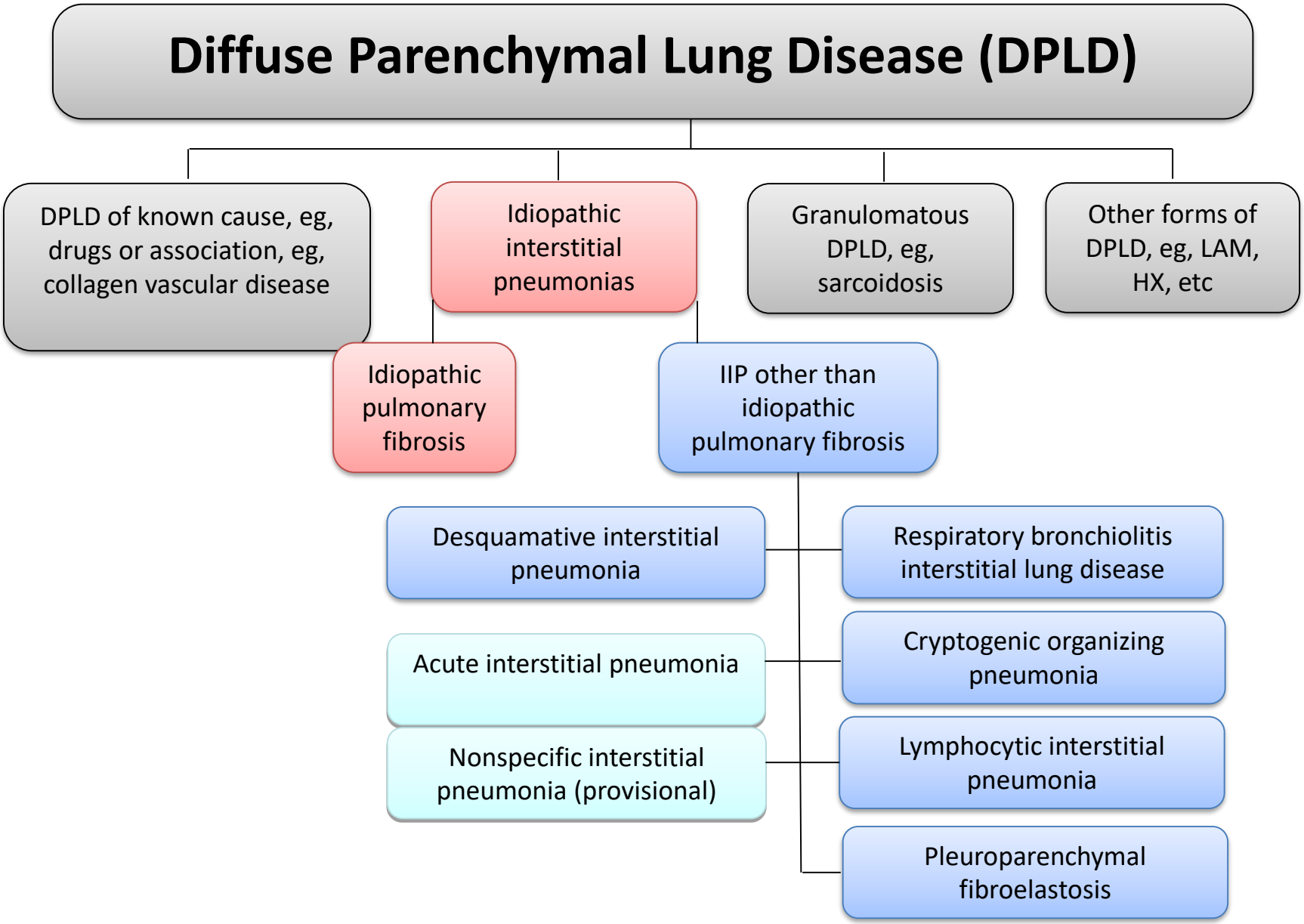
# Idiopathic pulmonary fibrosis

MUDr Lisá




Travis WD, et al; ATS/ERS Committee on Idiopathic Interstitial Pneumonias. *Am J Respir Crit Care Med*. 2013;188(6):733-748.

# Idiopathic Pulmonary Fibrosis

- Peripheral lobular fibrosis of unknown cause
- Clinical impact
  - Exertional dyspnea
  - Cough
  - Functional and exercise limitation
  - Impaired quality-of-life
  - Risk for acute respiratory failure and death
- Median survival time of 3-5 years
- Two new drugs approved by the FDA in October 2014
  - Nintedanib (Ofev)
  - Pirfenidone (Esbriet)

# Idiopathic Pulmonary Fibrosis Definition

- IPF is defined as a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, limited to the lungs, and associated with the histopathologic and/or radiologic pattern of UIP.

- The definition of IPF requires the exclusion of other forms of interstitial pneumonia including other idiopathic interstitial pneumonias and ILD associated with environmental exposure, medication, or systemic disease.

# Idiopathic Pulmonary Fibrosis

- IPF should be considered in patients over 50 years of age with unexplained insidious- and subacute-onset shortness of breath on exertion.
- It commonly presents with dry cough and bibasilar inspiratory Velcro crackles.
- Finger clubbing may be present.
- Male gender and cigarette smoking are

  risk factors for IPF

# Epidemiology

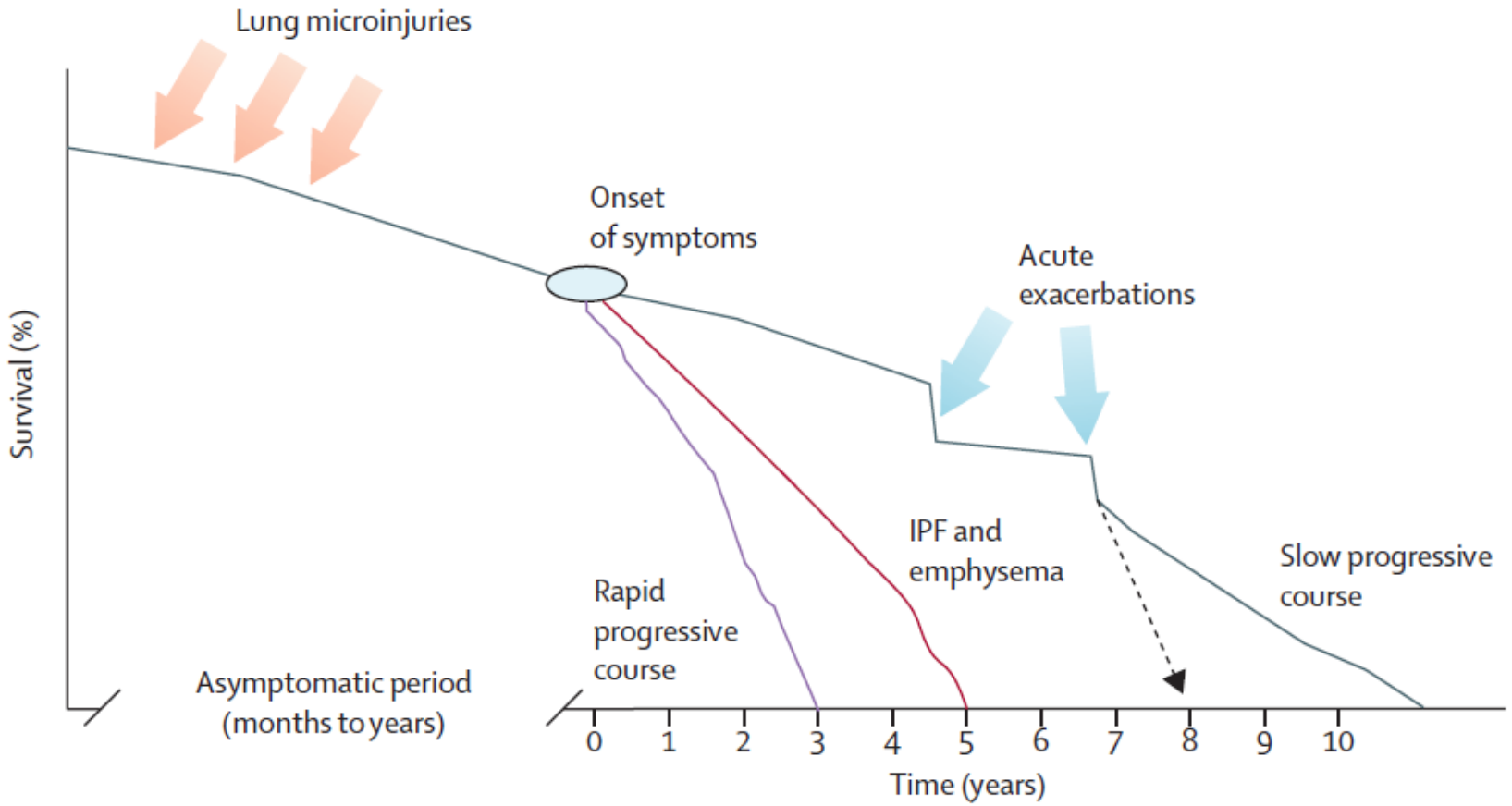
- Estimated prevalence of IPF is 14 to 43 cases per 100,000 people in the United states
- IPF is a disease of aging with a median age at diagnosis of 66
- IPF is more common in men
- Smoking is a risk factor for IPF
- A number of environmental associations have been identified, including metal dusts, wood dusts,stone/sand, and exposure to livestock

# Prognosis

- The prognosis of IPF is poor – patients have a median survival of 2 to 3 years from the time of diagnosis.

# The Disease Course of IPF is Extremely Variable

## "No one is average." – David Lederer, MD




King TE Jr, et al. *Lancet*. 2011;378(9807):1949-1961.

# IPF Radiographic Findings

***IPF cannot be ruled out by a normal chest radiograph***

| Typical Findings |
| --- |
| Reticular opacities at the bases and periphery |
| Reduced lung volume |
| Honeycombing |

| Atypical Findings |
| --- |
| Confluent alveolar opacities |
| Pleural disease |
| Significant lymphadenopathy |

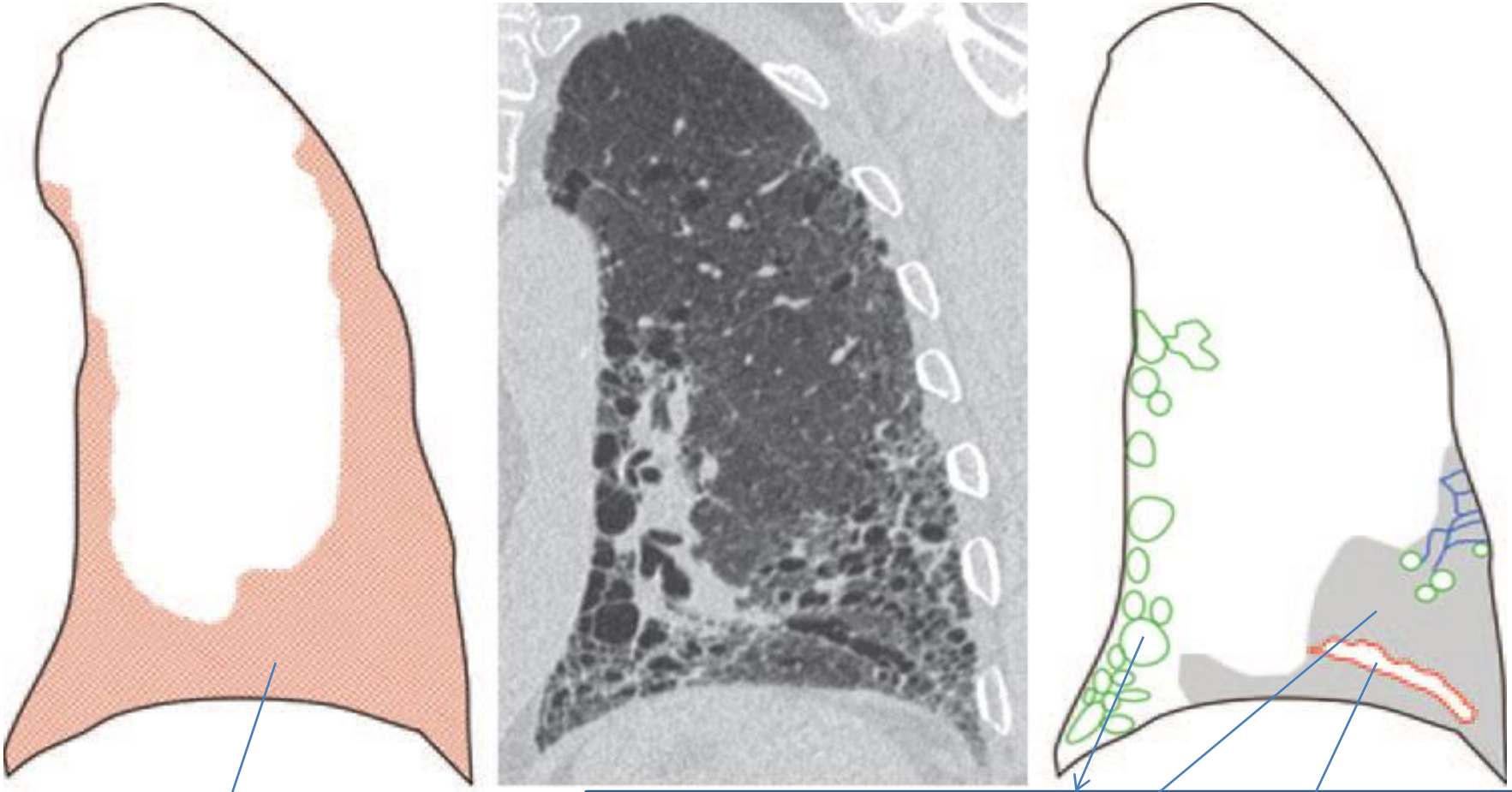
***16% of patients with ILD have normal chest radiographs***

# HRCT Criteria for UIP

| | UIP Pattern | Possible UIP Pattern |
|---|---|---|
| Subpleural, basal predominance | + | + |
| Reticular abnormality | + | + |
| Honeycombing (+/- traction bronchiectasis) | + | - |
| Absence of “inconsistent” features | + | + |

Raghu G, et al. *Am J Respir Crit Care Med*. 2011;183:788-824.

# Distribution **(a), CT image (b), and CT pattern (c) of UIP**



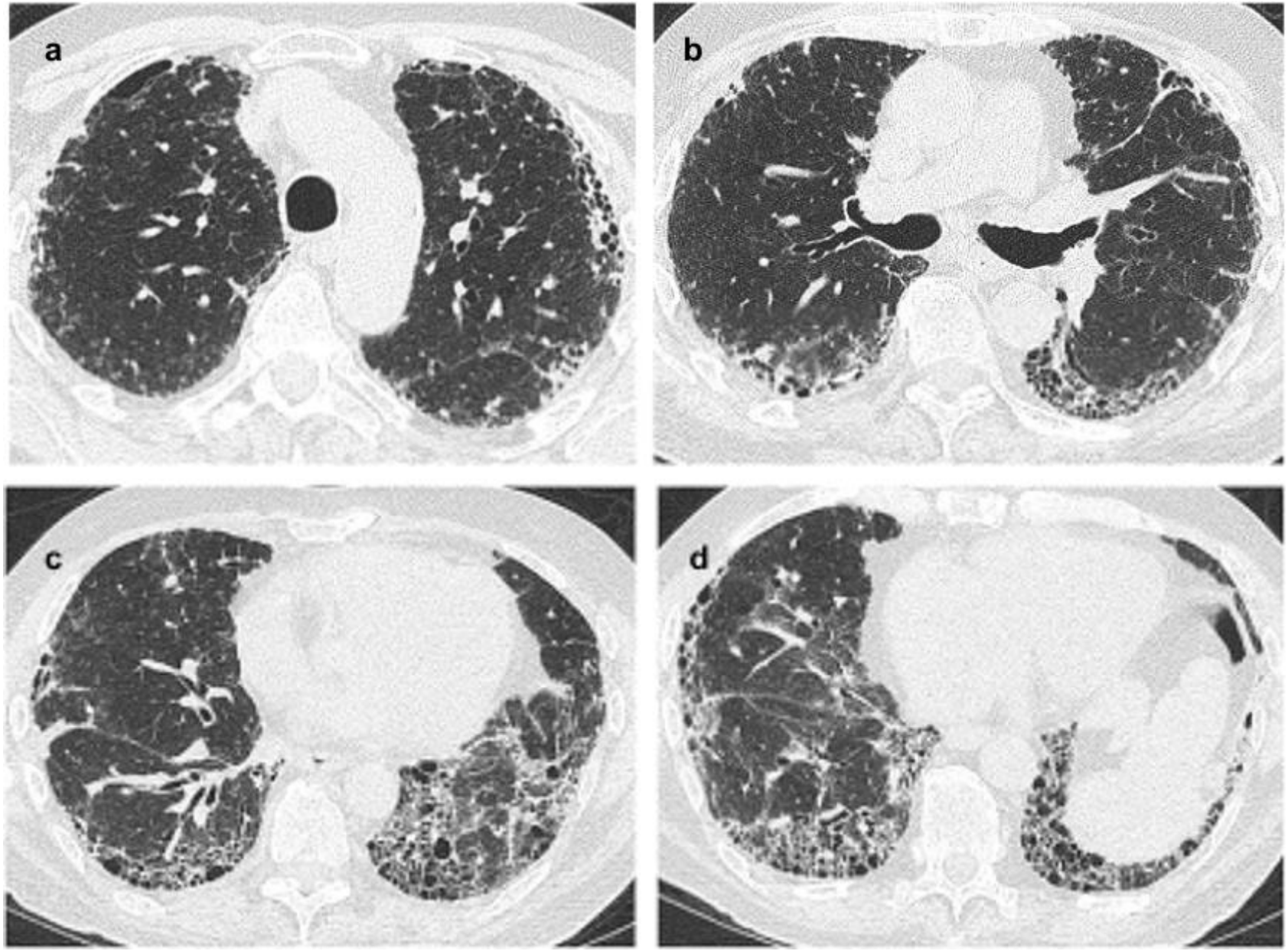
b.

- **The distribution is** subpleural with an apicobasal gradient (red area in **a**)

**CT shows honeycombing (green areas in c), reticular opacities (blue areas in c), traction bronchiectasis (red area in c), and focal** ground-glass opacity (gray area in **c**).

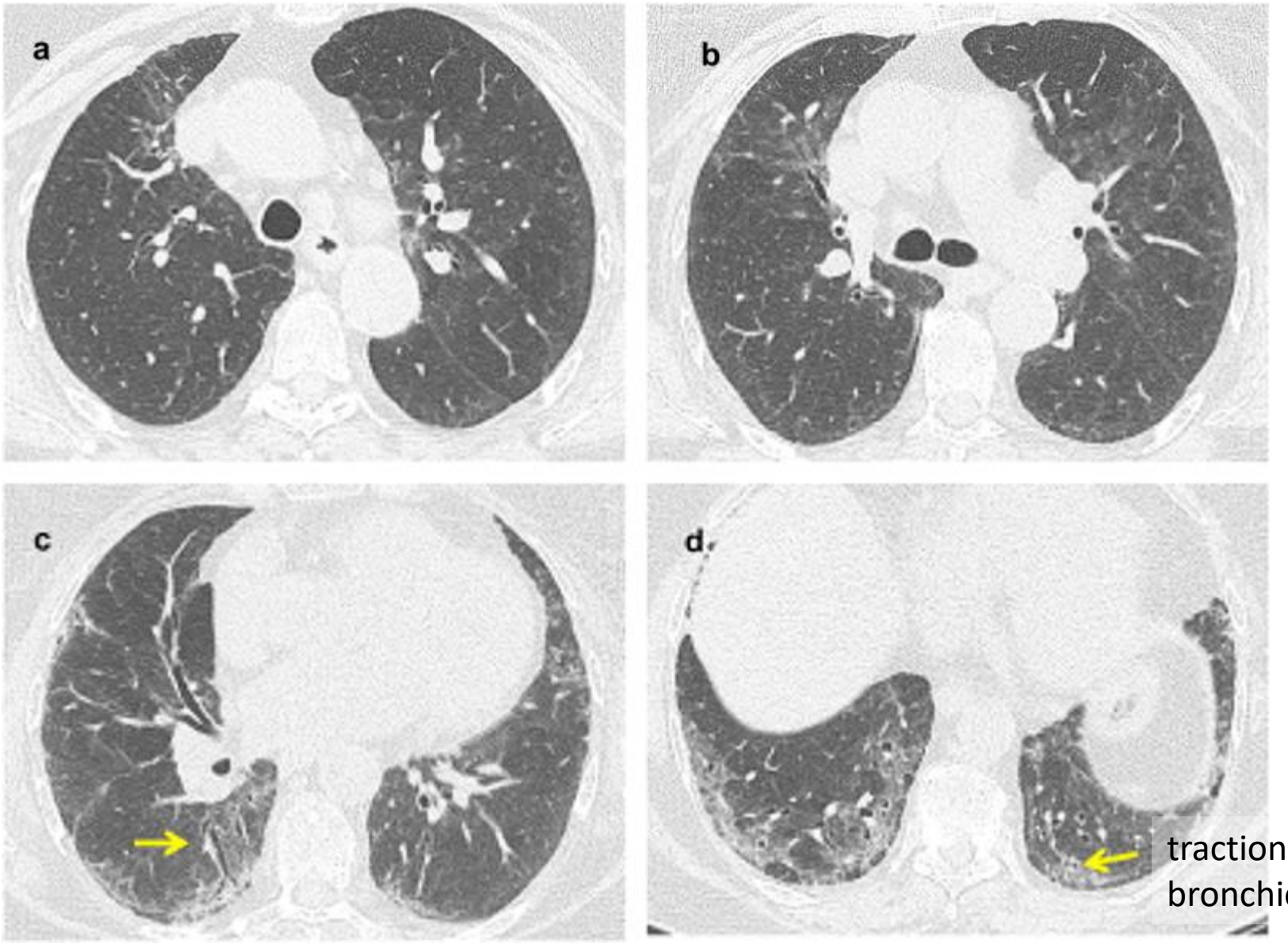
*Mueller-Mang CH. et al.What every radiologist schould konw about Idiopathic interstitial pneumonias. RadioGraphics 2007, 27: 595-615*

# UIP Pattern




Hodnett PA, et al. *Am J Respir Crit Care Med.* 2013;188:141-149.

# Possible UIP Pattern




traction bronchiectasis

Hodnett PA, et al. *Am J Respir Crit Care Med.* 2013;188:141-149.

# HRCT features *inconsistent* with IPF

| Inconsistent Features |
| --- |
| Upper lobe predominant |
| Peribronchovascular predominance |
| Ground-glass > extent of reticular abnormality |
| Profuse micronodules |
| Discrete cysts |
| Diffuse mosaic attenuation/gas-trapping |
| Consolidation |

Raghu G, et al. *Am J Respir Crit Care Med*. 2011;183:788-824.

# Histopathology

- Usual interstitial pneumonia (UIP) is characterized by a patchy, subpleural, and paraseptal pattern of involvement with areas of scarring alternating with uninvolved lung parenchyma.

  The involved lung shows dense collagen with scattered foci of proliferating fibroblasts (called fibroblastic foci)

# Pathofysiology

- Current theory holds that the development of UIP is extremely complex and involves, among other features, epithelial cell injury and death,aberrant wound-healing including failure of myofibroblasts to undergo apoptosis,excessive extra-cellular matrix deposition, and failure of alveolar epithelial cells to normally repopulate denuded alveolar basement membrane.

# Pathofysiology

- several environmental factors might contribute to epithelial injury and apoptosis

– Cigarette smoking
– Chronic silent microaspiration
– Chronic viral infection, mainly herpes
– Inhaled dusts

# Treatment IPF

-lung transplantation in appropriate patients

-long-term oxygen therapy in patients with IPF and clinically significant resting hypoxemia

-pulmonary rehabilitation

-treatment of symtomatic gastroesophageal reflux

- Two new drugs approved by the FDA
  - Nintedanib (Ofev)
  - Pirfenidone (Esbriet)