

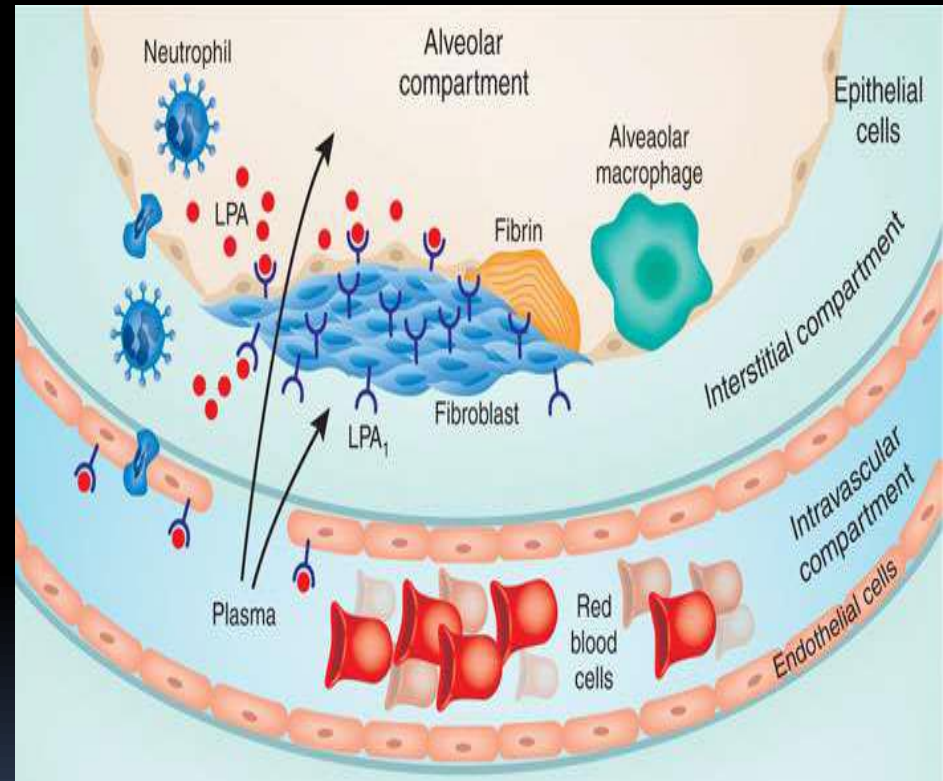
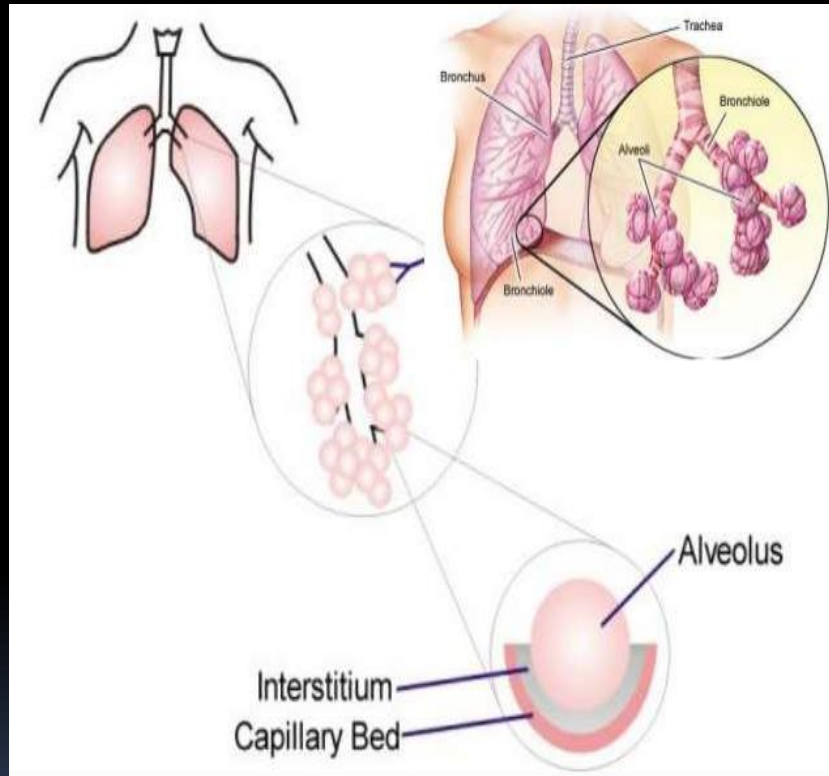


DIFFUSE PARENCHYMAL LUNG DISEASES/ INTERSTITIAL LUNG DISEASES



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INTRODUCTION- The interstitium





INTRODUCTION

- Interstitial is a misleading terminology because most of these disorders are associated with extensive alteration of airway and alveolar architecture in addition to changes in interstitial compartment.
 - For this reason Diffuse Parenchymal Lung Disease or DPLD is the better term.
- 



DEFINITION

- The idiopathic interstitial pneumonias (IIPs) are a group of diffuse parenchymal lung diseases of unknown etiology with varying degrees of inflammation and fibrosis
- 



In 1969, Liebow and Carrington published histopathologic classification schema for the IIPs

- **Usual interstitial pneumonia (UIP)**
- **Bronchiolitis obliterans interstitial pneumonia and diffuse alveolar damage**
- **Desquamative interstitial pneumonia**
- **Lymphocytic interstitial pneumonia (LIP)**
- **Giant cell interstitial pneumonia**



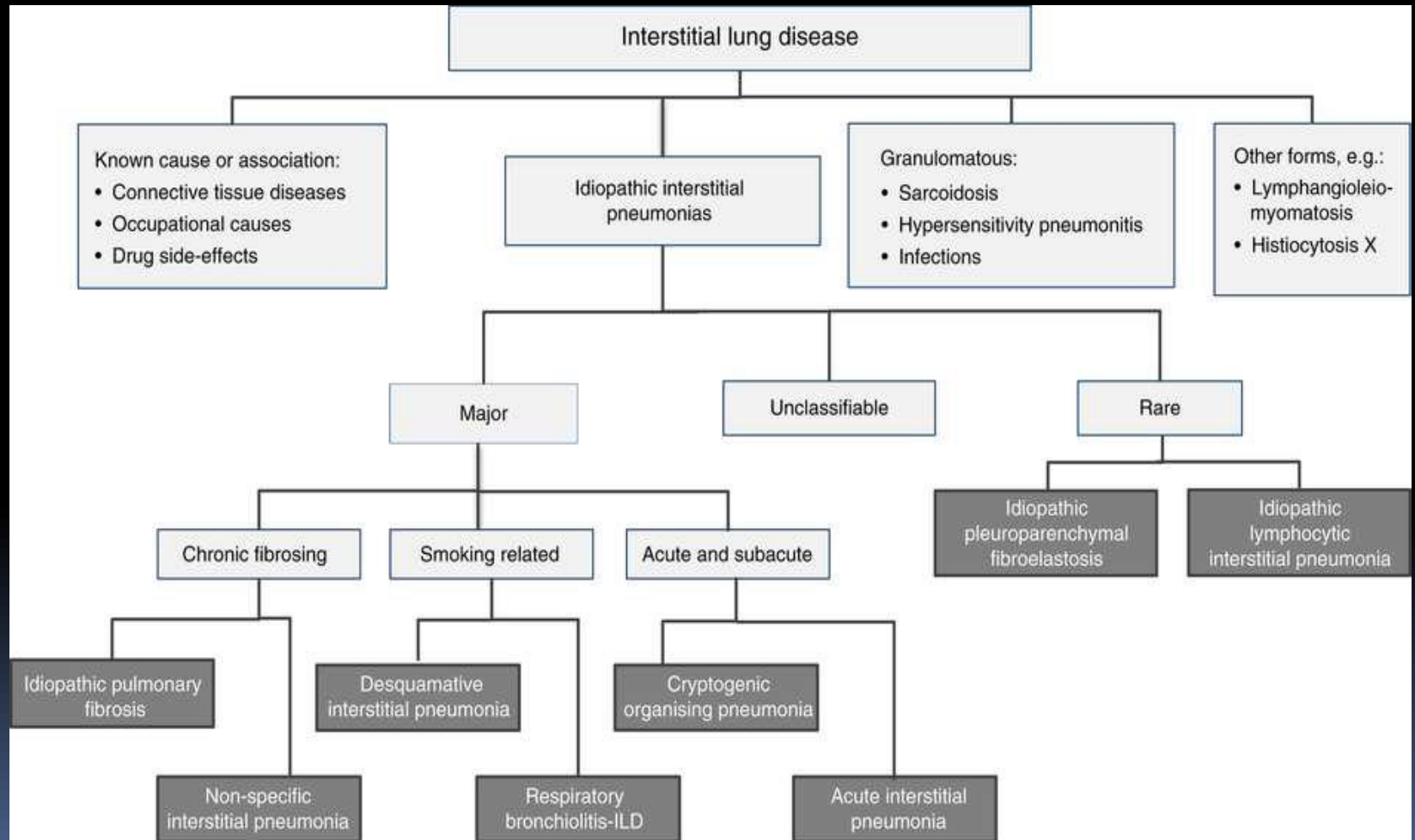
Katzenstein and Myers incorporated several key distinguishing features: the temporal heterogeneity of inflammation and fibrosis, the extent of inflammation, the extent of fibroblastic proliferation, the extent of accumulation of intraalveolar macrophages, and the presence of honeycombing or hyaline membranes .



CLASSIFICATION

- (1) Idiopathic DPLD (no known cause)-IIPs
 - (2) Those with identifiable cause or occurs secondary to other diseases
- 

CLASSIFICATION



CLASSIFICATION

TABLE 1. REVISED AMERICAN THORACIC SOCIETY/EUROPEAN RESPIRATORY SOCIETY CLASSIFICATION OF IDIOPATHIC INTERSTITIAL PNEUMONIAS: MULTIDISCIPLINARY DIAGNOSES

Major idiopathic interstitial pneumonias

Idiopathic pulmonary fibrosis

Idiopathic nonspecific interstitial pneumonia

Respiratory bronchiolitis–interstitial lung disease

Desquamative interstitial pneumonia

Cryptogenic organizing pneumonia

Acute interstitial pneumonia

Rare idiopathic interstitial pneumonias

Idiopathic lymphoid interstitial pneumonia

Idiopathic pleuroparenchymal fibroelastosis

Unclassifiable idiopathic interstitial pneumonias*

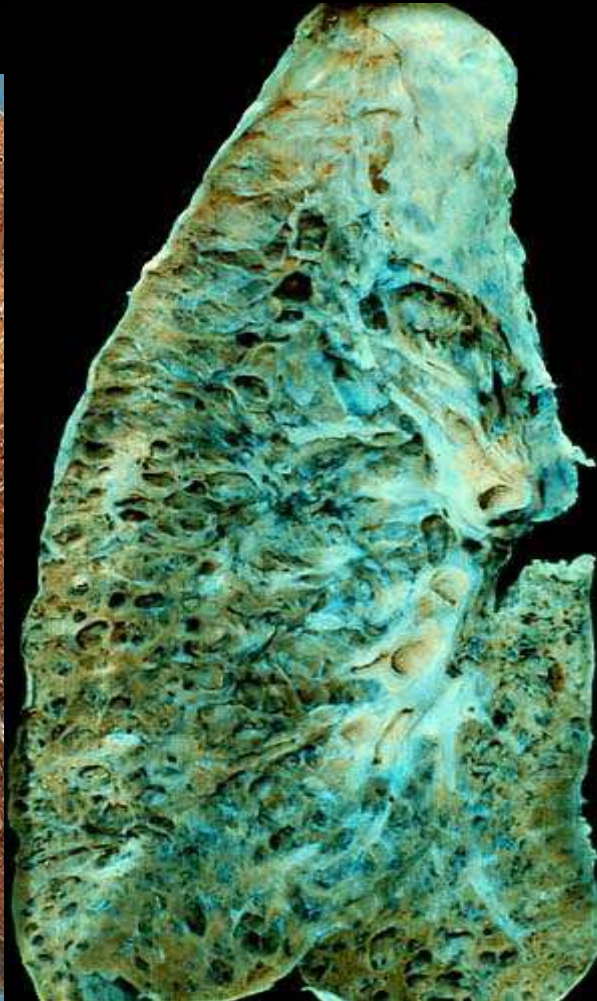
* Causes of unclassifiable idiopathic interstitial pneumonia include (1) inadequate clinical, radiologic, or pathologic data and (2) major discordance between clinical, radiologic, and pathologic findings that may occur in the following situations: (a) previous therapy resulting in substantial alteration of radiologic or histologic findings (e.g., biopsy of desquamative interstitial pneumonia after steroid therapy, which shows only residual nonspecific interstitial pneumonia [153]); (b) new entity, or unusual variant of recognized entity, not adequately characterized by the current American Thoracic Society/European Respiratory Society classification (e.g., variant of organizing pneumonia with supervening fibrosis) (79); and (c) multiple high-resolution computed tomography and/or pathologic patterns that may be encountered in patients with idiopathic interstitial pneumonia.



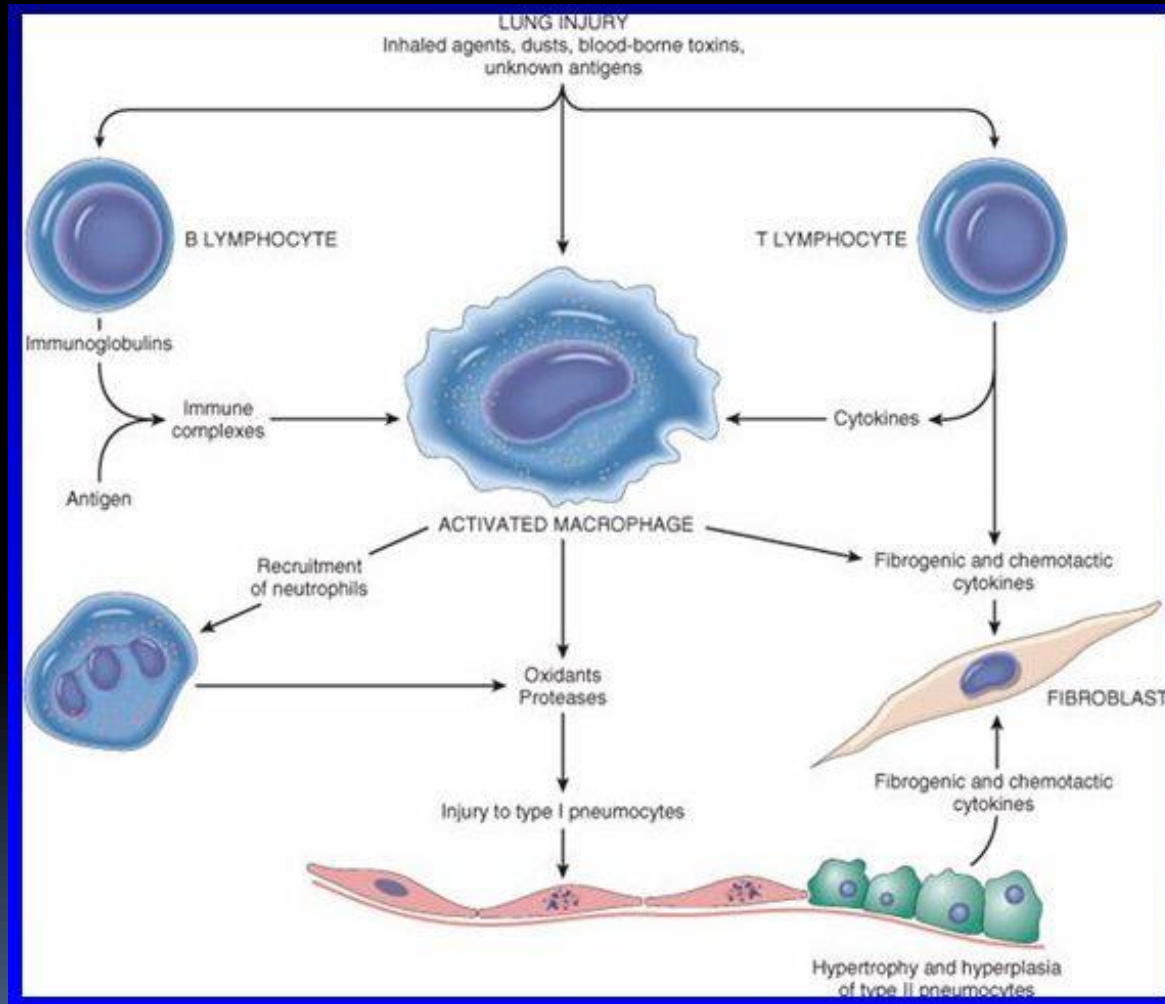
EPIDEMIOLOGY

- Incidence ranges from 3-26/1,00,000 per year.
- • Prevalence of preclinical and undiagnosed ILD is estimated
- to be 10 times that of clinical recognized disease.
- • IPF is the most common form representing at least 30 percent of the incident cases
- In India hypersensitivity pneumonitis and NSIP are more common

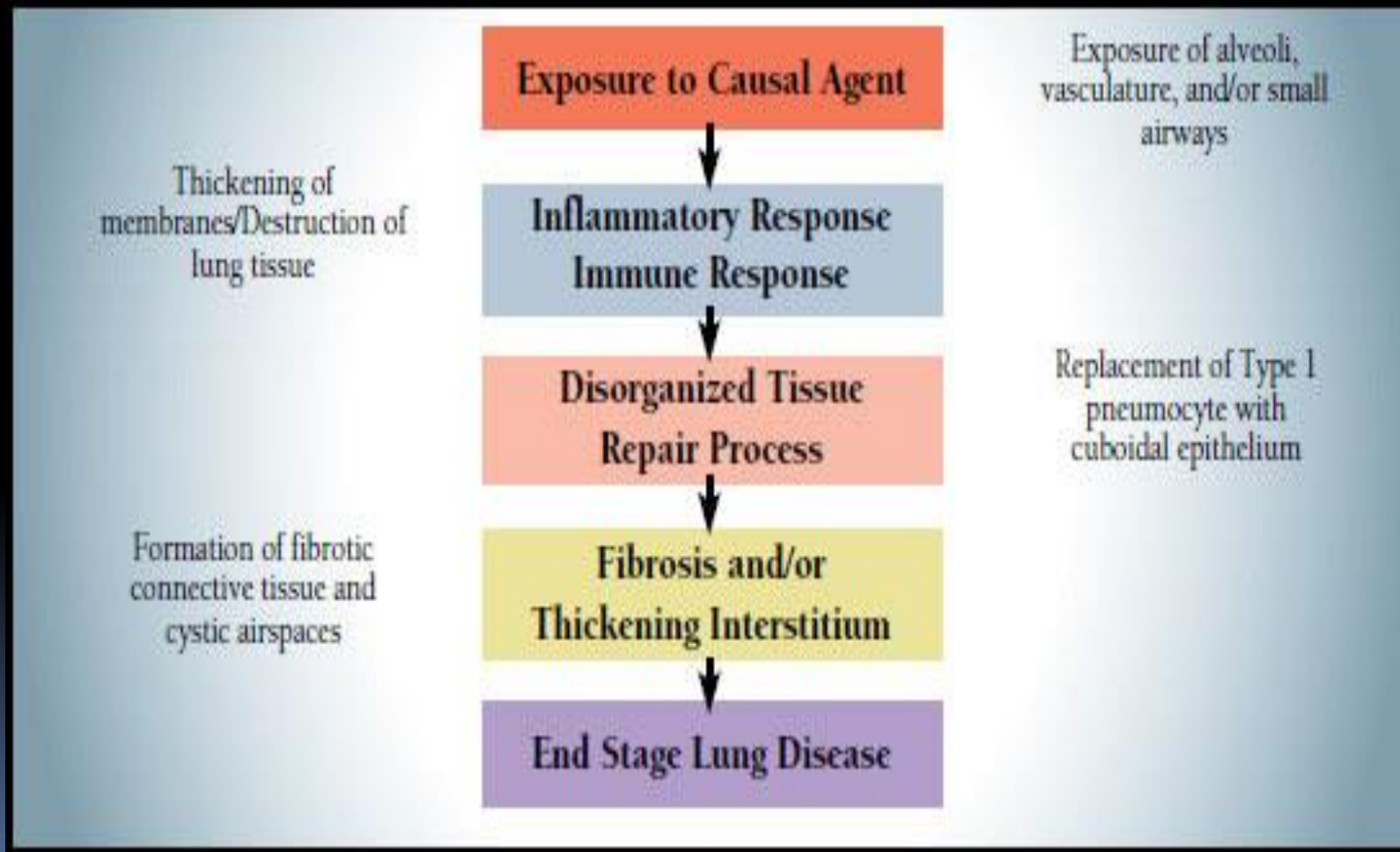
PATHOLOGY



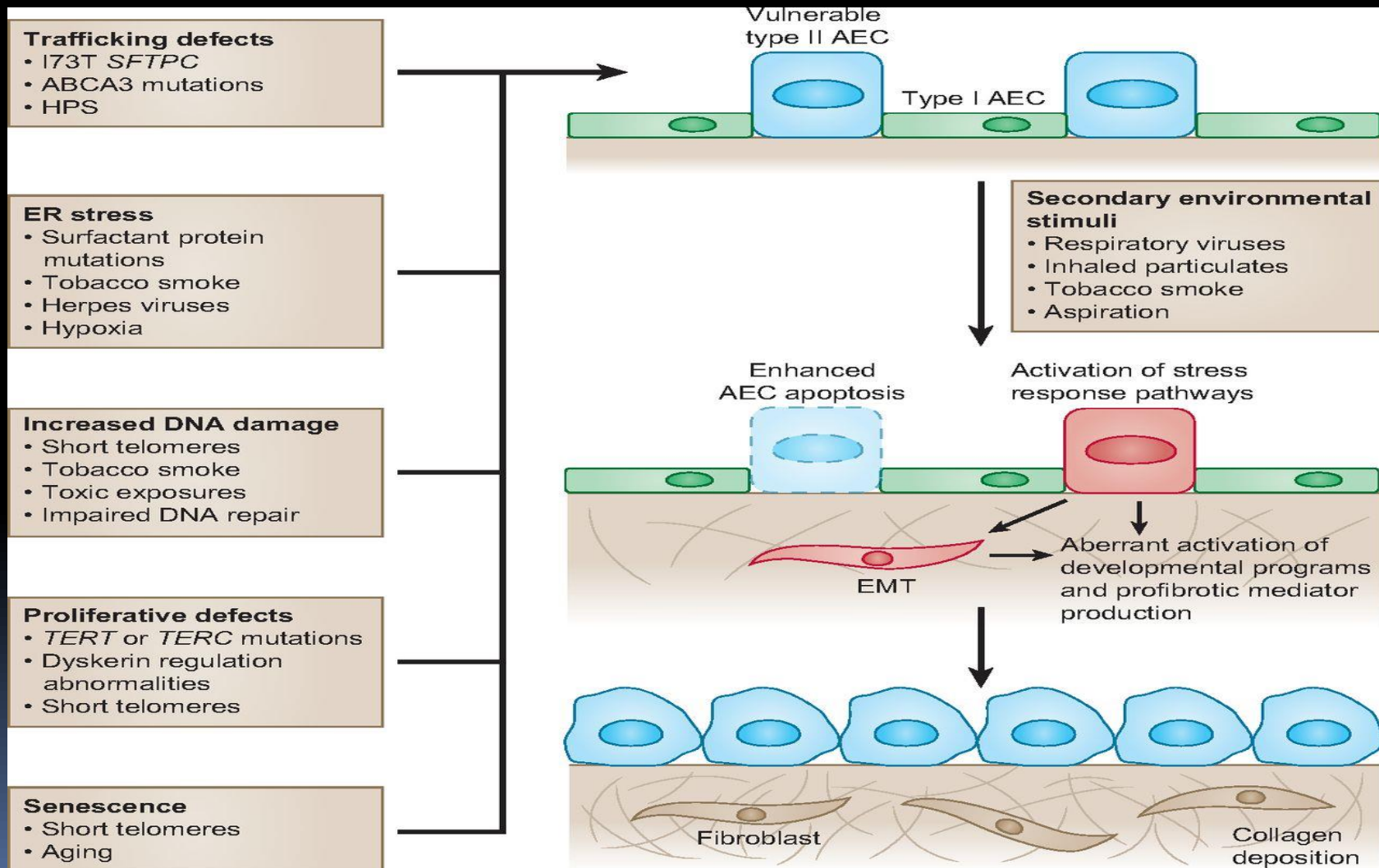
PATHOGENESIS



PATHOGENESIS OF ILD



PATHOGENESIS OF IPF





CLINICAL FEATURES

- BREATHLESSNESS
 - COUGH- DRY
 - WHEEZE+-
 - CHEST PAIN
 - NON RESPIRATORY SYMPTOMS
- 



CLINICAL FEATURES

- **NON RESPIRATORY SYMPTOMS**
- **ASSOCIATED WITH DIFFERENT DPLDS**
- • Arthritis
- • Ocular
- • Skin and muscle
- • GERD
- • Lower GI symptoms
- • Recurrent sinusitis
- • Neurological symptoms
- • Epilepsy & mental retardation
- • Diabetes insipidus

CLINICAL FEATURES

- Acute presentation (days to weeks)- eg: Acute idiopathic
- interstitial pneumoni,Hypersensitive pneumonitis
- 2. Sub-acute presentation (weeks to months)- eg:
 - Sarcoidosis,Drug induced ILD ,Alveolar hemorrhage
 - syndromes
- 3. Chronic presentation (months to years) – eg: IPF



CLINICAL FEATURES- HISTORY

- Smoking
 - •Medication history- Amiodorane,
Methotrexate,
 - •Occupational history
 - •Environmental exposure history
- 



CLINICAL FEATURES

- OCCUPATIONAL HISTORY
- Pneumoconioses – miners
- Silicosis – sand blasters & granite workers
- Asbestosis – welders, electricians, mechanics, workers with brakes, shipyard workers
- Berylliosis – aerospace, nuclear, computer & electronic industries
- Hypersensitive pneumonitis – farm workers, poultry workers, bird breeders
- The degree of exposure, duration, latency of exposure, and the use of protective devices should be elicited



CLINICAL FEATURES- HISTORY

- Exposures to pets (especially birds)
 - Air conditioners
 - Humidifiers
 - Hot tubs
 - Evaporative cooling systems
- 



CLINICAL FEATURES- EXAMINATION

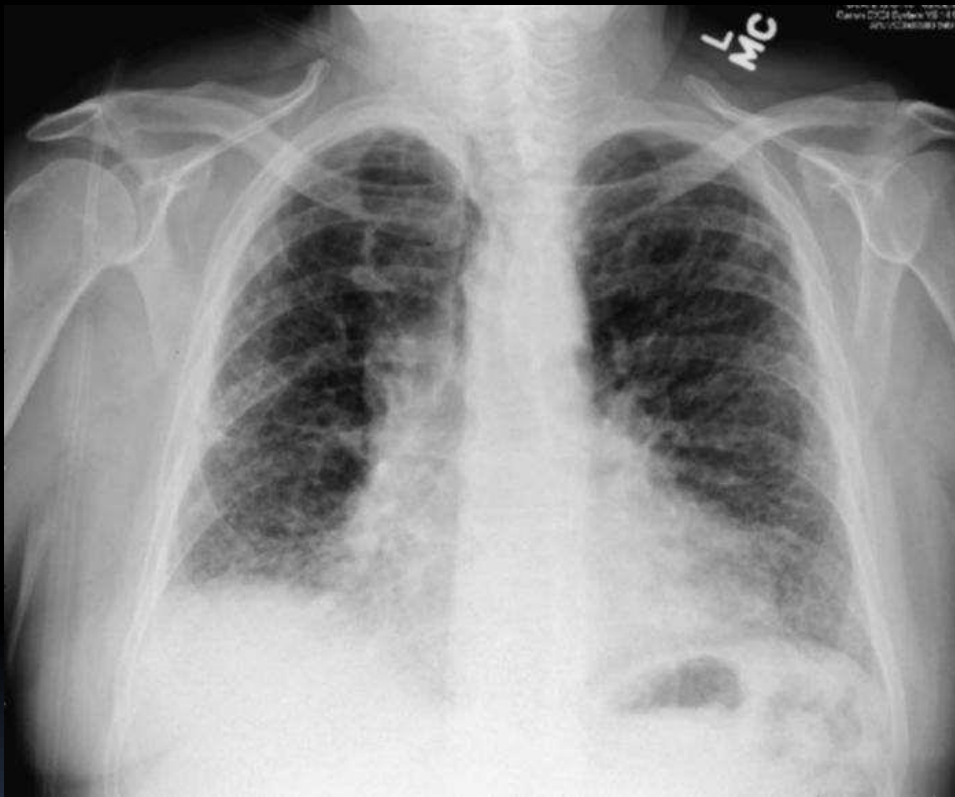
- Crackles - Dry, velcro , end inspiratory, predominantly bibasilar
 - Inspiratory squeaks- Mid inspiratory, high pitched- Seen in Airway centred pathologies
 - Cor pulmonale features
 - Clubbing – IPF, Fibrosing ILDs, DIP
- 



CLINICAL FEATURES-EXAMINATION

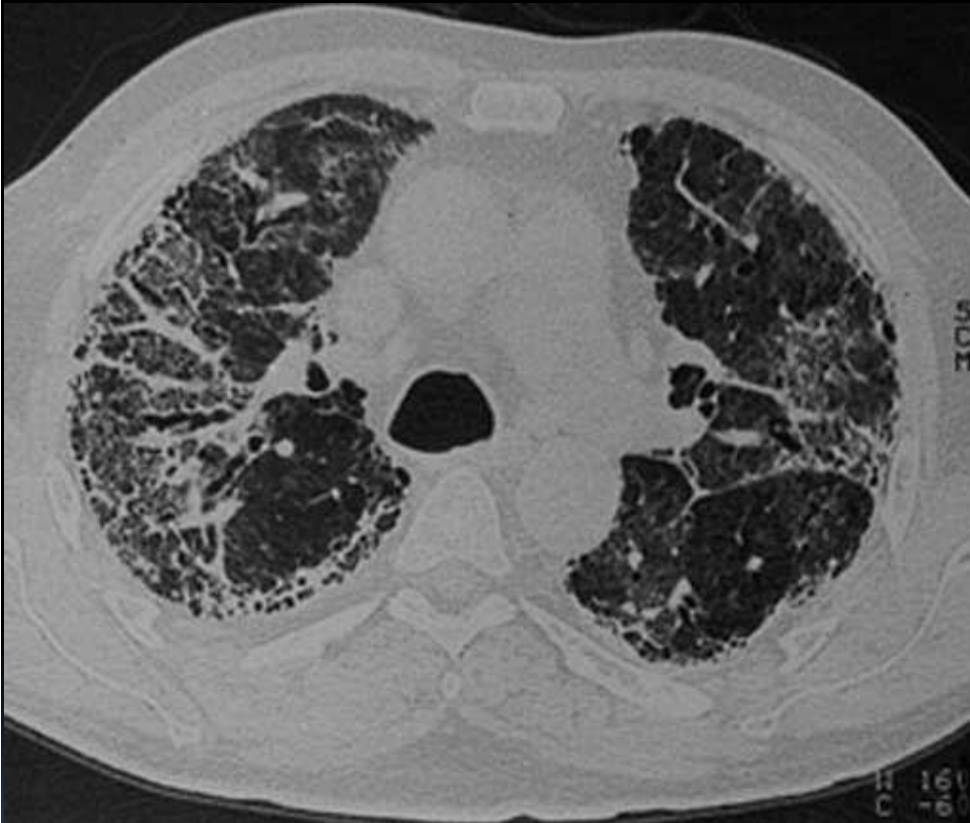
- Skin abnormalities
 - Lymphadenopathy
 - Hepatosplenomegaly
 - Maculopapular skin rashes
 - Erythema nodosum
 - Subcutaneous nodules
 - Proximal muscle weakness
 - Arthritis
- 

INVESTIGATIONS- CHEST XRAY



Typically small
“shrunk” lungs with
Reticular, Nodular, or
RETICULONODULAR
shadow. Shaggy heart
and diaphragm, cystic
shadows pleural and
basal zones

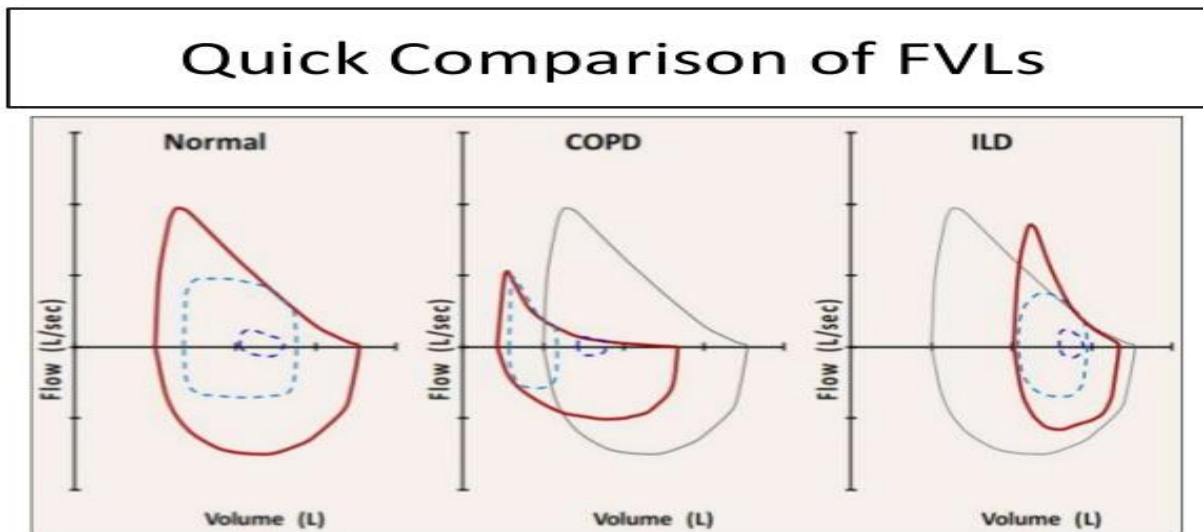
INVESTIGATIONS- HRCT CHEST



Combinations of ground glass changes, reticulonodular shadowing, honeycomb cysts and traction bronchiectasis

INVESTIGATIONS-PFT

- Restrictive Defect
 - • ↓ Lung volumes (TLC, FRC, RV < 80%)
 - • ↓ FEV₁, FVC With Normal or ↑ FEV₁/FVC
 - • Reduced diffusing capacity (DLCO)





INVESTIGATIONS

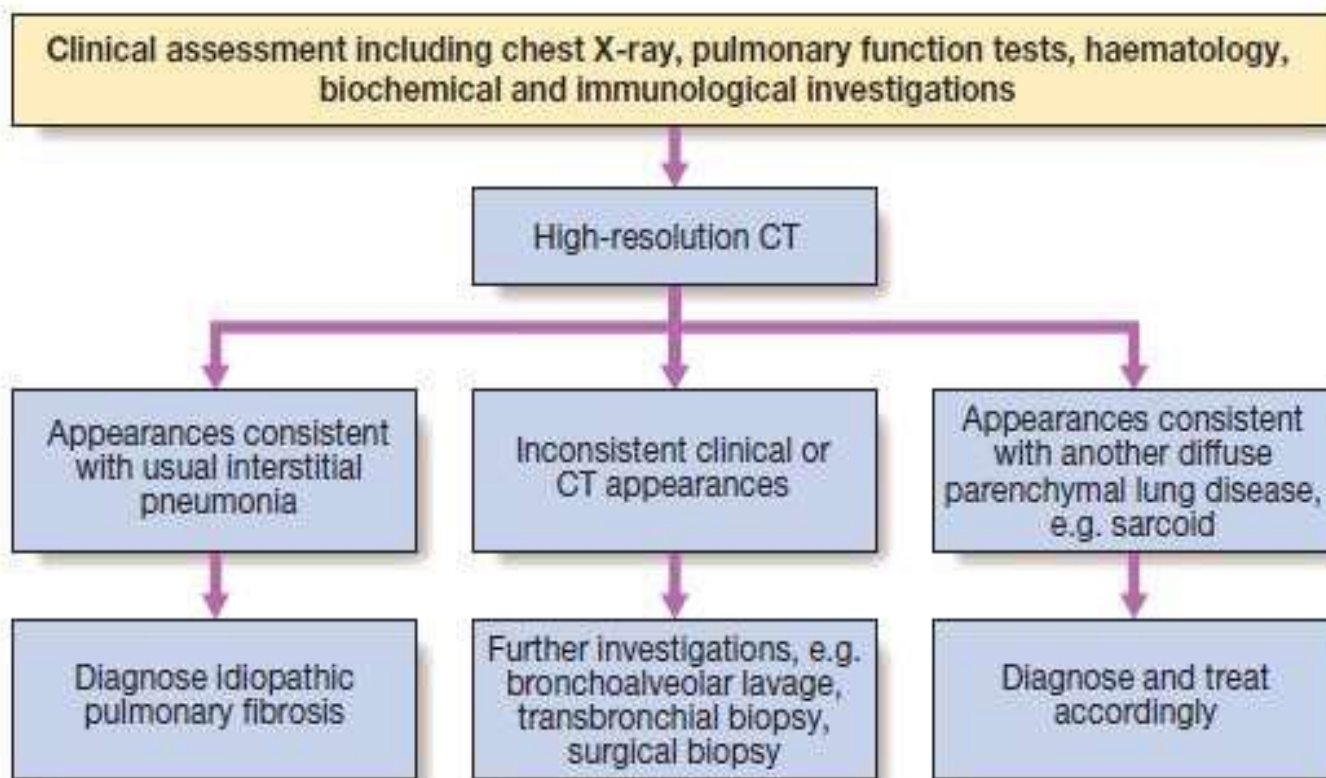
- Full blood count: lymphopenia in sarcoid; eosinophilia in pulmonary eosinophilias and drug reactions; neutrophilia in hypersensitivity pneumonitis
- Ca^{2+} : may be elevated in sarcoid
- Serum angiotensin-converting enzyme: non-specific indicator of disease activity in sarcoid
- ESR and CRP: non-specifically raised
- Autoimmune screen: autoantibodies may suggest connective tissue disease



INVESTIGATIONS

- ARTERIAL BLOOD GAS ANALYSIS
 - Tuberculin test – negative in 2/3 of sarcoidosis patients
 - BRONCHOALVEOLAR LAVAGE- cellular profile , special
 - stains or studies
 - Lung biopsy- TRANS BRONCHIAL BIOPSY , OPEN LUNG BIOPSY
- 

APPROACH TO A CASE OF ILD



Algorithm for the investigation of patients with interstitial lung disease following initial clinical and chest X-ray examination.



DIFFERENTIAL DIAGNOSIS

- HEART FAILURE
 - BRONCHIECTASIS
 - ADULT ONSET BRONCHIAL ASTHMA
 - ARDS
 - IDIOPATHIC PULMONARY HYPERTENSION
 - TROPICAL PULMONARY EOSINOPHILIA
 - CHRONIC EOSINOPHILIC PNEUMONIA
- 



MANAGEMENT

- PRINCIPAL AIMS

- (1) To remove exposure to injurious agents
 - (2) To suppress inflammation to prevent further destruction of the pulmonary parenchyma
 - (3) To palliate the manifestations of these diseases
- 

MANAGEMENT- IPF

- **Strong recommendation**
 - Oxygen
 - Pulmonary rehabilitation
 - Proton pump inhibitors- GERD
 - Vaccination- pneumococcal and influenza
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- **Conditional recommendation**
 - Pirfenidone- antifibrotic agent
 - Nintedanib – Tyrosine kinase inhibitor

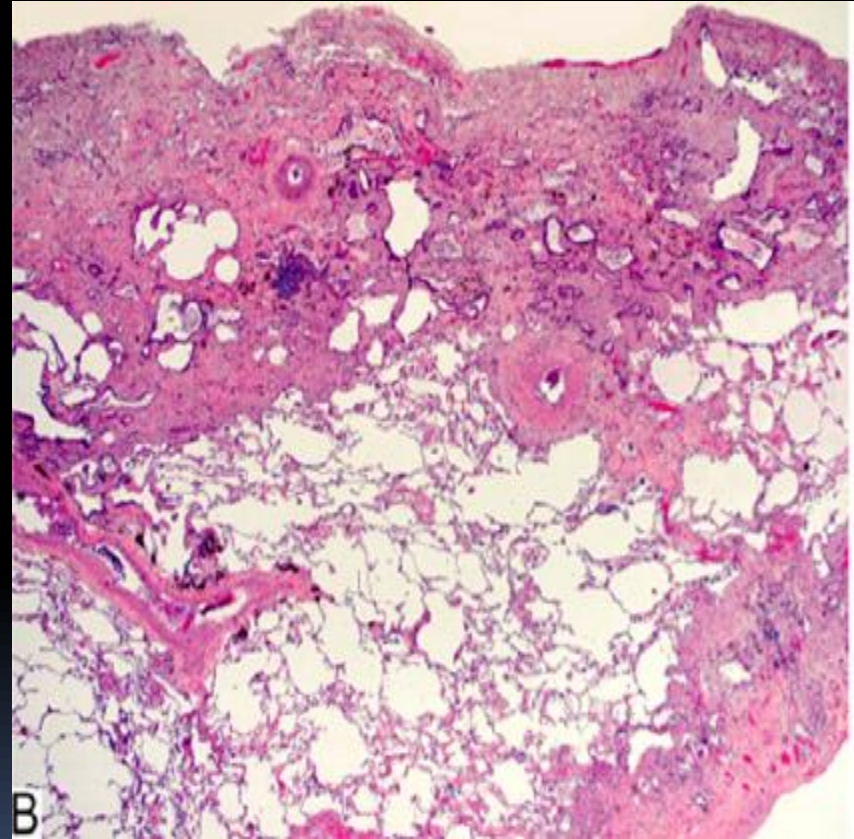
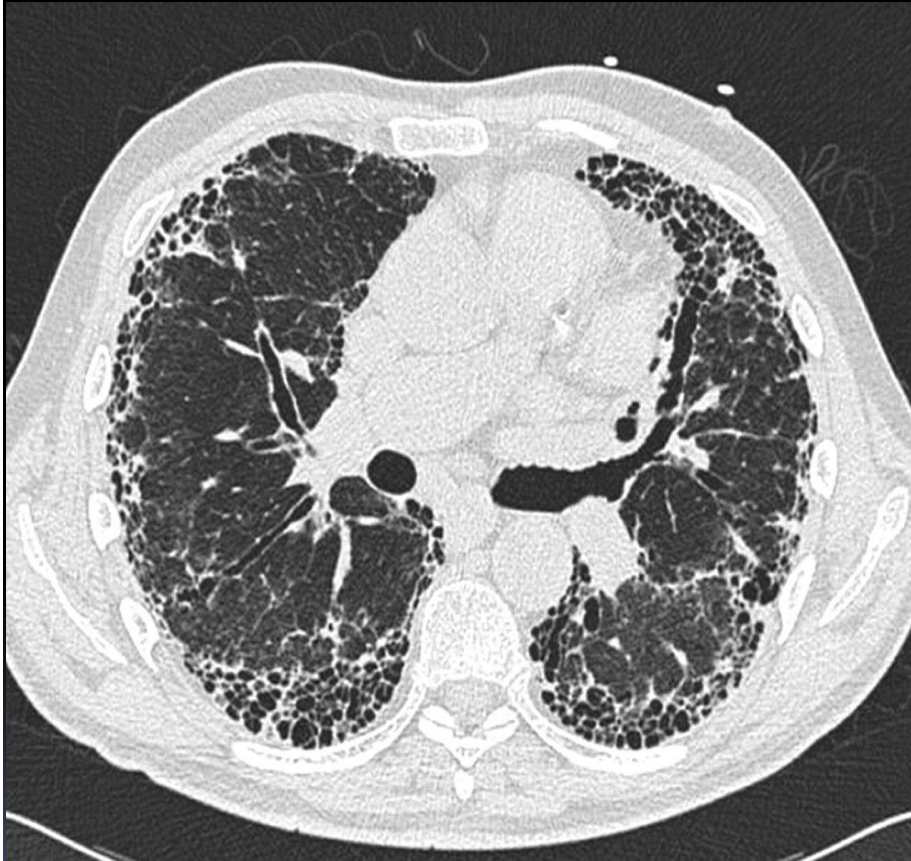
MANAGEMENT- other ILDS

- Steroids
- Immunosuppressive agents- azathioprin, cyclophosphamide, methotrexate etc...
- Inhaled corticosteroids
- Diuretics
- Treatment of Pulmonary hypertension

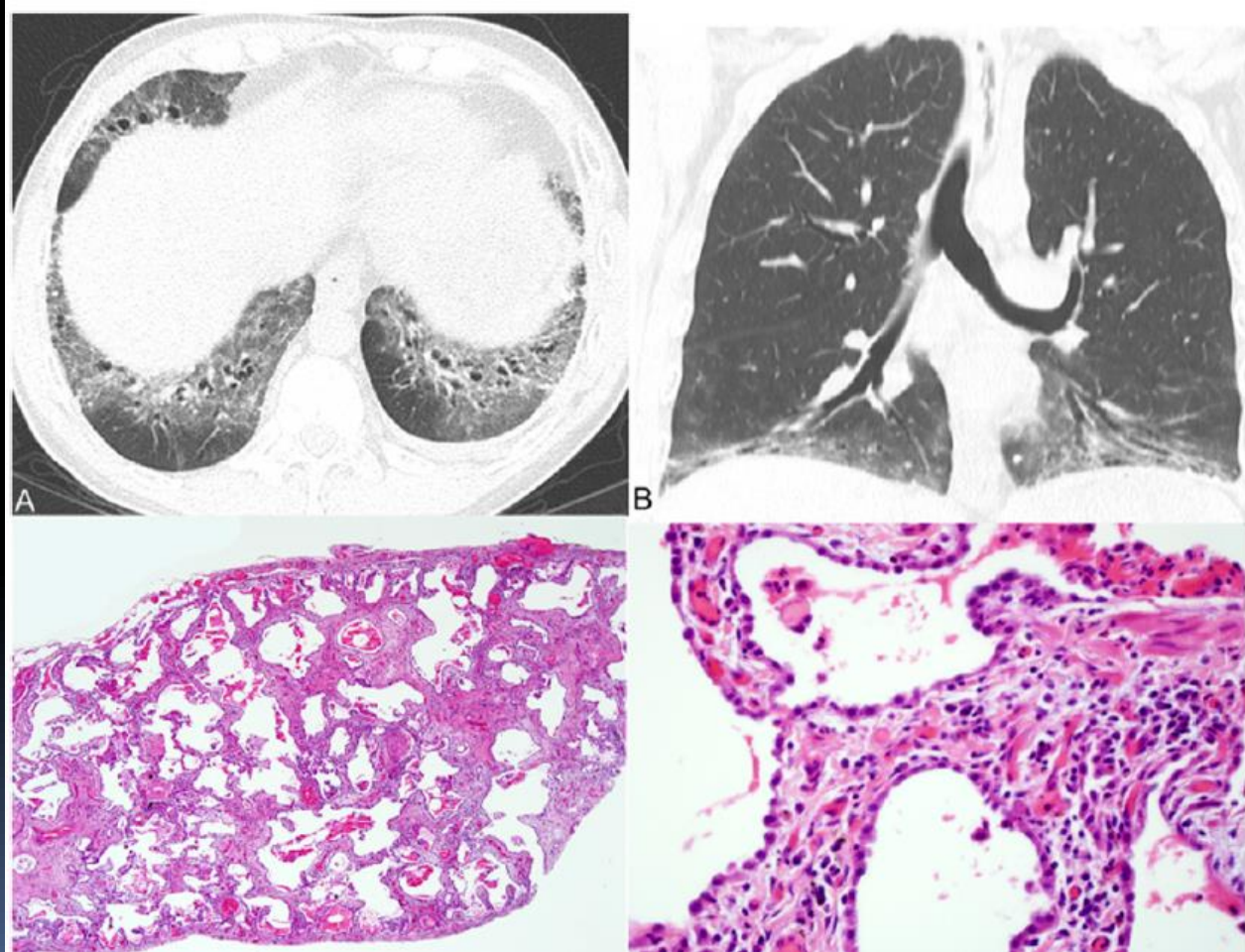
SUMMARY

	Idiopathic Pulmonary Fibrosis	NSIP	DIP, RB-ILD	Cryptogenic Organizing Pneumonia	Acute Interstitial Pneumonia	Lymphocytic Interstitial Pneumonia
Duration of illness	Chronic (> 12 mo)	Subacute to chronic (mo to yr)	Subacute (wk to mo);	Subacute (< 3 mo)	Abrupt (1 to 2 wk)	Chronic (> 12 mo)
HRCT	• Peripheral, subpleural, basal predominance • Reticular opacities • Architectural distortion • Traction bronchiectasis/bronchiolectasis • Honeycombing	• Peripheral, subpleural, basal, symmetric • Ground-glass attenuation • Consolidation (uncommon) • Lower lobe volume loss • Subpleural sparing may be seen	• DIP: diffuse ground-glass opacity in the middle and lower lung zones • RB-ILD: bronchial wall thickening; centrilobular nodules; patchy ground-glass opacity	• Subpleural or peribronchial • Patchy consolidation • Nodules	• Diffuse, bilateral • Ground-glass opacities often with lobular sparing	• Diffuse • Centrilobular nodules, • Ground-glass attenuation, • Septal and bronchovascular thickening, • Thin-walled cysts
Treatment	Poor response to corticosteroid or cytotoxic agents	Corticosteroid responsiveness	Smoking cessation, effectiveness of corticosteroid unknown	Corticosteroid responsiveness	Effectiveness of corticosteroid unknown	Corticosteroid responsiveness
Prognosis	5-yr mortality, 80% (median survival 2–3 yr)	Cellular NSIP: 5-yr mortality < 10% (median survival > 10 yr) Fibrotic NSIP: 5-yr mortality 10% (median survival 6–8 yr)	RB-ILD: no deaths reported DIP: 5-yr mortality < 5%	5-yr mortality < 5% (deaths rare)	60% mortality in < 6 mo	Limited data, not well defined

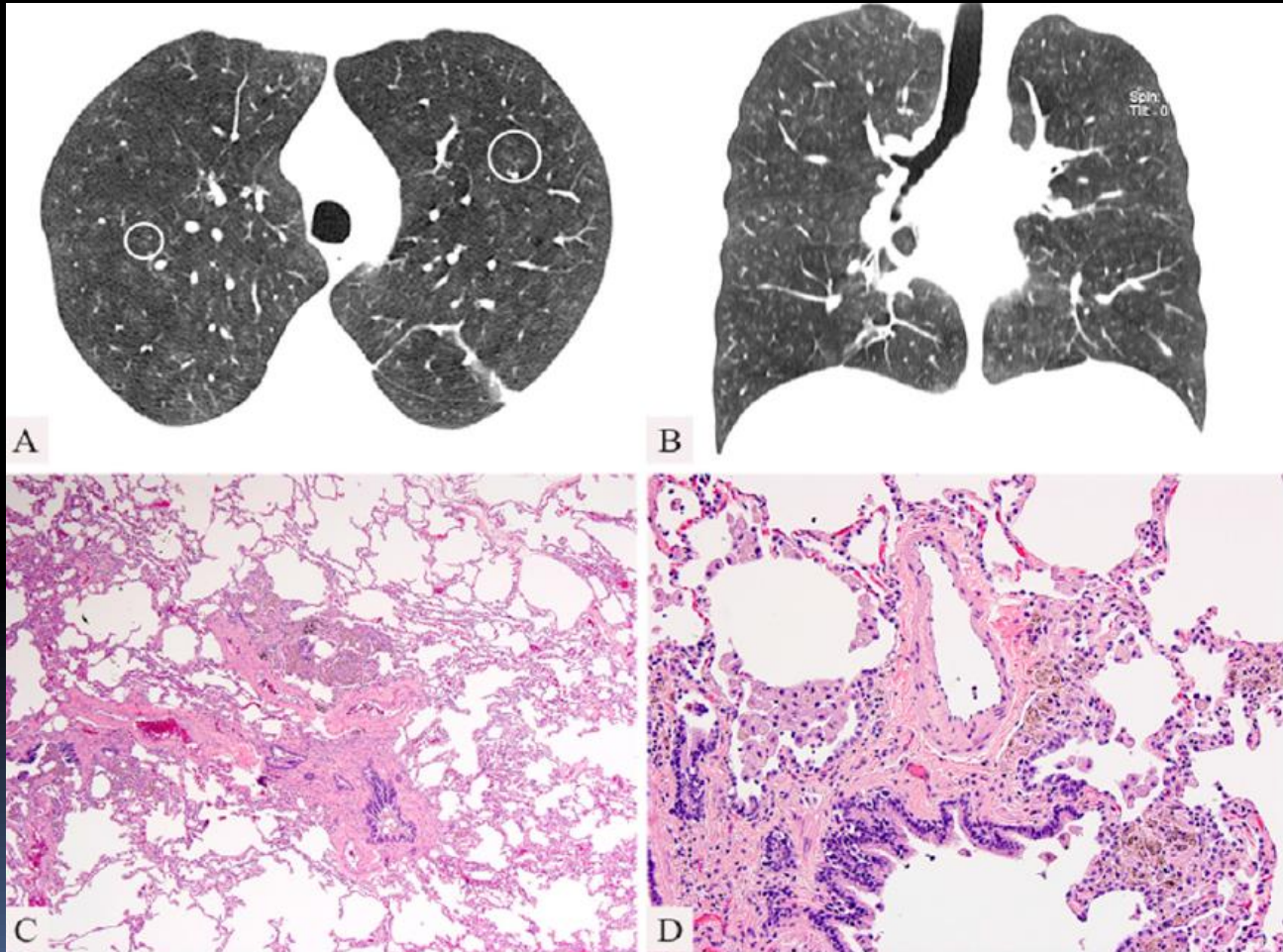
IPF



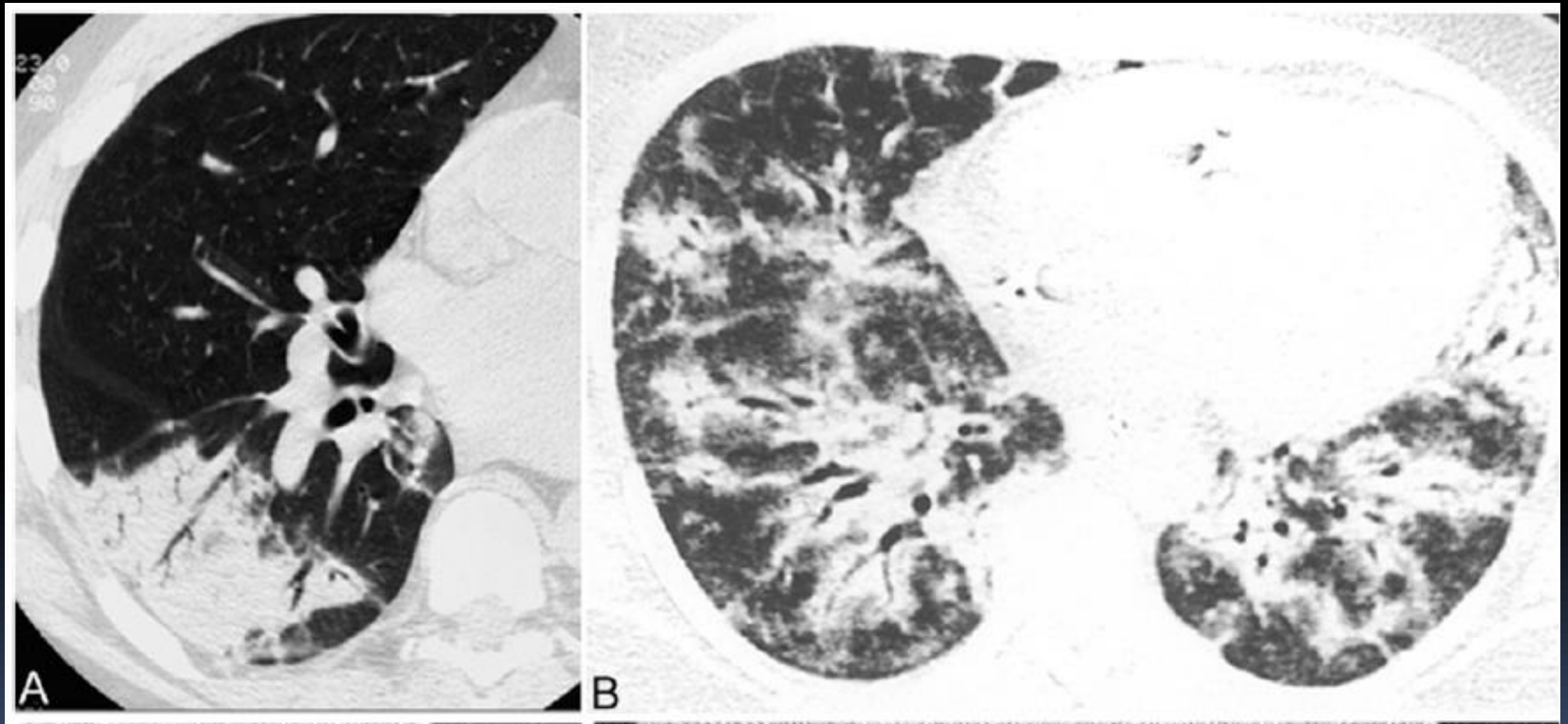
NSIP



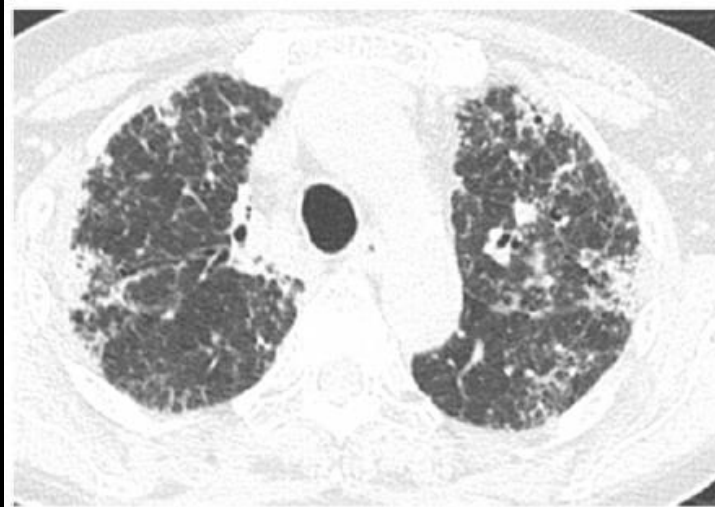
RBILD



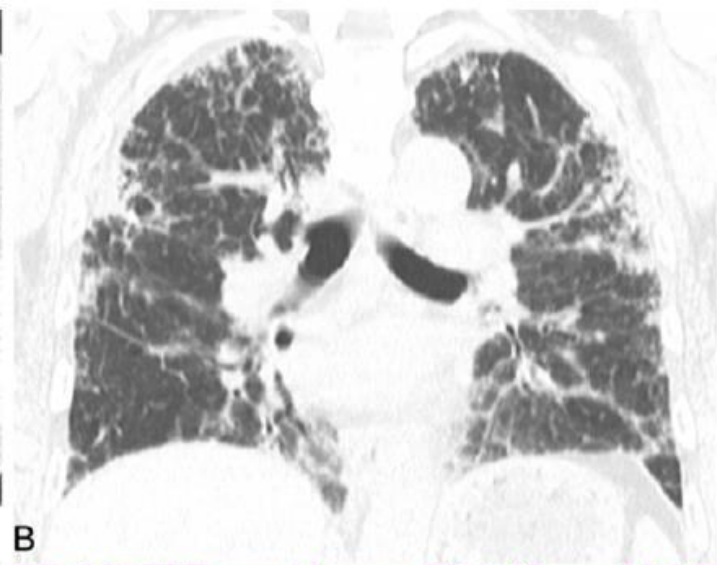
COP



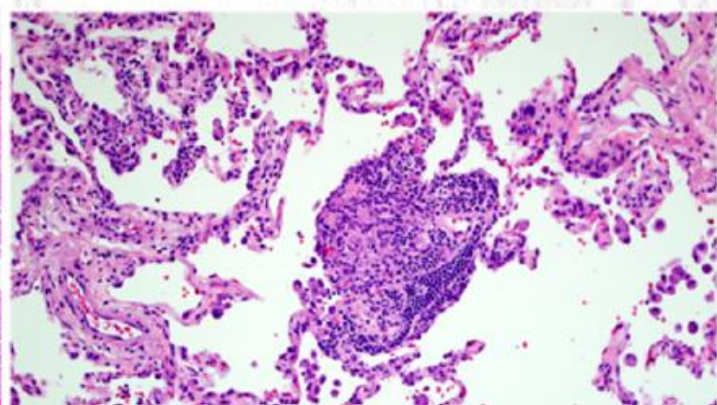
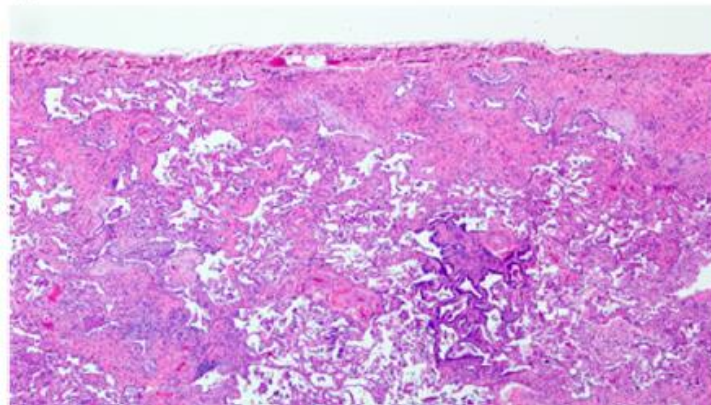
CHRONIC HYPERSENSITIVITY PNEUMONITIS



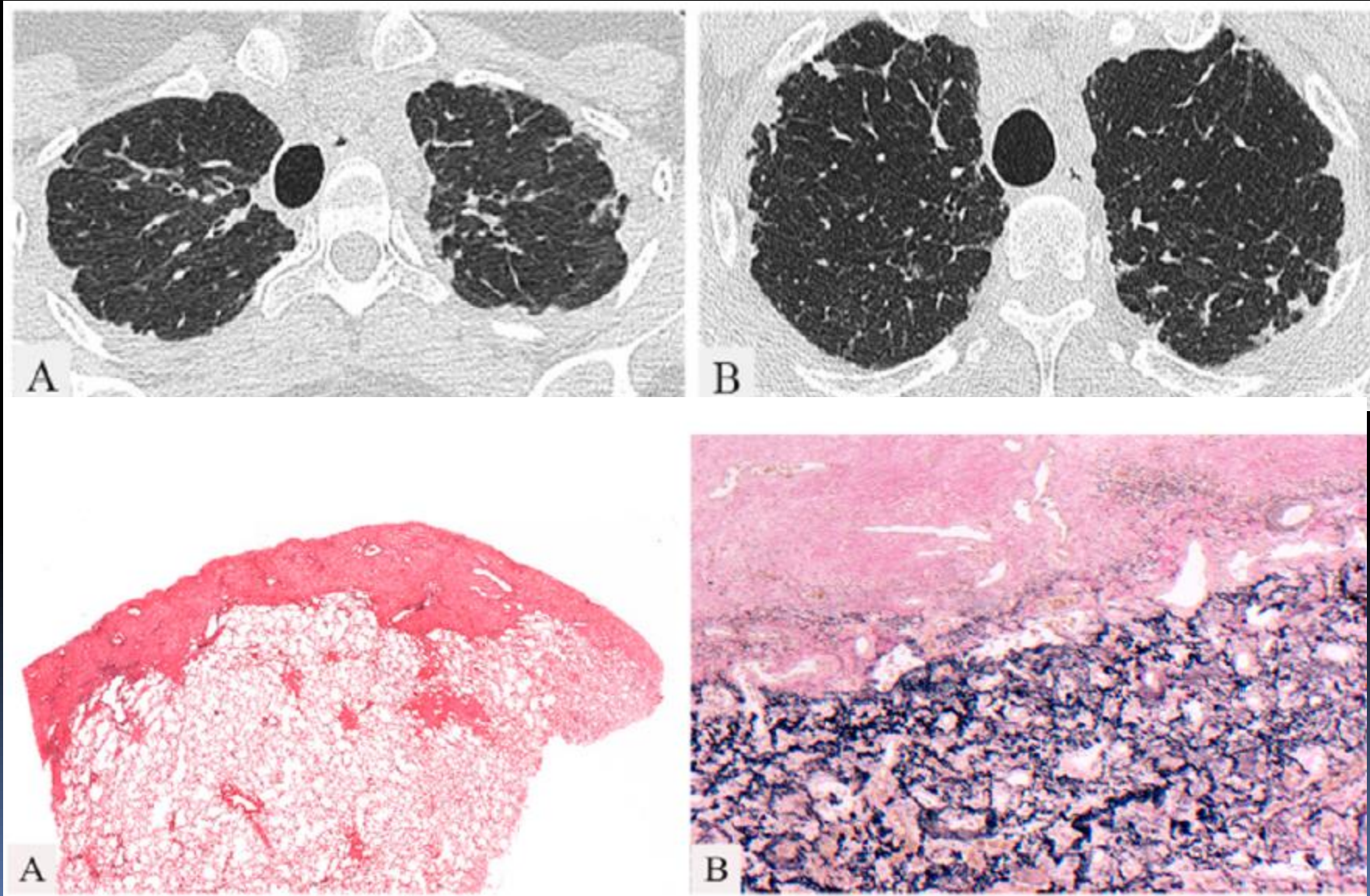
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PLEUROPARENCHYMAL FIBROELASTOSIS



Thank you !!

