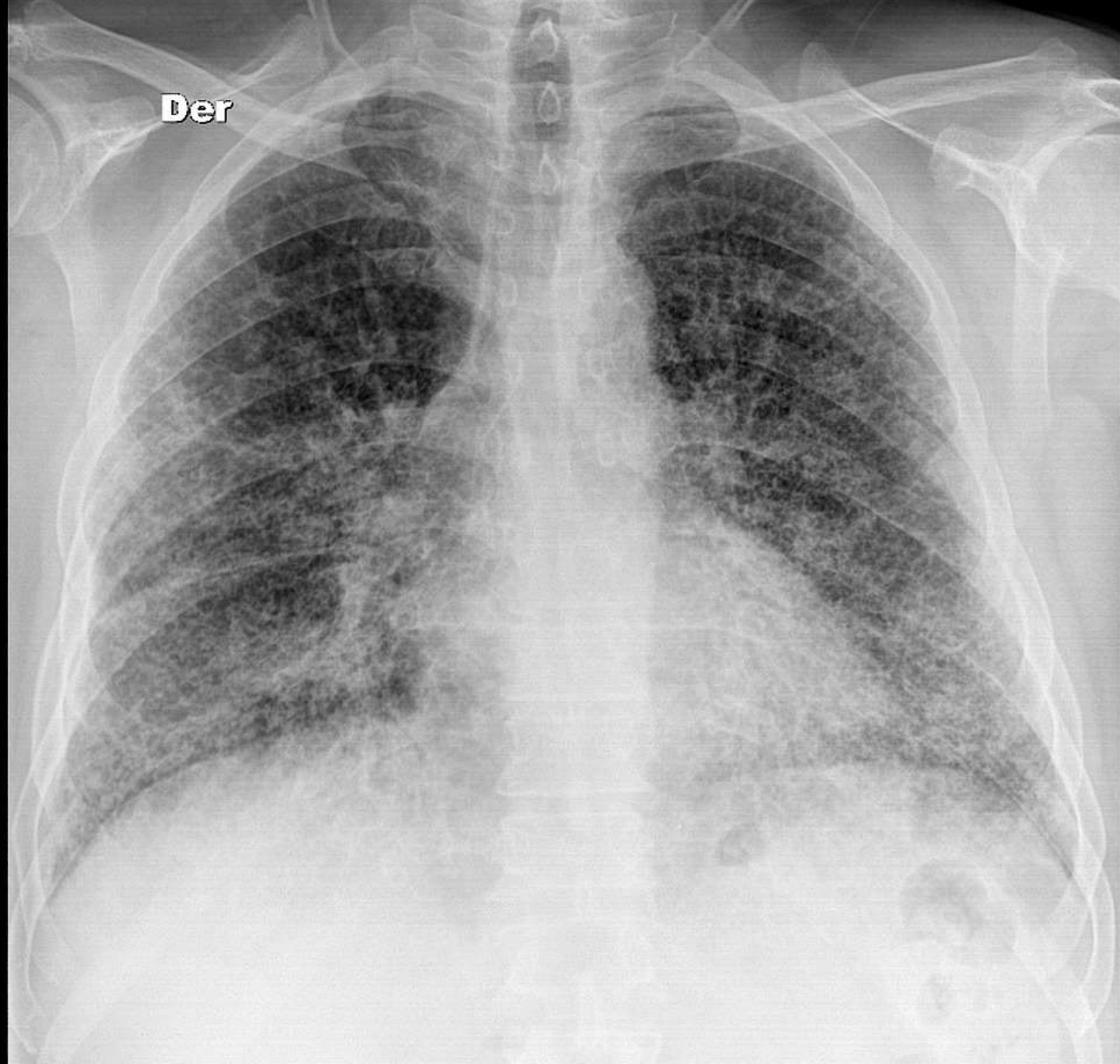

A Rapid Overview

IDIOPATHIC PULMONARY FIBROSIS (IPF)

Also Known As

CRYPTOGENIC FIBROSING ALVEOLITIS (CFA)



- **Idiopathic Interstitial Pneumonias (IIP)** makes up major portion of **Diffuse Parenchymal Lung Diseases (DPLD)** *or* **Interstitial Lung Disease**
- They have no known cause
- **Idiopathic Pulmonary Fibrosis (IPF)** is the main disease in Interstitial Pulmonary Pneumonia (IIP)

SYMPTOMS

- > 50 Years
- Dry Cough
- Progressive Shortness of Breath
- Malaise
- Weight Loss
- Arthralgia

SIGNS

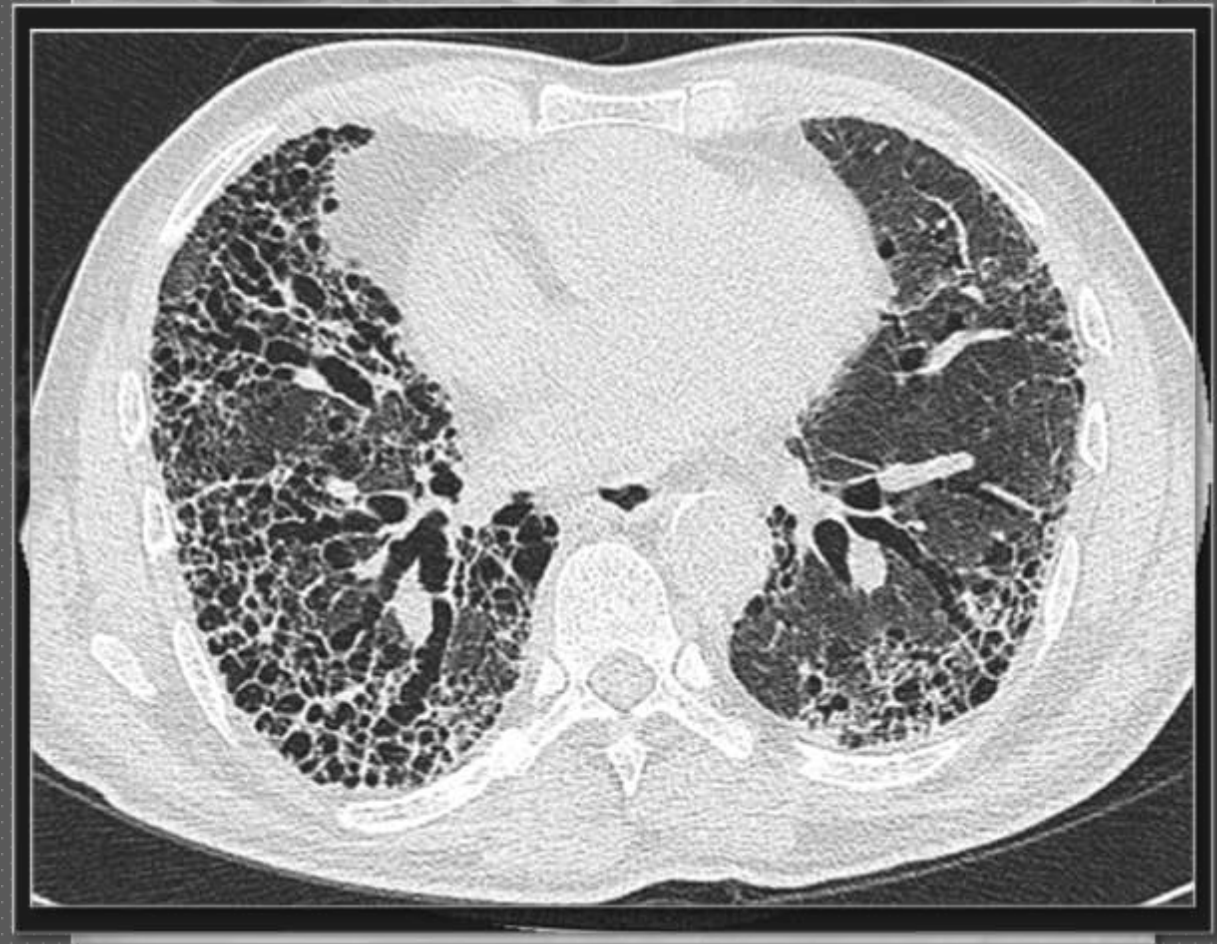
- Cyanosis
- Clubbing
- Fine End-Inspiratory Crepitations

LABORATORY

- CRP – Raised
- Immunoglobulins – Raised
- ABGs – Low pO_2 , +/- High pCO_2
- ANA - + in 30%
- RF - + 10%
- Anti CCP2 Ab

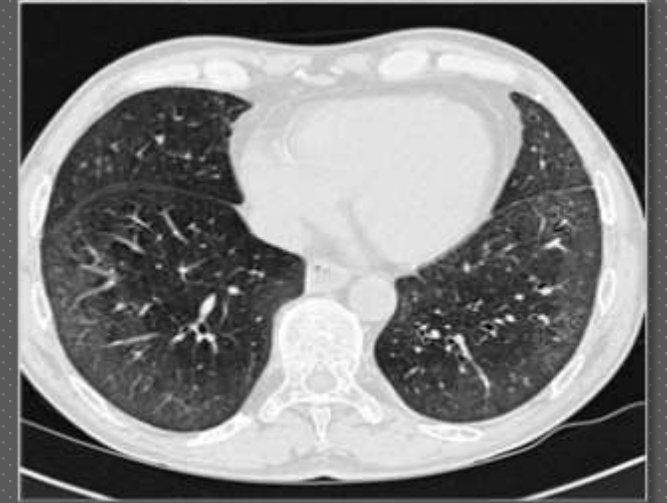
IMAGING

- Chest X-ray
- HRCT scan
 - Reduced Lung Volume
 - Reticular shadowing
 - *Bilateral Lower Lobes*
 - *Subpleural*
 - Traction Bronchiectasis
 - Honeycombing



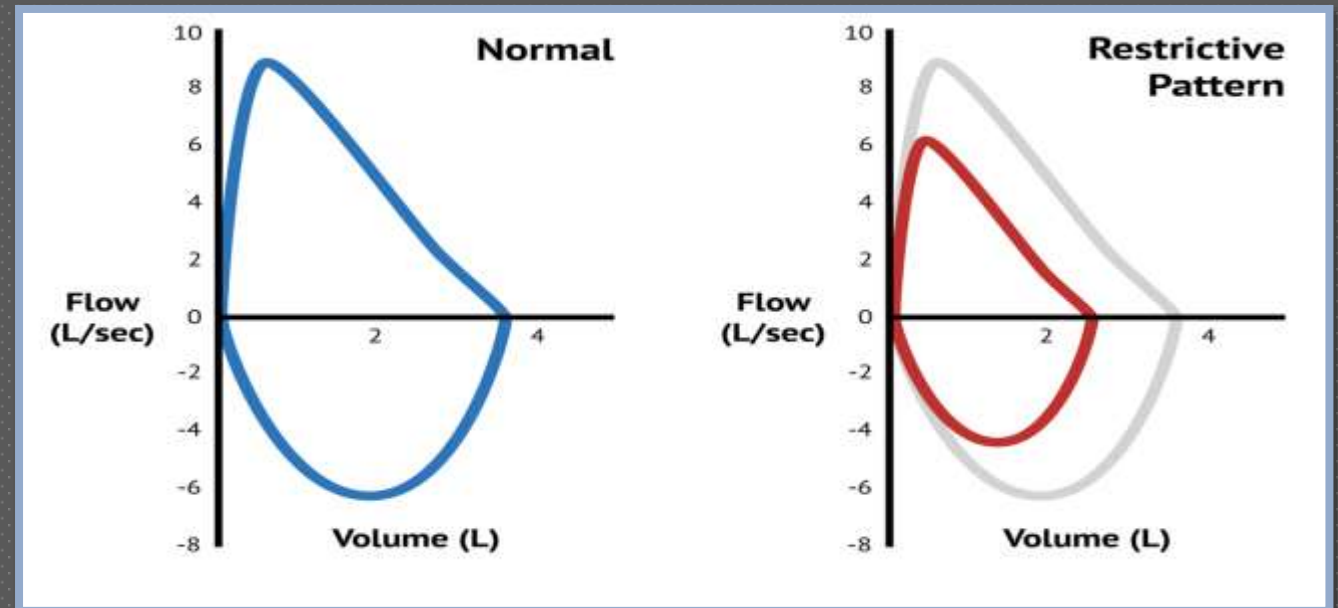
IMAGING

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SPIROMETRY

- Lung Volume
 - *Total Lung Capacity (TLC) - Reduced*
- Restrictive Lung Defect
 - *FVC - Reduced*
 - *FEV1 - Reduced/Normal*
 - *FEV1/FVC - Normal/Increased*
- Impaired Gas Transfer
 - *DLCO - Reduced*



BRONCHOSCOPY & BAL

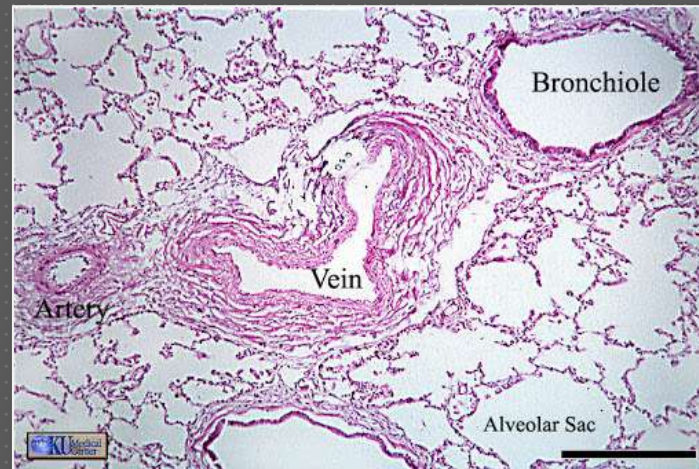
- Seldom Indicated
- D/D of Infection/Malignancy
- BAL Differential Cell Count
 - *Alveolitis Activity*
 - *Lymphocytes > **Good** Response & Prognosis*
 - *Neutrophils/Eosinophils > **Poor** Response & Prognosis*

LUNG BIOPSY

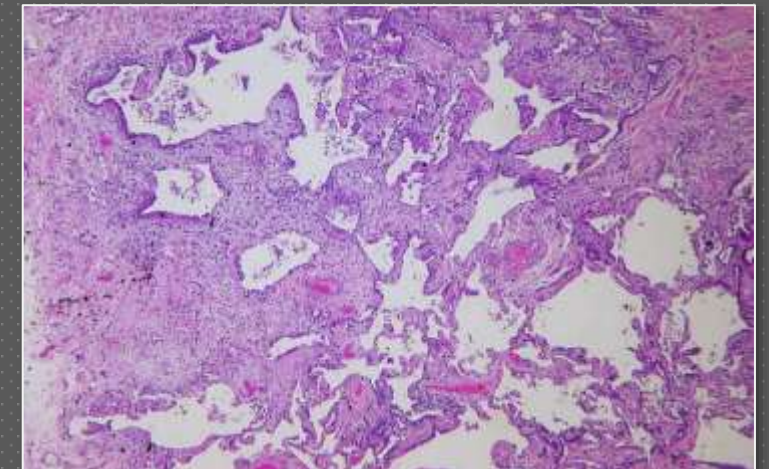
- Seldom Indicated
- Diagnostic Uncertainty or Atypical Feature
- Transbronchial – *Insufficient*
- Open Lung Biopsy - *Preferred*

HISTOPATHOLOGY

Usual Interstitial Pneumonia (UIP) pattern



Normal



UIP

- **Occupational lung diseases** - Asbestosis
- **Medications** – Nitrofurantoin, Busulphan, Amiodarone, Sulfasalazine
- **Connective Tissue Diseases** – SLE, Polymyositis, Dermatomyositis, RA

- **Pirfenidone** (Antibrotic & Immunosuppressant agent) or **Nintedanib** (Tyrosine Kinase Inhibitor) – If Vital Capacity between 50 – 80% predicted
- **Anti-Reflux** – Improve cough
- **Exercise & Pulmonary** Rehabilitation
- **Smoking Cessation**
- **Vaccination** – Influenza & Pneumococcal
- **Domiciliary Oxygen**
- **Lung Transplantation**
- **Exacerbations** – Supportive

