# ILD-INTERSTITIAL LUNG DISEASES

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RIP…

# CONTENT

- Introduction
- Classification
- Pathogenesis
- Clinical history
- Lab investigations
- Management
- Hrct features of different idiopathic interstitial pneumonias(IIPs)

# INTERSTITIAL LUNG DISEASES (ILD)

## DEFINITION

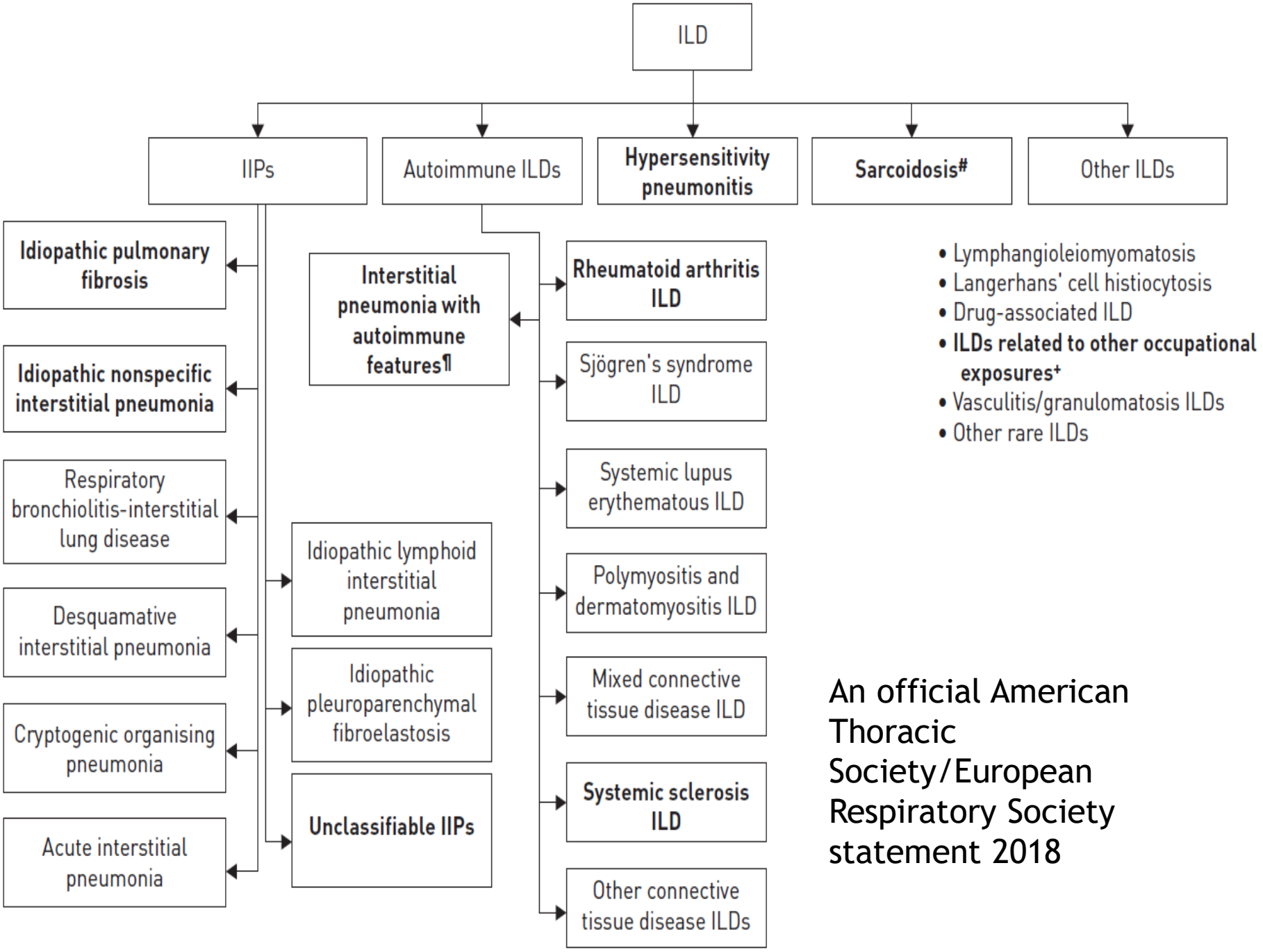
Interstitial lung diseases are a group of pulmonary disorders characterized by:

1. Radiologically diffused infiltrates.
2. Histologically by distortion of the gas exchanging units.
3. Physiologically by restriction of lung volumes and impaired oxygenation.

- ILD encompasses a large group of > 200 parenchymal pulmonary disorders, of which the majority are classified as rare.

- Given their overlapping clinical, radiological and pathological presentations, the terminology used to describe patients with fibrosing ILDs presenting with progressive phenotype is “progressive-fibrosing ILD (PF-ILD)”

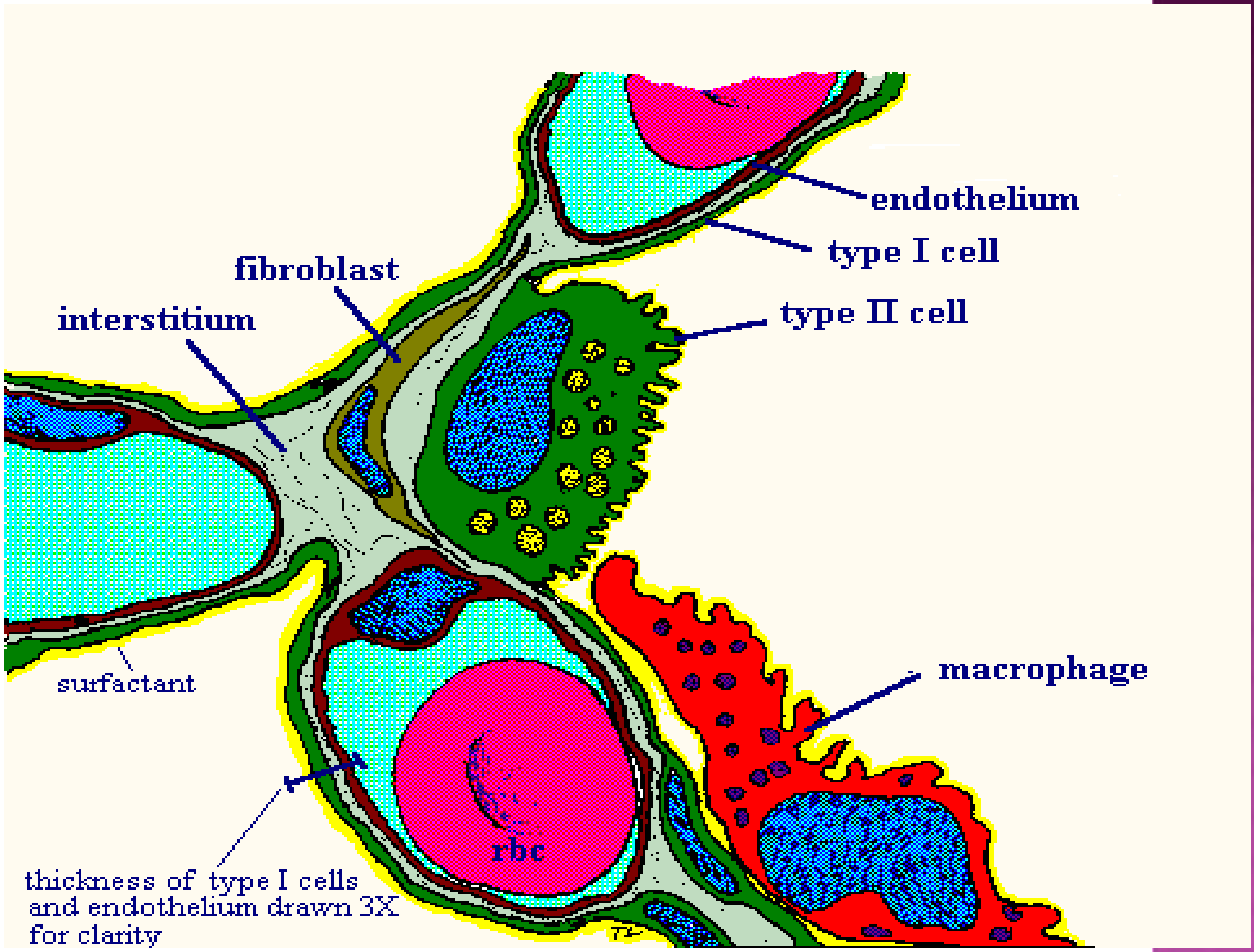
- Diagnosis based on comprehensive history, physical examination, radiography and MDT - important process with significant impact




An official American Thoracic Society/European Respiratory Society statement 2018

# WHAT DOES THE TERM “INTERSTITIAL’ MEAN?

- It implies that the inflammatory process is limited specifically to the area between the alveolar epithelial and capillary endothelial basement membranes.


endothelium
type I cell
fibroblast
interstitium
type II cell
surfactant
macrophage
rbc
thickness of type I cells
and endothelium drawn 3X
for clarity

# PATHOGENESIS OF INTERSTITIAL DISEASE:

Often at the end stage most disease processes end in fibrosis from different unknown causes.

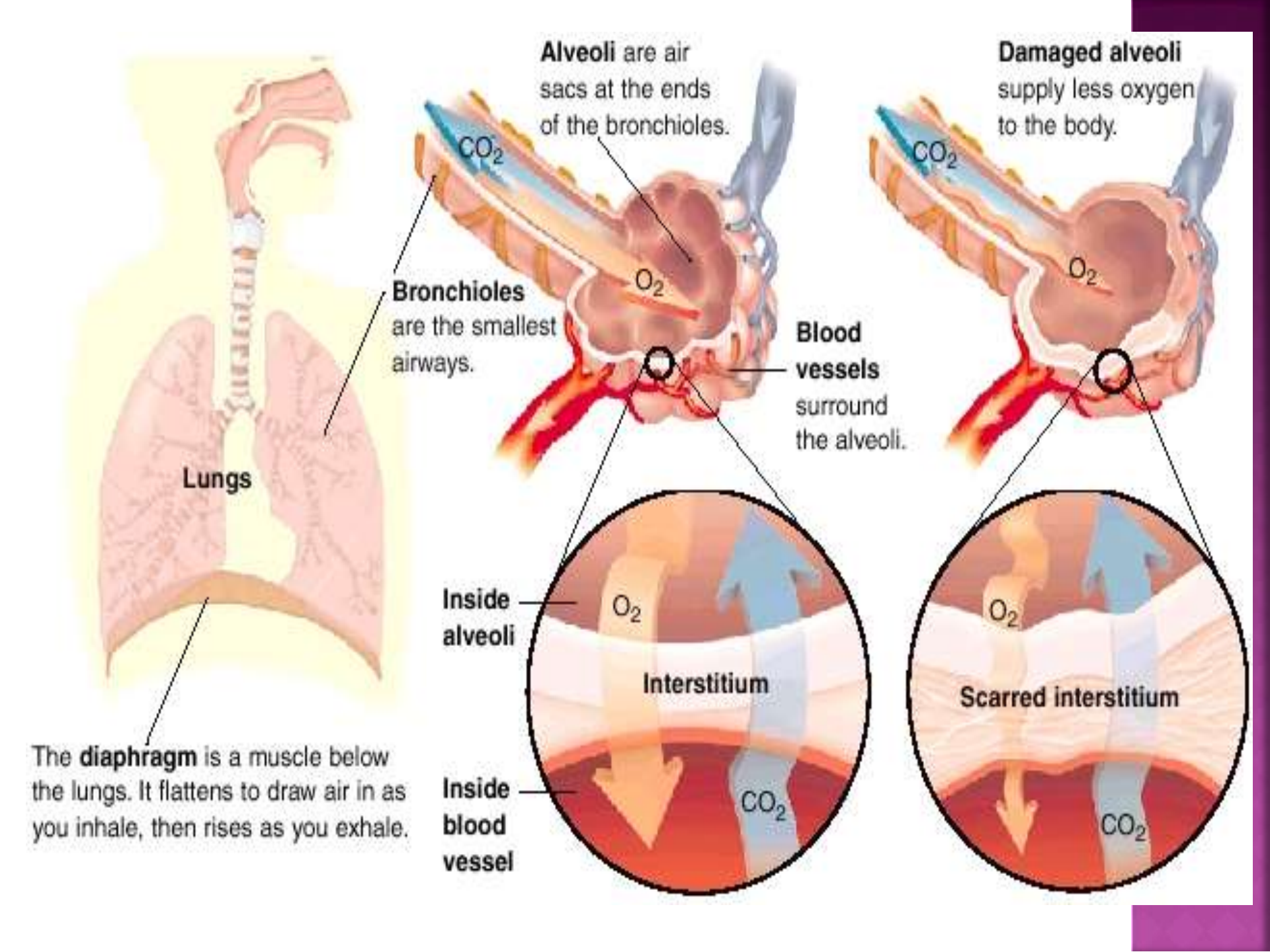
**Pathogenesis of Interstitial Fibrosis** – fibrous widening of interstitium is hallmark of chronic interstitial lung disease

Two Mechanisms:

- ***(1) Primary interstitial widening*** – edema and fibrosis formation directly within the interstitial compartment

  eg: interstitial edema, sarcoidosis (thickening of interstitium initially by granuloma formation)

- ***(2) Accretion*** – organization of exudate within the alveolar space that is converted to fibrous connective tissue and is incorporated into the interstitium; in some cases the exudate is cleared with resolution. eg: organizing pneumonia


Alveoli are air sacs at the ends of the bronchioles.
Damaged alveoli supply less oxygen to the body.
$CO_2$
$O_2$
Bronchioles are the smallest airways.
Blood vessels surround the alveoli.
Lungs
Inside alveoli
Interstitium
Scarred interstitium
Inside blood vessel
The diaphragm is a muscle below the lungs. It flattens to draw air in as you inhale, then rises as you exhale.

# ACUTE STAGE: (GENERAL INFLAMMATORY CONDITION)

- **Inflammation & Edema secondary to:**
- Infiltration of WBCs (Leukocytes)
  - Into the alveolar walls and interstitial spaces
  - neutrophils, eosinophils, basophils, macrophages, monocytes, and lymphocytes
- **Sometimes:** Bronchial inflammation and smooth muscle constriction (obstructive)

# CHRONIC STAGE

- Increased numerous WBCs (including some fibroblasts)
- Extensive inflammation:
  - Interstitium continues to thicken
  - Fibrosis and granulomas proliferate
  - Honeycombing and cavity formation ensue
  - Pleural effusions may occur
    - (end stage Pulmonary Fibrosis)

# Major Anatomic Alterations:

- Fibrotic thickening of the bronchioles and alveolar units
- Destruction of the alveoli and pulmonary capillaries
- Honeycombing and cavity formation
- Granulomas
- Airway obstruction: secondary to inflammation and bronchial constriction

# *PATHOGENESIS:*

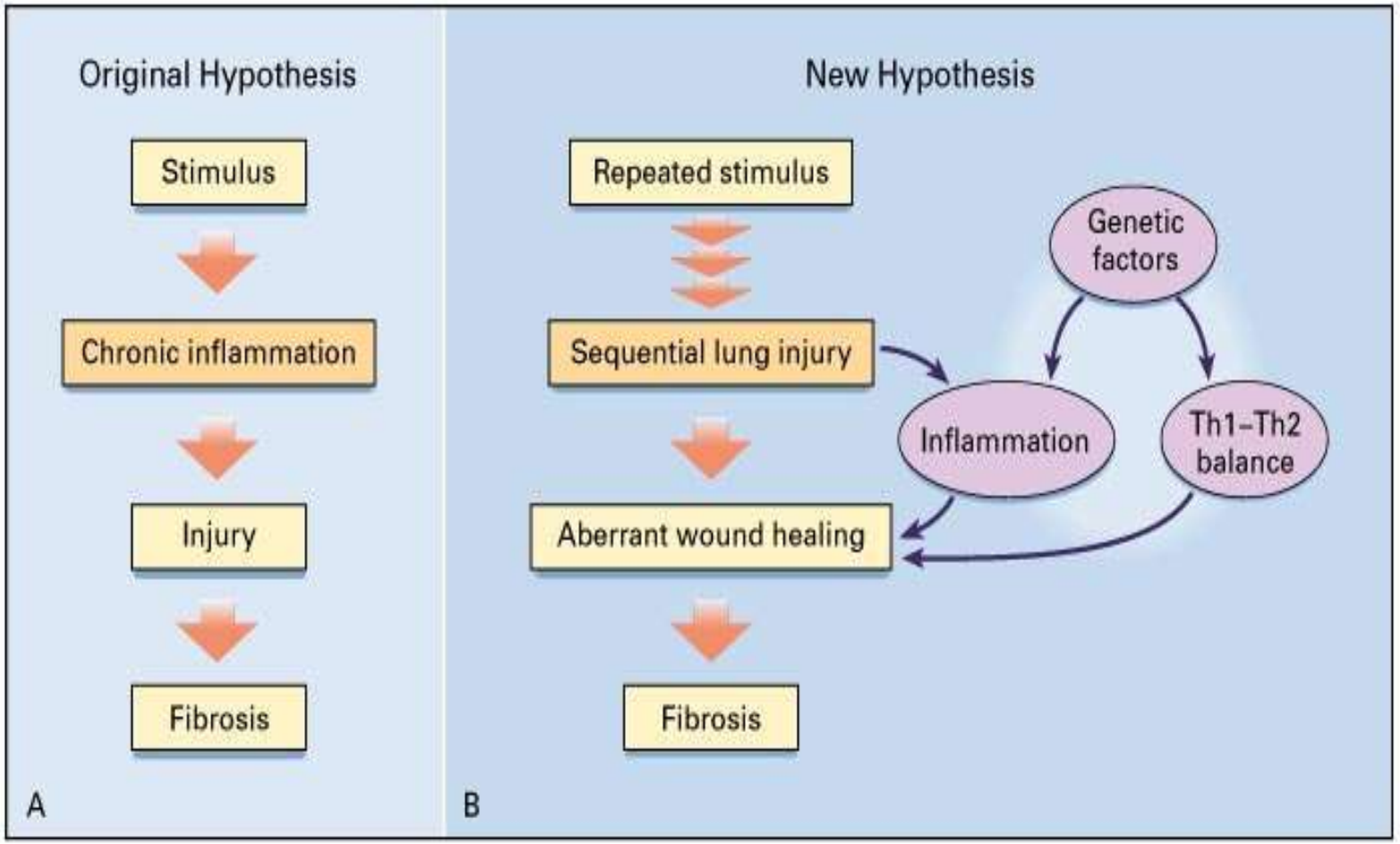
*The previous theory*

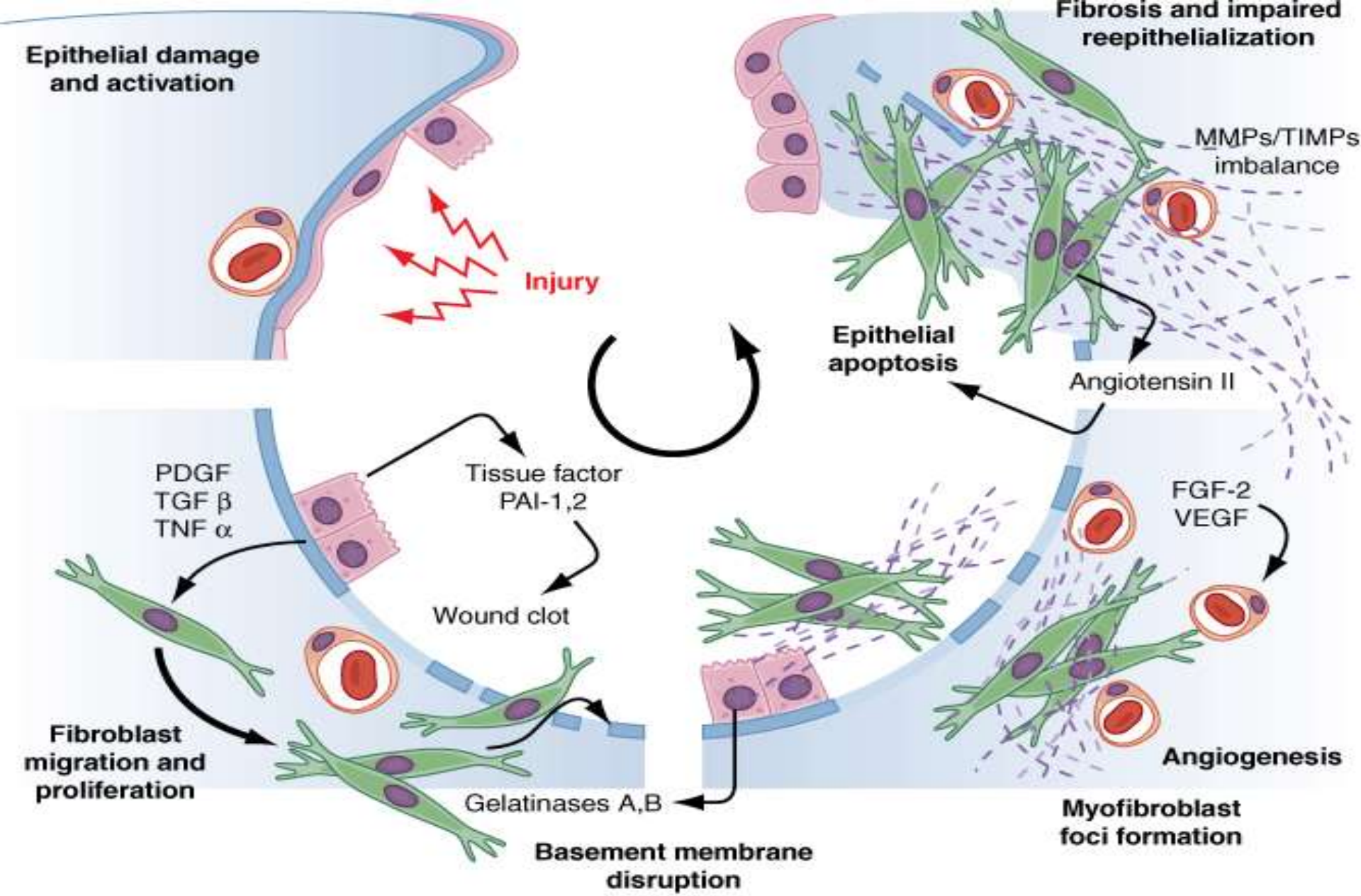
*generalized inflammation progressed to widespread parenchymal fibrosis. However, anti-inflammatory agents and immune modulators failed in modifying the natural course of the disease*

The current theory

unknown endogenous or environmental stimuli disrupt the homeostasis of alveolar epithelial cells, resulting in diffuse epithelial cell activation and aberrant epithelial cell repair.

# PATHOGENESIS






Source: Fauci AS, Kasper DL, Braunwald E, Hauser SL, Longo DL, Jameson JL, Loscalzo J: *Harrison's Principles of Internal Medicine*, 17th Edition: http://www.accessmedicine.com

# CLINICAL HISTORY

- **Fever-**collagen vascular disease, cryptogenic organizing pneumonia, acute interstitial pneumonia, sarcoidosis, hypersensitivity pneumonitis such as farmer's lung , tropical pulmonary eosinophilia, chronic eosinophilic pneumonia
- **Chest Pain/Pleurisy-**pulmonary vasculitis and collagen vascular diseases- systemic lupus erythematosus, mixed connective tissue disease , Wegener's granulomatosis , and rheumatoid arthritis. Sarcoidosis

# CLINICAL HISTORY

- **Smoking History-**idiopathic pulmonary fibrosis, Respiratory bronchiolitis-associated interstitial lung disease (RB-ILD), desquamating interstitial pneumonitis (DIP), and eosinophilic granuloma (EG, or Langerhans' cell granulomatosis), Goodpasture's syndrome

# EXPOSURE AND OCCUPATIONAL HISTORY

| Exposure | Associated Lung Disease |
|---|---|
| Bird breeders and fanciers | Hypersensitivity pneumonitis |
| Automotive mechanics<br>Electricians<br>Pipefitters<br>Shipyard workers | Asbestosis |
| Electronic and computer industry workers | Berylliosis |
| Farmers | Farmer's lung (hypersensitivity pneumonitis) |

| | |
|---|---|
| Hot tub, sauna, humidifiers | Hypersensitivity pneumonitis |
| Metal workers (tool and die) | Metal-induced pneumoconioses |
| Miners<br>Sandblasters<br>Ceramic workers | Silicosis |
| Miners (specifically coal) | Coal workers' pneumoconiosis |
| Bark strippers<br>Woodworkers | Hypersensitivity pneumonitis |

# DRUGS CAUSING ILD

**Antibiotics**
- Nitrofurantoin
- Minocycline
- Cephalosporins

**Antiarrhythmic**
- Amiodarone
- Tocainide

**Anti-inflammatory**
- Azathioprine
- Etanercept
- Gold salts
- Infliximab
- Methotrexate
- NSAIDs
- Penicillamine
- Sulfasalazine

**Neurologic/Psychiatric**
- Carbamazepine
- Phenytoin

**Drugs of Abuse**
- Cocaine
- Heroin
- Talc

**Chemotherapeutic**
- All-trans retinoic acid (ATRA)
- Alpha-interferon
- Antithymocyte globulin
- Bleomycin
- Busulfan
- Carmustine (BCNU)
- Chlorambucil
- Colony-stimulating factors (GM-CSF)
- Cyclophosphamide
- Cytosine arabinoside
- Docetaxel
- Geftinib
- Gemcitabine
- Interleukin-2
- Irinotecan
- Melphalan
- Mitomycin C
- Paclitaxel
- Procarbazine
- Vinorelbine

**Other**
- Bacille Calmette-Guérin (BCG)
- Mineral oil
- Radiation

# FAMILY HISTORY

- Family history of lung disease or connective tissue diseases
- Familial pulmonary fibrosis
- Sarcoidosis
- Tuberous sclerosis (hamartomas, epilepsy, or mental retardation)-lymphangioleiomyomatosis or multifocal micronodular pneumocyte hyperplasia

# CLINICAL HISTORY

- Symptoms of chronic sinusitis -diffuse panbronchiolitis and Wegener's granulomatosis
- Syndrome of uveitis, parotiditis, and erythema nodosum -self-limited sarcoidosis
- Travel history- parasitic infection as the cause of an eosinophilic pneumonia.
- Raynaud's phenomenon, proximalmuscle weakness, and joint swelling/pain-collagen vascular disease
- Esophageal reflux-idiopathic pulmonary fibrosis

# PHYSICAL EXAMINATION

- “Velcro rales,” or inspiratory crackles.
- Mid inspiratory squeaks- airway-centered diseases, including cryptogenic organizing pneumonia, constrictive bronchiolitis, hypersensitivity Pneumonitis, nonspecific interstitial pneumonia.
- Expiratory wheeze- Constrictive bronchiolitis

# PHYSICAL EXAMINATION

- Clubbing- Idiopathic pulmonary fibrosis (25 to 50 percent of patients), Desquamative interstitial pneumonia (50 percent of patients), Interstitial lung disease from rheumatoid arthritis (75 percentof patients)
- proximal muscle weakness (Myositis)- Polymyositis/ dermatomyositis, Sjogren's syndrome, scleroderma, and mixed connective tissue disease.

# LABORATORY EVALUATION

- Initial tests in all cases of suspected ILD should include a urine dipstick, full differential blood cell count, serum urea, electrolytes and creatinine, and liver function tests

# PULMONARY PHYSIOLOGY TESTING

- Restrictive changes- reduced total lung capacity, reduced residual volume, decreased static compliance, and a reduced VC, often with an increased FEV1/FVC ratio
- Reduced diffusing capacity for carbon monoxide (DLCO).
- Airflow obstruction- sarcoidosis, lymphangioleiomyomatosis, Langerhans' cell histiocytosis (eosinophilic granuloma), constrictive bronchiolitis, respiratory bronchiolitis–interstitial lung disease, and hypersensitivity pneumonia
- LCH and LAM- Mixed

# PULMONARY PHYSIOLOGY TESTING

- A TLCO level of ,40% is indicative of advanced disease in fibrotic IIP.
- In IPF a fall from baseline of >10% in FVC or >15% in TLCO in the first 6–12 months identifies patients witha much higher mortality.
- Desaturation during the 6 minute walk test at presentation is a stronger prognostic determinant in IPF than resting lung function.

# RECOMMENDATIONS FOR SURGICAL LUNG BIOPSY IN ILD

- It is recommended that the precise biopsy sites are based on HRCT appearances .
- In patients with suspected IIP, areas of intermediate abnormality or comparatively normal lung adjacent to areas of established honeycombing should be targeted with the specific aim of identifying UIP if present

# General management strategies in ILD

- Communicating the diagnosis
- Smoking cessation
- Pulmonary rehabilitation

# MANAGEMENT OF SPECIFIC ILDS

- **Recommendations for the management and treatment of IPF**
- Symptomatic treatment -oxygen therapy, pulmonary rehabilitation, opiates, antireflux therapy, withdrawal of steroids and other immunosuppressants.

- **Recommendations for referral for lung transplant in patients with IPF**
- Referral to a transplant centre should be made if the disease is advanced (TLCO <40% predicted) or progressive (>10% decline in FVC or >15% decline in FVC during 6 months of follow-up).

# TREATMENT OF NSIP

- Corticosteroids, with or without immunosuppressive agents
- No validated recommendations exist on indications for treatment, duration of therapy or treatment regimens

# RECOMMENDATIONS FOR COP

- COP usually responds to corticosteroid therapy but the optimum dose and length of treatment is not known. Initial doses of 0.75–1 mg/kg, weaning over 6– 12 months, are reasonable.
- Prolonged corticosteroid therapy should be carefully considered in patients with relapsing COP.

- **Treatment of RBILD and DIP**
- Smoking cessation
- **Lymphoid interstitial pneumonia (lip) Treatment**
- Corticosteroids
- Antiretroviral therapy -LIP secondary to HIV infection

- **Recommendations for HP**
- Avoidance of the causative antigen, when identified, is the most important and effective aspect of management.
- Corticosteroids may have a role in treating severe or progressive disease.

# RECOMMENDATIONS FOR THE TREATMENT OF CTD-ASSOCIATED ILD

- For the majority of CTDs, with the exception of SSc -oral prednisolone at an initial dose of 0.5–1 mg/kg with the aim of tapering to a maintenance dose of 10 mg/day or association with an immunosuppressive agent (usually oral or intravenous cyclophosphamide or oral azathioprine).
- ILD associated with PM/DM -early treatment with oral prednisolone (0.75–1 mg/kg) and cyclophosphamide or other immunosuppressive therapy to prevent disease progression.

# RECOMMENDATIONS FOR THE TREATMENT OF CTD-ASSOCIATED ILD

- In SSc-associated ILD- low-dose oral steroids (10 mg/day) and/or cyclophosphamide (oral or intravenous)

# CXR

| Distribution of ILD | |
|---|---|
| **Upper lung zone** | **Lower lung zone** |
| Sarcoidosis | Usual interstitial pneumonia (UIP/IPF) |
| Silicosis | Nonspecific interstitial pneumonia (NSIP) |
| Coal worker's pneumoconiosis | Connective tissue disease–associated ILD |
| Hypersensitivity pneumonitis | Asbestosis |
| Langerhans cell histiocytosis | Desquamative interstitial pneumonia (DIP) |
| Berylliosis | |
| Chronic eosinophilic pneumonia | |

| Peripheral reticular |
| --- |
| Idiopathic pulmonary fibrosis/usual interstitial pneumonia |
| Nonspecific interstitial pneumonia |

| Cystic |
| --- |
| Lymphangioleiomyomatosis |
| Langerhans cell histiocytosis |
| Lymphocytic interstitial pneumonia |
| *Pneumocystis jiroveci* pneumonia (PCP) |

| Nodular |
| --- |
| Sarcoidosis |
| Berylliosis |
| Hypersensitivity pneumonitis |
| Langerhans cell histiocytosis |
| Silicosis |
| Metastatic disease |
| Talcosis |
| Granulomatous polyangiitis (formerly known as Wegener's granulomatosis) |
| Respiratory bronchiolitis ILD |

| Ground glass |
| --- |
| NSIP |
| Cryptogenic organizing pneumonia |
| Eosinophilic pneumonia (chronic or acute) |
| Pulmonary edema |
| Infection (opportunistic or viral) |
| Alveolar hemorrhage |
| Hypersensitivity pneumonitis |
| Desquamative interstitial pneumonia |
| Sarcoidosis |
| Pulmonary alveolar proteinosis |

# IDIOPATHIC PULMONARY FIBROSIS

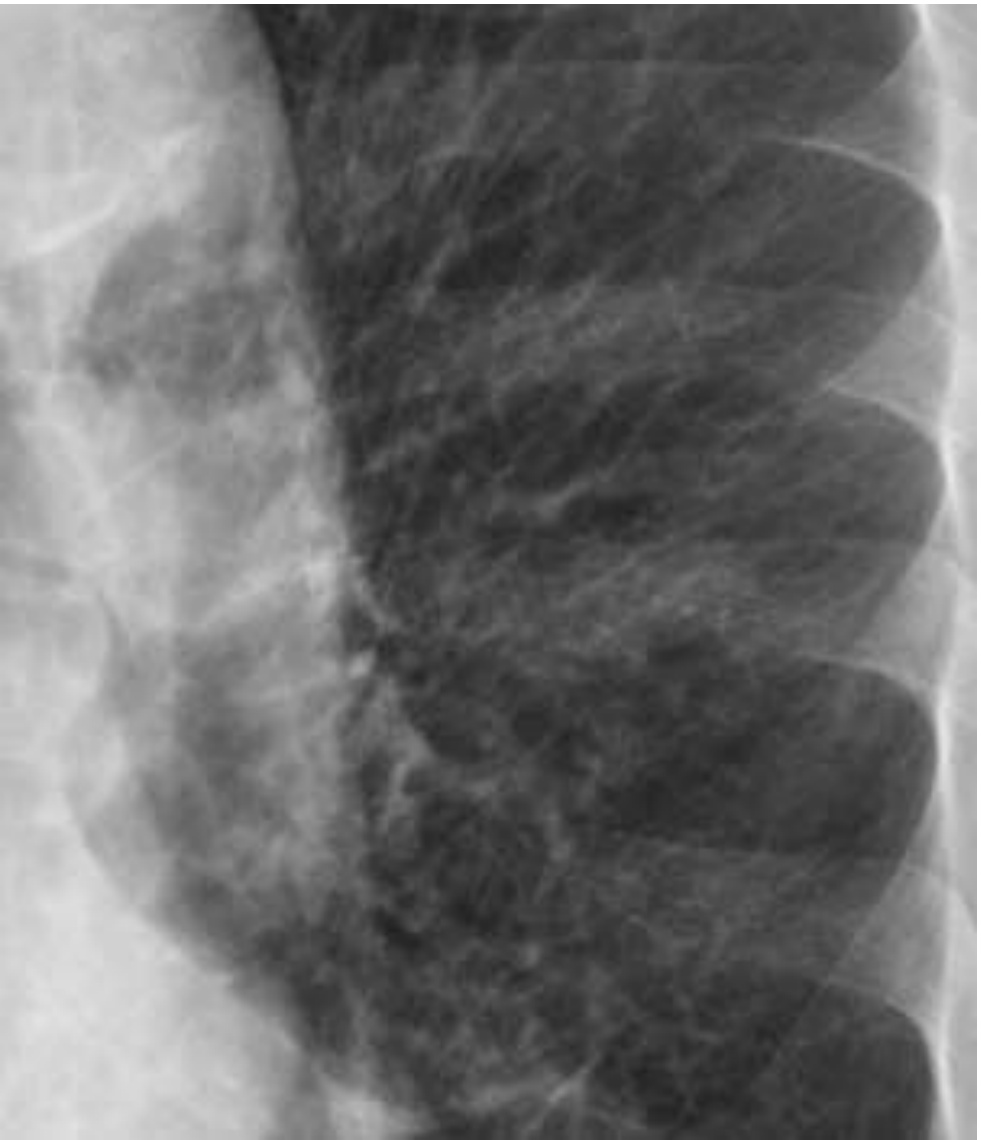
- best diagnostic clue- sub pleural and basal honeycombing and traction bronchiectasis
- Most affected portions of the lungs- subpleural and basilar lung
- Costophrenic angles should be most affected portions

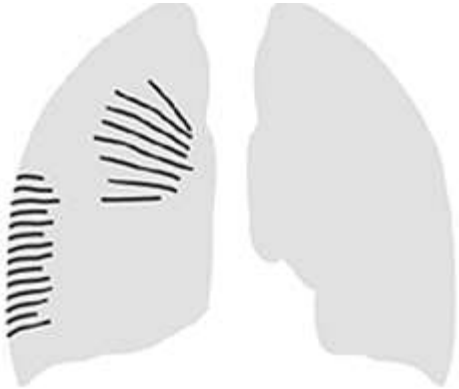
# HRCT

- Predominant reticular pattern and honeycombing
- Traction bronchiectasis is essential in diagnosis
- Reticular opacities> ground glass opacities
- Co-existant emphysema in 30%
- Distribution: basal and peripheral predominance [ not unilateral]

# PATTERNS ON CHEST X-RAY

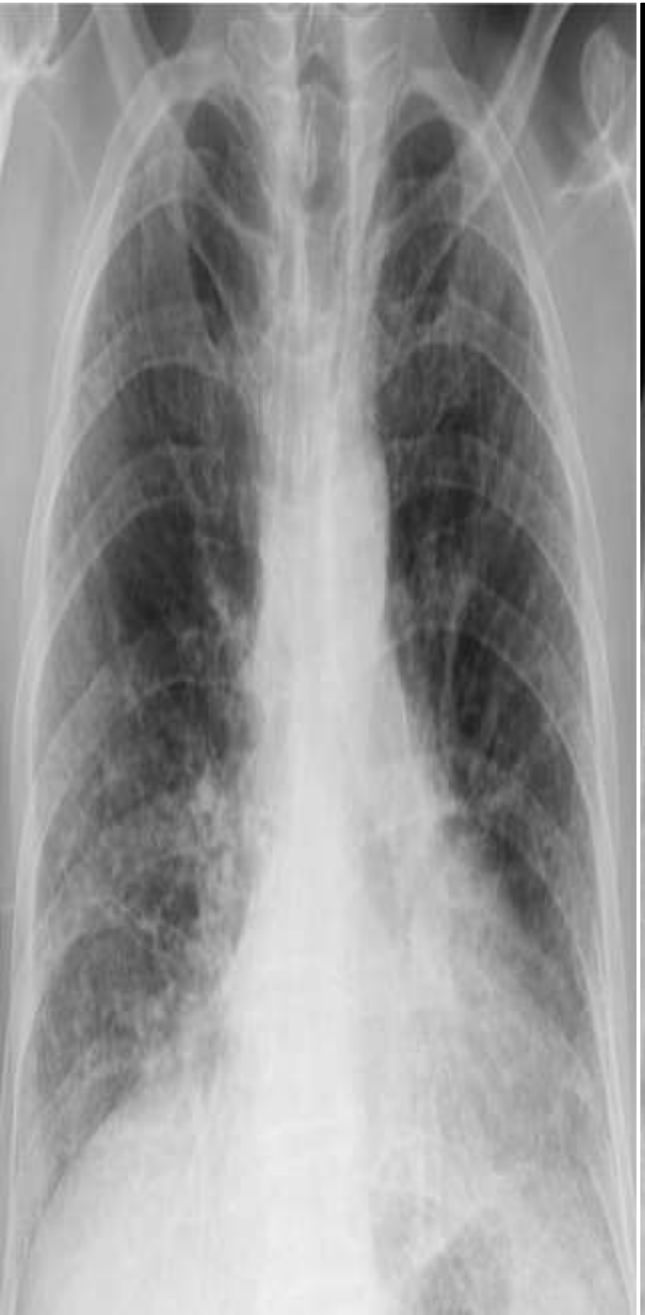
## LINEAR

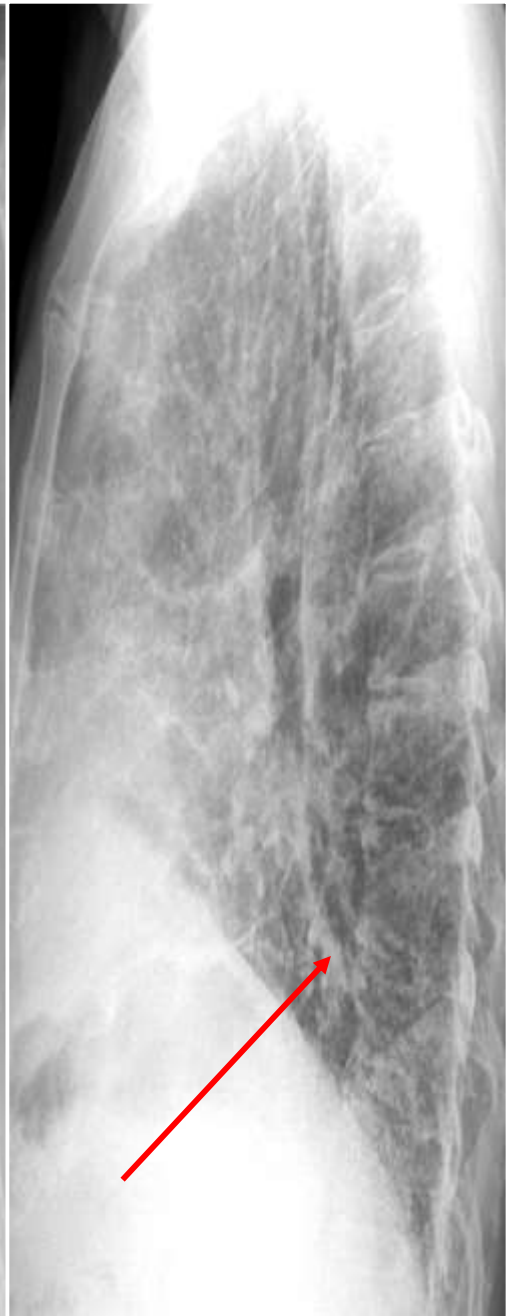


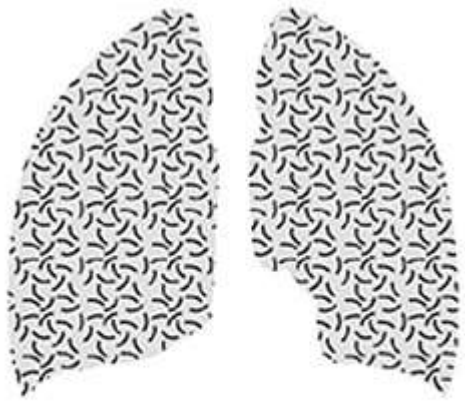


Linear

# RETICULAR

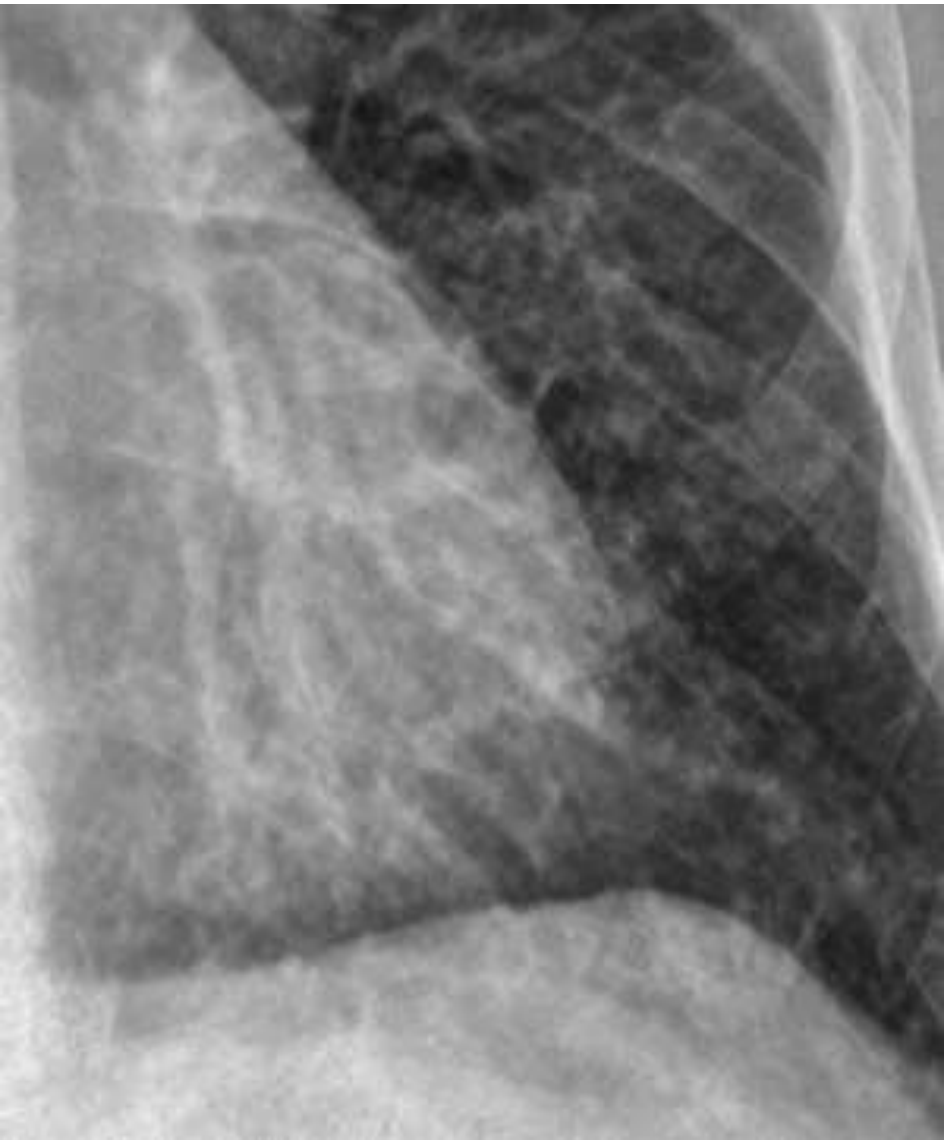


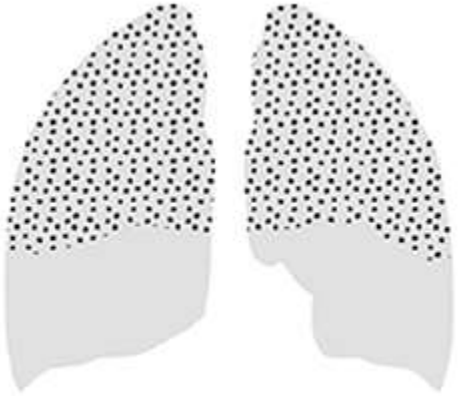




Reticular

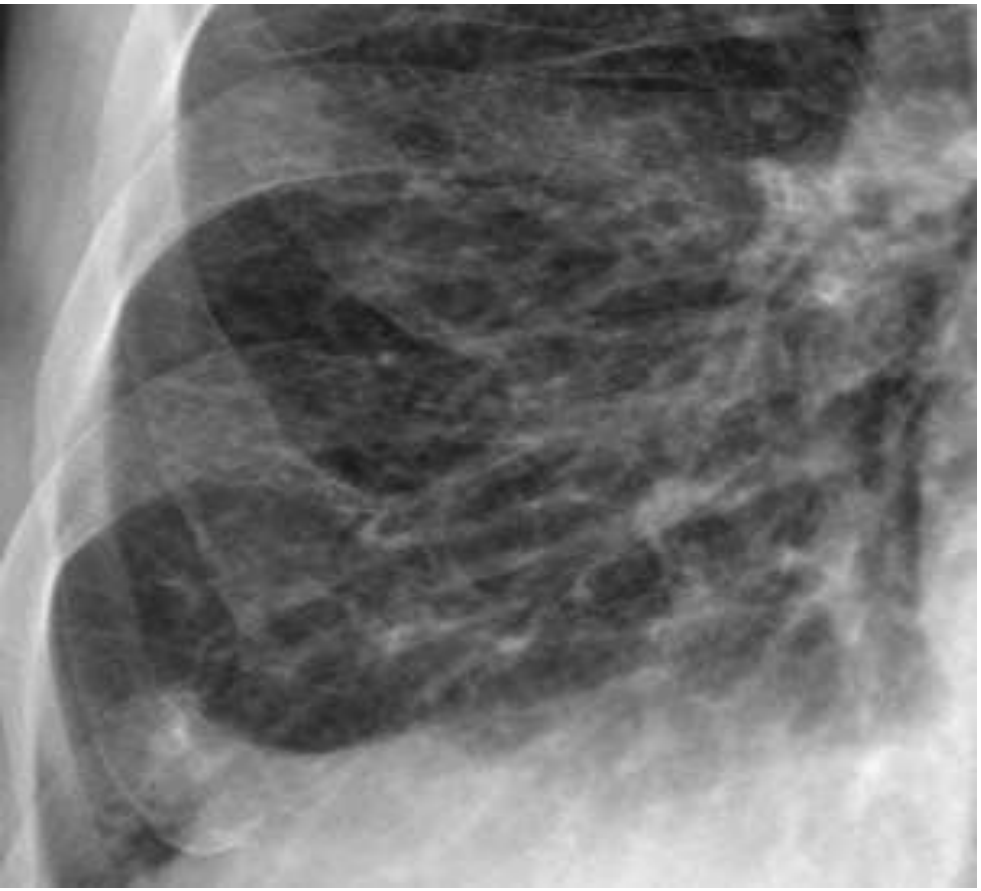
# NODULAR

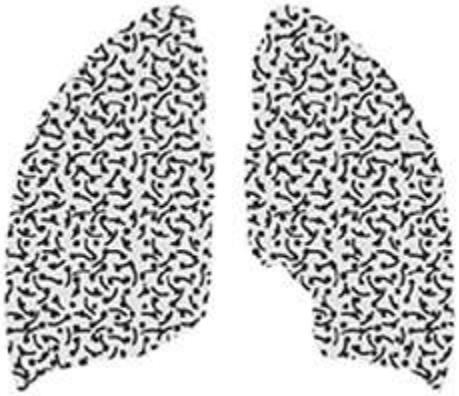




Nodular

# RETICULONODULAR





Reticulonodular

# HRCT CRITERIA FOR UIP PATTERN

| UIP | Probable UIP | Indeterminate for UIP |
|---|---|---|
| Subpleural and basal predominant; distribution is often heterogeneous* | Subpleural and basal predominant; distribution is often heterogeneous | Subpleural and basal predominant |
| Honeycombing with or without peripheral traction bronchiectasis or bronchiolectasis† | Reticular pattern with peripheral traction bronchiectasis or bronchiolectasis | Subtle reticulation; may have mild GGO or distortion ("early UIP pattern") |
| | May have mild GGO | CT features and/or distribution of lung fibrosis that do not suggest any specific etiology ("truly indeterminate for UIP") |

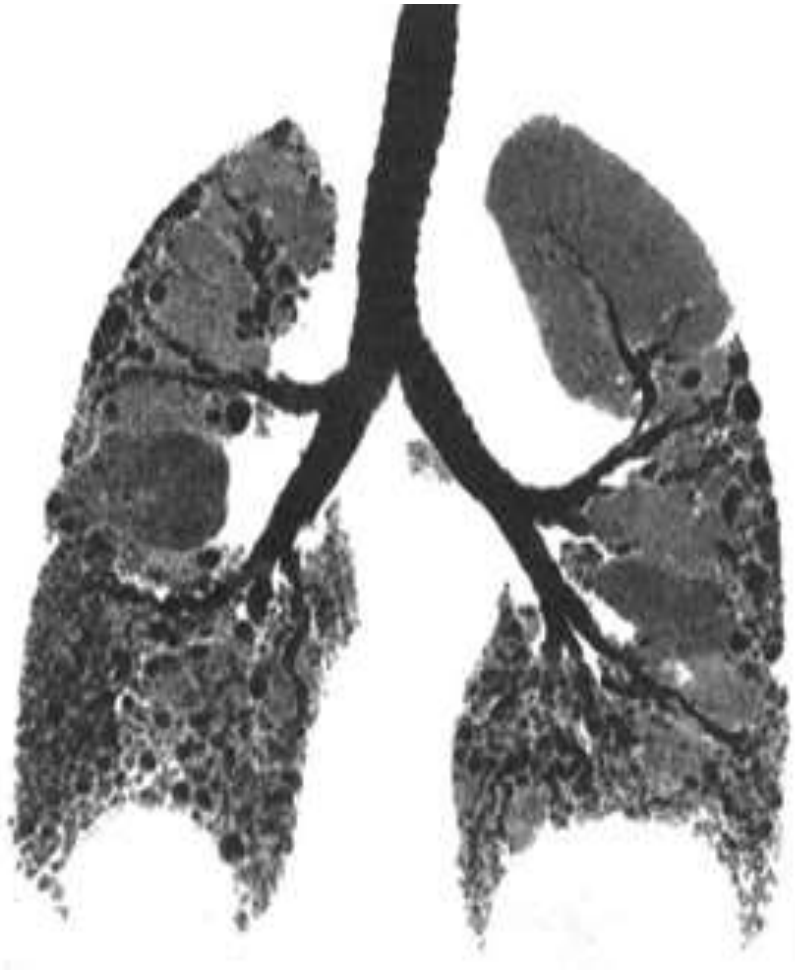
## Alternative Diagnosis

Findings suggestive of another diagnosis, including:

- CT features:
  - Cysts
  - Marked mosaic attenuation
  - Predominant GGO
  - Profuse micronodules
  - Centrilobular nodules
  - Nodules
  - Consolidation
- Predominant distribution:
  - Peribronchovascular
  - Perilymphatic
  - Upper or mid-lung
- Other:
  - Pleural plaques (consider asbestosis)
  - Dilated esophagus (consider CTD)
  - Distal clavicular erosions (consider RA)
  - Extensive lymph node enlargement (consider other etiologies)
  - Pleural effusions, pleural thickening (consider CTD/drugs)

# CRITERIA FOR DISEASE PROGRESSION

- Relative decline of ⩾10% in forced vital capacity (FVC)
- A relative decline of ⩾15% in diffusing capacity of the lung for carbon monoxide (DLCO)
- Worsening symptoms or a worsening radiological appearance accompanied by a ⩾5-<10% relative decrease in FVC



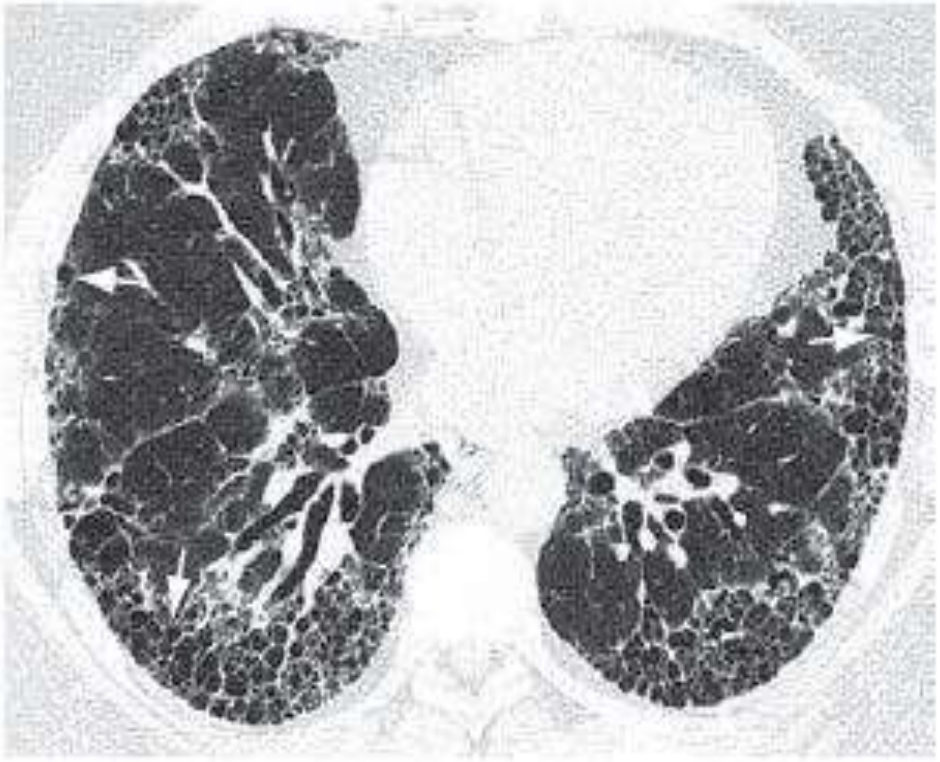


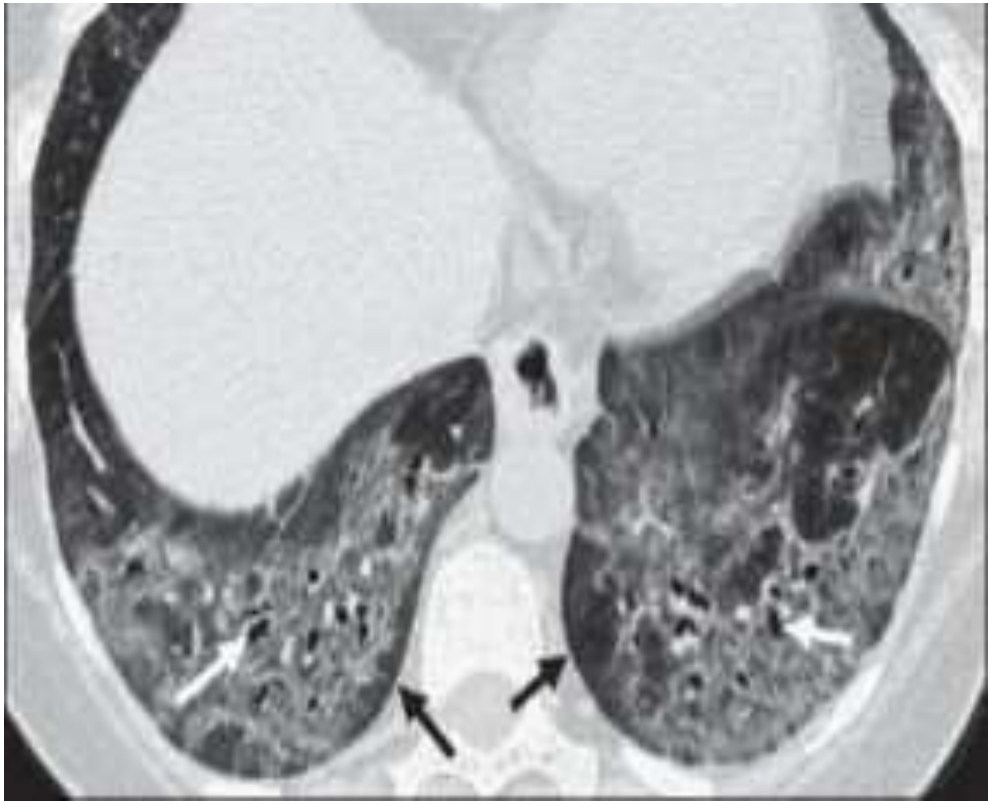
**Figure 27** – Honeycombing cysts (arrows) in a patient with pulmonary fibrosis.

# NONSPECIFIC INTERSTITIAL PNEUMONIA

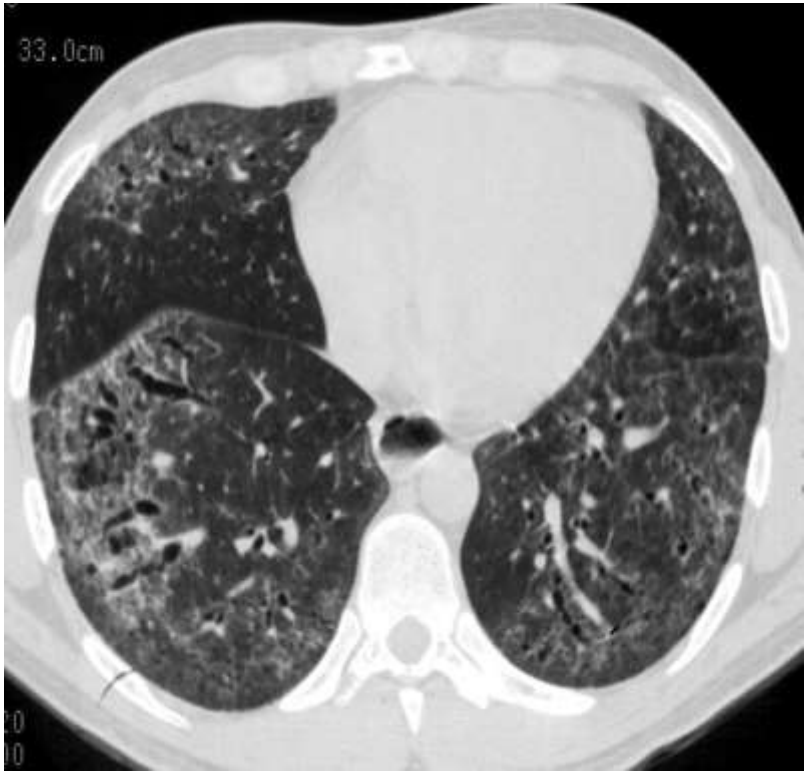
- Best diagnostic clue: traction bronchiectasis out of proportion to reticular opacities
- Location: peribronchovascular basilar distribution
- Reticular opacities: [80%], mixed with ground glass opacities, combined may cause crazy paving pattern
- Ground glass: [75%] , when present, often exceeds reticular opacities, 25-35% of lung volume is involved with it.

- Traction bronchiectasis: [80%], out of proportion to degree of reticular opacities, considerable lobar volume loss.
- Honeycombing: rare [5%]
- May have fine honeycombing-microcystic honeycombing
- Isolated enlarged airspaces may be present.
- Consolidation less common

- Distribution: symmetry
- Craniocaudal: lower 92%, diffuse 8%
- Axial: diffuse 60%, peripheral 35%
- Subpleural sparing 20%
- Mediastinal lymph node enlargement <2cm short axis diameter, MC- right paratracheal nodal station



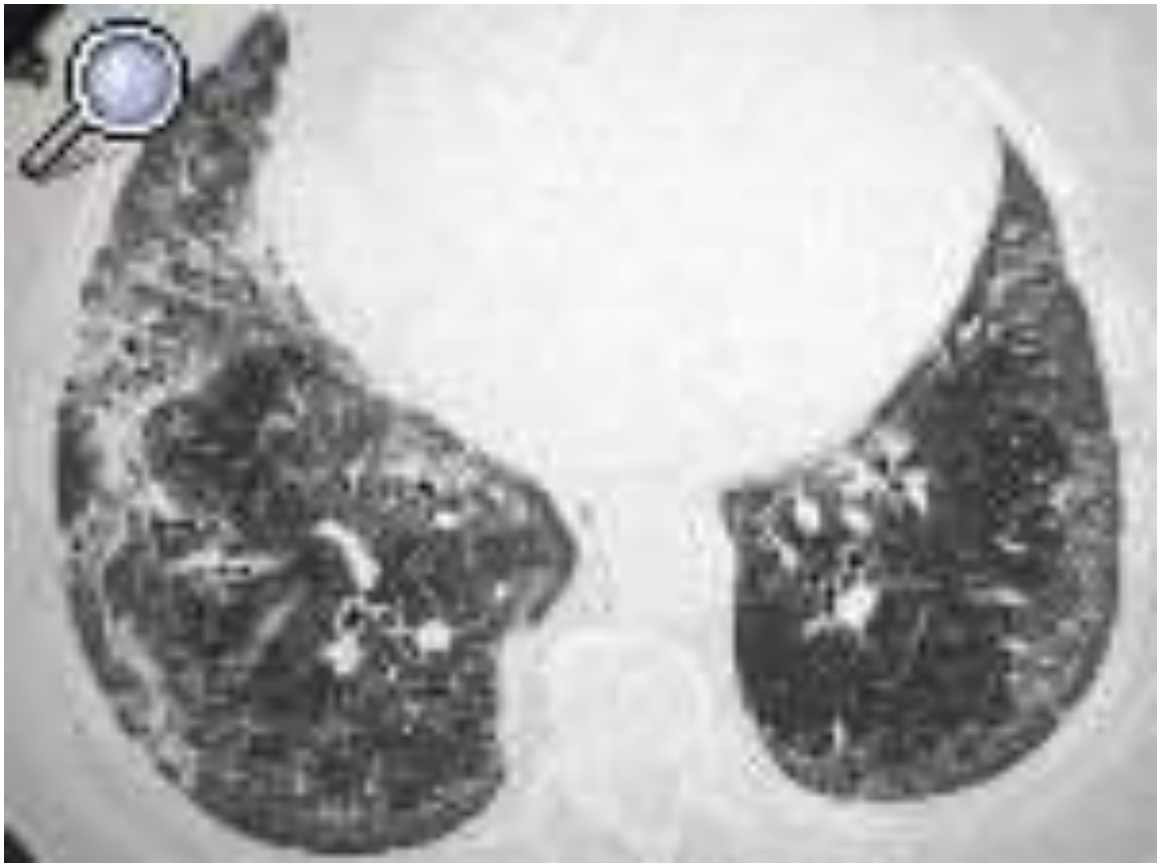
Peribronchovascular and basilar distribution of ground glass opacities with traction bronchiectasis, subpleural sparing is indicative of NSIP


33.0cm

# DIFFUSE INTERSTITIAL PNEUMONIA/DESQUAMATIVE INTERSTITIAL PNEUMONIA [DIP]

- Best diagnostic clue: diffuse ground glass opacities with scattered cysts in heavy smoker
- Location: lower lung predominance [70%], peripheral subpleural distribution [60%]
- Ground glass: 100%, panlobular, sharply demarcated from normal lung by interlobular septa, extent is correlating with pack years

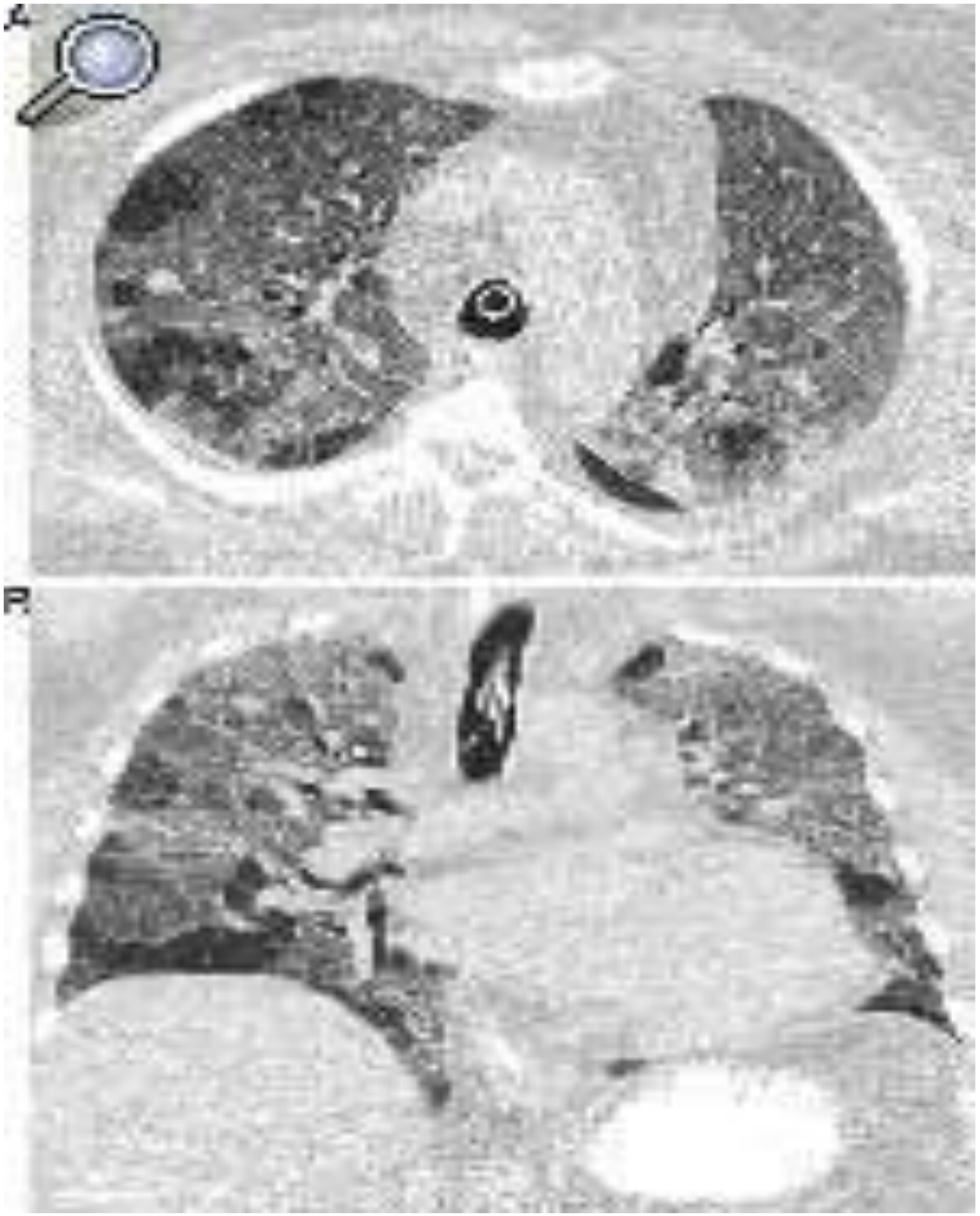
- Small well defined cysts [80%], <2cm in diameter, may resolve with treatment
- Superimposed emphysema common [60%].
- Honeycombing [10%]
- Nodules- uncommon, if present , consider superimposed respiratory bronchiolitis or langer hans cell histiocytosis [ all three can present together]



# ACUTE INTERSTITIAL PNEUMONIA/ HAMMAN RICH SYNDROME/ NON CARDIOGENIC PULMONARY EDEMA/ARDS/ DIFFUSE ALVEOLAR DAMAGE

- Best diagnostic clue: diffuse symmetrical air space opacification
- Location: lower lung zones , symmetry is the rule
- Ground glass opacities- predominant abnormality, >50% involvement, more extensive than consolidation, often sparing lobules [geographic pattern]
- More extensive ground glass opacity without traction bronchiectasis associated with better outcome

- Consolidation- in 2/3rd cases. Involves 25% of lung.
- Honeycombing –rare
- Traction bronchiectasis- combined with ground glass opacities, involves central airways more commonly than peripheral, out of proportion to reticular opacities

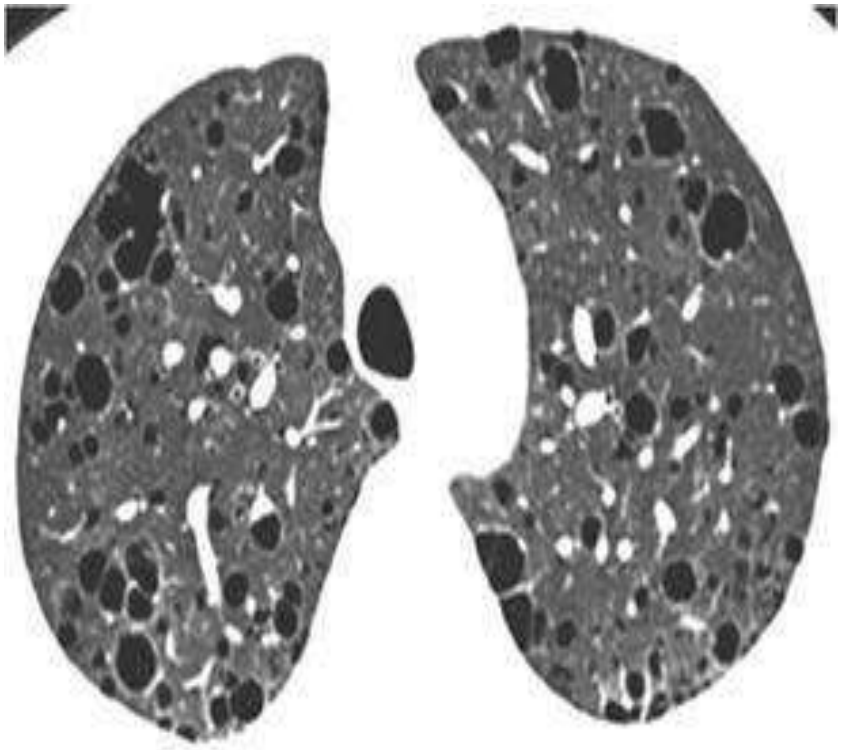

A.
P.

# LYMPHOCYTIC INTERSTITIAL PNEUMONIA [LIP]

- Best diagnostic clue- thin walled cysts and centrilobular nodules
- Location- centered on lymphatic pathways: peribronchovascular, centrilobular, septa and pleura

- Diffuse variety[referred as LIP]: ground glass opacities 100%, distributed bilateral [90%],
- Centrilobular nodules, poorly defined, 2-4mm
- Thin walled cysts- most distinctive finding [80%], range 1-30mm in diameter, involve <10% of lung, may be isolated finding
- Combination of ground glass opacities, centrilobular nodules and thin walled cysts

- Focal [ pseudolymphoma]: air-space mass, consolidation with air bronchograms, nodules >5mm [peribronchial], no lobar predilection, cavitation –rare
- All findings mat resolve except for cysts, centrilobular nodules may evolve into cysts, air space consolidation may evolve into honeycombing
- Lymph node may be enlarged [70%]- mc in AIDS, not large enough to be seen on x ray.
- Pleural effusions rare.



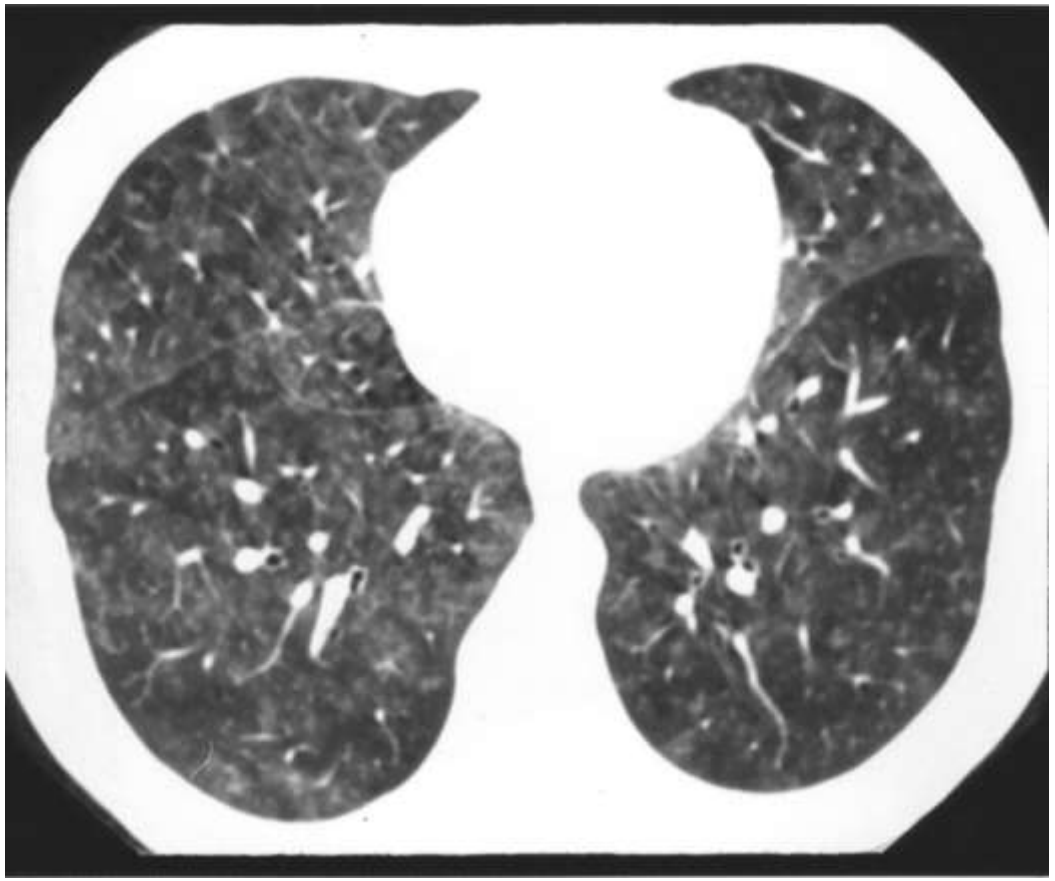
Lymphocytic interstitial pneumonia in a female patient with Sjogren's disease. HRCT at the level of the upper lobes exhibits a mixed "ground-glass and cystic pattern"

# HYPERSENSITIVITY PNEMONITIS/EXTRINSIC ALLERGIC ALVEOLITIS/FARMER'S LUNG

- Best diagnostic clue- ground glass centrilobular nodules & mosaic perfusion [ or lobular air trapping]
- Location : diffuse mid lung most common, typically spares costophrenic angles
- Ground glass- 100%, central and peripheral portions, non specific
- Centrilobular nodules [70%]-pleural surfaces usually spared
- Mosaic perfusion [80%] –due to air trapping

- Ground glass + normal lung+mosaic perfusion + air trapping = head cheese sign
- Ground glass centrilobular nodules +mosaic perfusion [ or lobular hyperinflation ] = HP until proven wrong
- Acute stage - diffuse ground glass opacities, small ill defined centrilobular nodules, nodules more likely to be found in less severely involved lung, air trapping common

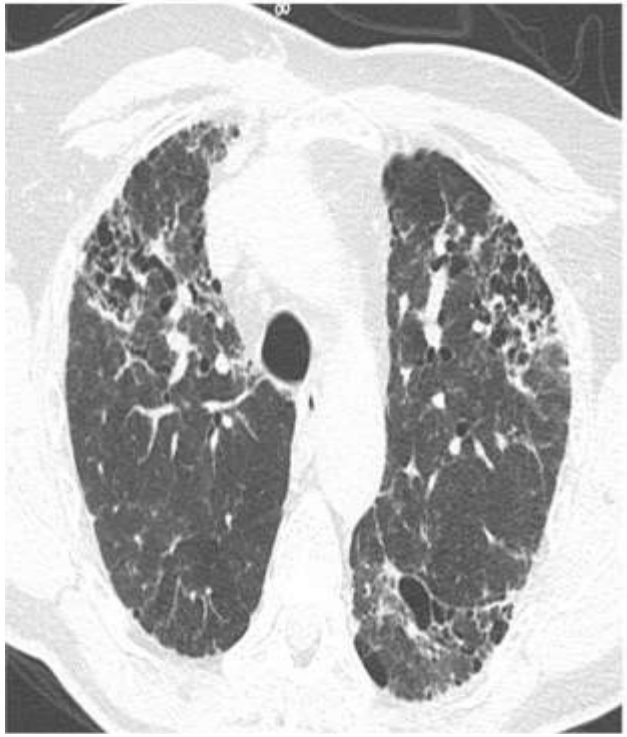
- sub acute stage: ground glass opacities [ patchy distribution] to mosaic perfusion, ill defined centrilobular nodules [ more common than in acute stage], LUNG CYSTS [10%] .
- Mediastinal adenopathy – 50%
- Pleural effusions- rare
- Resolution- lung may return to normal if avoidance of antigen or steroid therapy



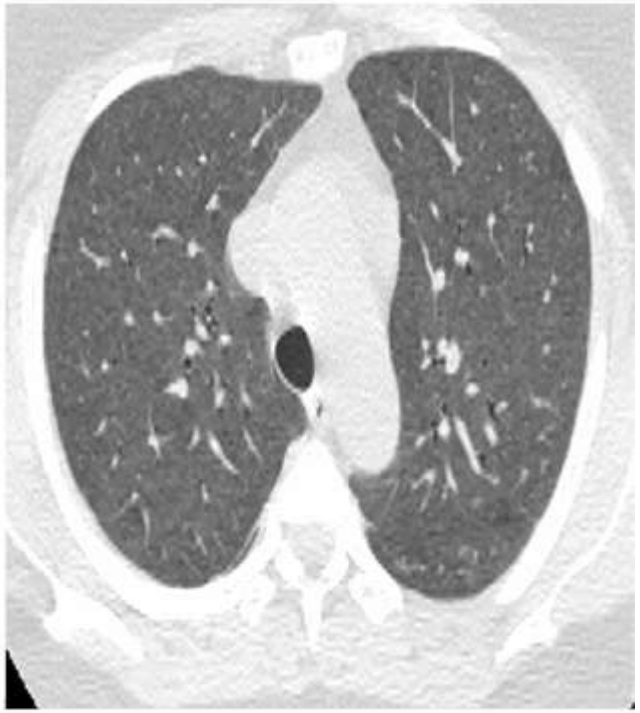
# CHRONIC HYPERSENTIVITY PNEUMONITIS

- Best diagnostic clue: ground glass [100%] + centrilobular opacities [60%] + lobular hyperinflation + signs of fibrosis [ traction bronchiectasis 40%, irregular reticular lines 20% , honeycombing 50% ]
- Location :
- Mid lung – in bird breeders and those with continuous exposure
- Upper lung – in farmers [ intermittent exposure ]
- Costophrenic angles less involved in contrast to IPF [ MC mimics IPF AND NSIP]

Chronic HP



Subacute HP

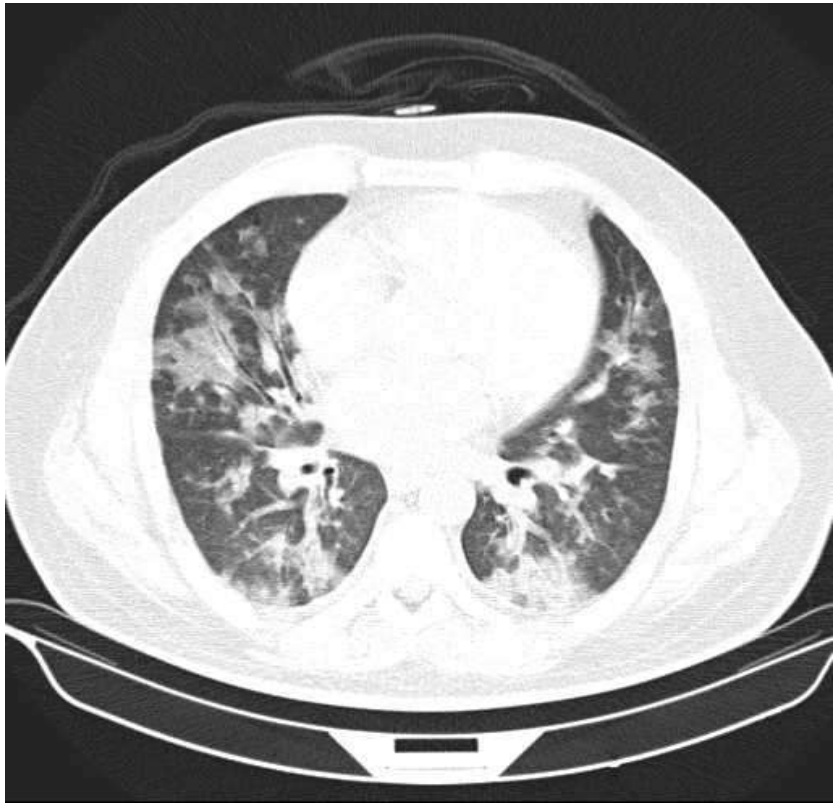


# CRYPTOGENIC ORGANISING PNEUMONIA/ BOOP

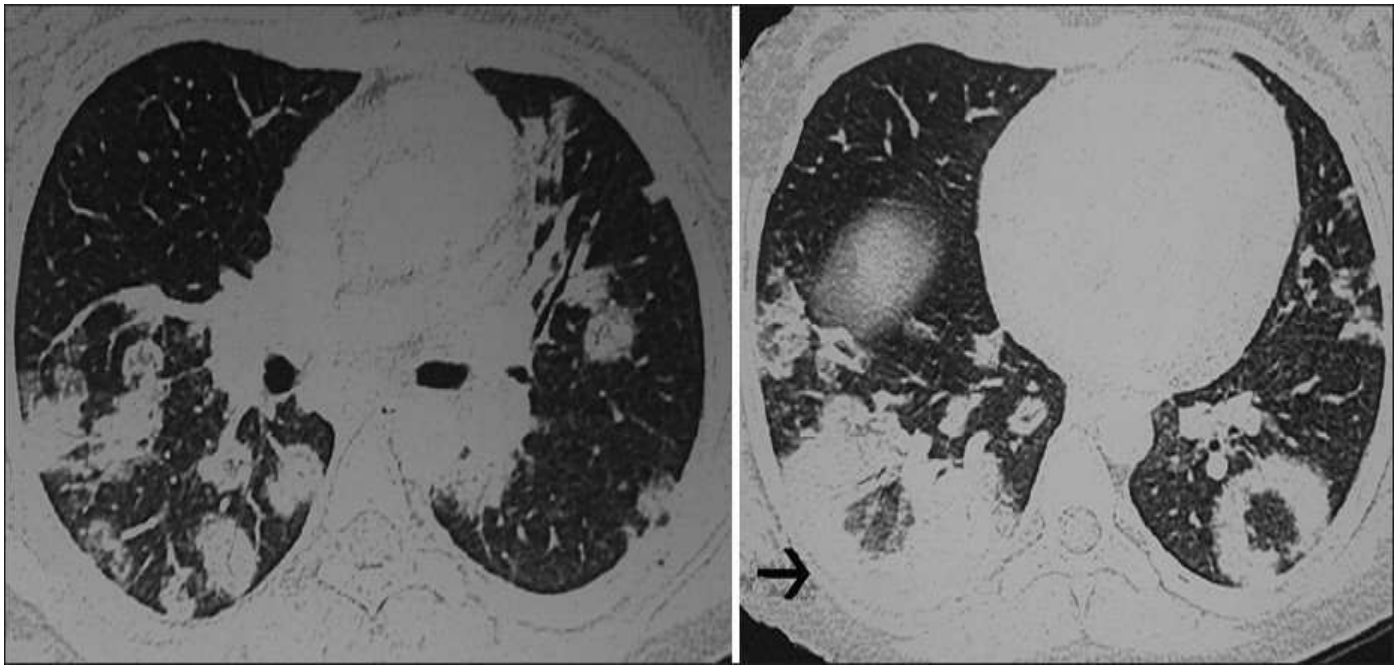
- Best diagnostic clue: bilateral, peripheral,basal nodular consolidation
- Location: typically in mid and lower zones
- Size: tiny nudoles to whole lobes
- Multiple alveolar opacities [90%]-
- Size of consolidation from few cm to whole lobe, admixed with ground glassing, air-bronchograms common, bilateral lower zones, lung volumes preserved, may be migratory, wax and wane over weeks, MC in immunocompetent.

- Presence of consolidation – likelihood to respond to steroids.
- Multiple nodules [10%]
- Single alveolar opacity [10%]- mimics bronchogenic carcinoma, <3cm [60%], >3cm [40%]. , mc in upper zones, pleural tag [50%], irregular margin [95%], satellite nodules [55%], vessels converge at the end of lesion [80%], may be cavitary.

- Plerual effusions – less common, when present small
- Mediastinal adenopathy- 20%.
- Reverse halo sign [20%]



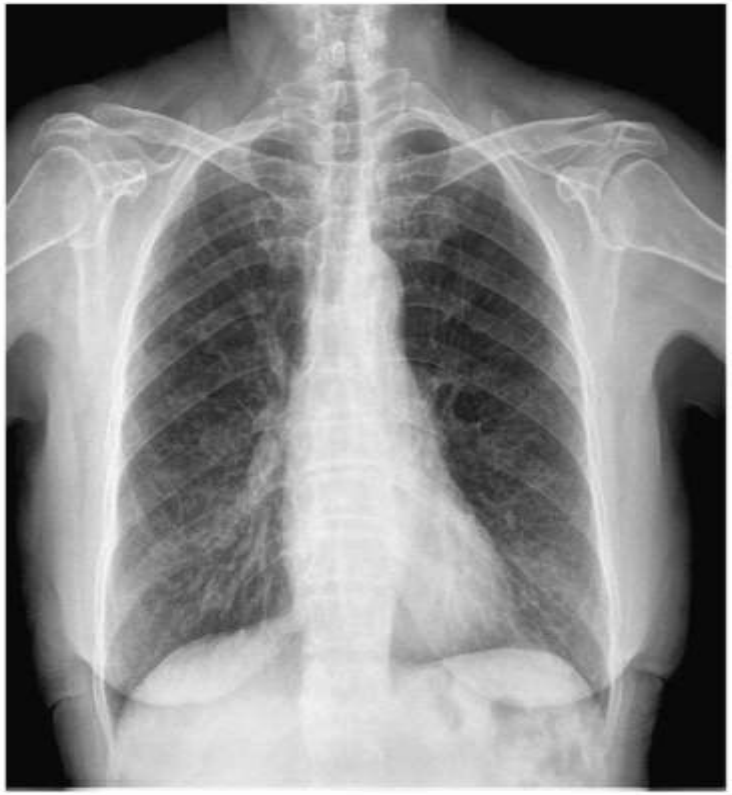
# REVERSE HALO SIGN

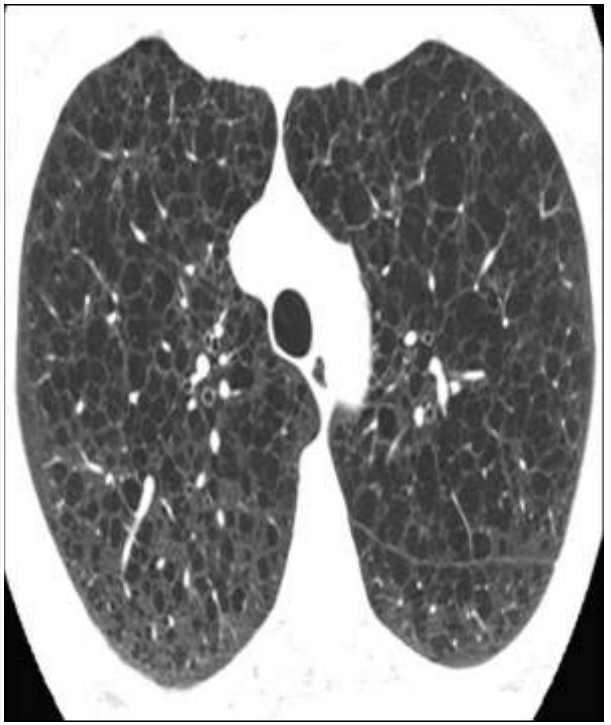


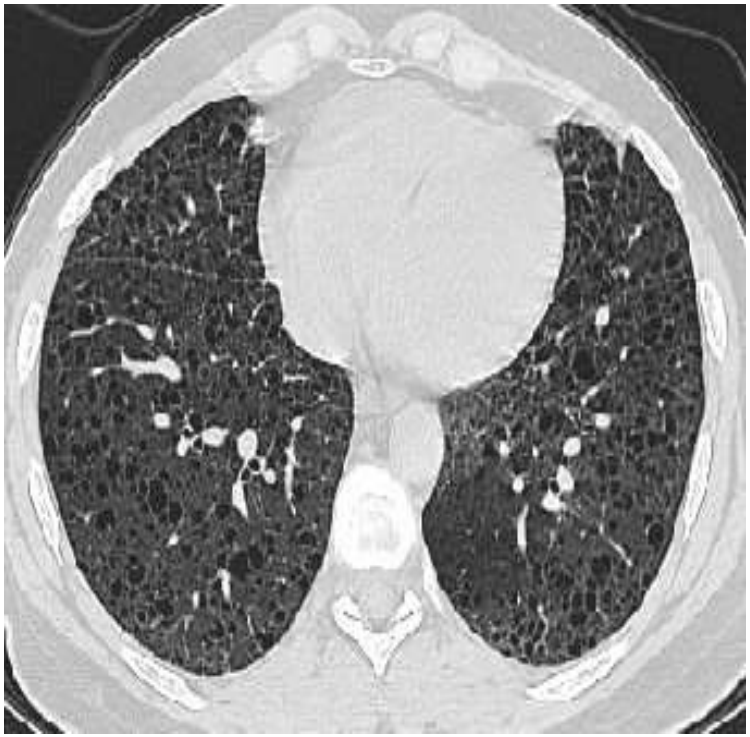
# LYMPHANGIOLEIOMYOMATOSIS

- Best diagnostic clue- woman of childbrearing age, progressive dyspnea
- Large volumes, reticular opacities, chylothorax or spontaneous pneumothorax
- Diffuse bilateral thin walled air-filled cysts with intervening normal lung
- Location: diffuse bilateral involvement
- Size: variable cysts 2-5mm, also 6-12mm possible
- Spherical cysts with smooth thin walls

- Relative sparing of lung apices and lung bases
- Round, ovoid or polygonal cysts
- Cysts wall – range from barely perceptible to 2-4 mm
- Pleura- pleural effusion, chylothorax, pneumothorax, hydropneumothorax.
- Lymphadenopathy- thoracic, abdominal, pelvic
- Renal angiomyolipomas [20-54%]





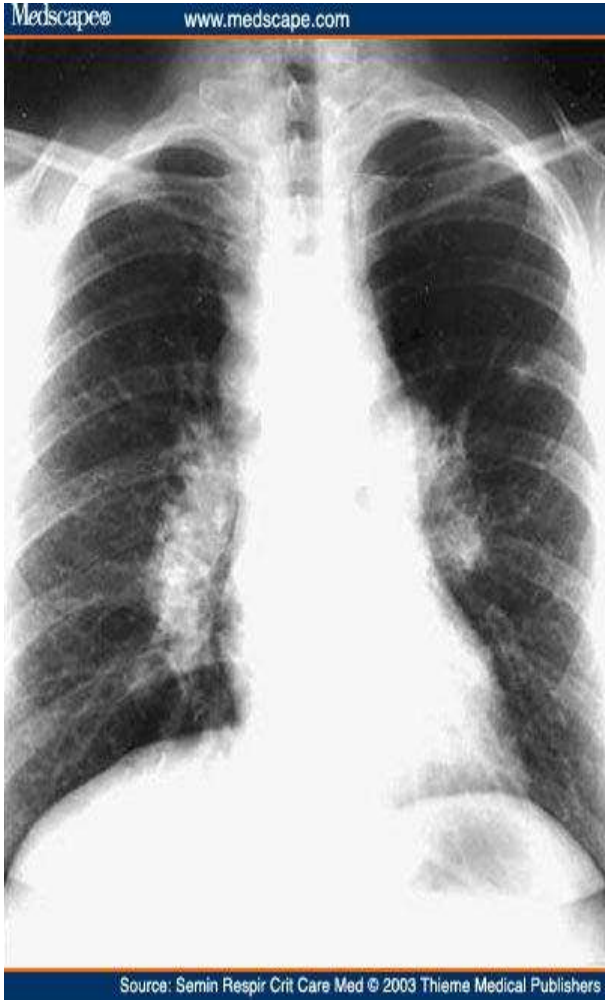


# SARCOIDOSIS

- Best diagnostic clue- young patient with symmetrical lymphadenopathy± bilateral small lung nodules
- Location: bilateral hilar paratracheal and AP window
- Upper lobe predominance
- Size: lymphadenopathy variable size, may be bulky
- Pulmonary opacities- small nodules to large masses

- Perilymphatic nodules [90-100%]: axial peribronchovascular interstitium, centrilobular/septal/pleural, small, well-defined with nodular thickening of vessel margins and airway walls, nodular interlobular septal thickening, subpleural nodules and centriobular nodules.
- Multifocal patchy air space opacities: nodular consolidation, ground glass opacities, intrinsic air bronchograms, can cause pneumothorax
- Both nodules and air space- in mid and upper lung zones

- Lymphadenopathy:
- Symmetric, bilateral [hilar/paratracheal, AP window, subcarinal
- Calcification with chronic disease: amorphous, punctate, egg shell
- Pleural effusion –rare
- Nuclear scan: GALLIUM 67 SCINTIGRAPHY:
- Lamba pattern in paratracheal and hilar nodes
- Panda sign in lacrimal and parotid uptake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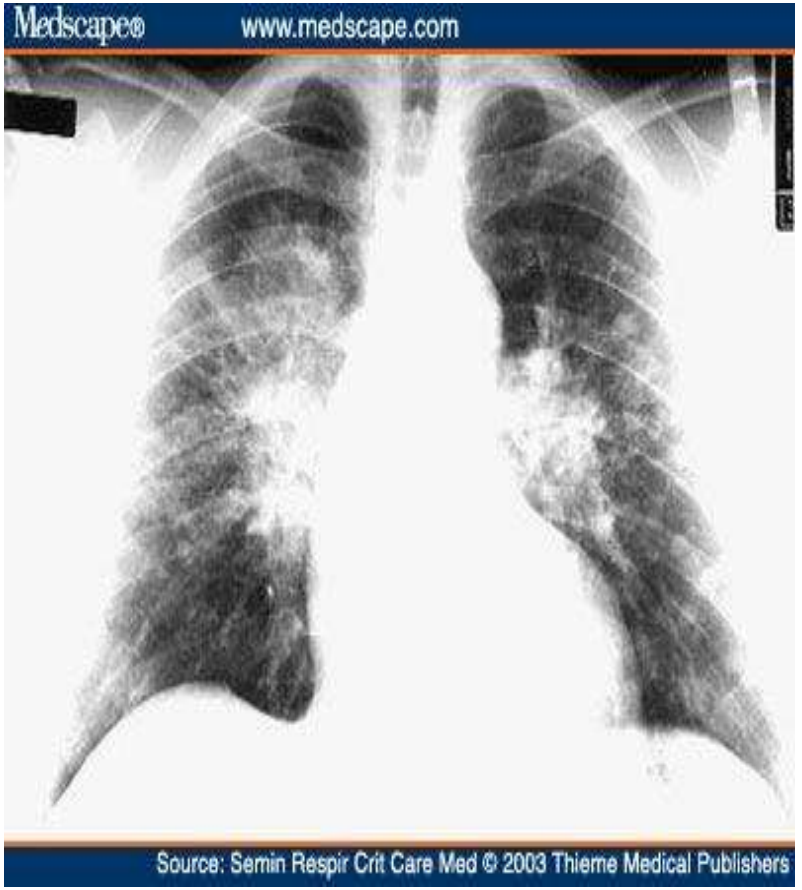
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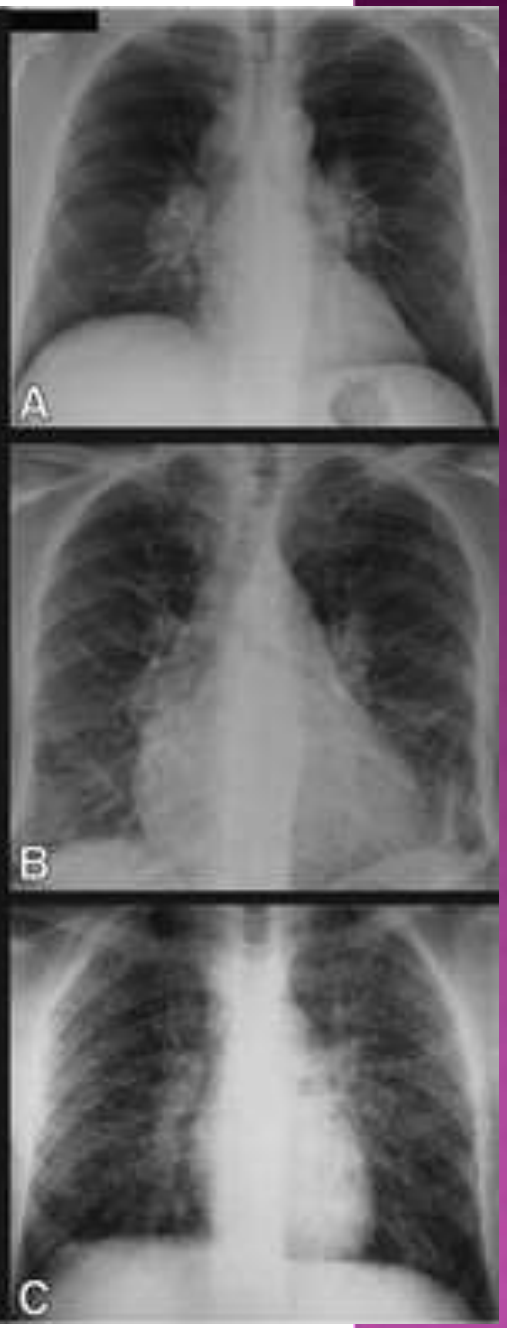
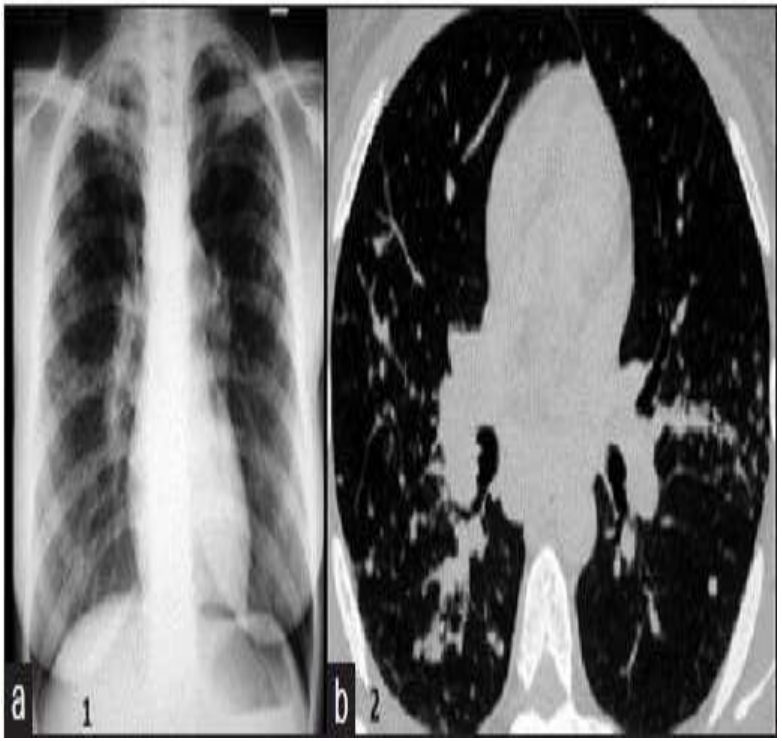
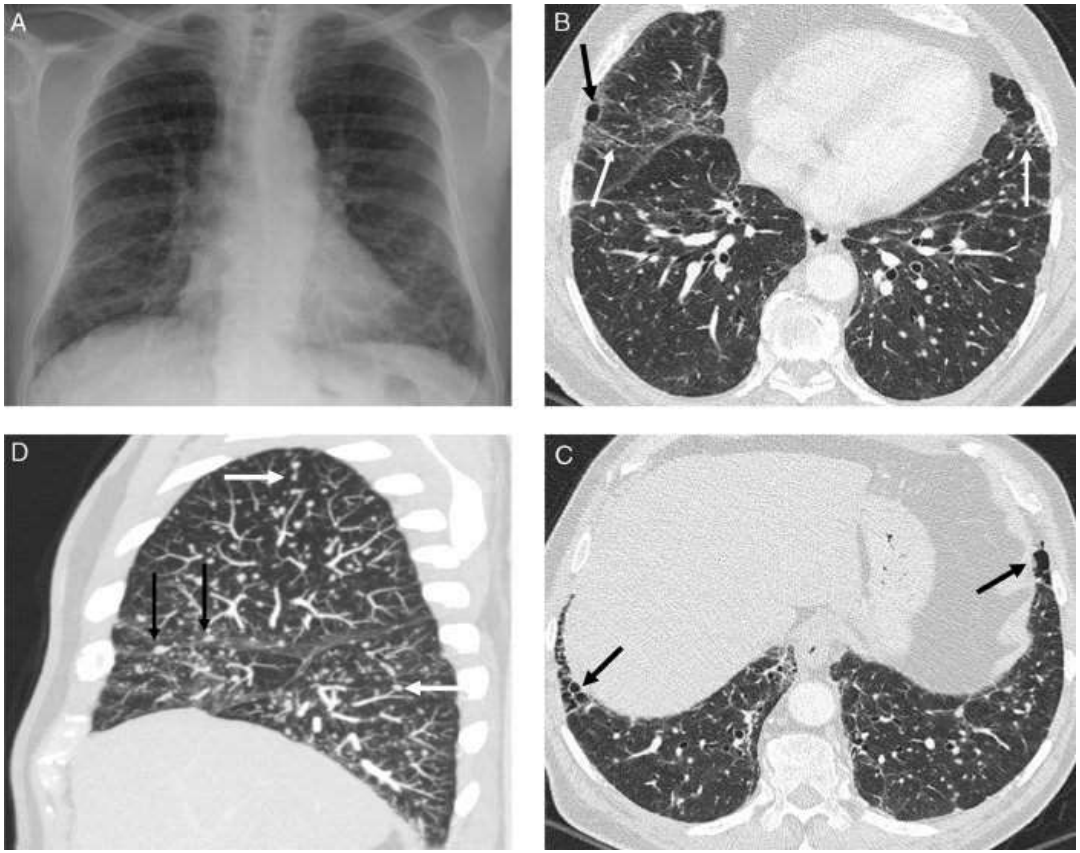
## Staging of Sarcoidosis on the Basis of Chest Radiographs

| | | |
|---|---|---|
| STAGE 0 | No abnormalities | 5%–10% |
| STAGE 1 | Lymphadenopathy (fig. A) | 50% |
| STAGE 2 | Lymphadenopathy + pulmonary infiltration (fig. B) | 25%–30% |
| STAGE 3 | Pulmonary infiltration (fig. C) | 10%–12% |
| STAGE 4 | Fibrosis | 5% (up to 25% during the course of the disease) |

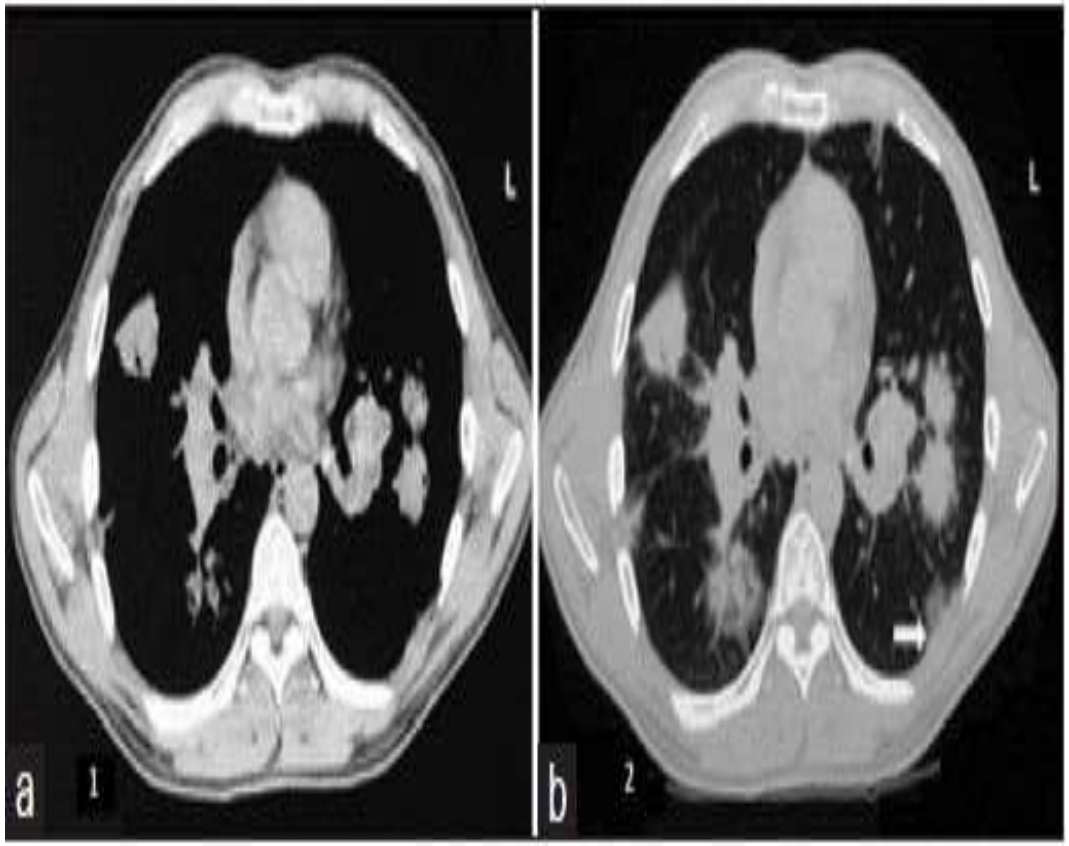




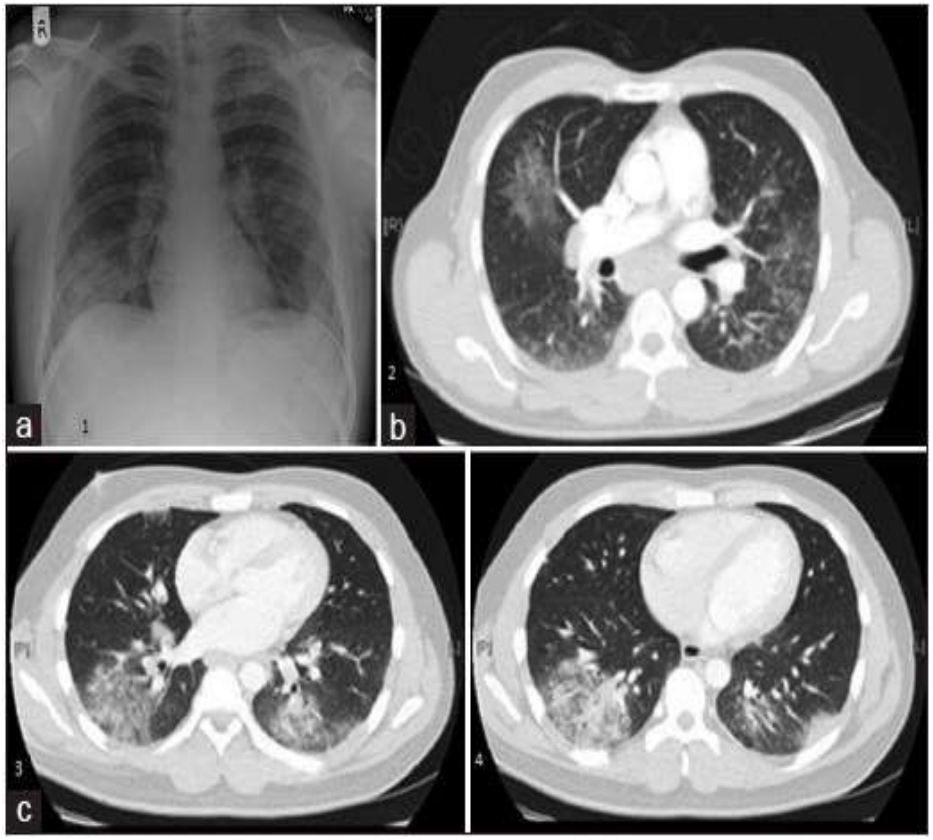

small nodules mostly along the bronchovascular bundles, giving the bronchi and vessels a beaded appearance. This distribution along the bronchovascular bundles accounts for the fact that transbronchial biopsy is usually successful for obtaining tissue for diagnosis.



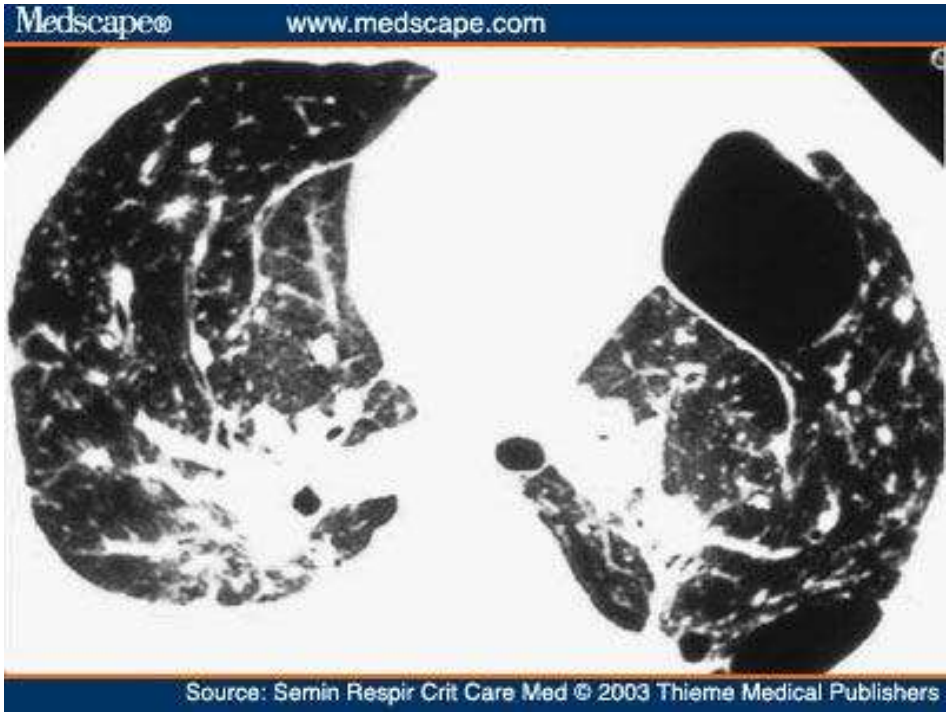

Periplymphatic nodules




mediastinal lymphadenopathy. Several large, ill-defined nodules and areas of consolidation resulting from the confluence of lung nodules are noted in both lung fields. small low-attenuation spaces that correspond to the spaces between partially coalescent small nodules are visible peripherally giving rise to the so-called sarcoid galaxy sign




ground-glass opacities and mosaic attenuation pattern. Patient was 27-year-old with an angiotensin-converting-enzyme level of 90 IU/L




stage IV sarcoidosis demonstrates bulla formation, central bronchovascular thickening, and architectural distortion with rotation of the hila toward the spine. The lung interface with the pleura is irregular because of subpleural nodules

# INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES (IPAF)

- Clinical features that suggest an underlying autoimmune process but do not meet established criteria for a connective tissue disease (CTD).

# CLASSIFICATION CRITERIA

1. Presence of an interstitial pneumonia (by HRCT or surgical lung biopsy) *and*,
2. Exclusion of alternative aetiologies *and*,
3. Does not meet criteria of a defined connective tissue disease *and*,
4. At least one feature from at least two of these domains:
   A. Clinical domain
   B. Serologic domain
   C. Morphologic domain

A. Clinical domain

1. Distal digital fissuring (*i.e.* "mechanic hands")
2. Distal digital tip ulceration
3. Inflammatory arthritis *or* polyarticular morning joint stiffness ⩾60 min
4. Palmar telangiectasia
5. Raynaud's phenomenon
6. Unexplained digital oedema
7. Unexplained fixed rash on the digital extensor surfaces (Gottron's sign)

B. Serologic domain

1. ANA ⩾1:320 titre, diffuse, speckled, homogeneous patterns *or*
   a. ANA nucleolar pattern (any titre) *or*
   b. ANA centromere pattern (any titre)
2. Rheumatoid factor ⩾2× upper limit of normal
3. Anti-CCP
4. Anti-dsDNA
5. Anti-Ro (SS-A)
6. Anti-La (SS-B)
7. Anti-ribonucleoprotein
8. Anti-Smith
9. Anti-topoisomerase (Scl-70)
10. Anti-tRNA synthetase (*e.g.* Jo-1, PL-7, PL-12; others are: EJ, OJ, KS, Zo, tRS)
11. Anti-PM-Scl
12. Anti-MDA-5

C. Morphologic domain

1. Suggestive radiology patterns by HRCT (see text for descriptions):
   a. NSIP
   b. OP
   c. NSIP with OP overlap
   d. LIP
2. Histopathology patterns or features by surgical lung biopsy:
   a. NSIP
   b. OP
   c. NSIP with OP overlap
   d. LIP
   e. Interstitial lymphoid aggregates with germinal centres
   f. Diffuse lymphoplasmacytic infiltration (with or without lymphoid follicles)
3. Multi-compartment involvement (in addition to interstitial pneumonia):
   a. Unexplained pleural effusion or thickening
   b. Unexplained pericardial effusion or thickening
   c. Unexplained intrinsic airways disease[#] (by PFT, imaging or pathology)
   d. Unexplained pulmonary vasculopathy

# MCQs

1)PFT in ILDs will show

a)Reduction in TLC

b)Increase in functional residual capacity

c)Increase residual volume

d)All of the above

2)Which of the following is false about DIP

a)Found exclusively in cigarette smokers

b)Macrophages in intraalveolar spaces

c)Minimal interstitial fibrosis

d)Worse prognosis than IPF

3)Hamman rich syndrome is name given to

a)Acute interstitial pneumonia

b)Hypersensitivity pneumonitis

c)DIP

d)Respiratory bronchiolitis

4)Most common form of pulmonary involvement in connective tissue disorders

a)Cryptogenic organizing pneumonia

b)Desquamative interstitial pneumonia

c)Respiratory bronchiolitis

d)Nonspecific interstitial pneumonia

5)Which of the following is known as BOOP

a)Cryptogenic organizing pneumonia

b)Desquamative interstitial pneumonia

c)Respiratory bronchiolitis

d)Lymphocytic interstitial pneumonia

6)Variety of ILD associated with smoking is

a)Acute interstitial pneumonia

b)Respiratory bronchiolitis

c)Idiopathic pulmonary fibrosis

d)Non specific interstitial pneumonia

# THANQ...