



# Interstitial Lung Disease

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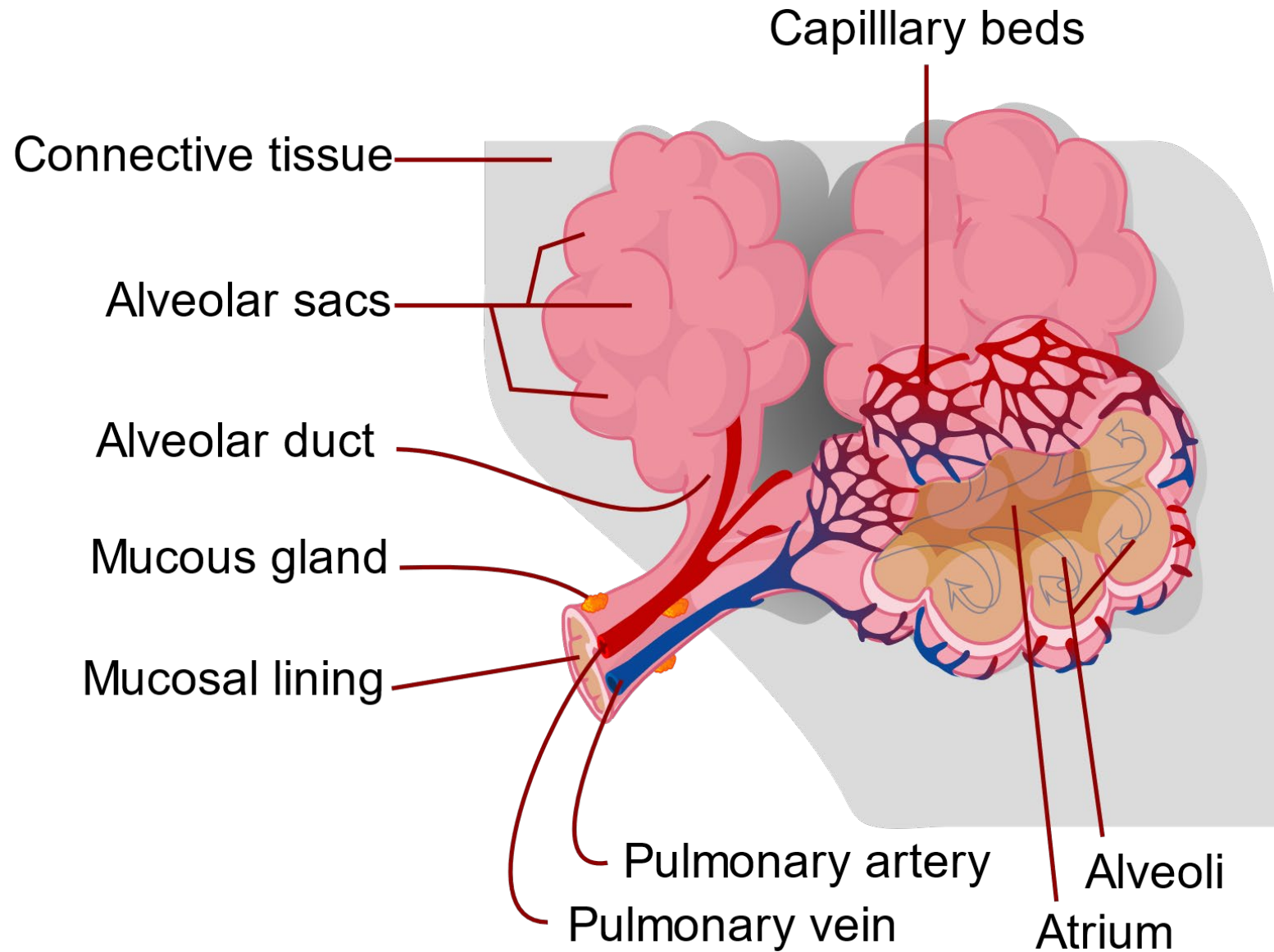


# ILD-General Concepts

- ▶ Also known as diffuse parenchymal lung diseases
- ▶ Heterogenous group of disorders with similar manifestations
  - ▶ Radiographic
  - ▶ Physiologic
  - ▶ Clinical
  - ▶ Histopathology

# Pulmonary Interstitium

- ▶ Space between functional lung units (alveoli)
- ▶ Includes vascular structures, connective tissue, lymphatics
- ▶ Essentially everything outside the airways and alveoli



# ILD-Classification

- ▶ Known causes
  - ▶ Exposure to occupational and environmental factors
  - ▶ Drug induced injury
  - ▶ Radiation induced injury
  - ▶ ILD associated with connective tissue diseases
- ▶ Idiopathic
  - ▶ Sarcoidosis
  - ▶ Cryptogenic organizing pneumonia
  - ▶ Idiopathic interstitial pneumonias (IPF)

# ILD-Clinical Presentation

- ▶ Progressive dyspnea on exertion
- ▶ Non-productive cough
- ▶ Pulmonary symptoms associated with another disease
- ▶ Family history of ILD
- ▶ History of occupational exposure
- ▶ Abnormal radiograph (CXR, CT)
- ▶ PFT abnormality

# ILD-Clinical Evaluation

- ▶ History
  - ▶ Age and gender
  - ▶ Onset and duration of symptoms
  - ▶ Past medical history
  - ▶ Smoking history
  - ▶ Family history
  - ▶ Medication use and irradiation
  - ▶ Occupational and environmental exposures



# ILD-Clinical Evaluation

- ▶ Physical exam
  - ▶ Lung exam-Rales, late inspiratory squeaks
  - ▶ Cardiac exam-Normal early, signs of pulmonary HTN or cor pulmonale late
  - ▶ Clubbing
  - ▶ Findings of systemic disease (amyloidosis, neurofibromatosis, sarcoidosis, tuberous sclerosis)

# ILD-Diagnostic Testing

- ▶ Lab studies
  - ▶ CBC with differential (eosinophilia)
  - ▶ Serologic testing (ANA, RF, etc.)
- ▶ Imaging
  - ▶ Chest X-Ray
  - ▶ CT High resolution technique, inspiratory and expiratory, prone and supine
- ▶ Pulmonary function testing
  - ▶ Spirometry, lung volumes, diffusing capacity
  - ▶ Rest and exercise pulse oximetry

# ILD-Diagnostic Testing

- ▶ Cardiac evaluation
  - ▶ Echocardiogram
  - ▶ Cardiopulmonary exercise testing
- ▶ Bronchoscopy
  - ▶ Bronchoalveolar lavage
  - ▶ Transbronchial biopsy
- ▶ Surgical lung biopsy
  - ▶ Thoracoscopic
  - ▶ Thoracotomy



# Specific ILD's aka Alphabet Soup

- ▶ UIP-Usual interstitial pneumonitis
- ▶ NSIP-Nonspecific interstitial pneumonitis
- ▶ DIP-Desquamative interstitial pneumonia
- ▶ AIP-Acute interstitial pneumonitis
- ▶ RB-ILD-Respiratory bronchiolitis associated ILD
- ▶ LIP-Lymphocytic interstitial pneumonitis
- ▶ LAM-Lymphangiomyomatosis
- ▶ COP-Cryptogenic organizing pneumonia
- ▶ HP-Hypersensitivity pneumonitis

# Specific ILDs

- ▶ Usual interstitial pneumonitis/Idiopathic pulmonary fibrosis
- ▶ Hypersensitivity pneumonitis

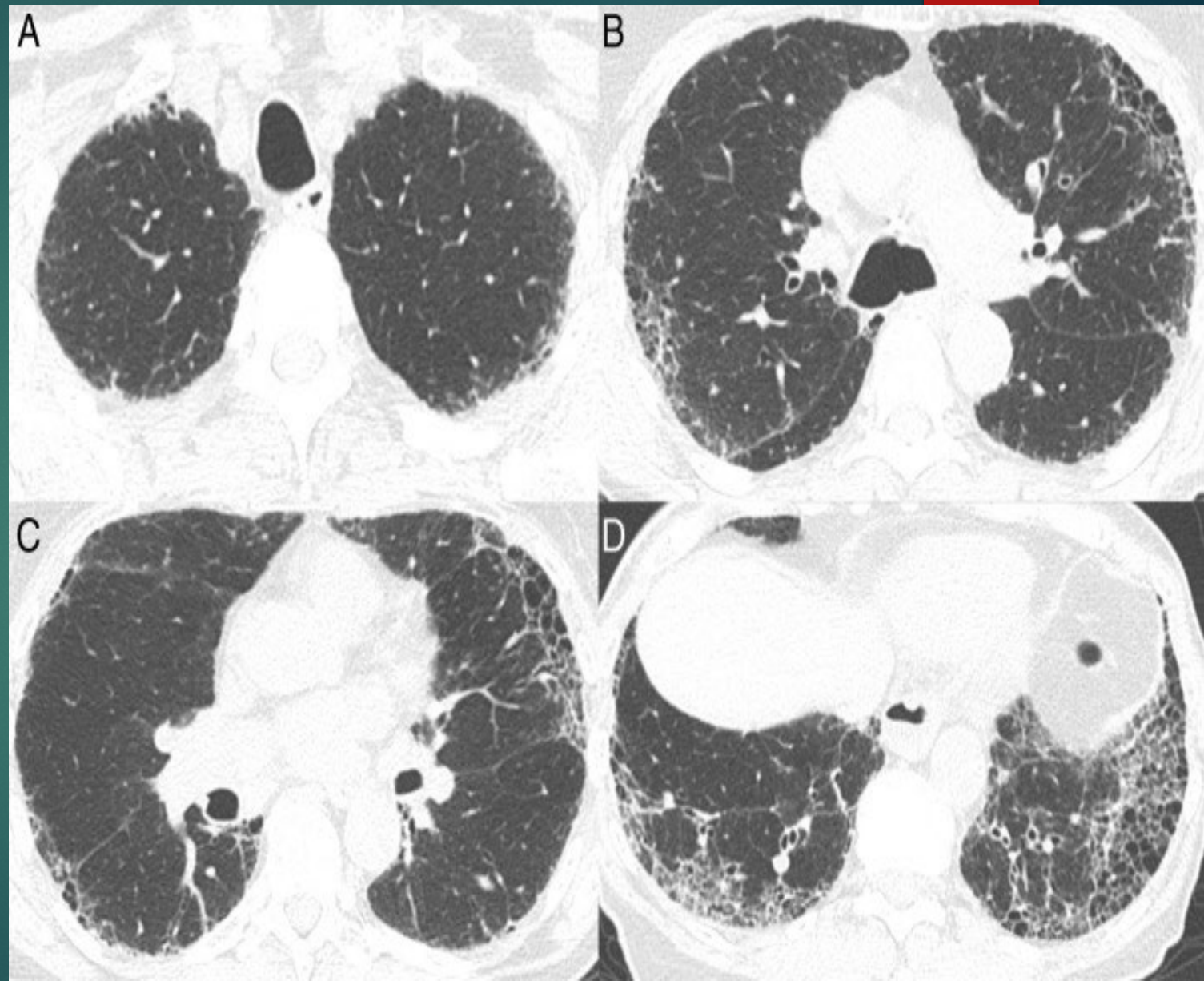
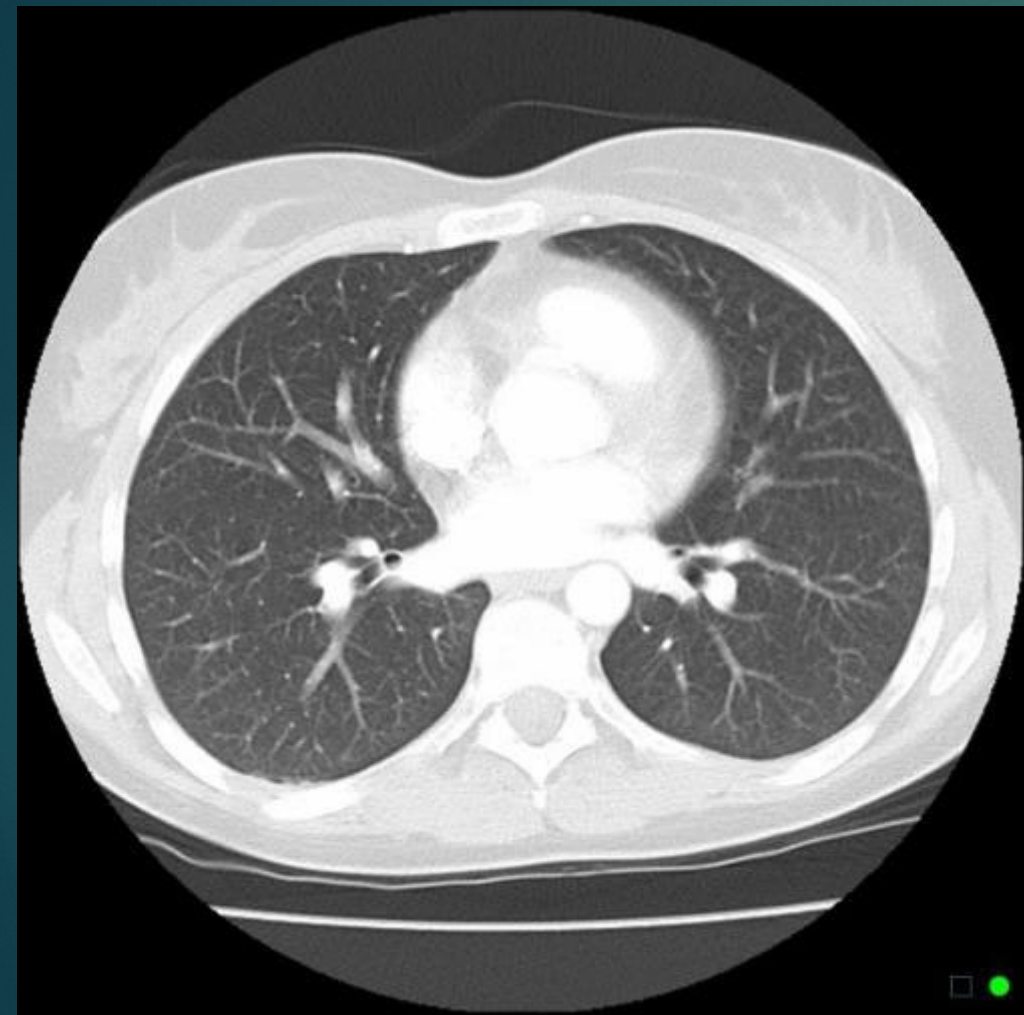
# Usual Interstitial Pneumonitis

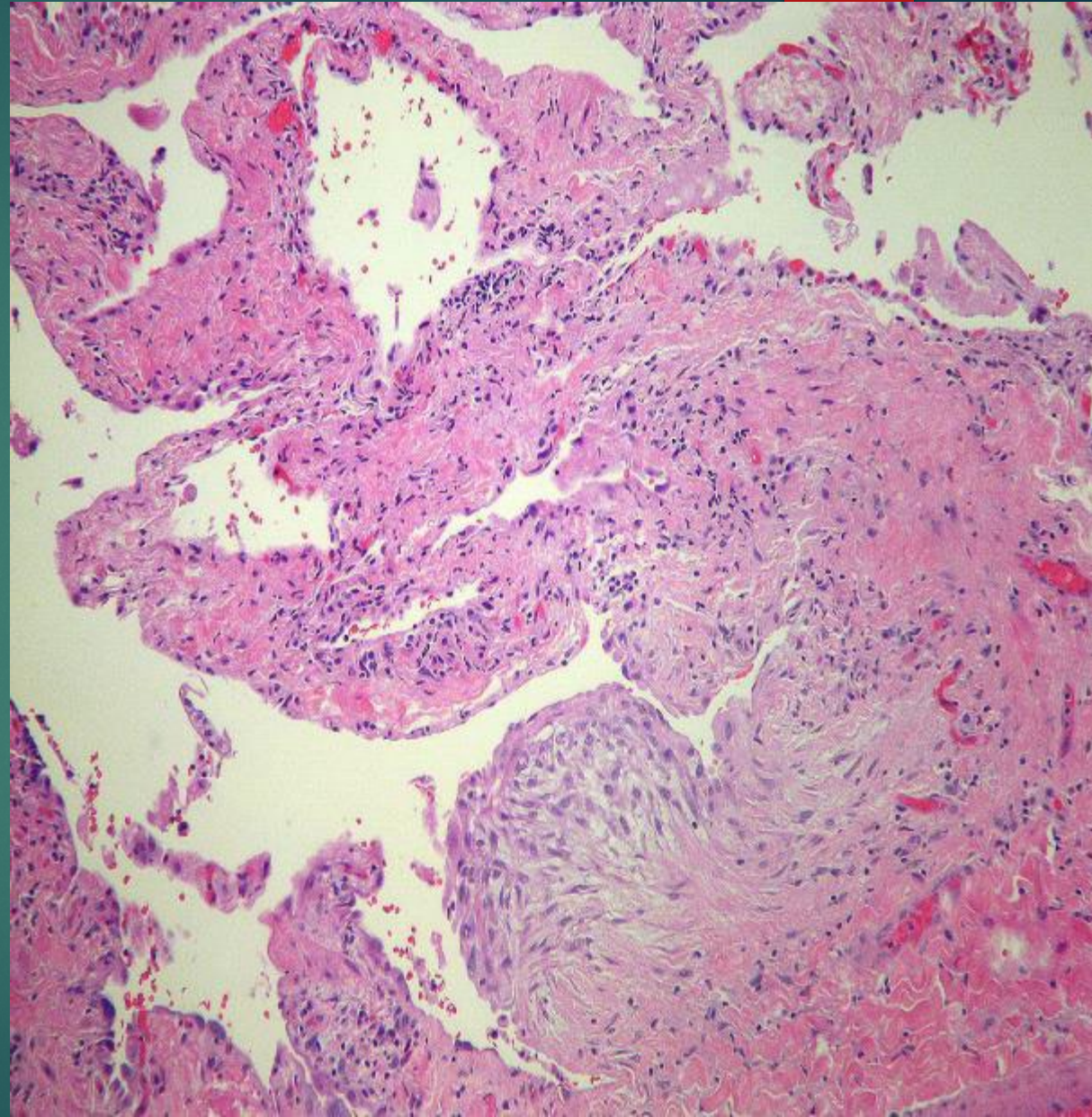
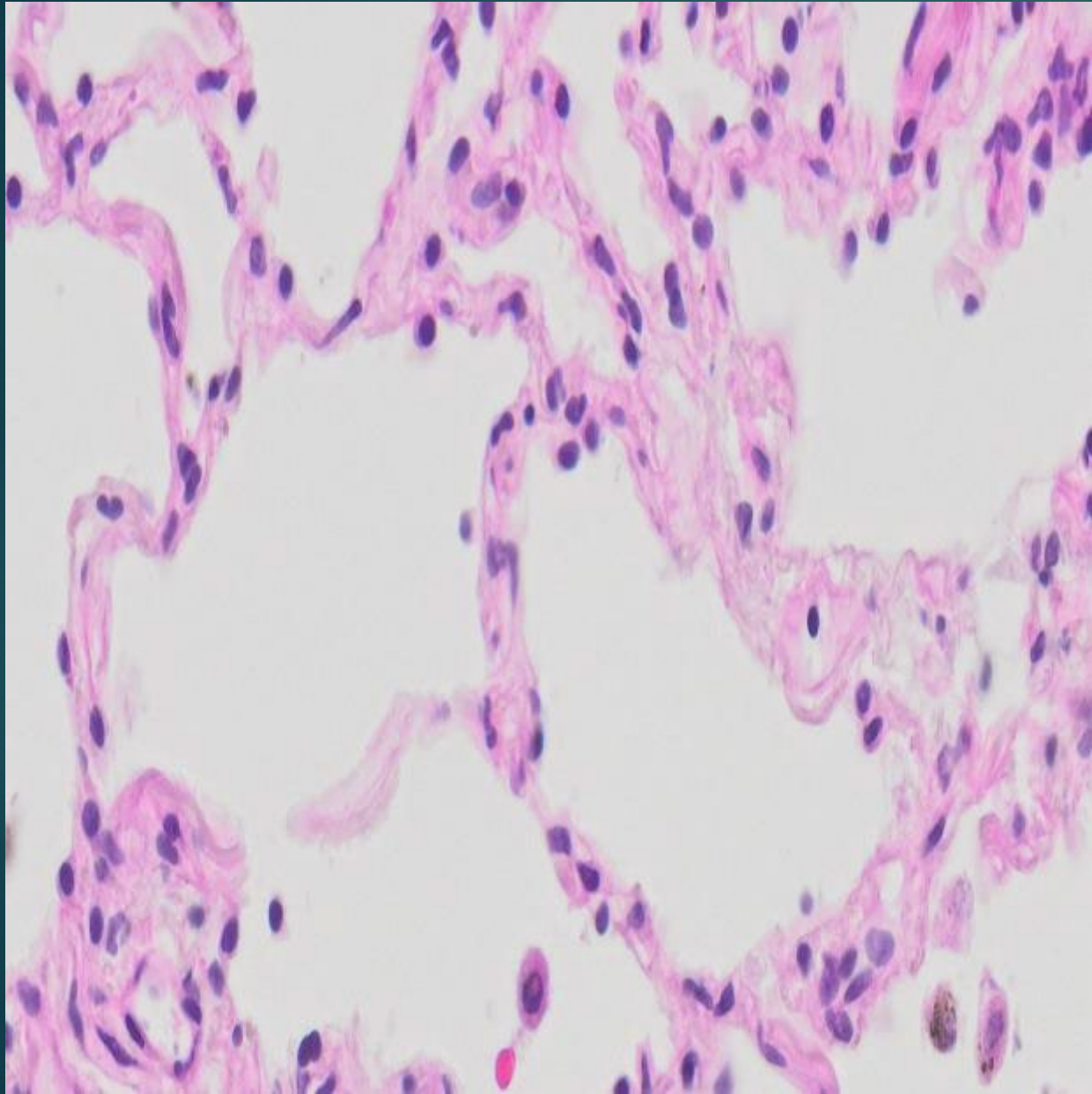
- ▶ Pathologic term for clinical entity idiopathic pulmonary fibrosis
- ▶ Most common idiopathic interstitial pneumonia
- ▶ Usually presents in 6<sup>th</sup> or 7<sup>th</sup> decade
- ▶ 3-9 cases per 100,000, higher among older populations
- ▶ Risk factors include smoking, male>female, industrial dust

# UIP-Clinical Features

- ▶ Gradual onset of dyspnea and nonproductive cough over several months
- ▶ Constitutional symptoms rare
- ▶ Crackles on exam
- ▶ Digital clubbing
- ▶ PFTs with restrictive pattern and reduced DLCO
- ▶ Biopsy not necessary if CT highly suggestive











# UIP-Treatment

- ▶ Supplemental oxygen
- ▶ Education
- ▶ Medications
  - ▶ Nintedanib (Ofev)
  - ▶ Perfenidone (Esbriet)
- ▶ Infection prevention/vaccinations
- ▶ Lung transplantation

# Hypersensitivity Pneumonitis

- ▶ Also known as Extrinsic Allergic Alveolitis
- ▶ Complex syndrome involving immunologic reaction to an inhaled agent, especially organic material
- ▶ Farmer's Lung, Bird Fancier's Lung, Baker's Lung, etc
- ▶ Affects as much as 0.4-7% of the farming population, more prevalent in humid areas
- ▶ Cigarette smoking associated with decreased risk

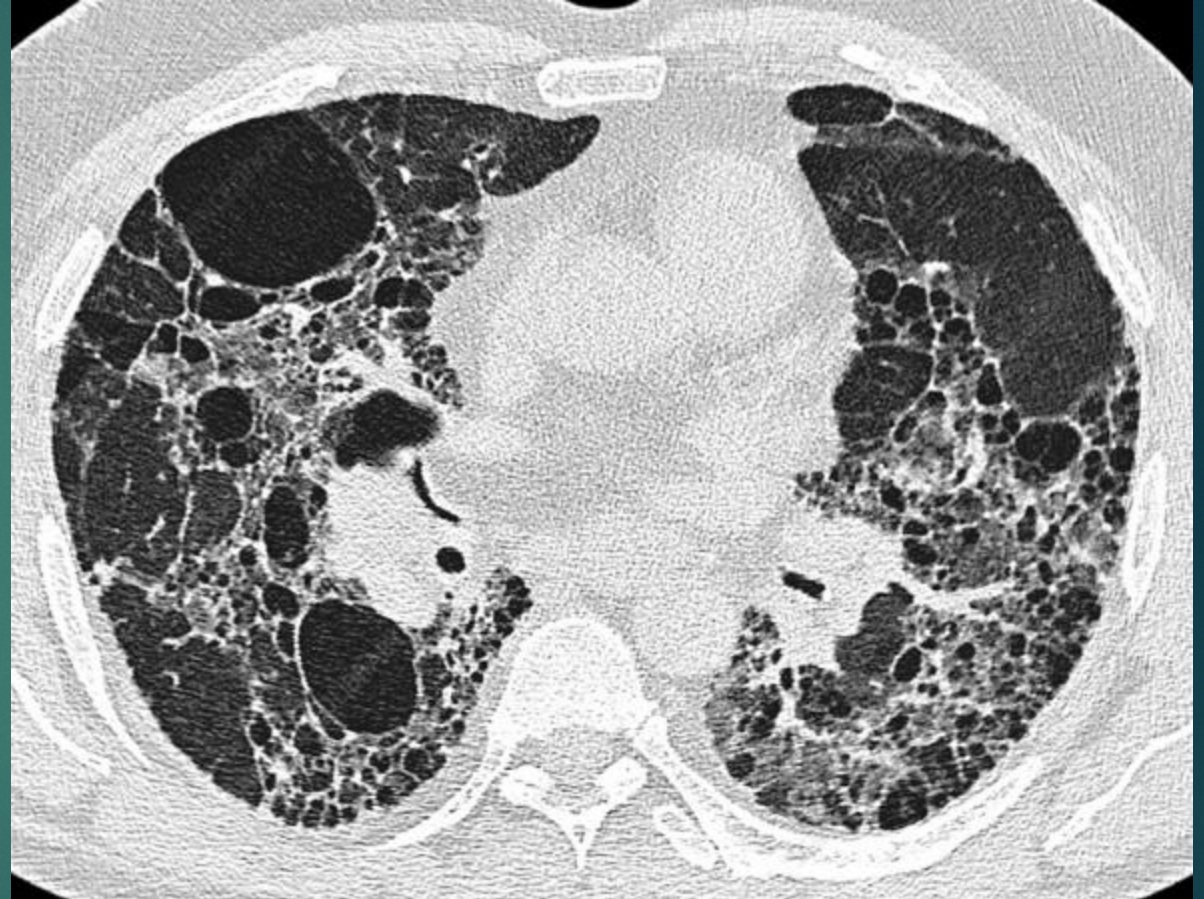
# HP-Clinical Presentation

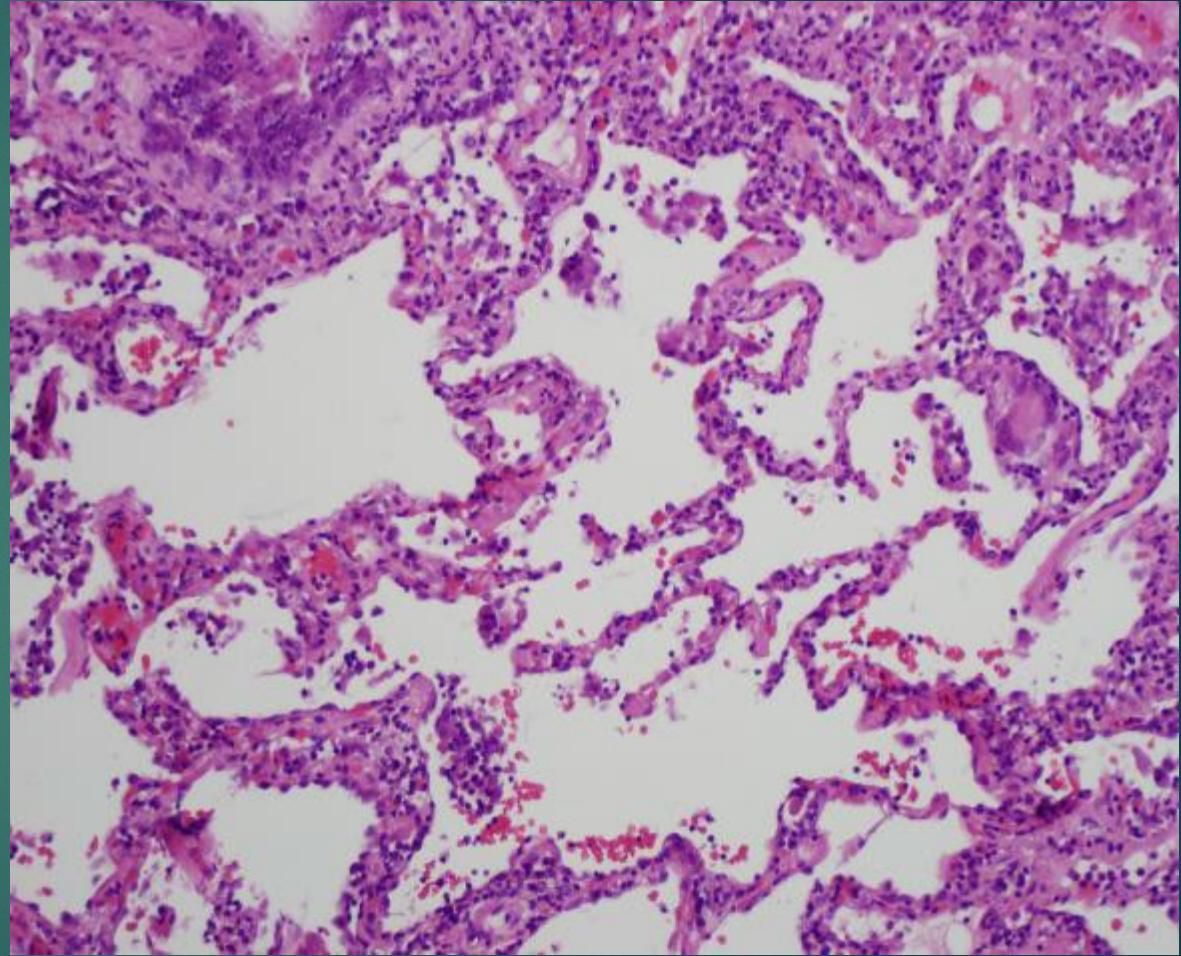
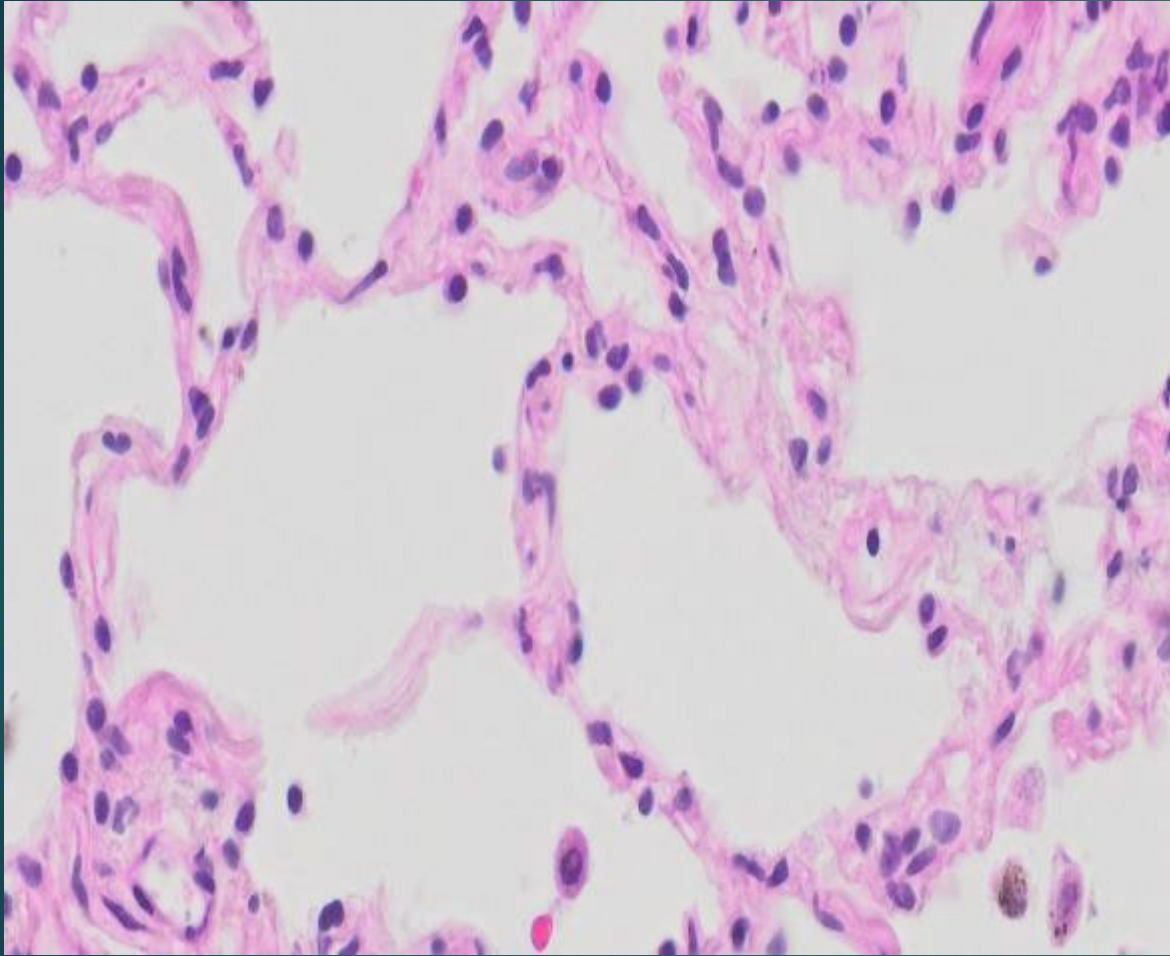
- ▶ Two phenotypes: Fibrotic and nonfibrotic
- ▶ Dyspnea and cough most common sx's.
- ▶ Fever, chills, weight loss, malaise
- ▶ May present over days-weeks or months-years
- ▶ Physical exam
  - ▶ Normal lung sounds to diffuse rales
  - ▶ Clubbing less common than in UIP

# HP-Clinical Evaluation

- ▶ Extensive occupational and recreational history
  - ▶ Hobbies, occupations, household exposures
- ▶ Lab tests-Not particularly helpful, can test for specific IgG antibodies but high false positive rate
- ▶ PFTs-No specific pattern but restriction and decreased DLCO common
- ▶ High resolution CT-Variable appearance, fibrotic vs. nonfibrotic
- ▶ Lung biopsy









# HP-Treatment

- ▶ Antigen avoidance
- ▶ Glucocorticoids
- ▶ Immunosuppressives
  - ▶ Azathioprine (Imuran)
  - ▶ Mycophenylate (Cellcept)
- ▶ Antifibrotics (nintedanib, pirfenidone)
- ▶ Lung transplantation

