Interstitial Lung Disease

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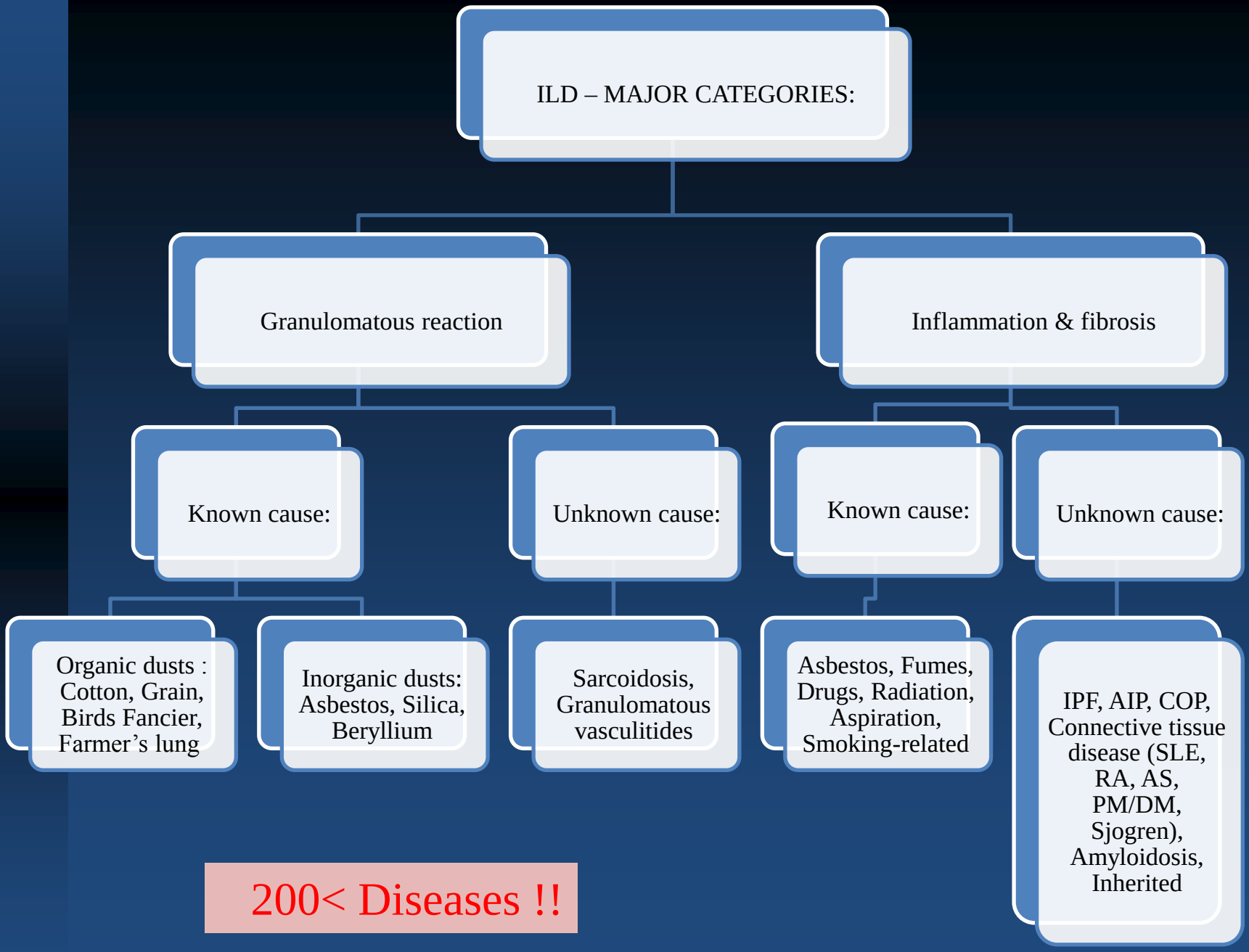
The Chaim Sheba Medical Center
at Tel Hashomer - Est. 1948
The Hospital of Israel

What will we talk about ?

- General classification
- Evaluation of ILD
- Occupational
- Idiopatic
- HRCT
- IPF: In depth look


ILD – MAJOR CATEGORIES:
Granulomatous reaction
Inflammation & fibrosis
Known cause:
Unknown cause:
Known cause:
Unknown cause:
Organic dusts : Cotton, Grain, Birds Fancier, Farmer's lung
Inorganic dusts: Asbestos, Silica, Beryllium
Sarcoidosis, Granulomatous vasculitides
Asbestos, Fumes, Drugs, Radiation, Aspiration, Smoking-related
IPF, AIP, COP, Connective tissue disease (SLE, RA, AS, PM/DM, Sjogren), Amyloidosis, Inherited
200< Diseases !!

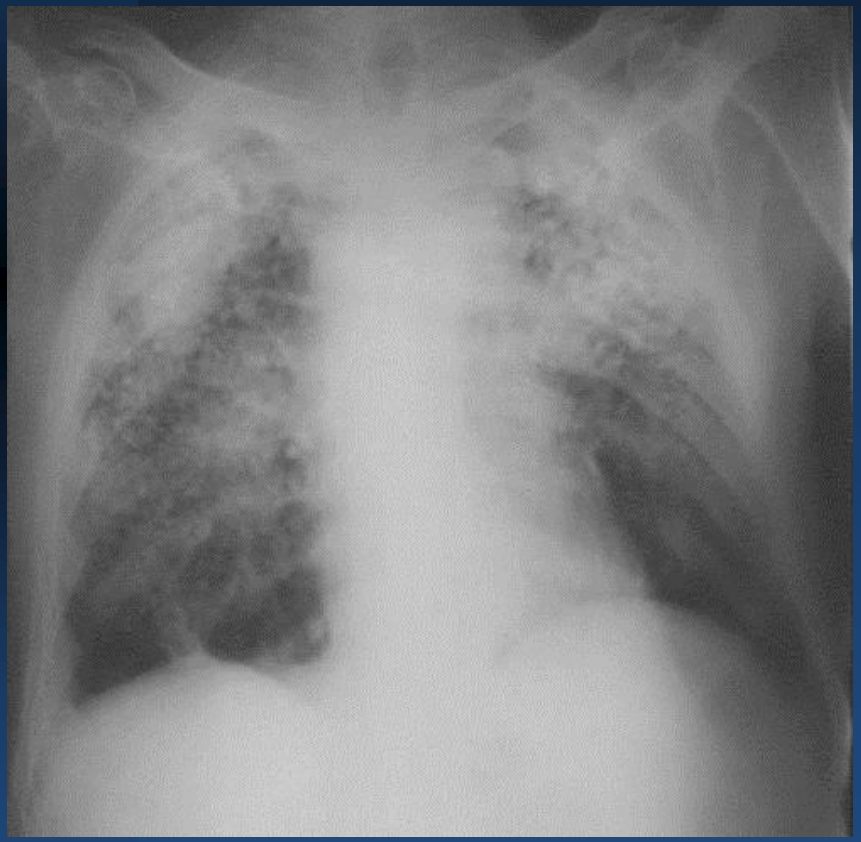
Evaluation of ILD

- EXTENSIVE HISTORY
- AGE, GENDER, UNDERLYING COMORBIDITIES, DRUGS, SMOKING, OCCUPATIONAL HISTORY, HOBBIES, PETS, FAMHX
- DURATION OF SYMPTOMS
- PHYSICAL EXAM
- LABORATORIES
- IMAGING
- SPIROMETRY, LUNG VOLUMES AND DLCO

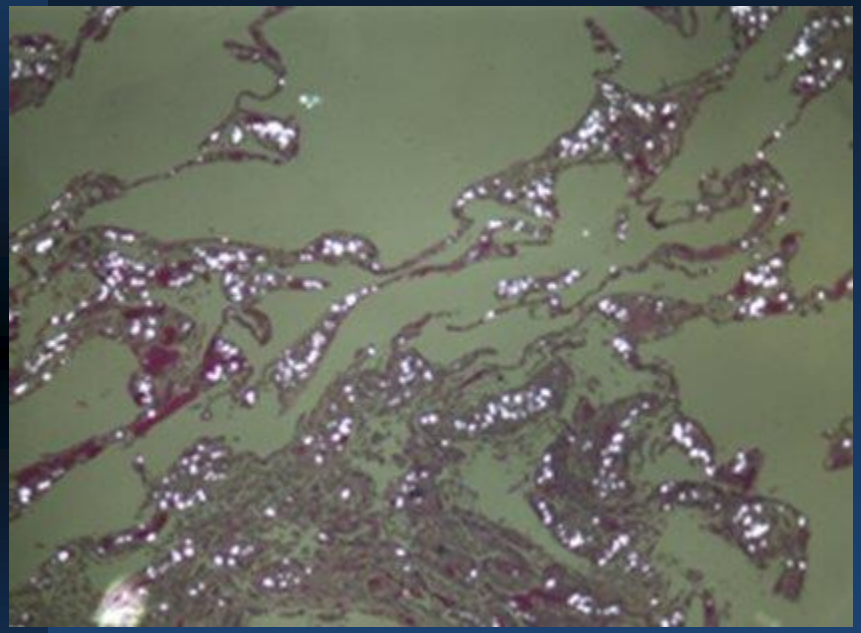
Table 42.2
Frequently encountered diffuse lung diseases with identifiable underlying cause

Cause	Differential diagnosis	
Occupational or other inhalant-related, inorganic	Coal worker's pneumoconiosis	Metal polisher's lung/hard metal fibrosis
	Asbestosis	Berylliosis
	Silicosis	Baritosis (barium)
	Talc pneumoconiosis	Siderosis (iron oxide)
	Aluminum oxide fibrosis	Stannosis
Occupational or other inhalant-related, organic	Bird fancier's lung	Mushroom worker's lung
	Farmer's lung	Maple bark stripper's lung
	Bagassosis (sugar cane)	Malt worker's lung
	Coffee worker's lung	Tea grower's lung
	Tobacco grower's lung	Pituitary snuff-taker's lung
	Fishmeal worker's lung	
Collagen vascular disease related	Systemic lupus erythematosus	Ankylosing spondylitis
	Rheumatoid arthritis	Mixed connective tissue disease
	Scleroderma	Primary Sjögren's syndrome
	Polymyositis	Behçet's syndrome
	Dermatomyositis	Goodpasture's syndrome
Drug related	Amiodarone	Bleomycin
	Propranolol	Busulfan
	Tocainide	Cyclophosphamide
	Nitrofurantoin	Chlorambucil
	Sulfasalazine	Melphalan
	Cephalosporins	Methotrexate
	Gold	Azathioprine
	Penicillamine	Cytosine arabinoside
	Phenytoin	Carmustine
	Mitomycin	Lomustine
	Bromocryptine	
Physical agents/ toxins	Radiation/radiotherapy	Cocaine inhalation
	High concentration oxygen	Intravenous drug abuse
	Paraquat toxicity	
Neoplastic disease	Lymphangitis carcinomatosis	
	Bronchoalveolar cell carcinoma	
Vasculitis related	Wegener's granulomatosis	
	Giant cell arteritis	
	Churg–Strauss syndrome	
Disorders of circulation	Pulmonary edema	
	Pulmonary veno-occlusive disease	
Chronic infection	Tuberculosis	Viruses
	Aspergillosis	Parasites
	Histoplasmosis	
Smoking induced	Emphysema	Desquamative interstitial pneumonia
	Langerhans cell histiocytosis	Respiratory bronchiolitis
	Alveolar cell carcinoma	Non-specific interstitial pneumonia
	Respiratory bronchiolitis with associated interstitial lung disease	

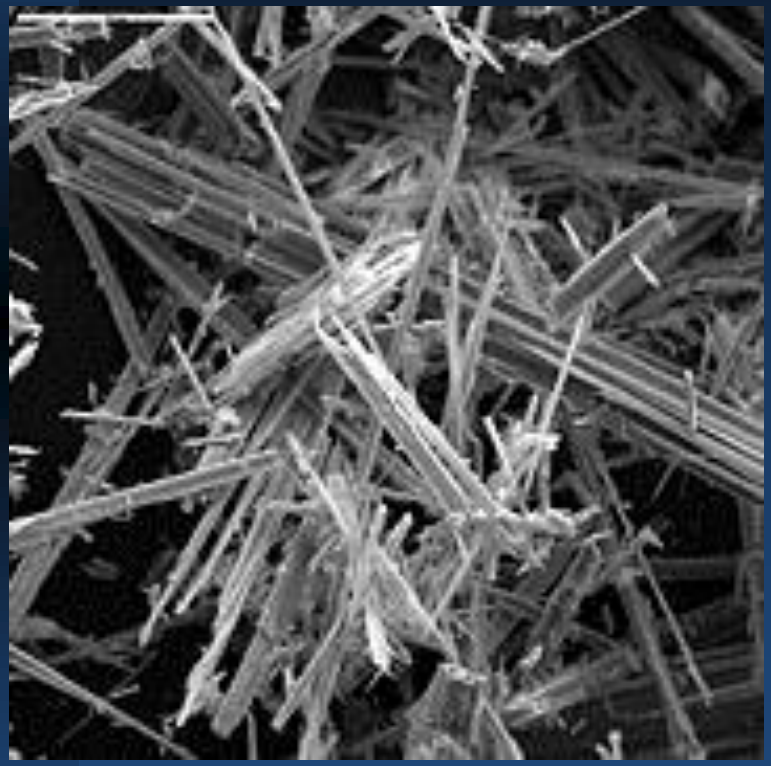
Silicosis

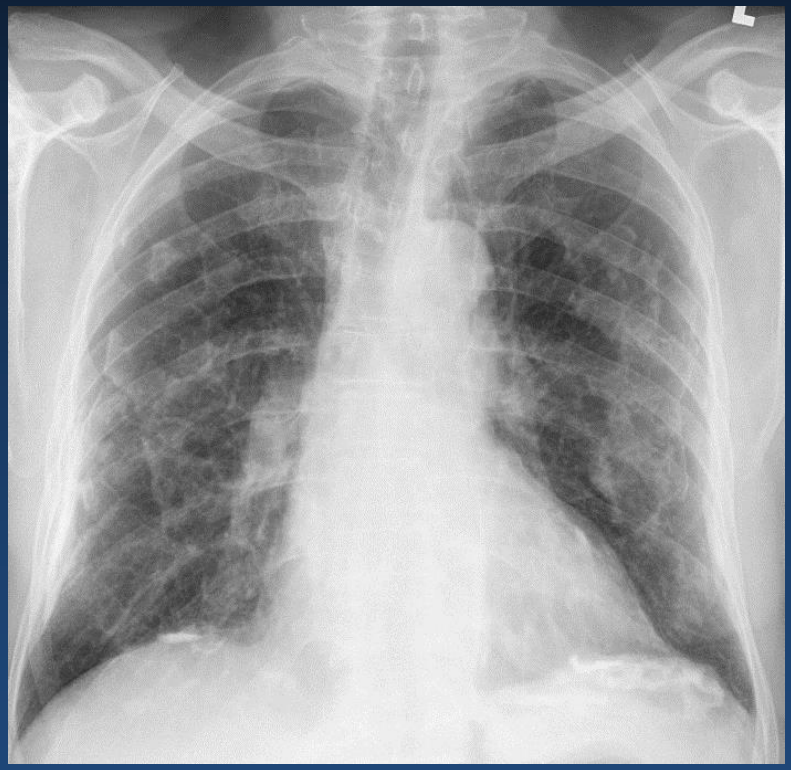


Talcosis

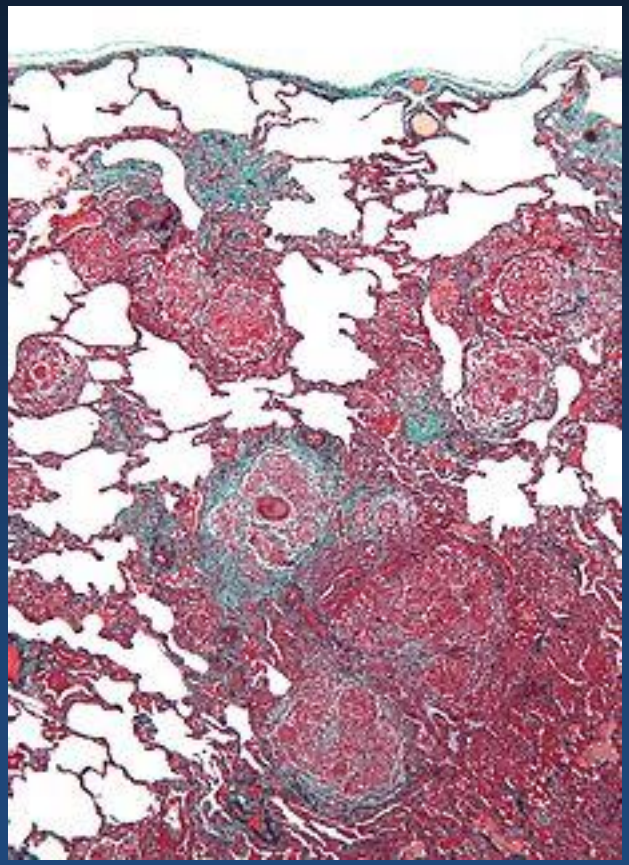


Asbestosis

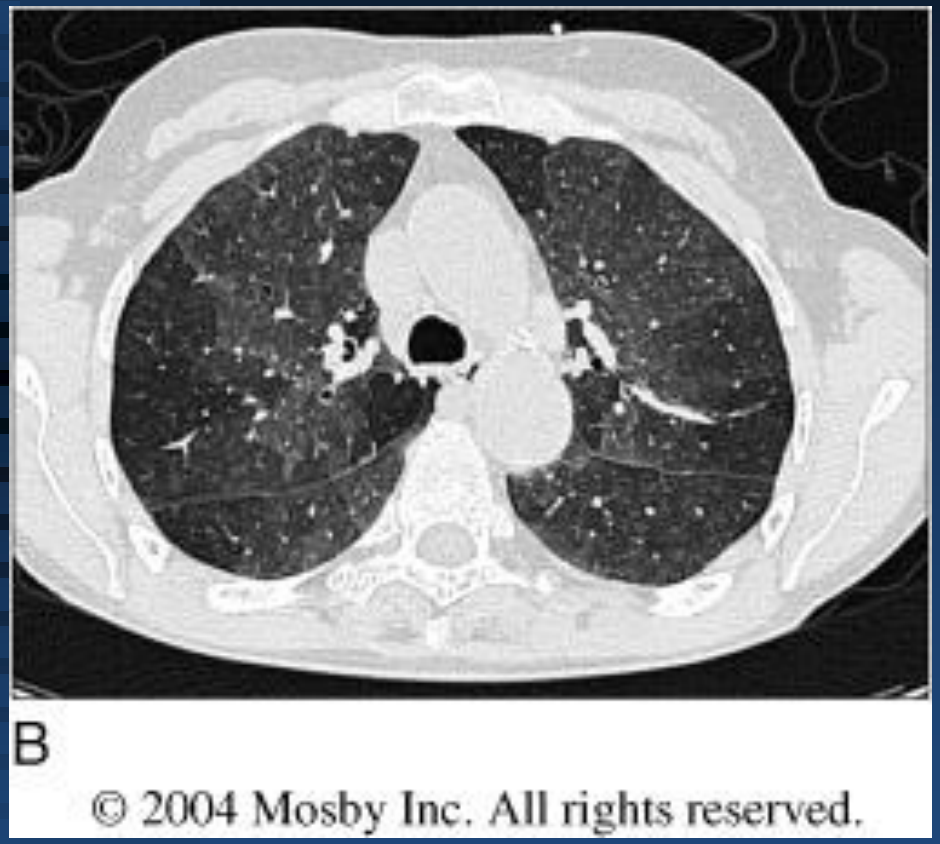




Birds fancier lung disease



Hypersensitivity Pneumonitis



B

- Organic dusts (Cotton, Grain, Birds Fancier, Farmer's lung)
- Ground-glass attenuation
- Poorly formed centrilobular nodules on expiratory HRCT

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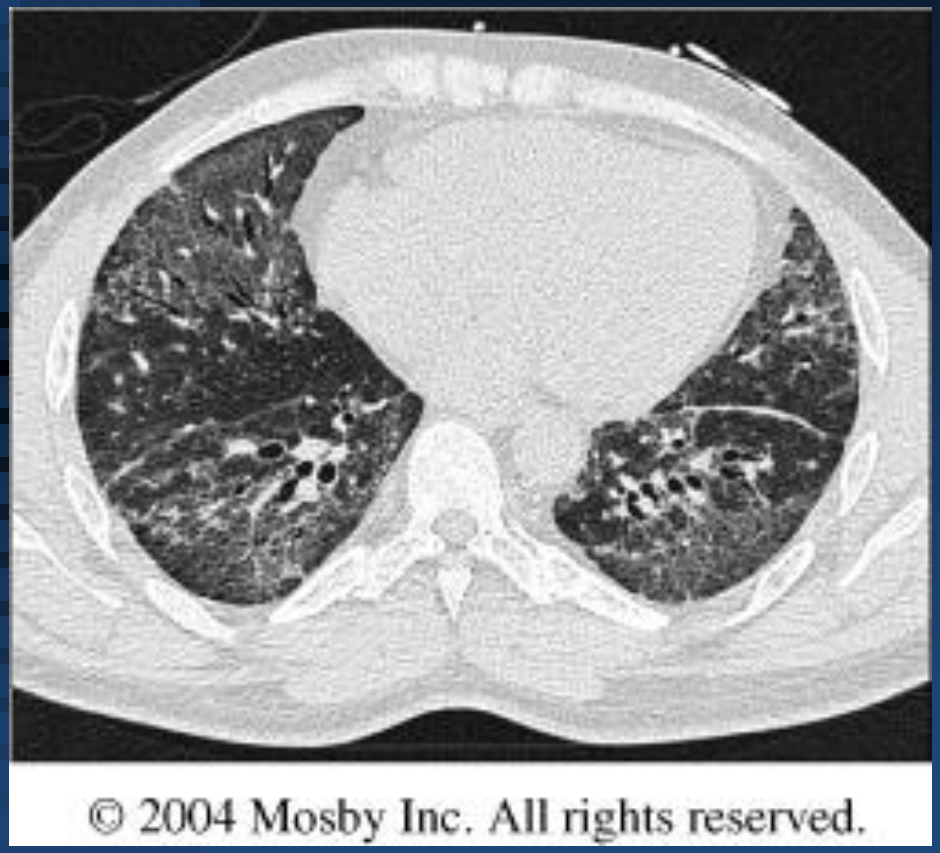
Table 42.5
ATS/ERS Consensus Classification of the Idiopathic Interstitial Pneumonias, and the Most Frequent Patterns of Longitudinal Behavior Associated with Individual Diagnoses

Clinicopathologic Diagnosis	Likely Longitudinal Behavior
Idiopathic pulmonary fibrosis/ cryptogenic fibrosing alveolitis	Inexorably progressive fibrosis
NSIP	Cellular NSIP: self-limited or major inflammation Fibrotic NSIP: stable or progressive fibrosis
Cryptogenic organizing pneumonia	Self-limited or major inflammation
Acute interstitial pneumonia	Explosive acute disease
Desquamative interstitial pneumonia	Self-limited or major inflammation
Respiratory bronchiolitis–associated interstitial lung disease	Self-limited inflammation
Lymphocytic interstitial pneumonia	Self-limited or major inflammation

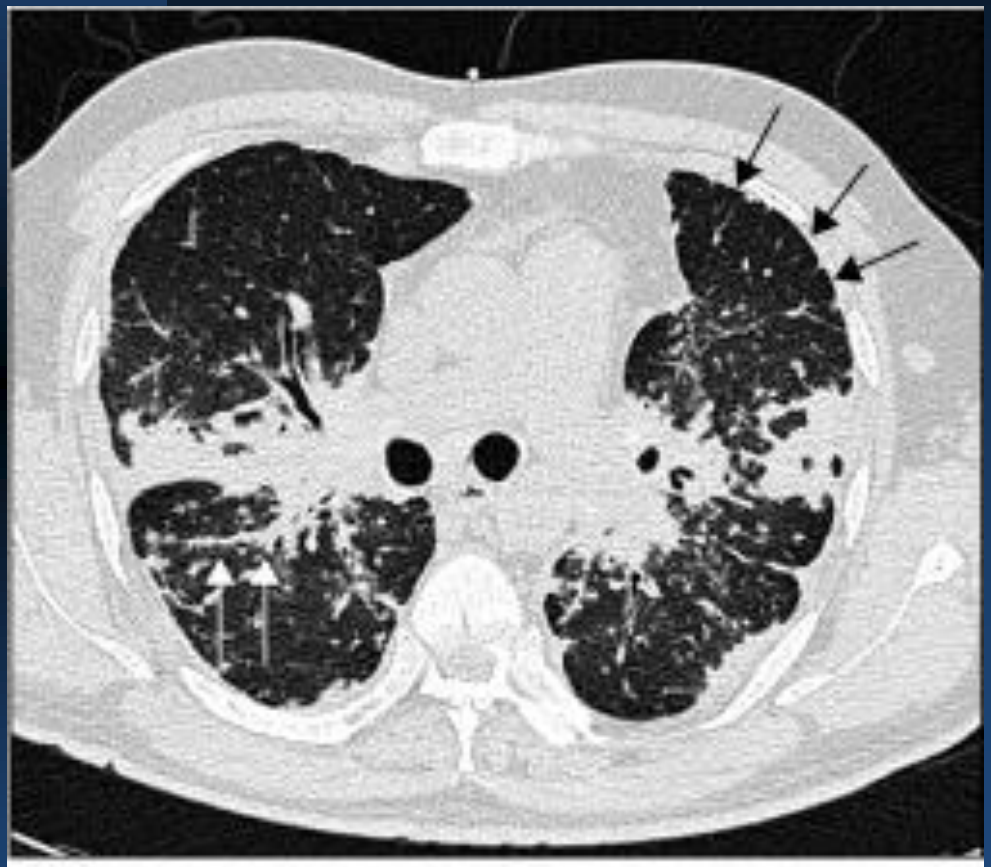
NSIP, nonspecific interstitial pneumonia.

NSIP



- Ground-glass attenuation, with variable fine reticular abnormalities
- There is no honeycomb change
- Good prognosis with GC treatment

Sarcoidosis



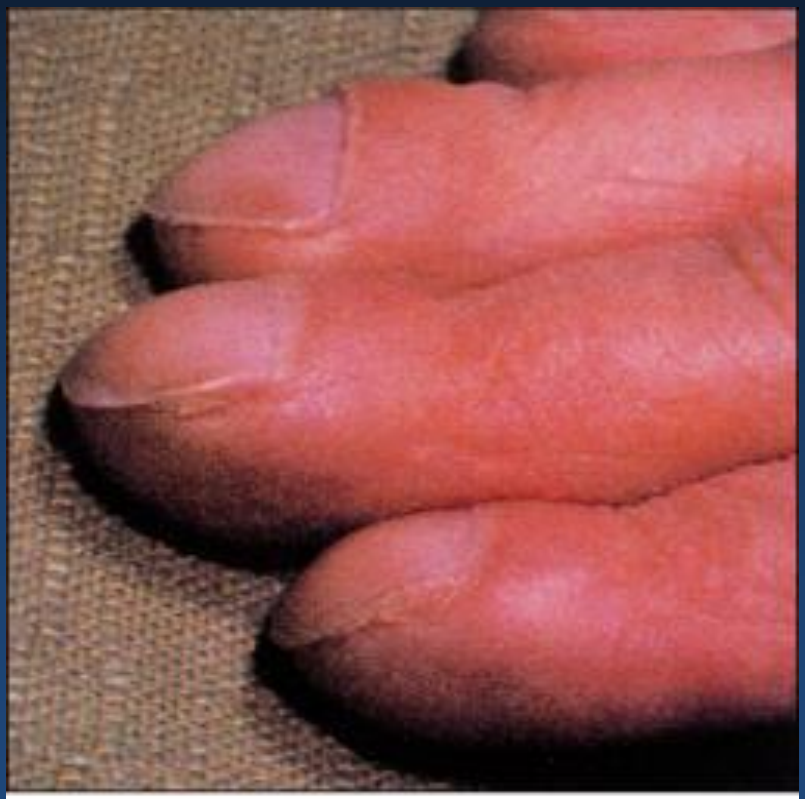
- Multisystem granulomas of unknown cause
- Mostly lung, liver, skin, and eye
- Peribronchial thickening and subpleural reticular nodular changes
- Majority resolve in 2-5 yrs with GC treatment

IPF-Clinical Symptoms

- Age: 50-60 yrs old
- Male:Female 3:1
- Progressive dyspnea
- Dry non-productive cough
- Non specific symptoms (weakness, loss of weight, arthralgia)

IPF-Physical Examination

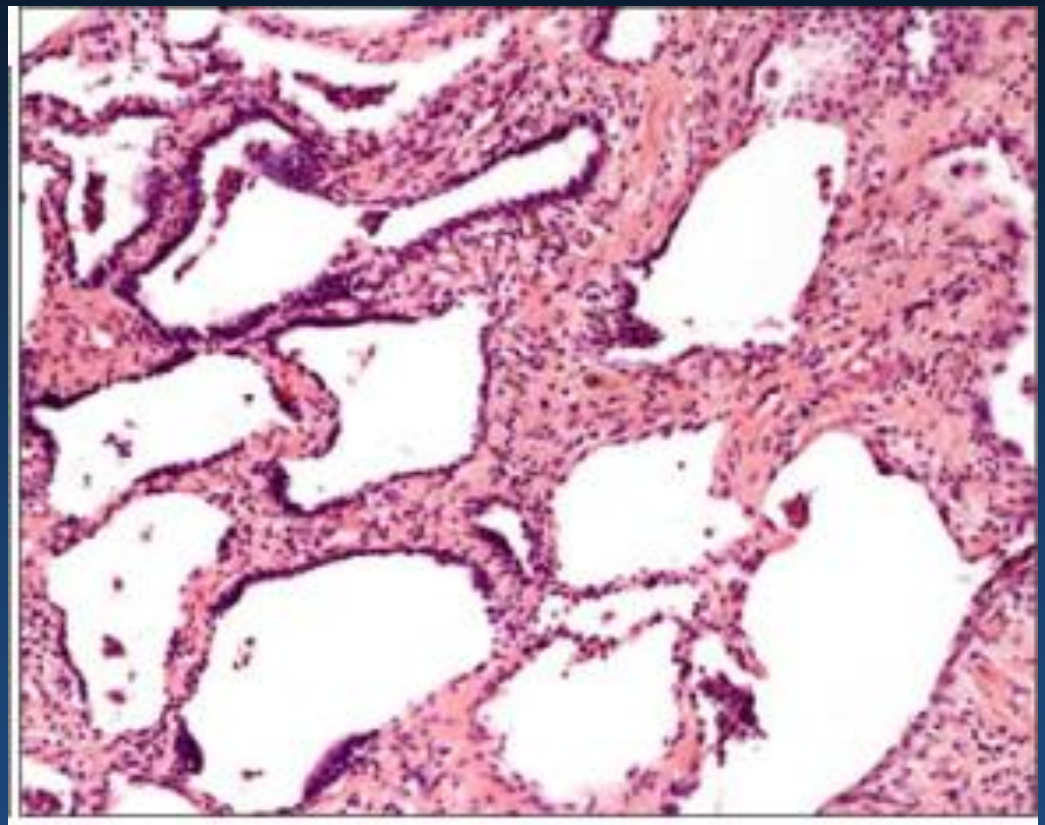
- Inspiratory crackles
- Tachypnea
- Cyanosis
- Clubbing
- Signs of cor pulmonale



IPF-Laboratory

- High ESR
- Positive ANA
- Positive RF in 10-30%

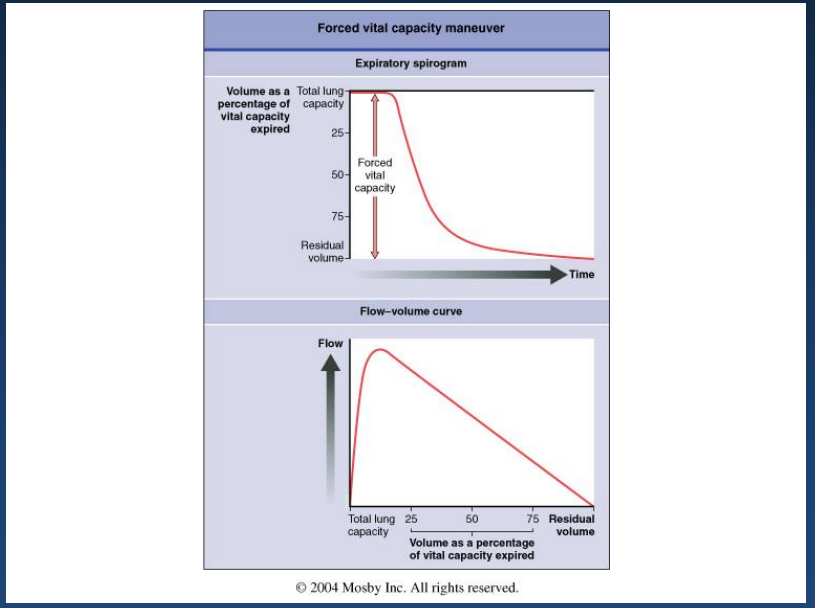
IPF- pathology



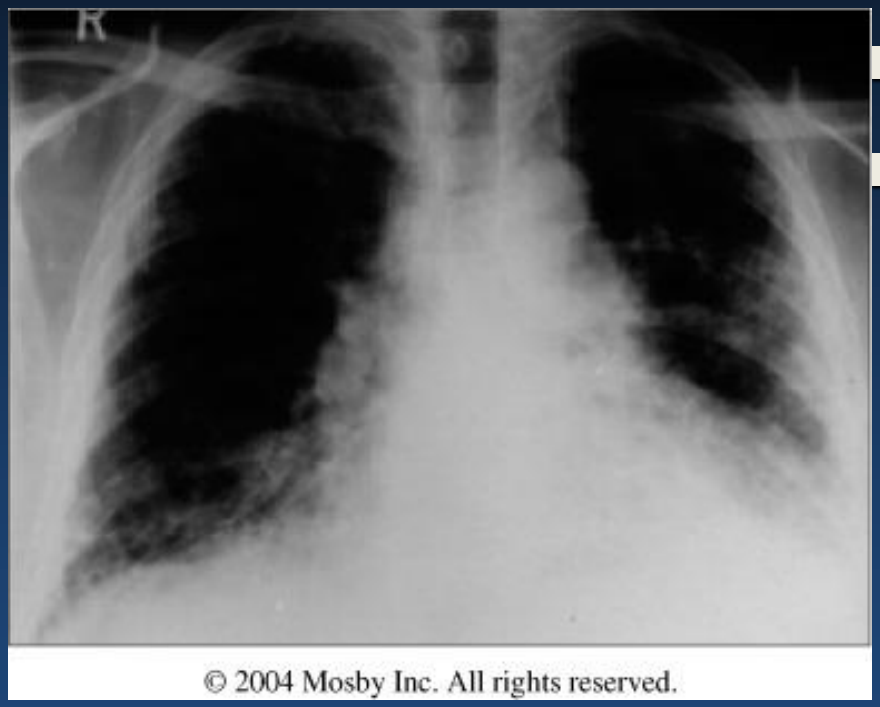
B2

IPF- Lung Function Test

- Low FEV1
- Normal FEV1/FVC
- Low TLC
- Low Dlco
- Hypoxemia (exercise)
- High A-a Diff

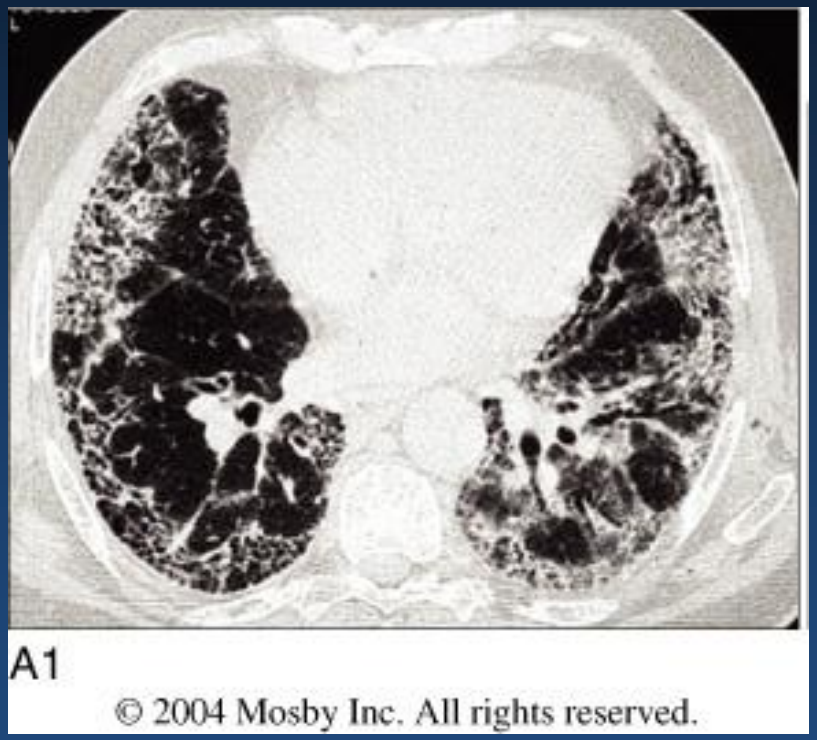


Chest X Ray: IPF



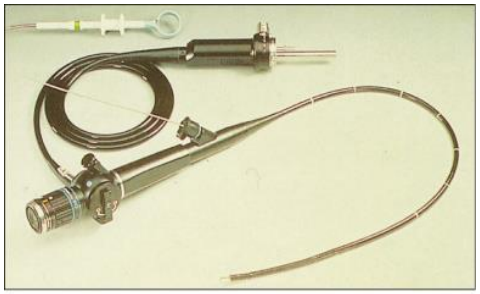
- Low lungs volume.
- Reticulonodular or reticular pattern in the base of the lungs.

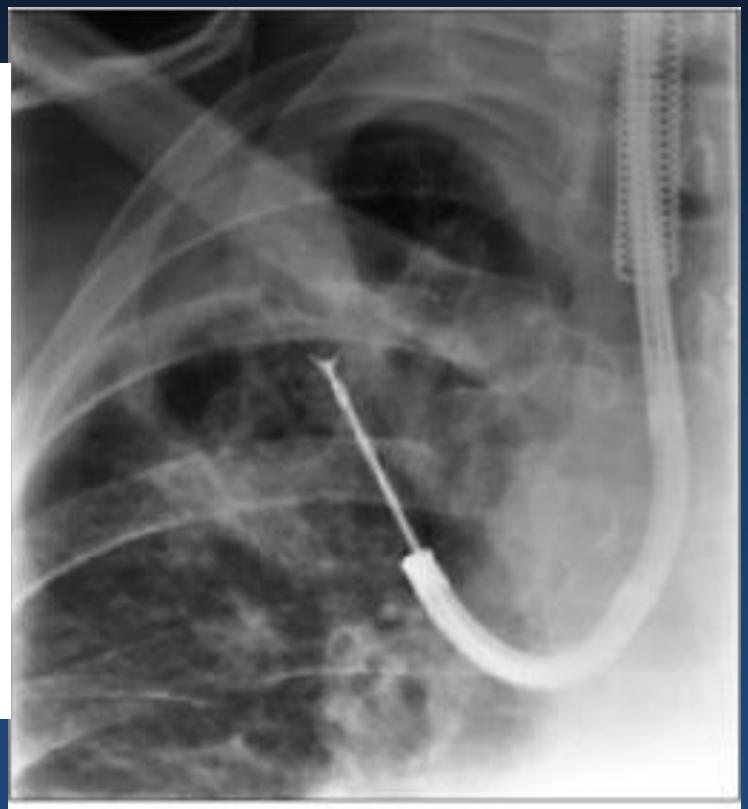
High Resolution CT- IPF



- Highly diagnostic
- Honey combing in late stage.
- Traction bronchiectasis.

Bronchoscopy





Bronchoscopy in Lung Fibrosis

- 4-8 transbronchial biopsies.
- Aim: To rule out infections, carcinoma (Bronchoalveolar carcinoma), sarcoidosis
- Bronchoalveolar lavage (BAL) –high eosinophils and neutrophils.
- BAL has no role in monitoring disease activity.

Open Lung Biopsy in ILD

- “Gold standard” diagnosis.
- When the disease deteriorate and bronchoscopy was non-diagnostic

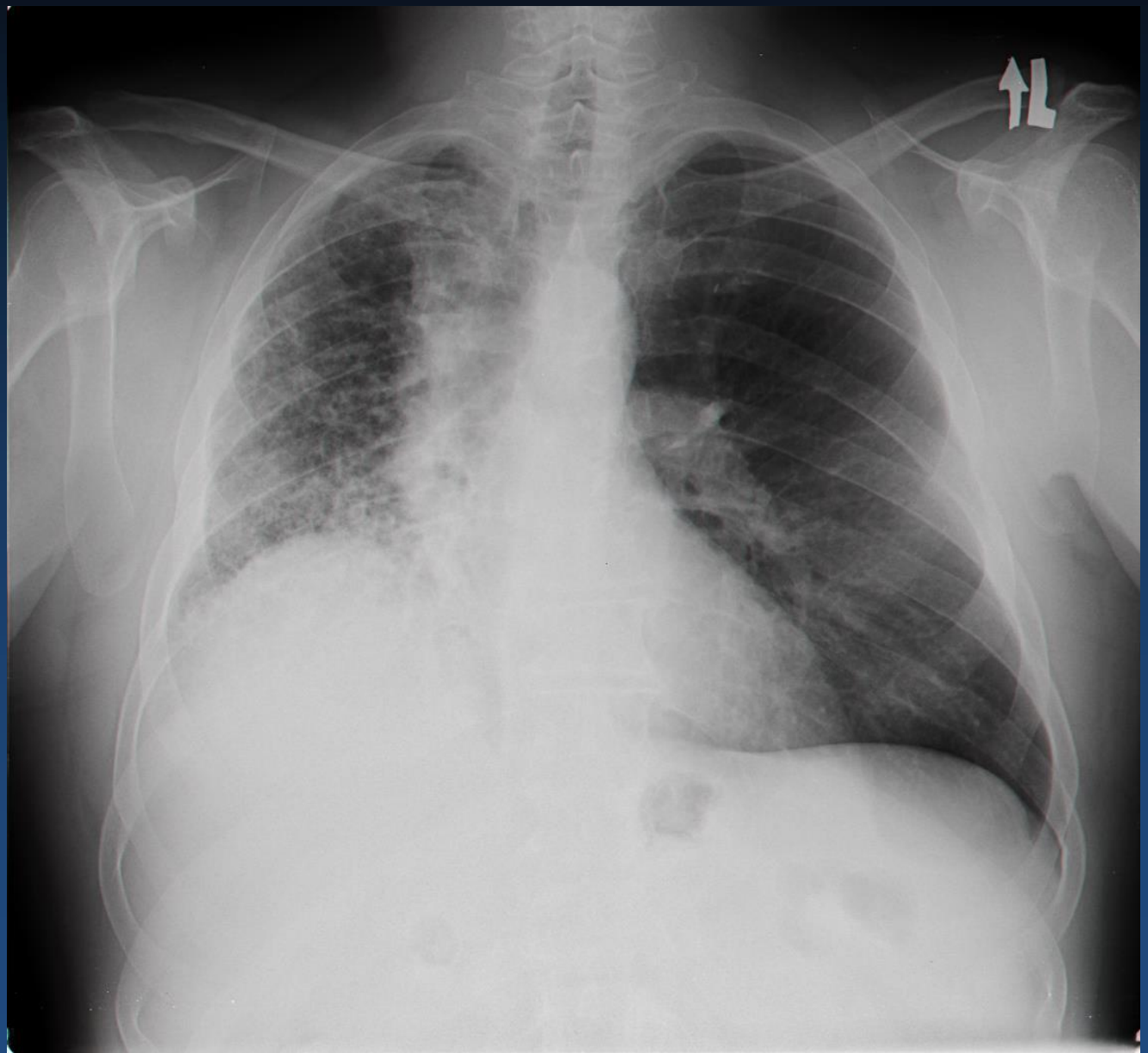
IPF-Therapy

- O2 therapy to all patients with hypoxemia
- Trial of oral steroids (predinsone) 0.5-1 mg/kg
- In cases of response- lower dose 0.25-0.5 mg/kg
- In unresponse cases- add immunosuppression: Cyclophosphamide (daily or I.V. every month).
- Only 20-30% response in IPF.

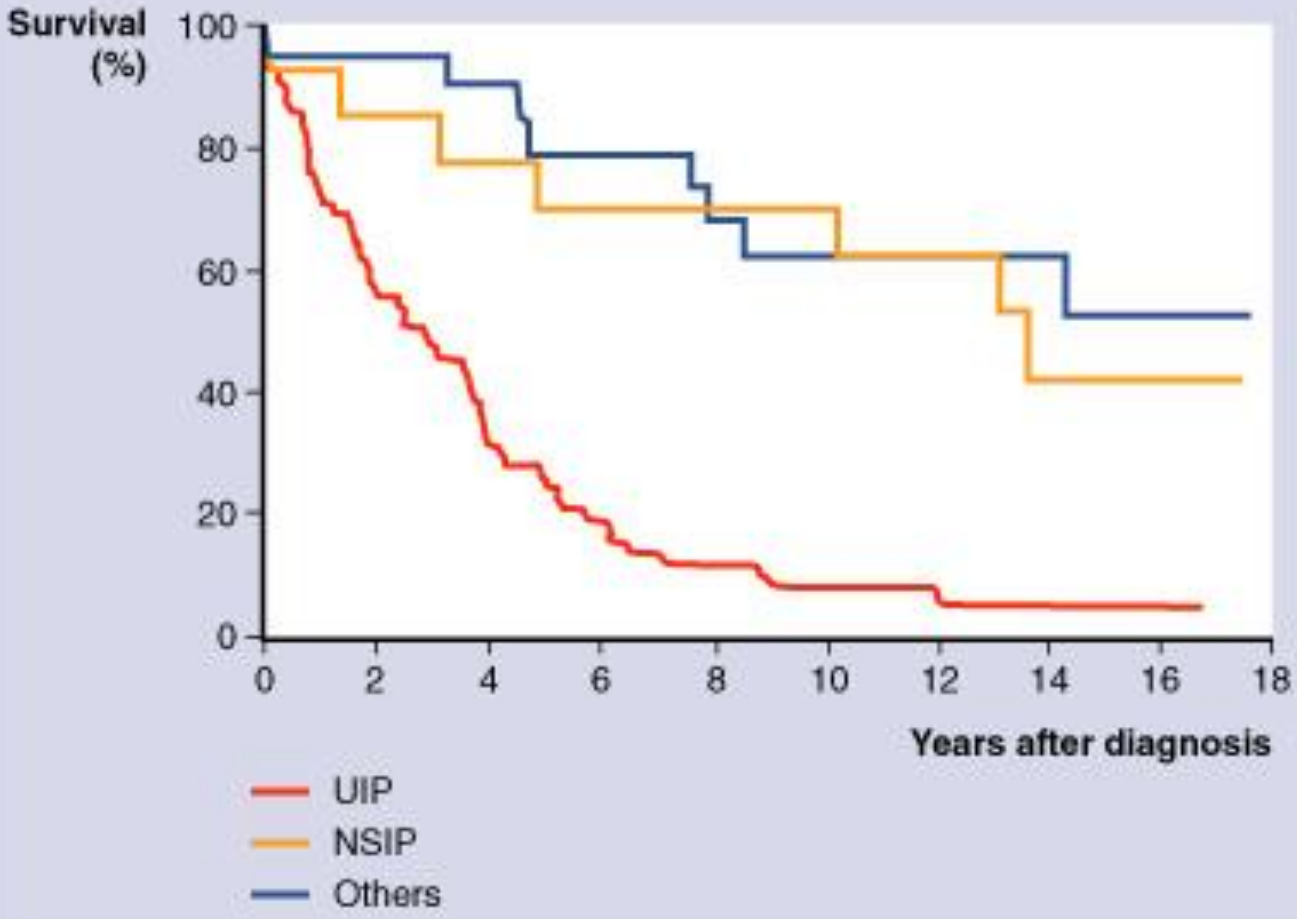
IPF-Therapy (2)

- Other therapeutic options:
  - Azathioprine (Immuran)
  - Colchicine
  - Methotrexate
  - Cyclosporine
  - Lung transplantation

Lung transplantation



Survival in Idiopathic Interstitial Pneumonias



Questions?