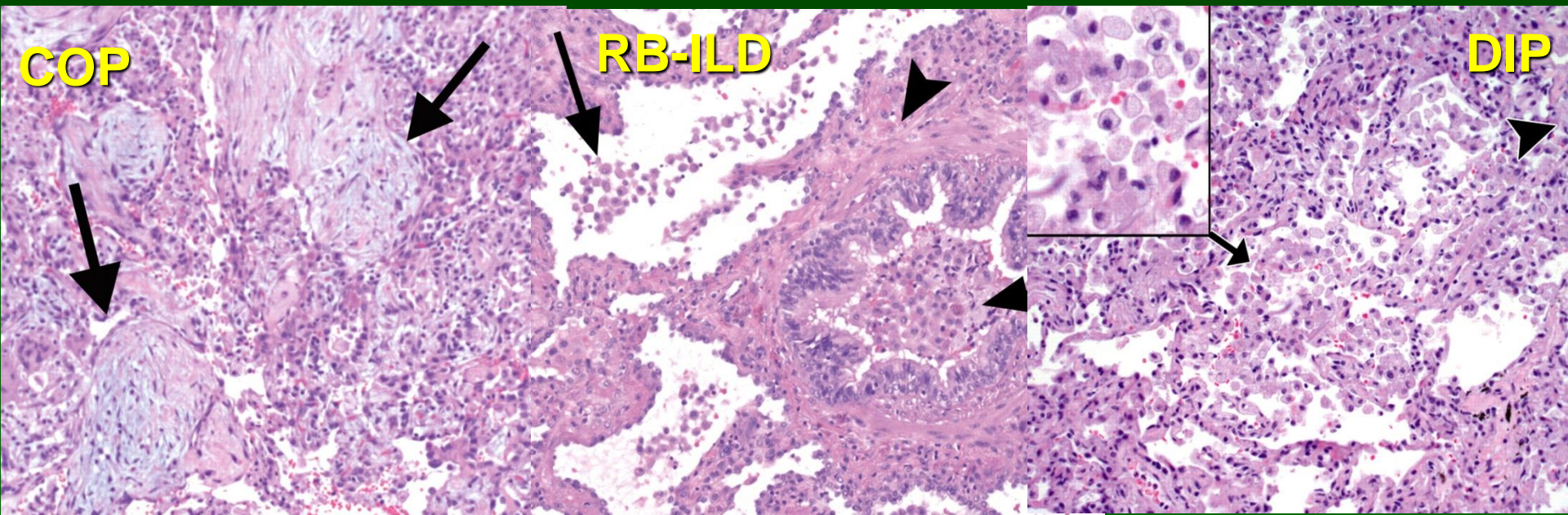
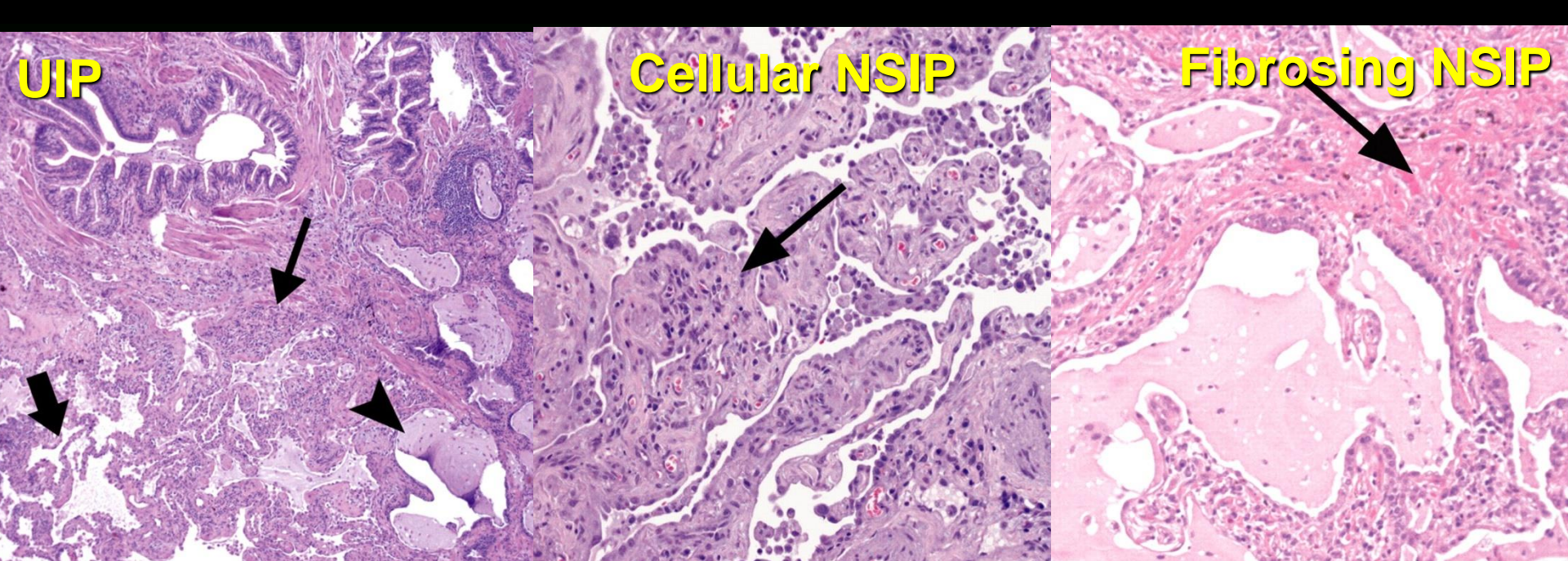
CATEGORIZATION OF MAJOR IDIOPATHIC INTERSTITIAL PNEUMONIAS

Category	Clinical–Radiologic–Pathologic Diagnoses	Associated Radiologic and/or Pathologic–Morphologic Patterns
Chronic fibrosing IP	Idiopathic pulmonary fibrosis Idiopathic nonspecific interstitial pneumonia	Usual interstitial pneumonia Nonspecific interstitial pneumonia
Smoking-related IP*	Respiratory bronchiolitis-interstitial lung disease Desquamative interstitial pneumonia	Respiratory bronchiolitis Desquamative interstitial pneumonia
Acute/subacute IP	Cryptogenic organizing pneumonia Acute interstitial pneumonia	Organizing pneumonia Diffuse alveolar damage

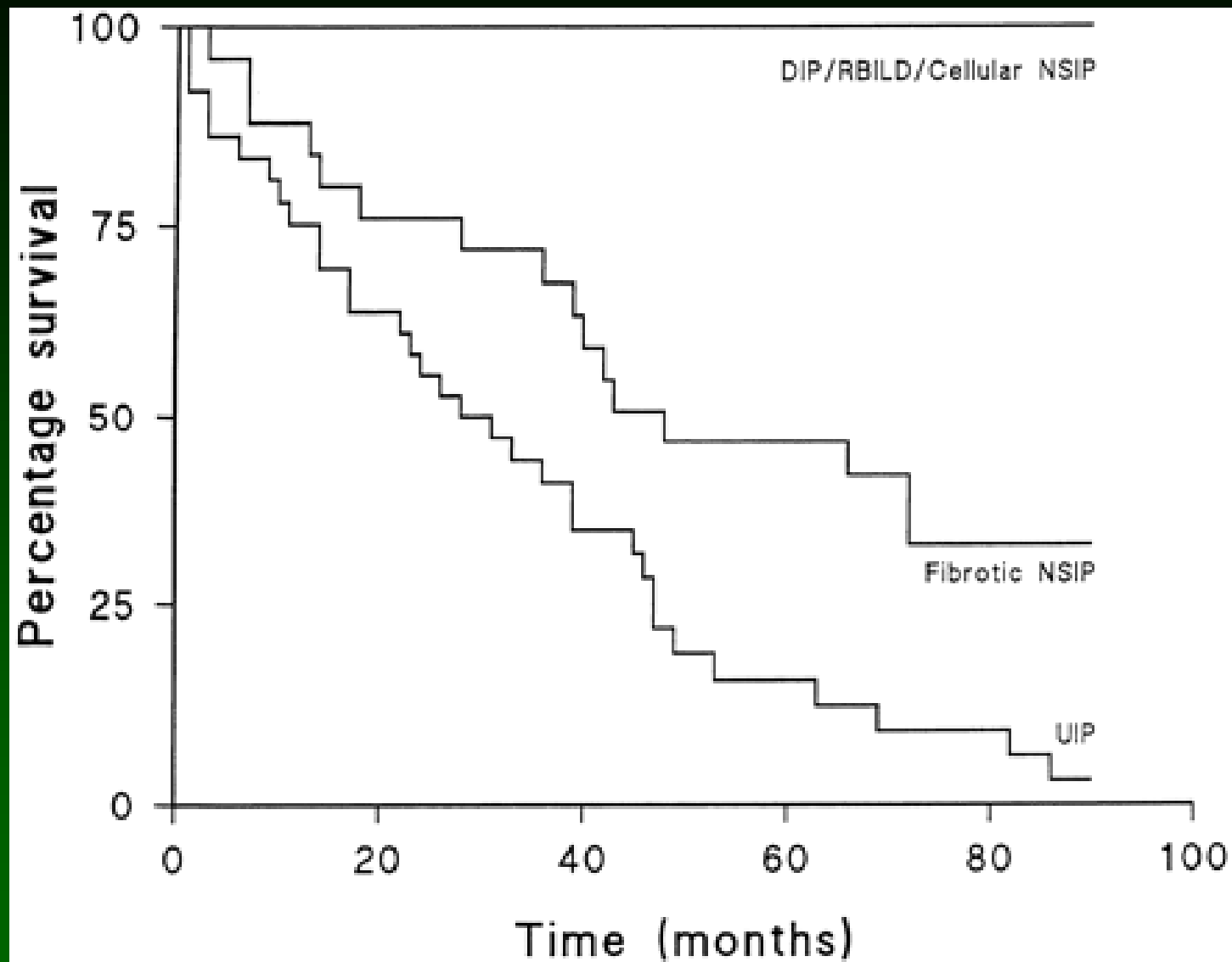
Definition of abbreviation: IP = interstitial pneumonia.

*Desquamative interstitial pneumonia can occasionally occur in nonsmokers.

Morphologic Pattern	Histopathologic Features	Distribution at CT	High-Resolution CT Features
UIP	Spatial and temporal heterogeneity, architectural distortion, fibroblastic foci	Apicobasal gradient, subpleural	Macrocystic honeycombing, reticular opacities, traction bronchiectasis, architectural distortion, focal ground-glass opacity
NSIP	Spatial and temporal homogeneity; cellular pattern shows mild to moderate interstitial chronic inflammation; fibrosing pattern shows dense or loose interstitial fibrosis	No obvious gradient, subpleural, symmetric	Ground-glass opacities, irregular linear or reticular opacities, micronodules, consolidation, microcystic honeycombing
COP	Patchy distribution of intraluminal organizing fibrosis in distal airspaces, preservation of lung architecture, uniform temporal appearance	Patchy, peripheral or peribronchial, basal predominance, sometimes sparing of subpleural space, migration tendency	Airspace consolidation, mild bronchial dilatation, ground-glass opacities, large nodules (rare)
RB-ILD	Bronchiolocentric accumulation of alveolar macrophages containing brown particles, mild bronchiolar fibrosis	Diffuse or upper lung predominance	Centrilobular nodules, patchy ground-glass opacities, bronchial wall thickening
DIP	Diffuse accumulation of macrophages in distal airspaces, mild interstitial fibrosis, mild chronic inflammation	Apicobasal gradient, peripheral predominance	Ground-glass opacities, irregular linear or reticular opacities, sometimes cysts
LIP	Diffuse infiltration of alveolar septa by lymphoid cells, lymphoid hyperplasia frequent	Basilar predominance or diffuse	Ground-glass opacities, perivascular cysts, septal thickening, centrilobular nodules
AIP	Diffuse alveolar damage; exudative phase shows hyaline membranes, diffuse alveolar infiltration by lymphocytes; organizing phase shows alveolar wall thickening due to fibrosis, pneumocyte hyperplasia	Lower lung predominance, symmetric, bilateral	Exudative phase shows ground-glass opacities, airspace consolidation; organizing phase shows bronchial dilatation, architectural distortion



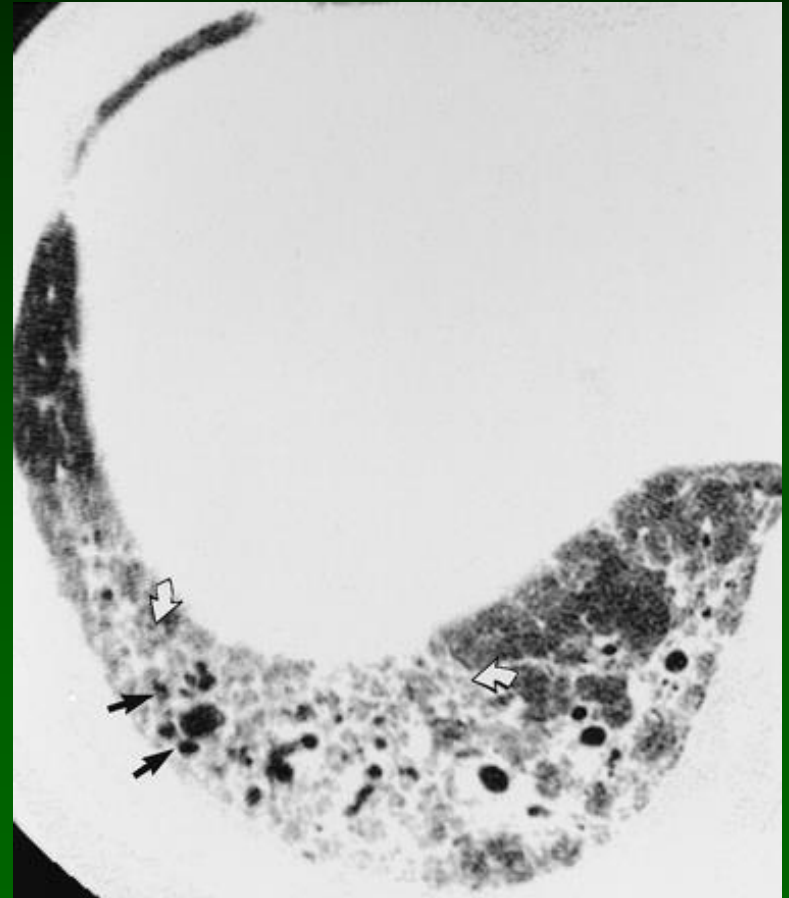
(Muller-Mang et al. RadioGraphics 2007;27:595)



(Nicholson et al. AJRCCM 2000;162:2213-7)

UIP/IPF

- Reticular pattern predominantly subpleural;
- usually lower-zone predominance;
- may show areas of GGO or consolidation, but intralobular reticular opacities (white arrow) predominate;
- may show honeycombing (black arrow)



(Radiology. 1999;211:555-560.)

High-resolution computed tomography criteria for UIP pattern

Probable UIP

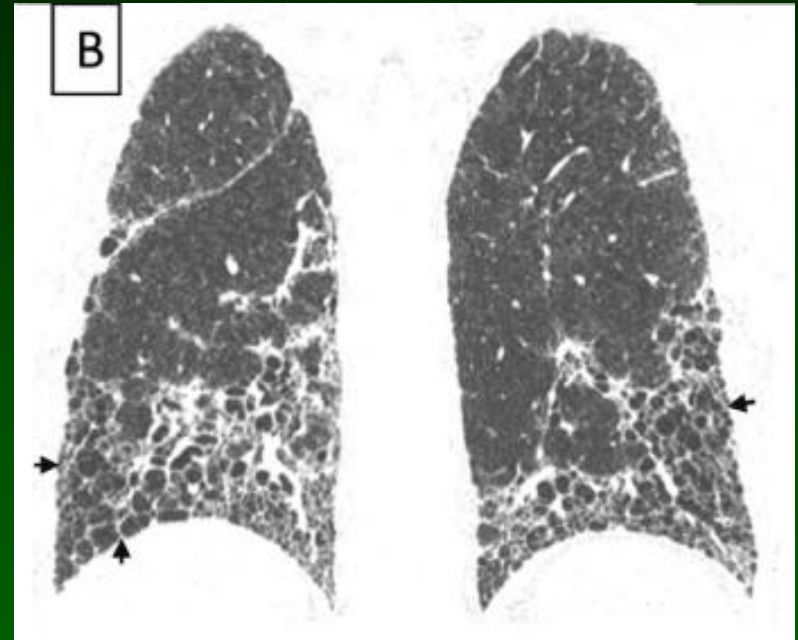
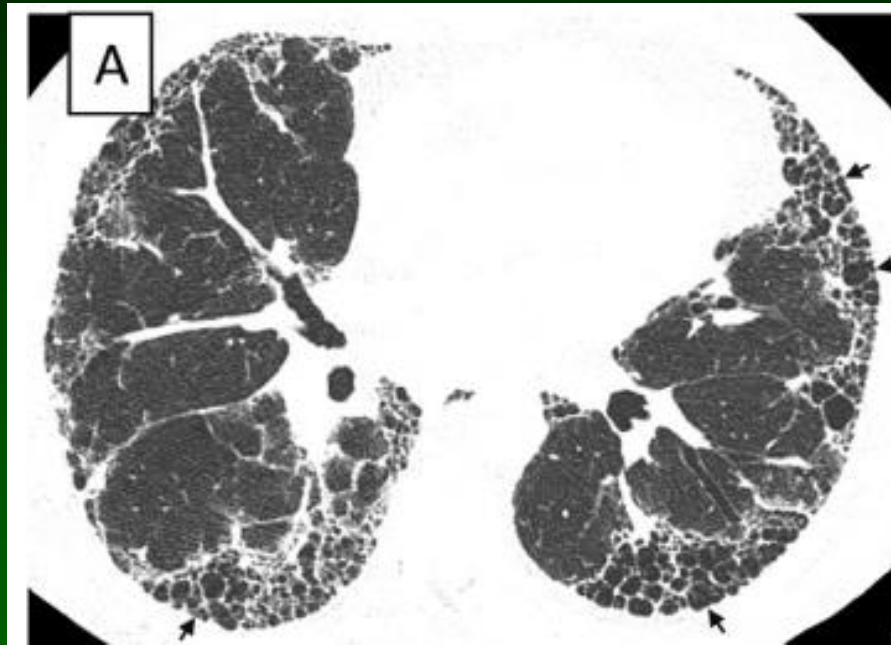
Alternative diagnosis

UIP Pattern (All Four Features)	Possible UIP Pattern (All Three Features)	Inconsistent with UIP Pattern (any of the Seven Features)
<ul style="list-style-type: none"> ■ Subpleural, basal predominance ■ Reticular abnormality ■ Honeycombing with or without traction bronchiectasis ■ Absence of features listed as inconsistent with UP <p>Variant distribution : occasional diffuse, maybe asymmetrical Superimposed CT features: mild GGO, reticular pattern, pulmonary ossification</p>	<ul style="list-style-type: none"> ■ Subpleural, basal predominance ■ Reticular abnormality ■ Absence of features listed as inconsistent with UIP pattern (see third column) 	<ul style="list-style-type: none"> ■ Upper or mid-lung predominance ■ Peribronchovascular predominance ■ Extensive ground glass abnormality (extent >reticular abnormality) ■ Profuse micronodules (bilateral, predominantly upper lobes) ■ Discrete cysts (multiple, bilateral, away from areas of honeycombing) ■ Diffuse mosaic attenuation/air-trapping (bilateral in three or more lobes) ■ Consolidation in bronchopulmonary segment(s)/lobe(s)

Respiratory Research 2013

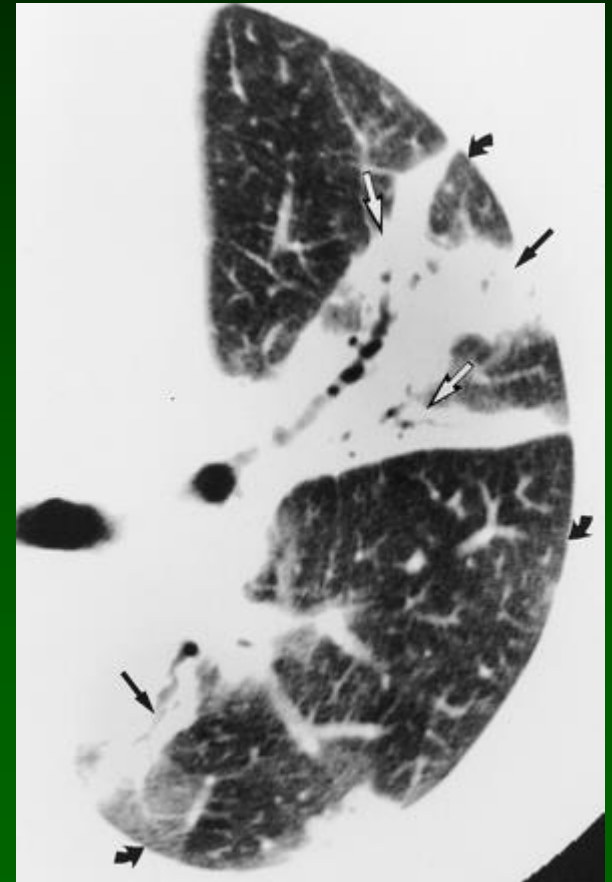
ATS/ERS/JRS/ALAT 2011 revised diagnostic criteria

ATS/ERS/JRS/ALAT 2018 revised diagnostic criteria



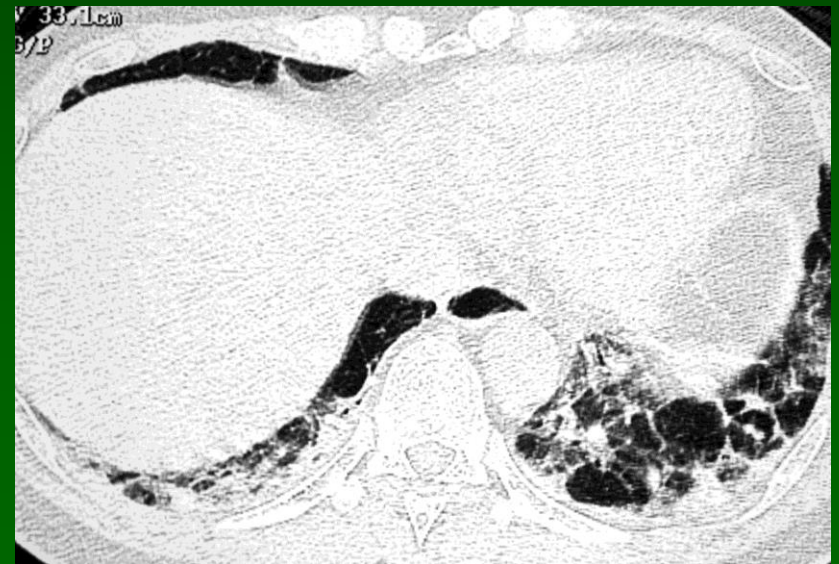
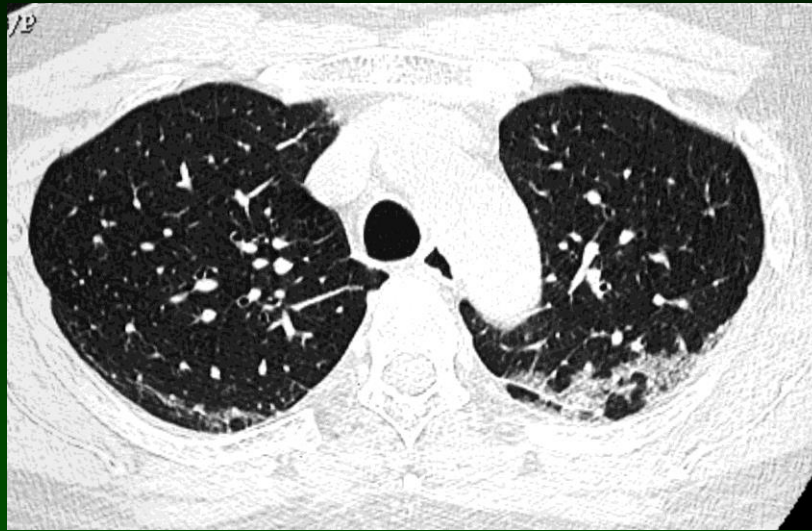
NSIP

- Patchy areas of ground-glass attenuation (curved arrow);
- Less apico-basal gradient;
- may show air-space consolidation (straight arrows) or intralobular reticular opacities
- Subpleural sparing

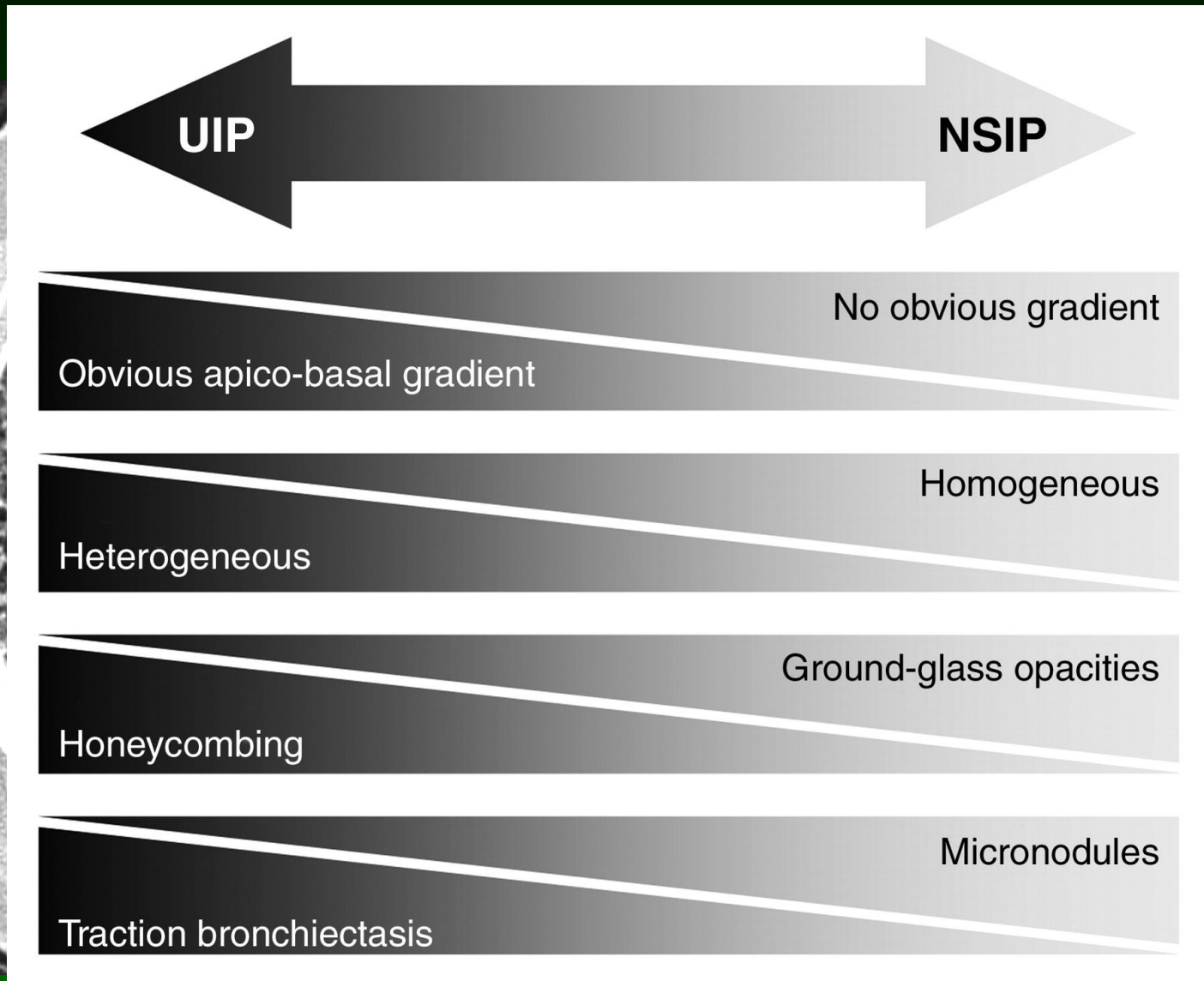


(Radiology. 1999;211:555-560)

NSIP



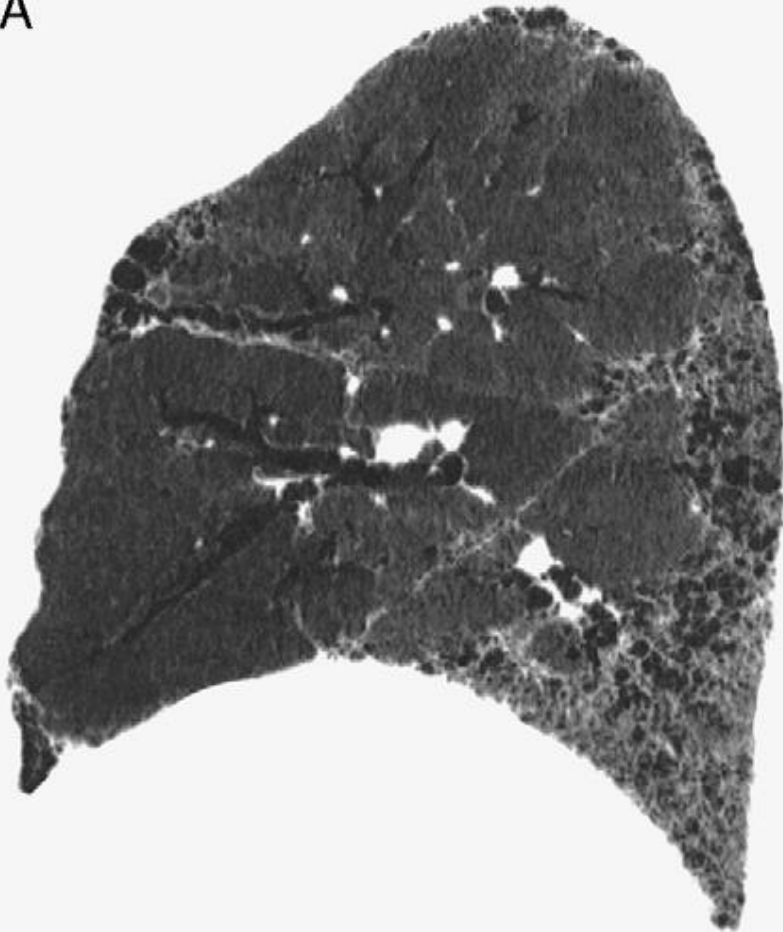
UIP vs. NSIP



(Muller-Mang et al. RadioGraphics 2007;27:595)

UIP

A



NSIP

B



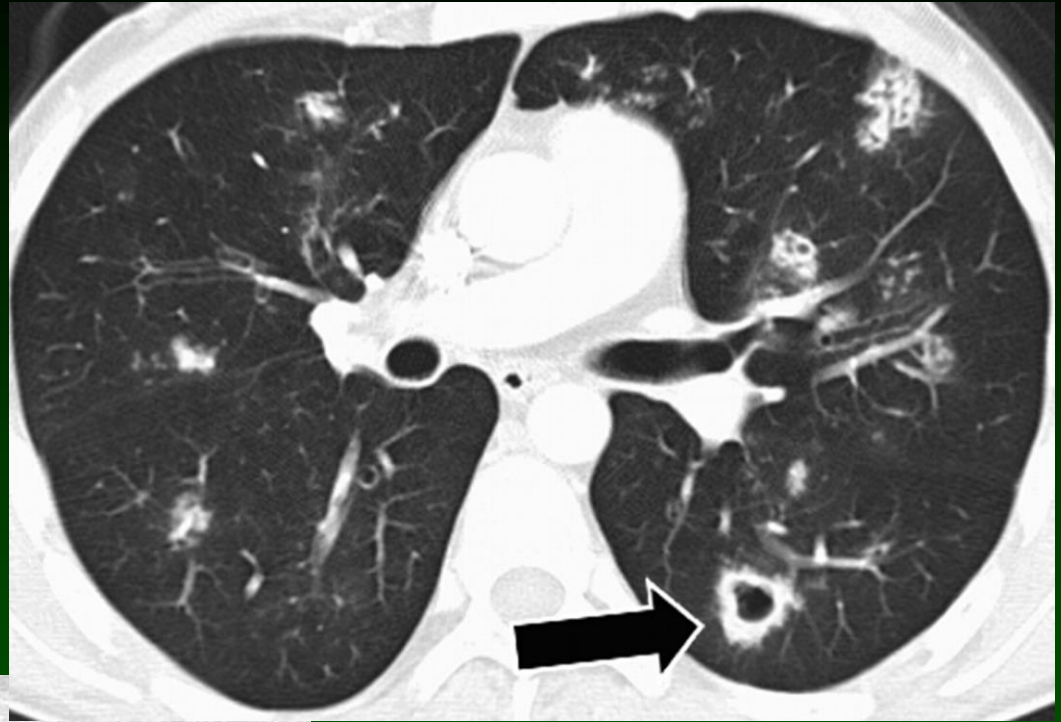
COP/BOOP

- Patchy subpleural and/or peribronchovascular air-space **consolidation** (open arrows)
- often associated with areas of ground-glass attenuation (curved arrows);
- may show **centrilobular nodules** (solid straight arrow) or bronchial wall thickening



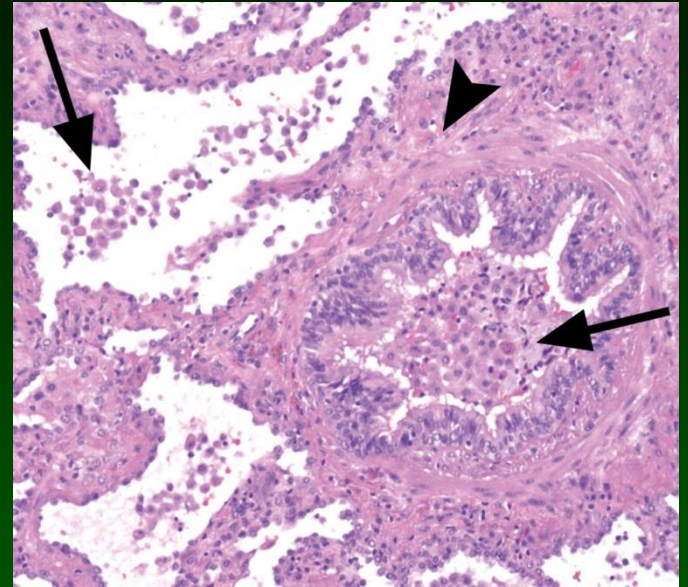
(Radiology. 1999;211:555-560)

Atypical appearance of COP



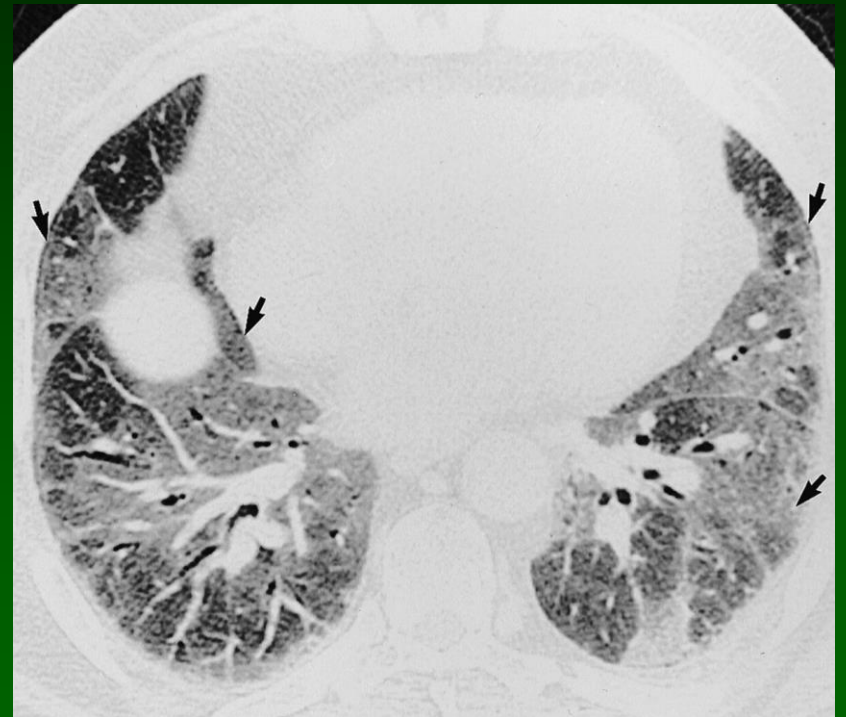
RB-ILD

- Smoking-related
- Upper-lung predominance
- Centrilobular nodules in combination with GGO and bronchial wall thickening
- Coexisting with centrilobular emphysema is common



DIP

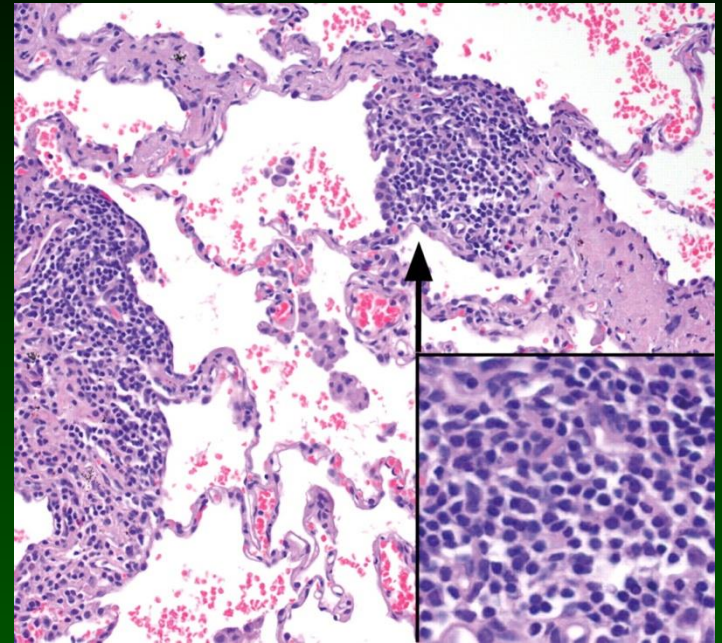
- Smoking-related
- Patchy areas of GGO (arrows) predominantly subpleural; usually lower zone predominance; may show intralobular reticular opacities or mild honeycombing



(Radiology. 1999;211:555-560.)

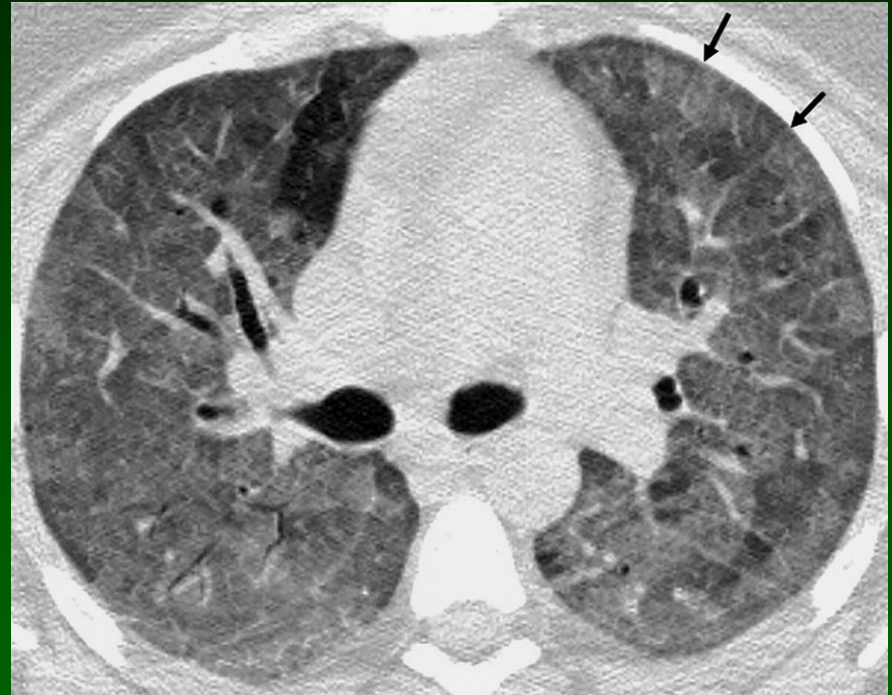
LIP

- More common with systemic disease (Sjögren syndrome, HIV infections, immunodeficiency, Castleman syndrome)
- GGO with diffuse or lower-zone predominance (arrow)
- Thin-wall perivascular cyst (arrowheads)
- Centrilobular nodules and septal thickening are occasionally seen



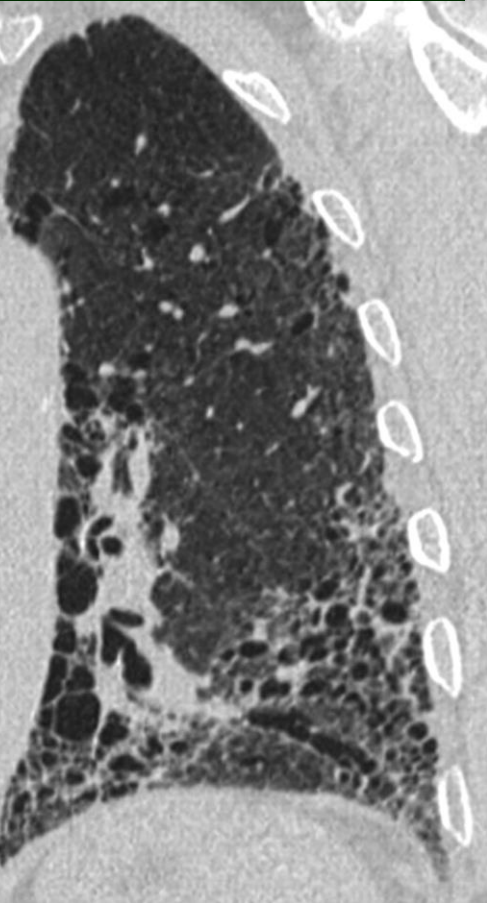
AIP

- Histology: DAD
- Image: similar to ARDS, but more likely to be symmetric, bilateral and lower lobe predominance
- GGO to consolidation to architectural distortion

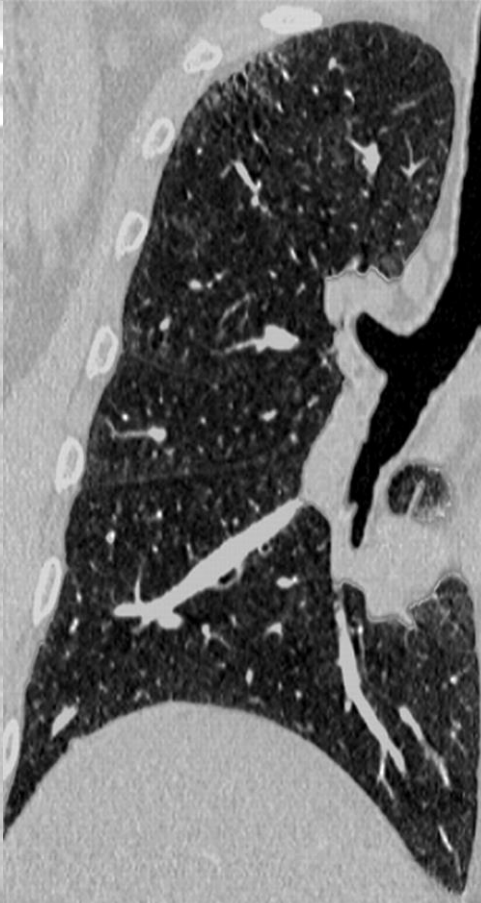


Spatial distribution of IIP

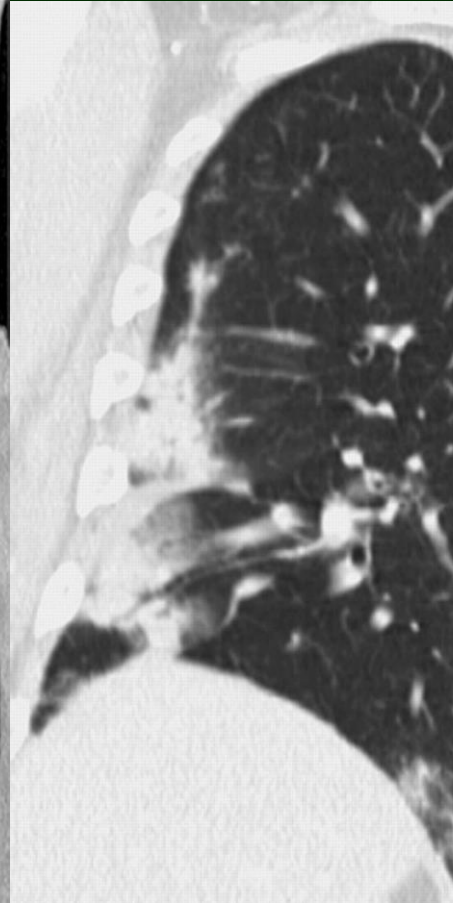
UIP



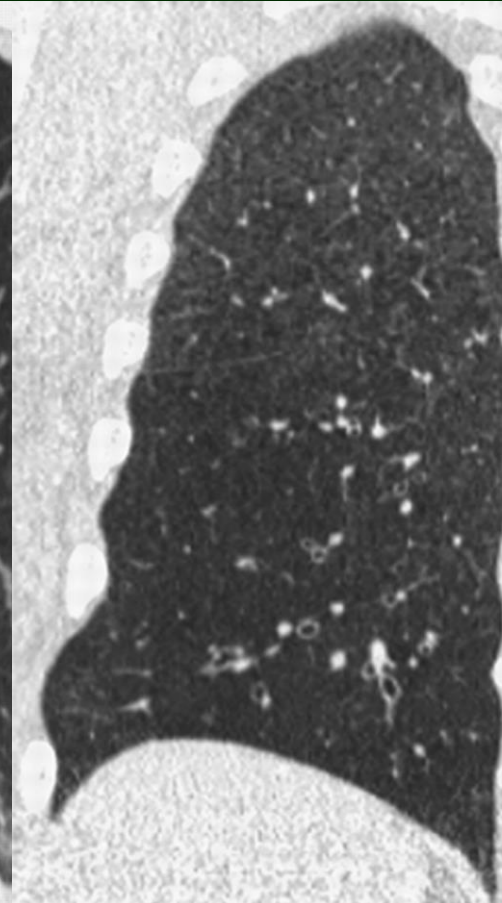
NSIP



COP



RBILD



(Muller-Mang et al. RadioGraphics 2007;27:595)

RARE IIP

Idiopathic LIP

Idiopathic pleuroparenchymal fibroelastosis
(PPFE)

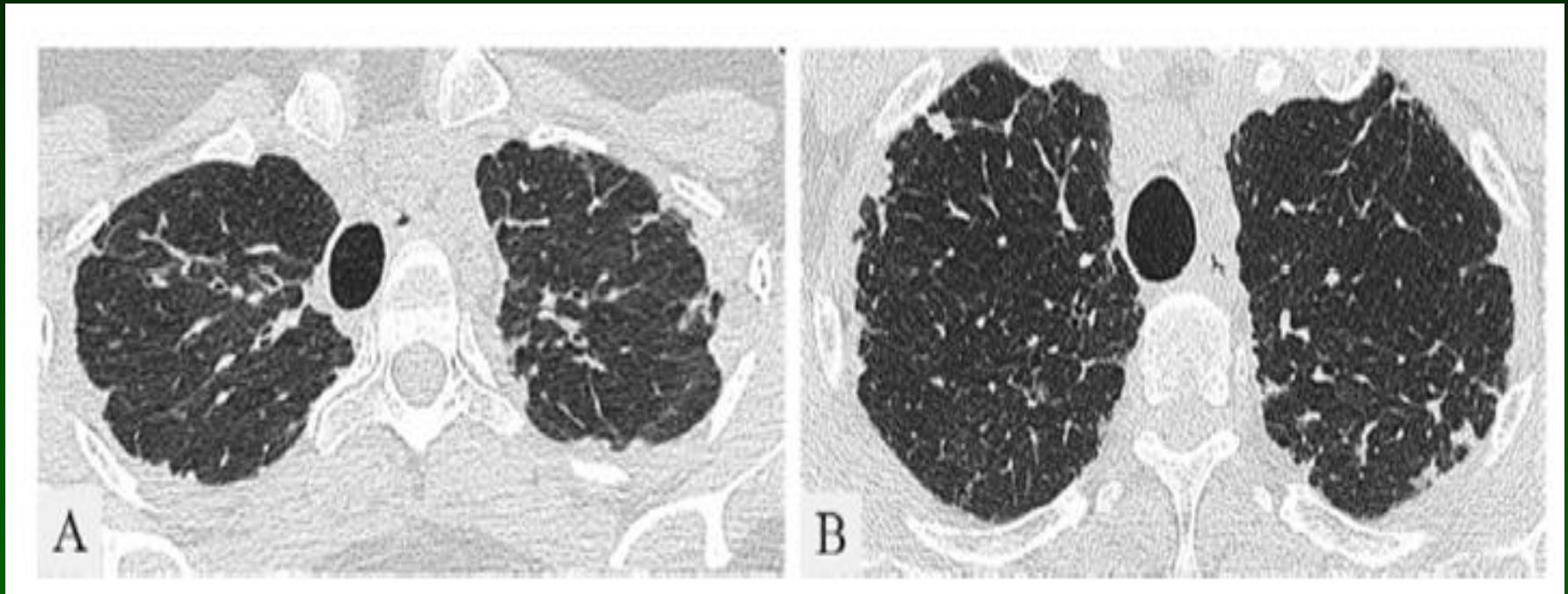
Acute fibrinoid organizing pneumonia(AFOP)

Group of bronchiolocentric patterns

CT findings in pleuropulmonary fibroelastosis.

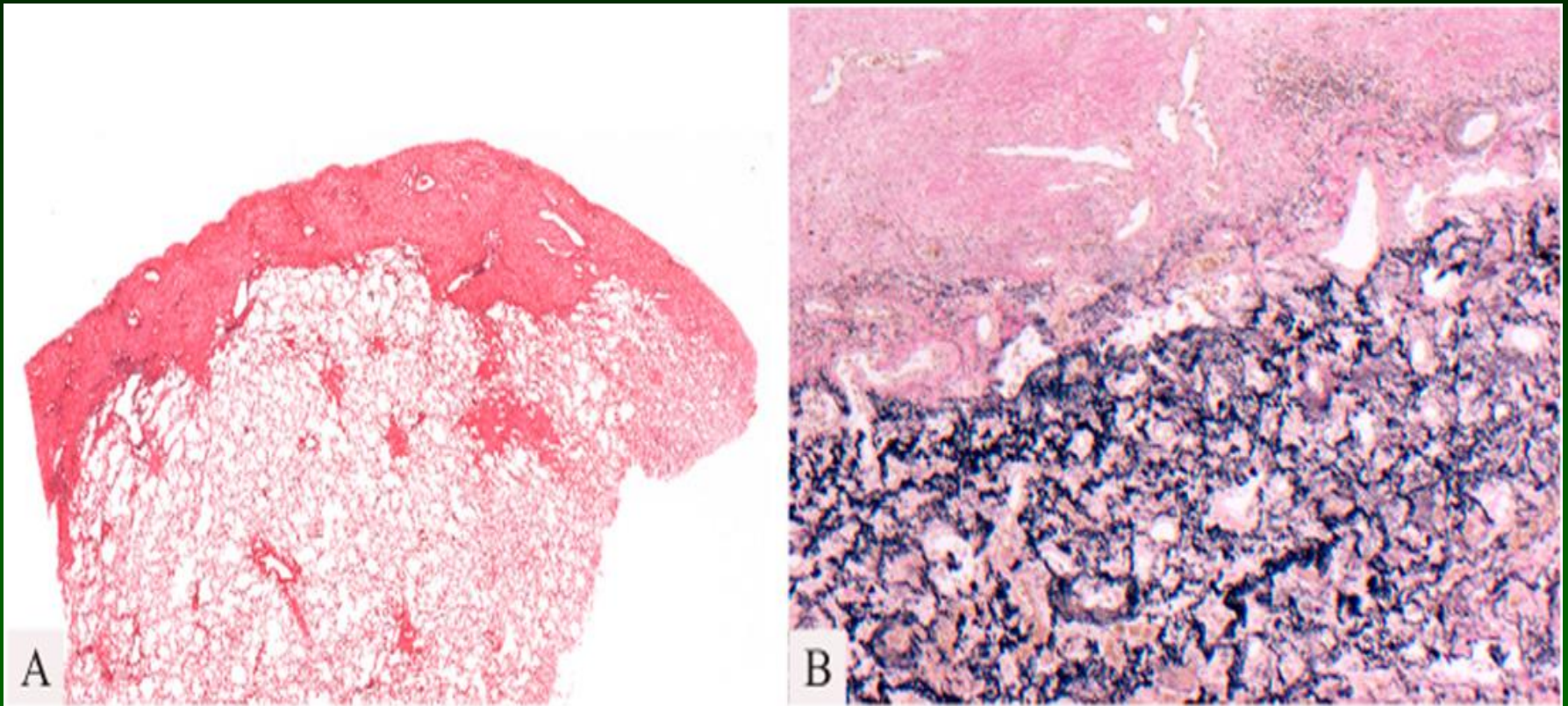
- Dense subpleural areas of airspace consolidation with traction bronchiectasis
- Architectural distortion
- Upper lobe volume loss
- Subpleural cysts (sometimes)
- Features of UIP or NSIP in almost half of all cases

Pleuroparenchymal fibroelastosis.



Travis WD. *Am J Respir Crit Care Med* 2013

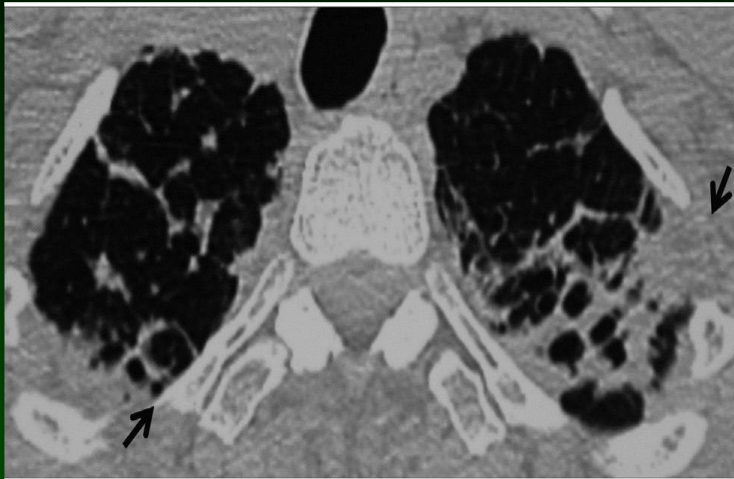
Pleuroparenchymal fibroelastosis.



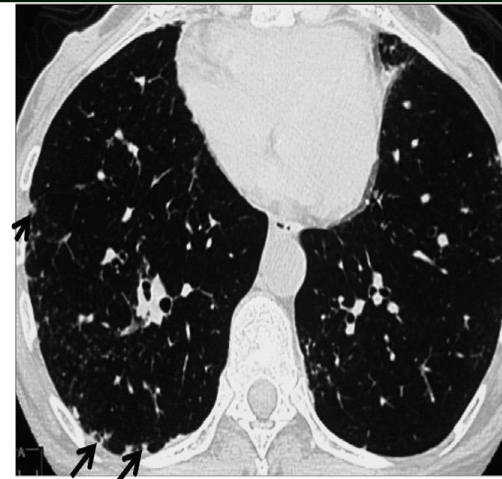
Pleural thickening
Subpleural fibrosis
Fibroelastic

Travis WD. *Am J Respir Crit Care Med* 2013

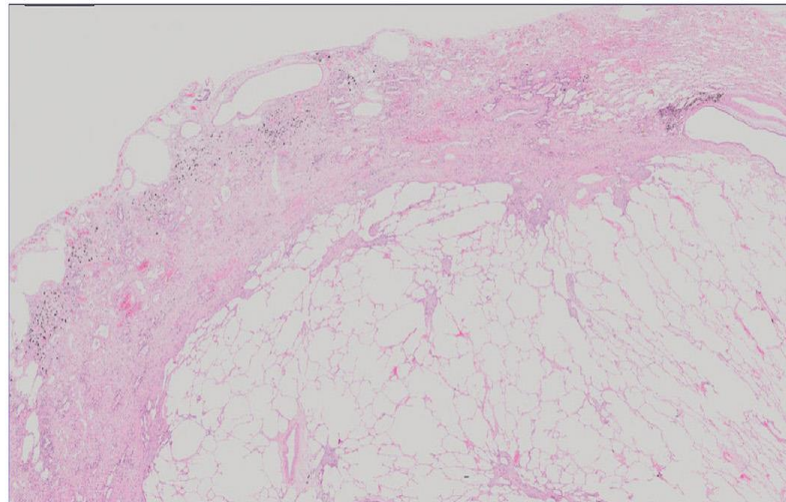
Pleuropulmonary fibroelastosis — pure.



A.CT



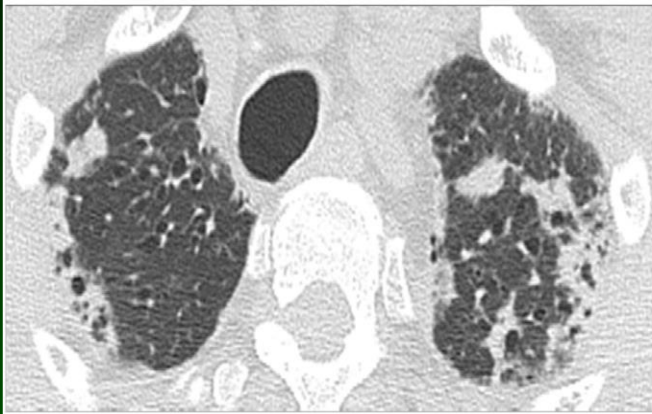
B.CT



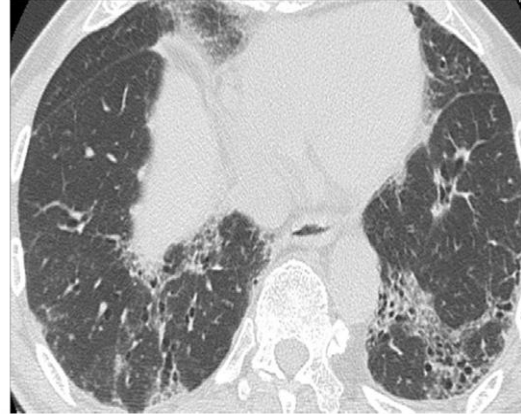
C.Histologic finding; HE x2

Takeshi J Eur J
Radiology 2015

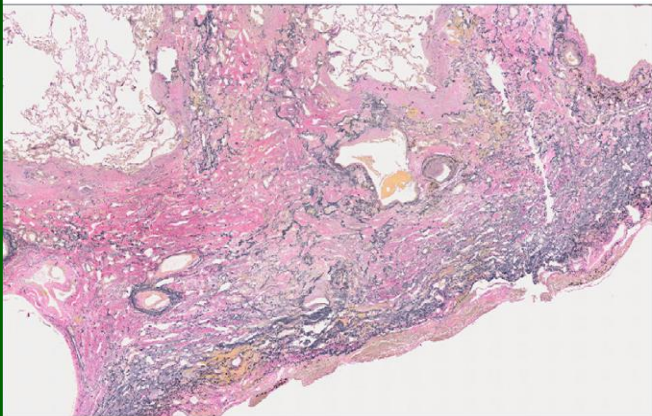
Idiopathic pleuropulmonary fibroelastosis with usual interstitial



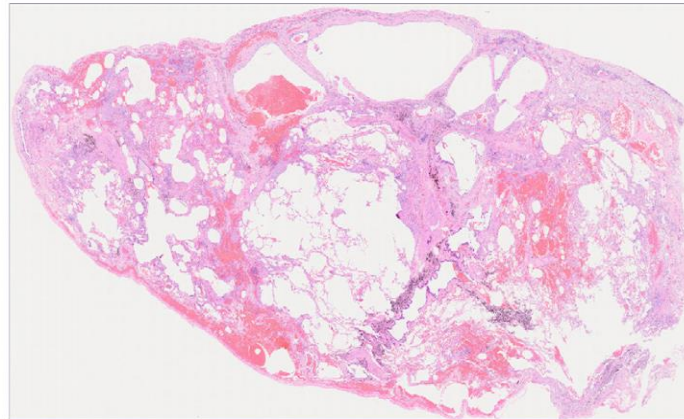
A.CT



B. CT



Histologic finding; EVG x2



Histologic finding; HE x2: UIP

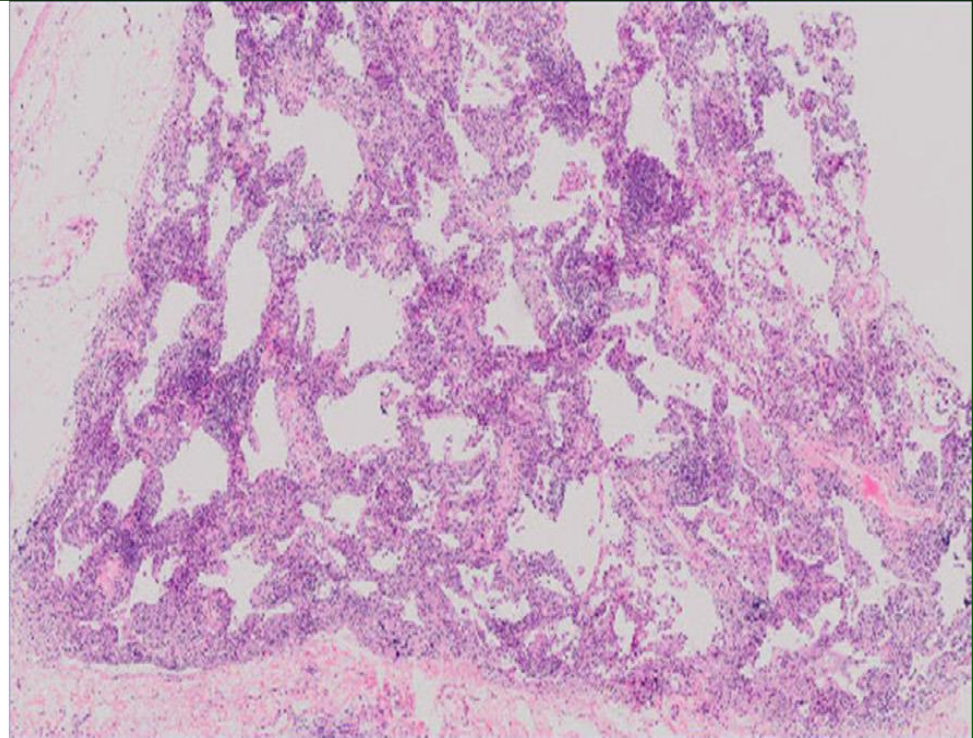
CT findings in lymphoid interstitial pneumonia.

- Lower lobe predominance
- Areas with ground-glass attenuation
- Cyst formation

Lymphoid interstitial pneumonia.



A.CT

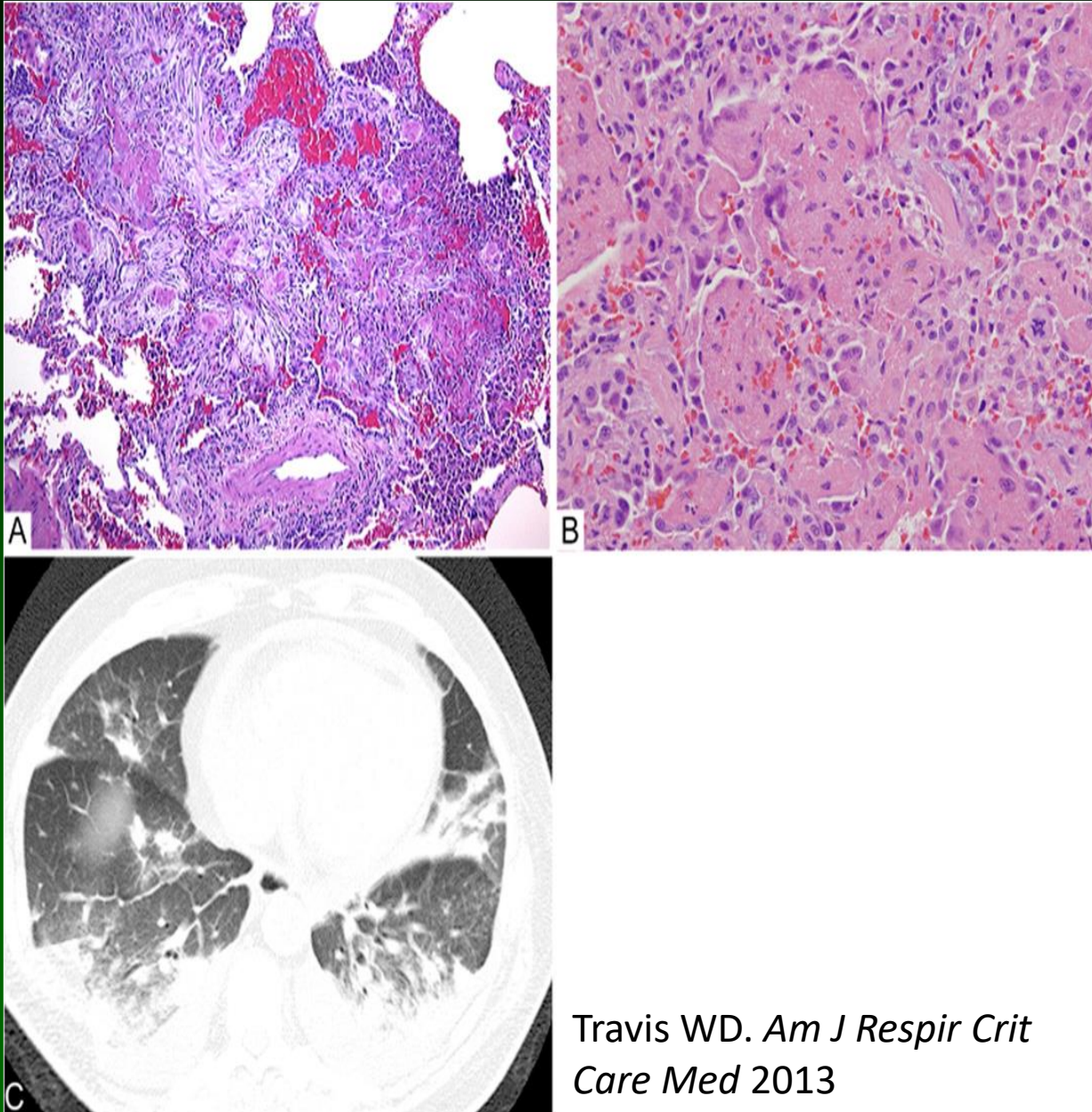


B.Histologic Finding; HEx2

CT findings of acute fibrinous organizing pneumonia.

- Bilateral basal opacities and areas of consolidation
- Diffuse areas of airspace consolidation along bronchovascular bundles (sometimes)
- Traction bronchiectasis in areas of airspace consolidation (sometimes)

Acute fibrinous and organizing pneumonia.

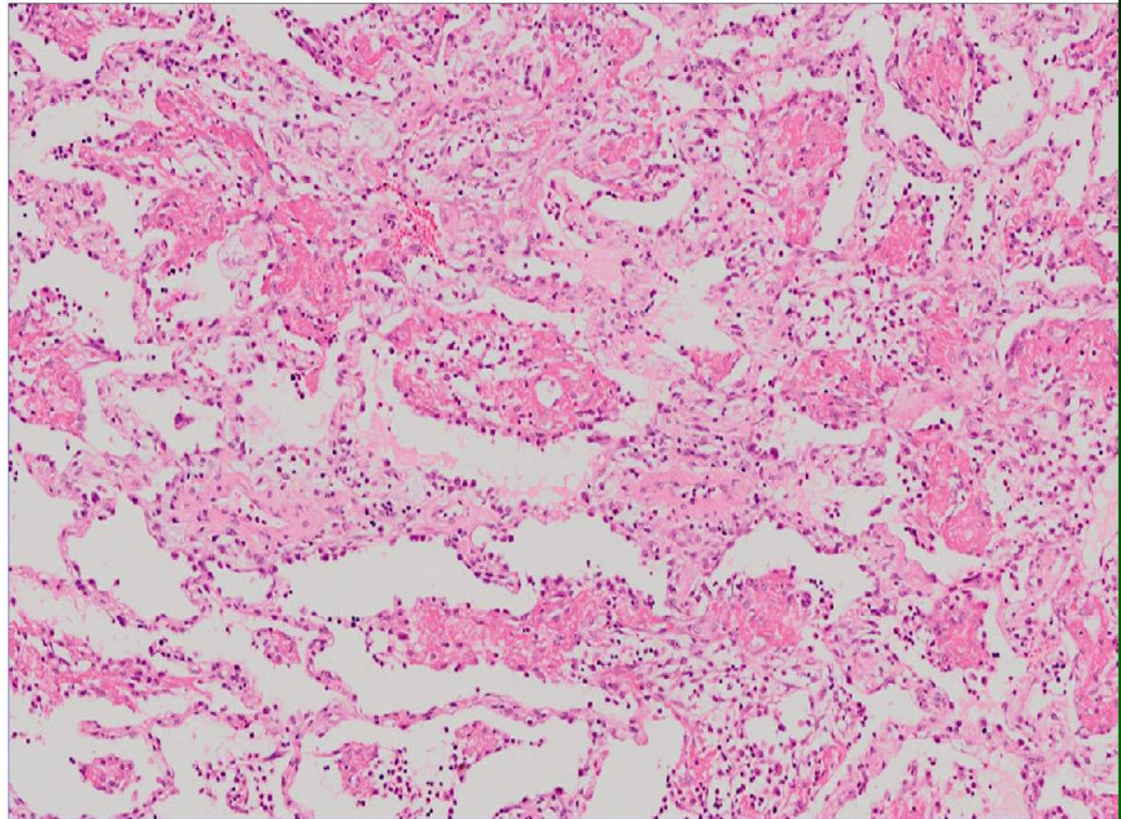


Travis WD. *Am J Respir Crit Care Med* 2013

Acute fibrinous organizing pneumonia.

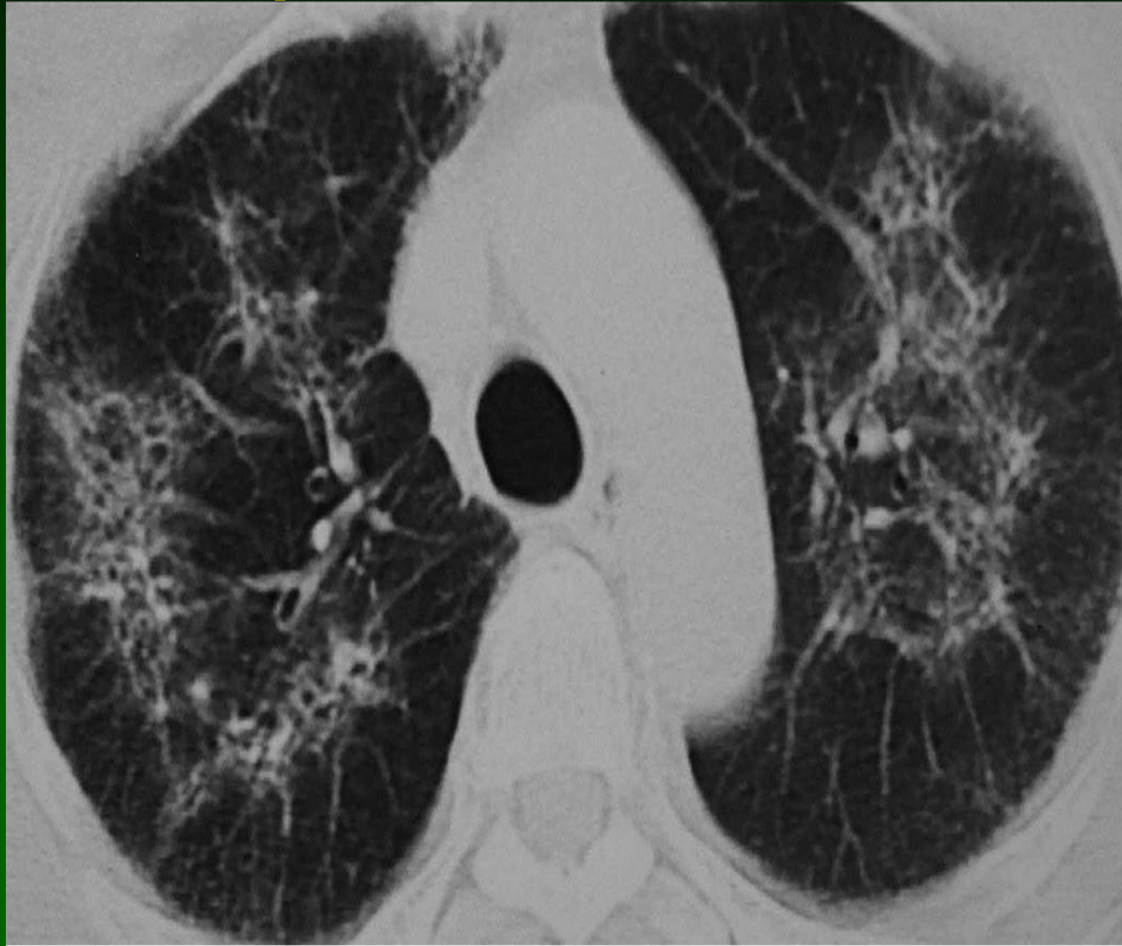


A. CT: Bilateral lower airspace consolidation



B. HE; X

Acute fibrinous organizing pneumonia.

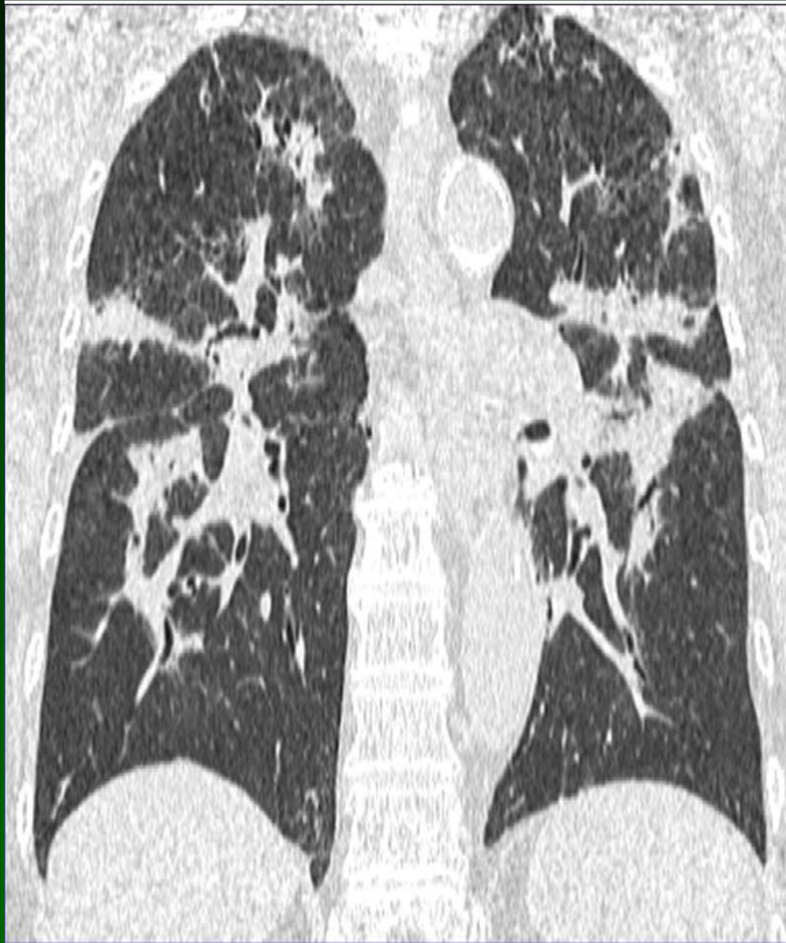


T. Diffuse peribronchovascular consolidation

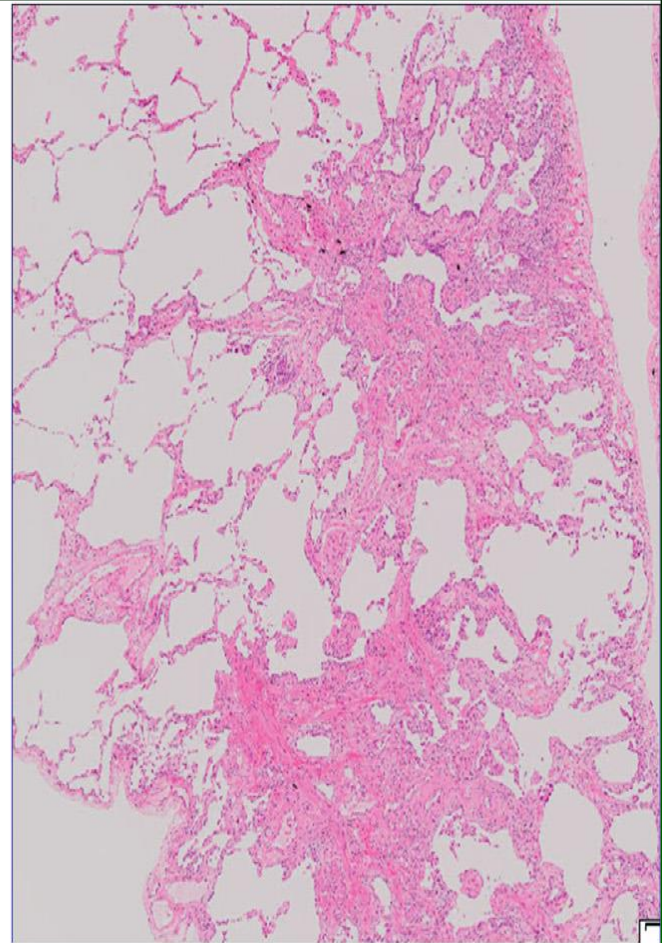
CT findings of bronchiolocentric patterns of interstitial pneumonia.

- Areas with airspace consolidation or ground-glass attenuation along bronchovascular bundles
- Centrilobular nodules
- Air trapping

Airway Centered interstitial fibrosis.

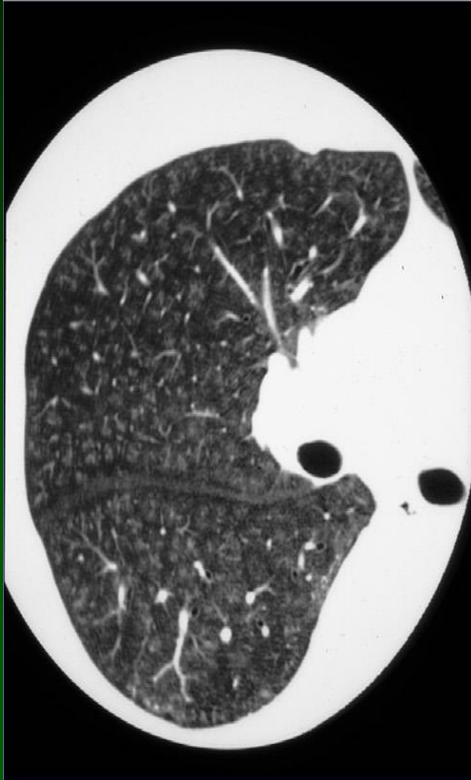


CT; Coronal MPR

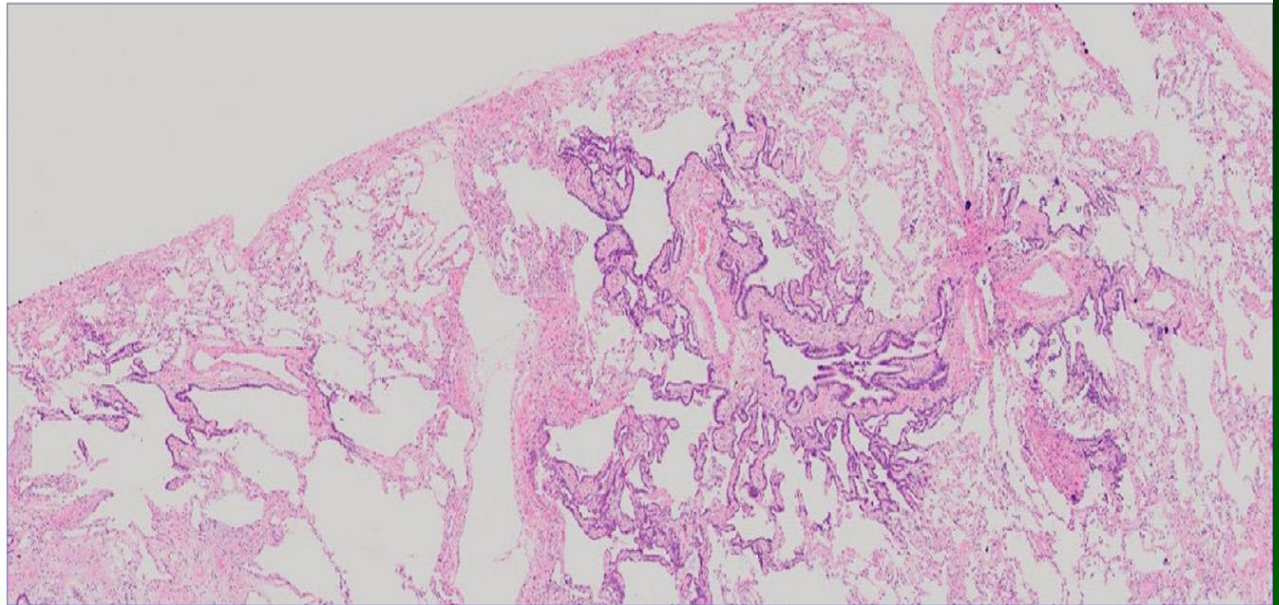


B.Histologic Findings:HE; x4

Peribronchial metaplasia interstitial lung disease.



A.CT



B.Histologic Findings:HE; x4

Hypersensitivity Pneumonitis

- An allergic lung disease caused by inhalation of antigens contained in a variety of organic dusts
 - Farmer's lung, pigeon breeder's lung, etc..
- A major imaging differential diagnosis of IIP
 - UIP, NSIP, DIP, RB-ILD, LIP

Examples of Hypersensitivity Pneumonitis

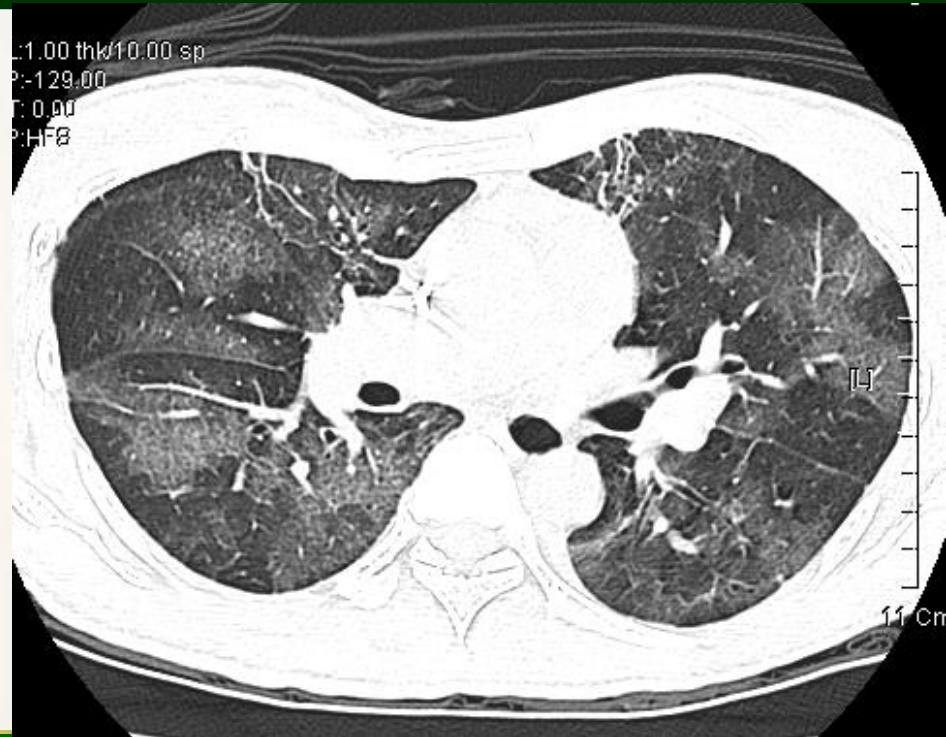
Disease	Antigen Source	Putative Antigen
Bird fancier's disease	Various birds	Protein avian feces, feathers
Cheese worker's lung	Moldy cheese	Penicillium species
Coffee worker's lung	Coffee bean	Unknown
Farmer's lung	Moldy hay	Thermophilic actinomycetes
Furrier's lung	Animal fur	Protein in animal fur
Hot tub lung	Warm water	Mycobacterium avium complex
Humidifier lung	Warm water	Thermophilic actinomycetes
Japanese summer disease	Moldy houses	Various fungi
Machine worker's lung	Metal-cutting fluid	Mycobacterium species, Gram-negative bacilli
Malt worker's lung	Moldy malt	Aspergillus species
Mushroom worker's lung	Mushrooms	Mushroom spores, various other fungi
Peat moss worker's lung	Moldy peat moss	Various fungi
Sauna bather's lung	Sauna water	Various fungi
Sequoiosis	Moldy redwood dust	Various fungi
Suberosis	Cork	Aspergillus species, cork dust

Imaging-based Diagnosis of Hypersensitivity Pneumonitis

Type of Disease	Diagnostic Imaging Features
Acute or insidious onset without fibrosis	Diffuse GGO, centrilobular GGO, air trapping, headcheese sign
Insidious onset with fibrosis	Reticulation, peribronchovascular interstitial thickening, architectural distortion, honeycombing, air trapping, sparing of extreme basal lung, ill-defined centrilobular GGO

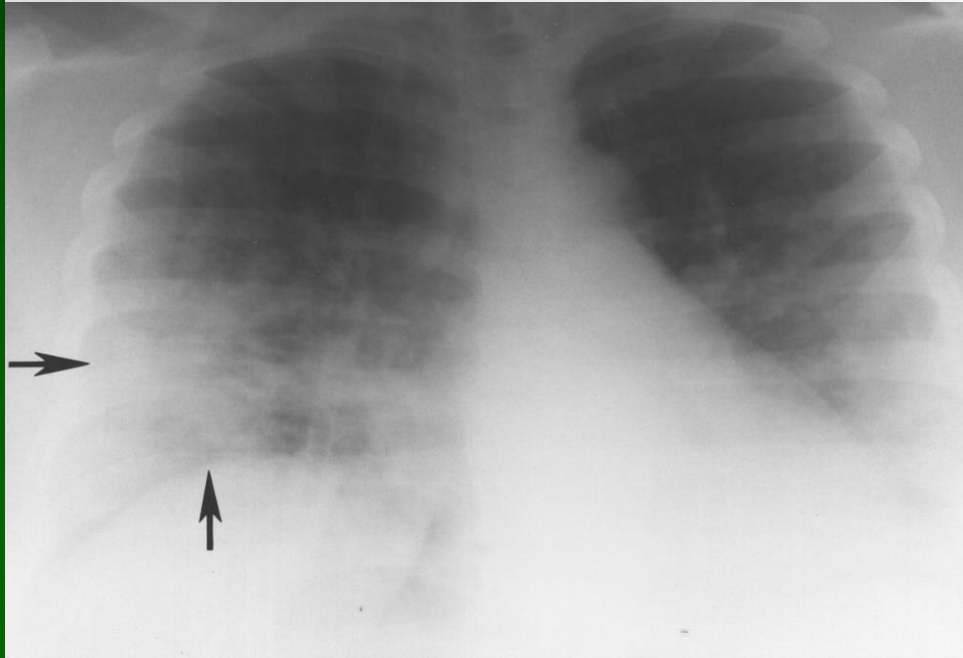
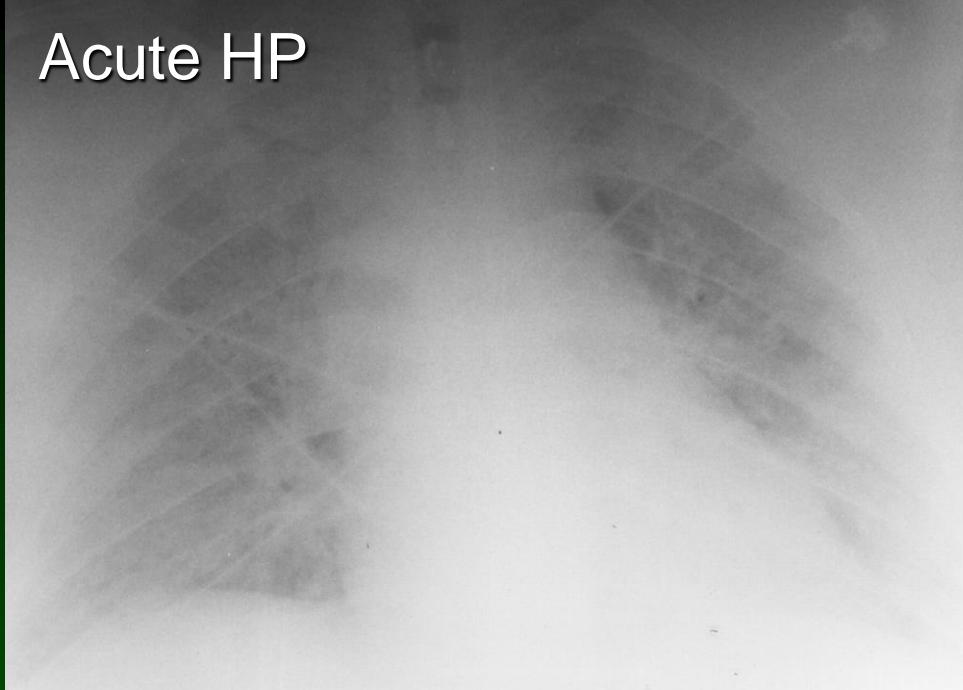
Head-cheese Sign

- Mixed GGO and mosaic perfusion, often indicates bronchiolitis



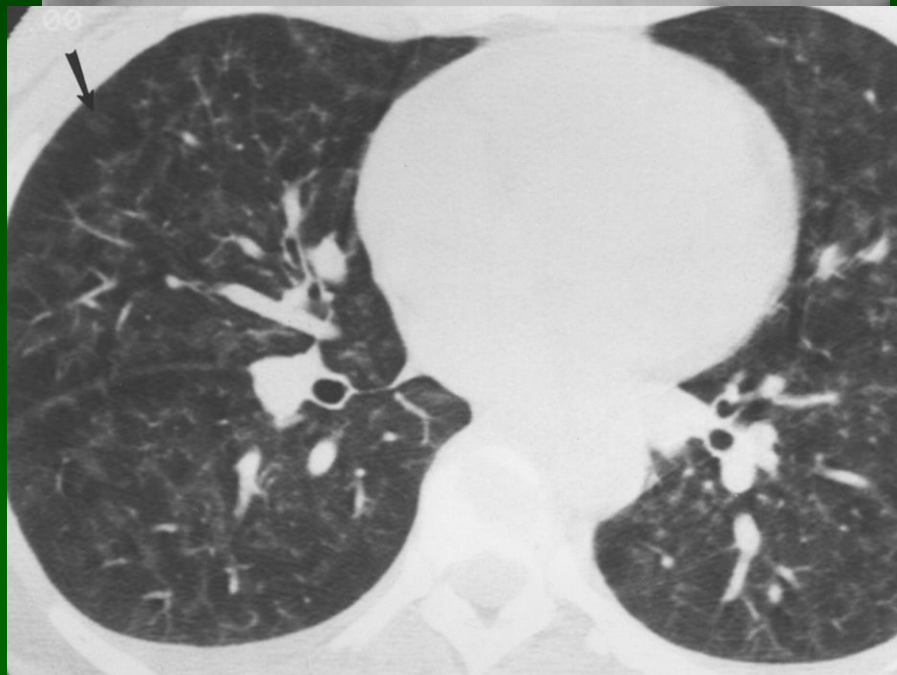
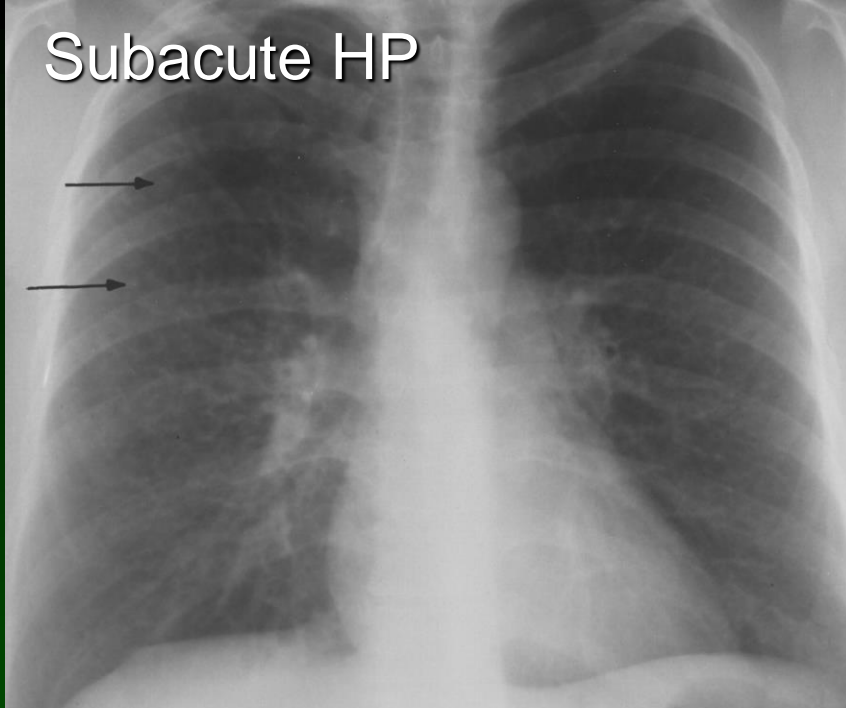
- HP, DIP, RBILD, sarcoidosis, atypical infection

Acute HP



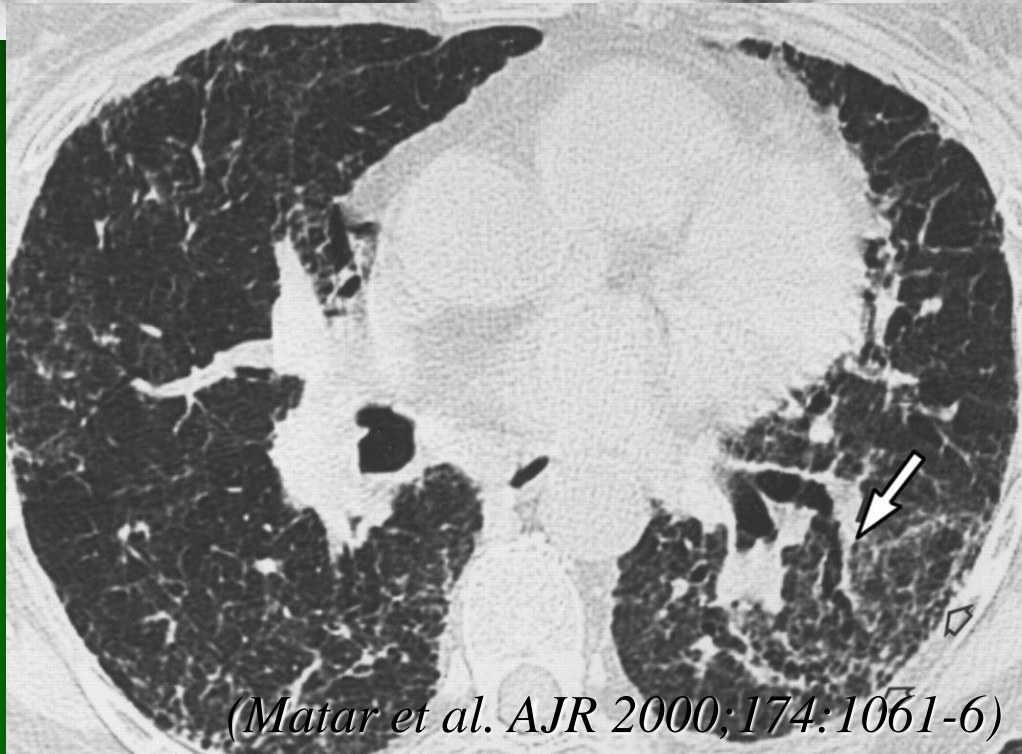
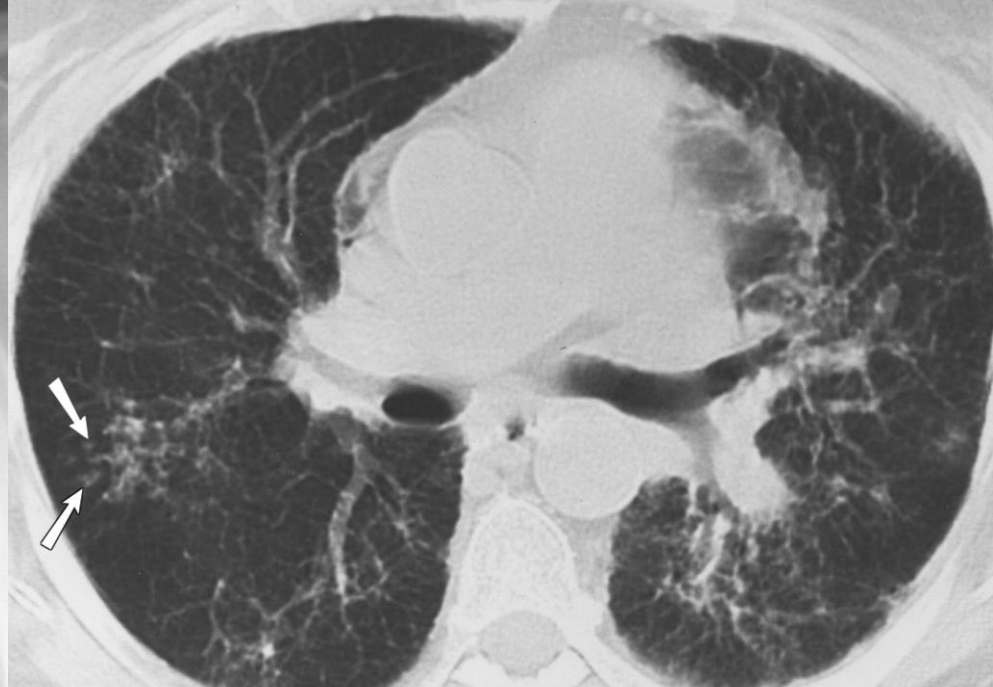
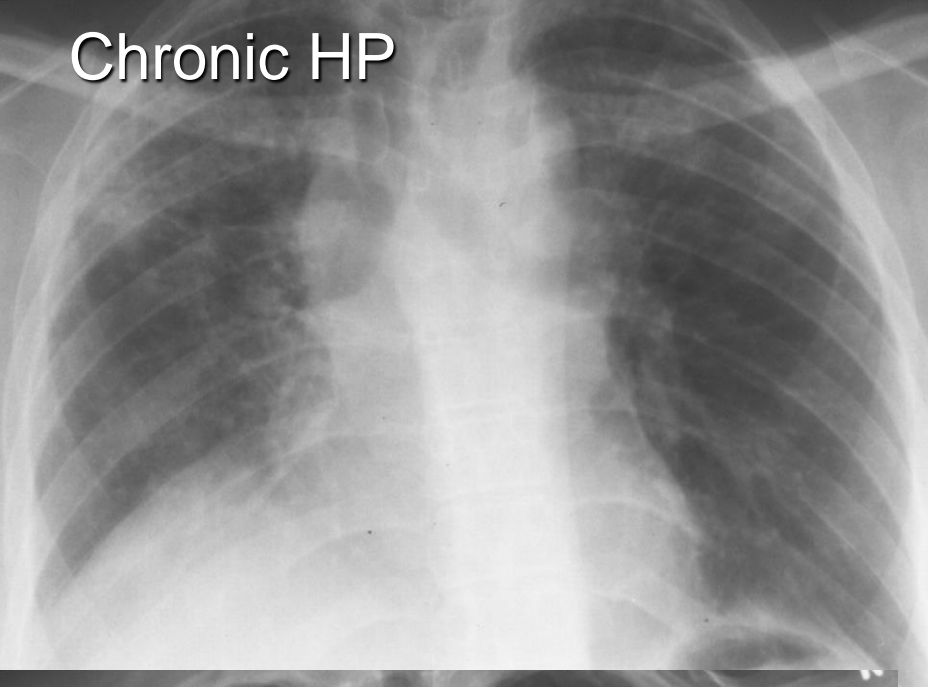
(Matar et al. AJR 2000;174:1061-6)

Subacute HP



(Matar et al. AJR 2000;174:1061-6)

Chronic HP



(Matar et al. AJR 2000;174:1061-6)

Differential Diagnosis of Diffuse Lung Disease

- First step
 - Airspace disease or
 - Interstitial disease

Differential Diagnosis of Diffuse Lung Disease

- Airspace disease
 - Acute
 - Edema: cardiogenic vs. noncardiogenic
 - Infection
 - Neoplastic
 - Blood: contusion, hemorrhage, infarction
 - Idiopathic: sarcoid, PIE

Differential Diagnosis of Diffuse Lung Disease

- Airspace disease
 - Chronic (TF DALLAS)
 - TB
 - Fungus
 - DIP
 - Alveolar cell ca
 - Lymphoma
 - Lipoid pneumonia
 - Alveolar proteinosis
 - Sarcoidosis

Differential Diagnosis of Diffuse Lung Disease

- Interstitial disease
 - Fine reticular or reticulonodular pattern
 - Granulomatous
 - Infection
 - Inhalational
 - Idiopathic
 - Neoplastic
 - Hemosiderosis

Differential Diagnosis of Diffuse Lung Disease

- Interstitial disease
 - Coarse reticular with fibrosis
 - Upper lung: cystic fibrosis, AS, Silicosis, Sarcoidosis, EG, TB, extrinsic allergic alveolitis (hypersensitive pneumonitis), neurofibromatosis
 - Lower lung: IPF/UIP, asbestosis, collagen vascular disease, chronic aspiration, drug

Thanks for Your Attention

