Interstitial Lung Disease

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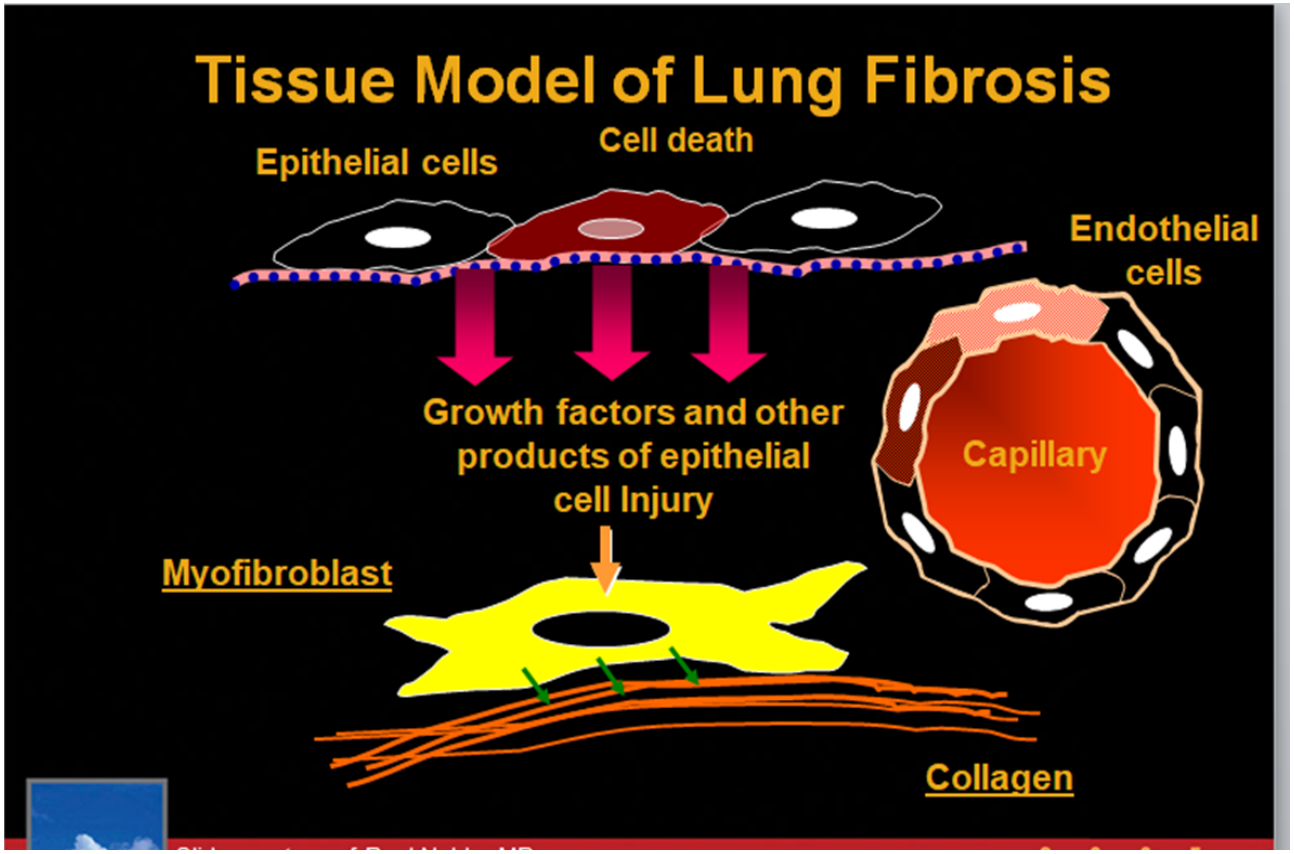
Respiratory and sleep medicine
JUH

Definition

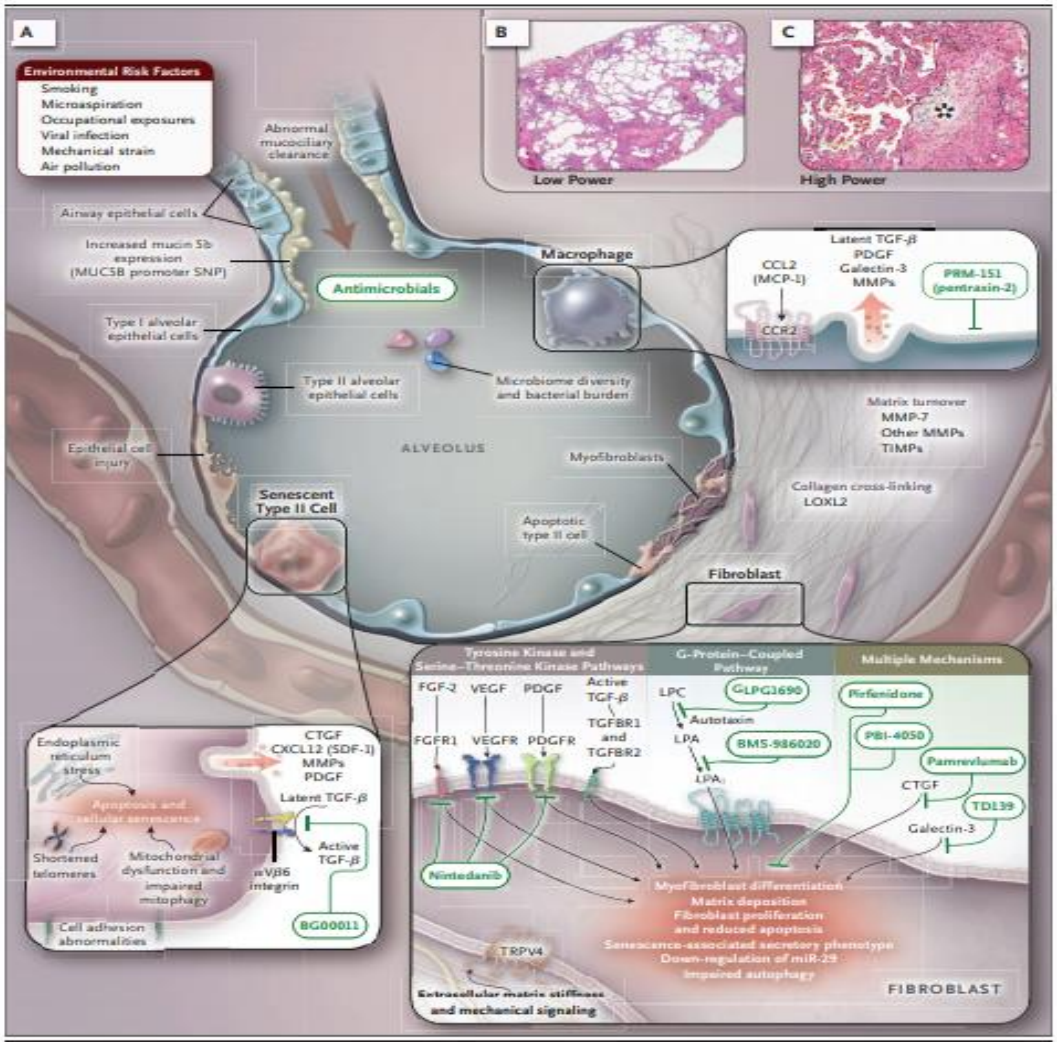
Group of pulmonary disorders characterized clinically by respiratory symptoms associated with :

- Radiologically diffused infiltrates.
- Histologically by distortion of the gas exchanging units.
- Physiologically by restriction of lung volumes and impaired oxygenation.

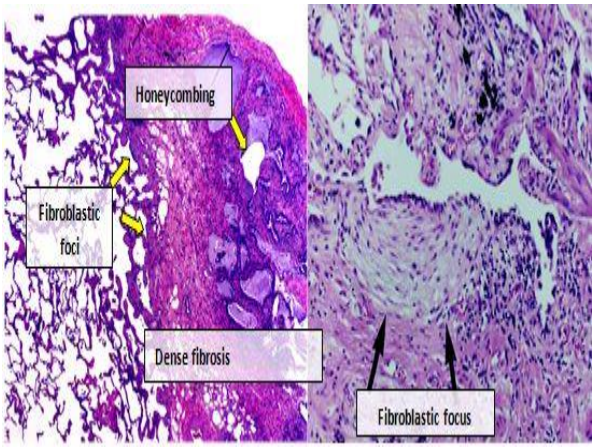
Pathophysiology

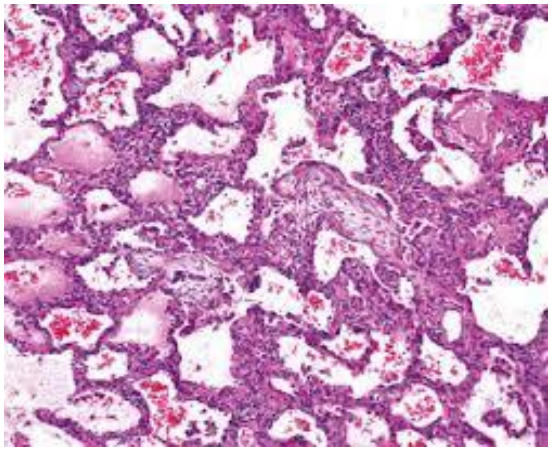


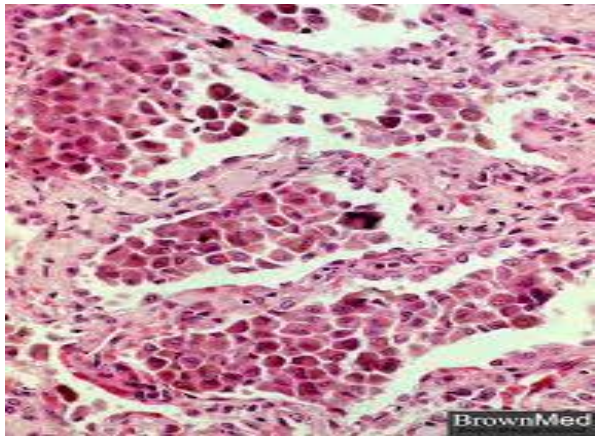
Pathophysiology

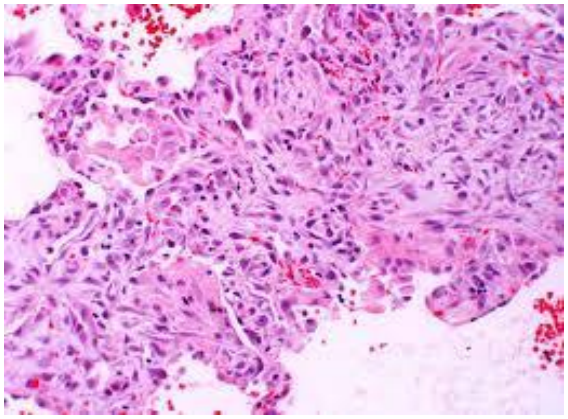


Evolution of ILDS
1970, TISSUES showed patterns and patterns given names

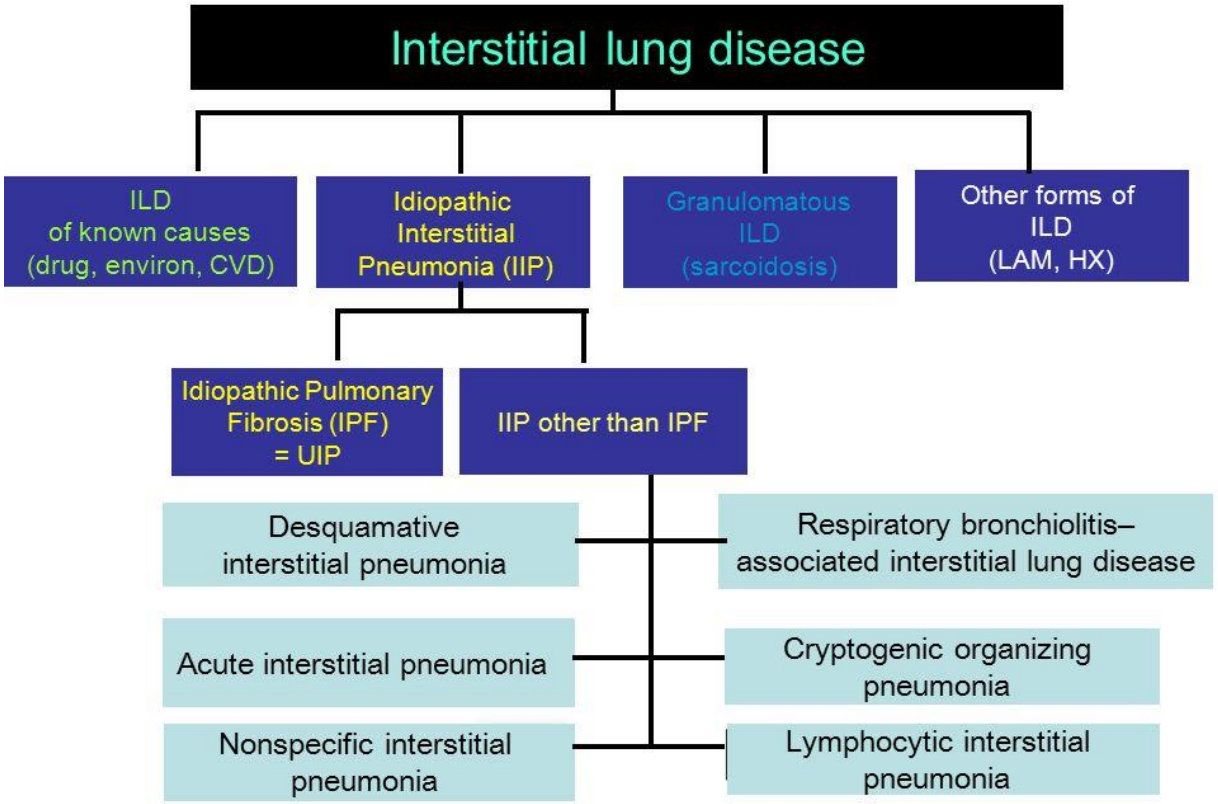






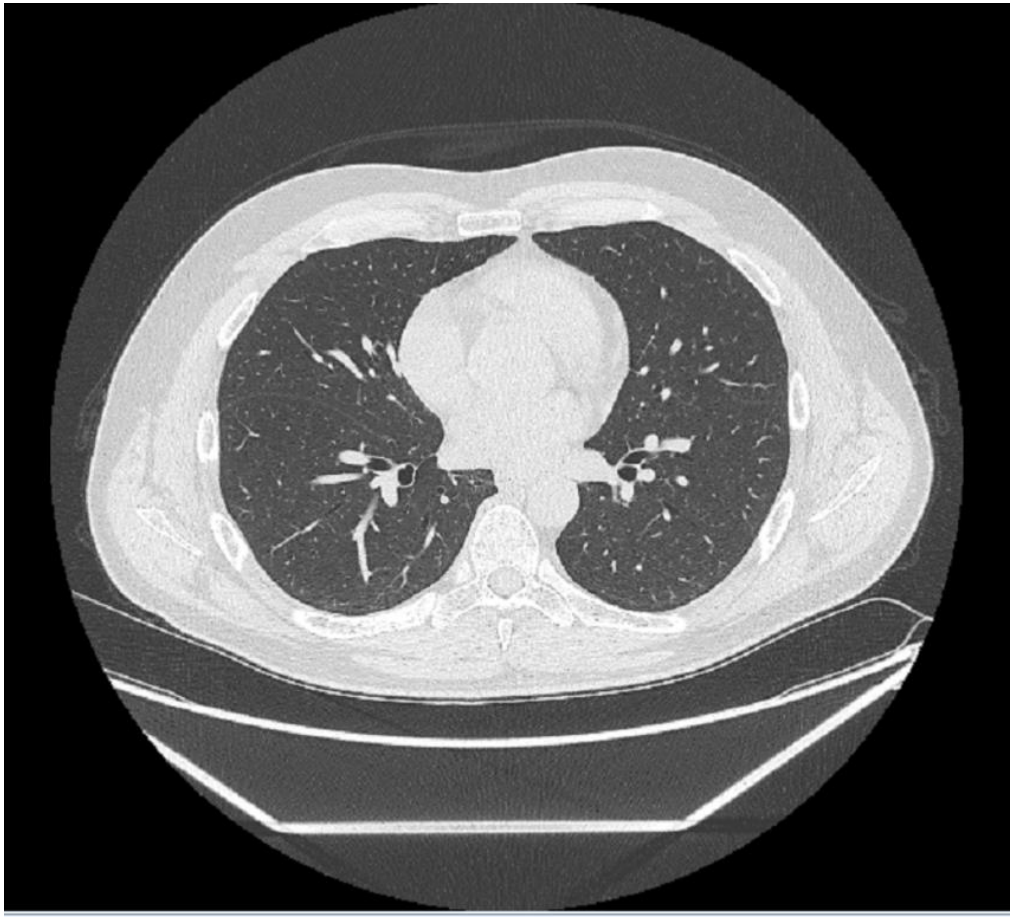


Classification ATS/ERS

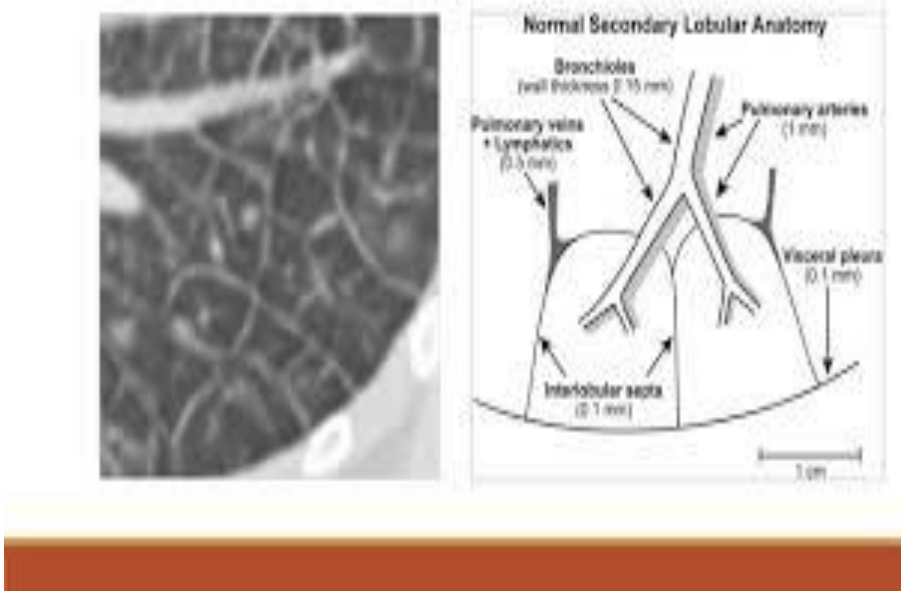


HRCT chest

Gold standard imaging modality for assessment of ILD



Secondary pulmonary lobule

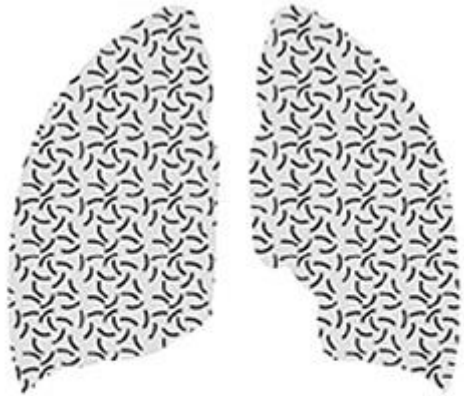


Patterns on Chest X-ray

Linear

Linear

RETICULAR

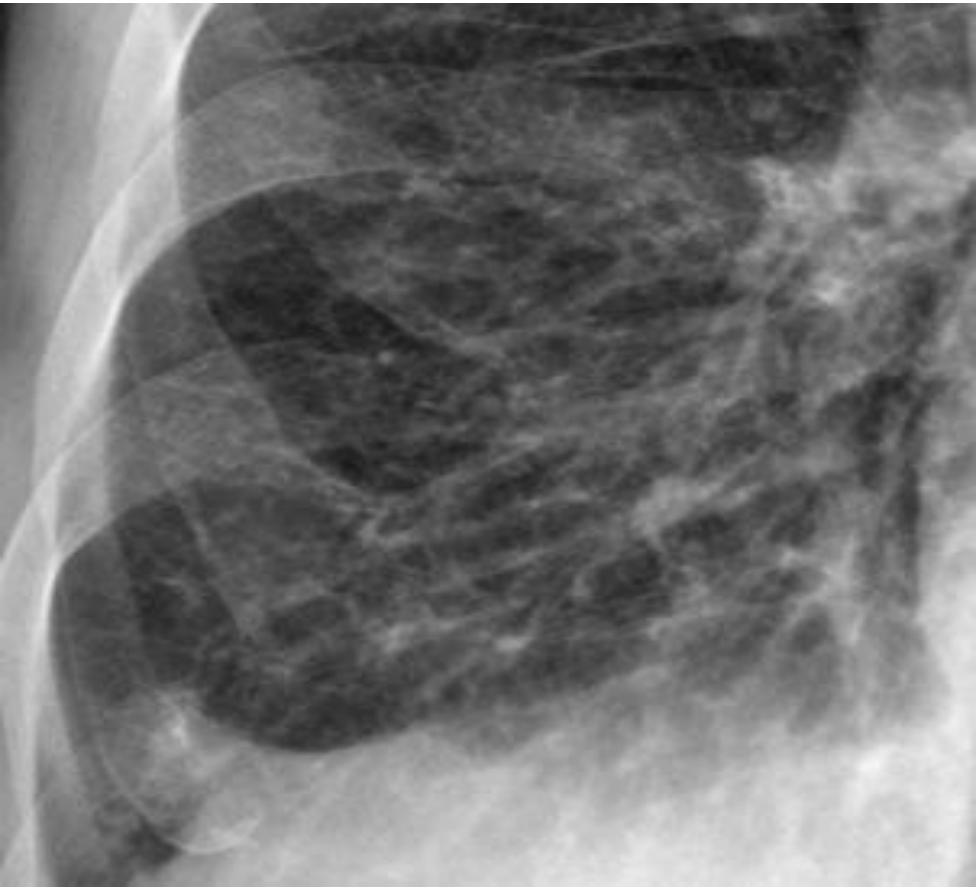


Reticular

NODULAR

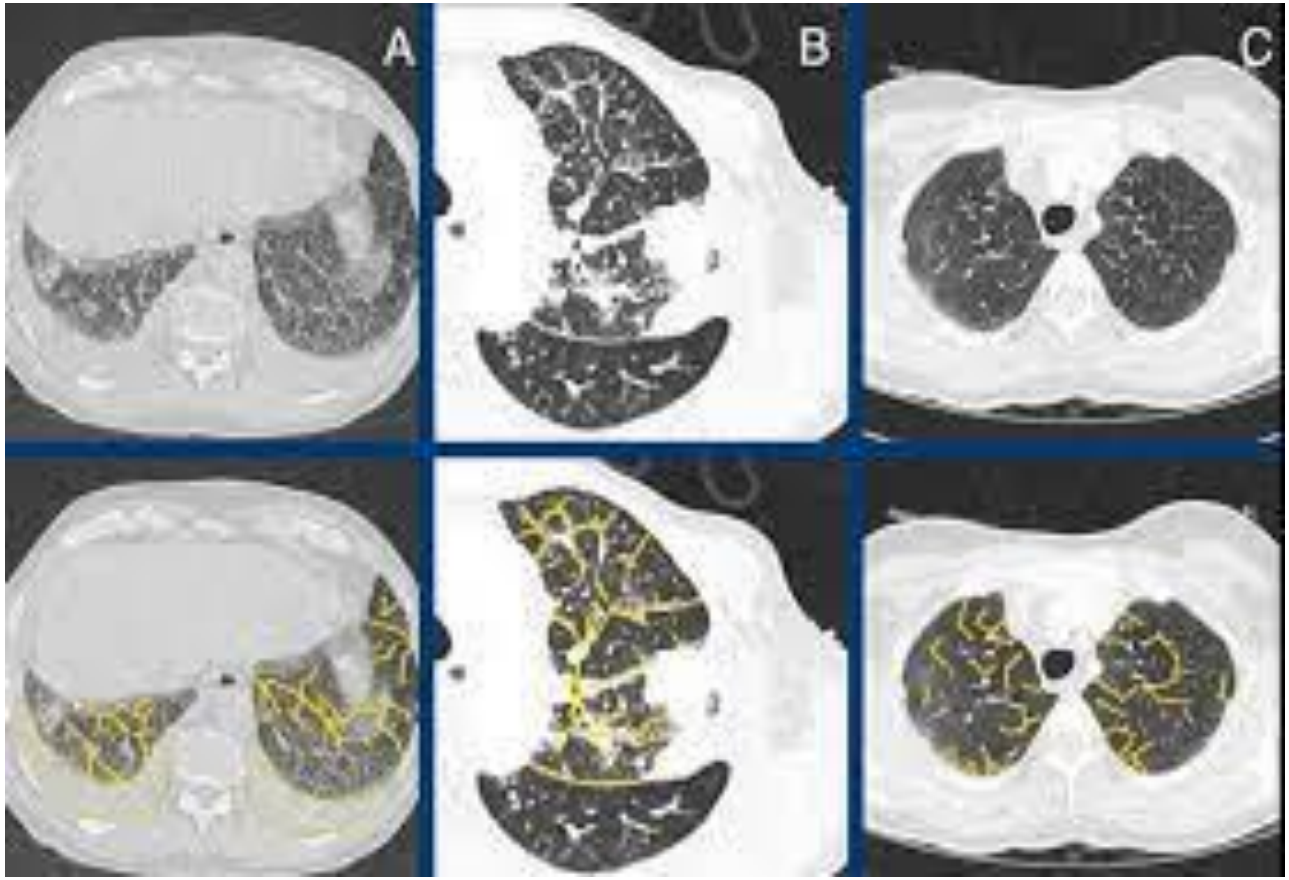
Nodular

RETICULONODULAR

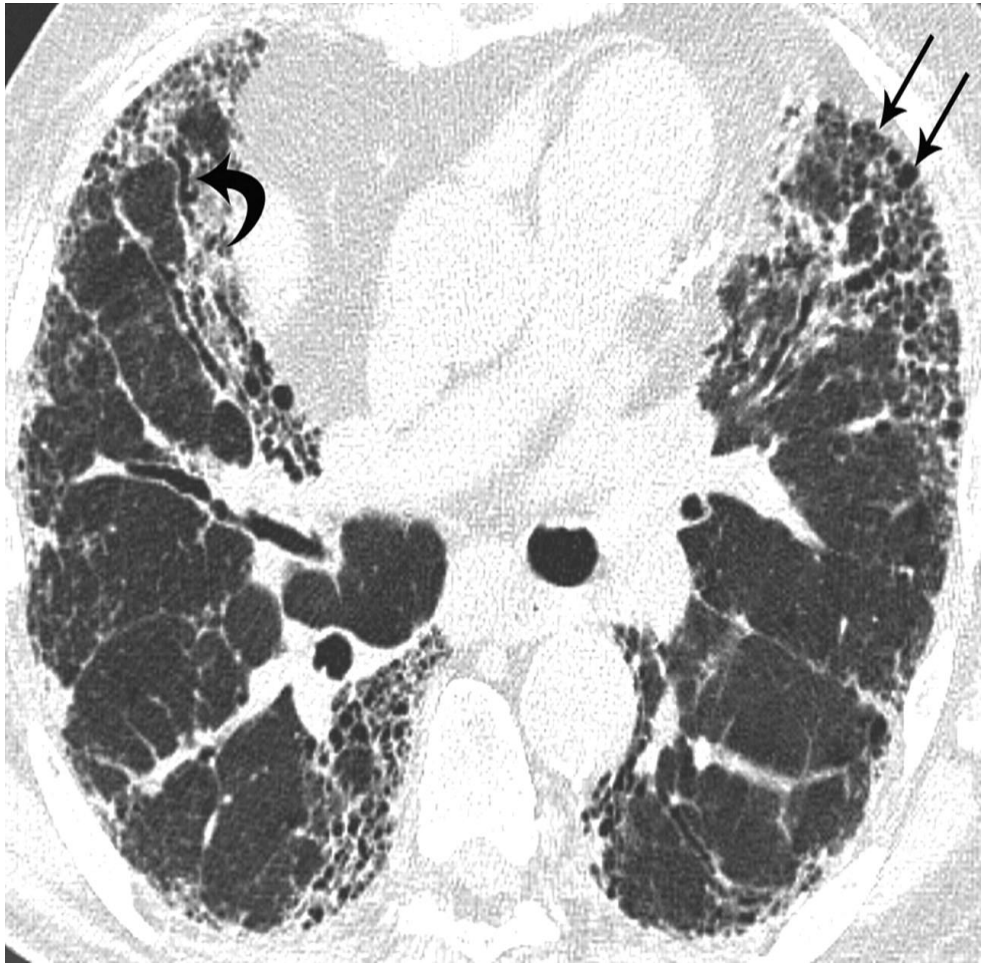


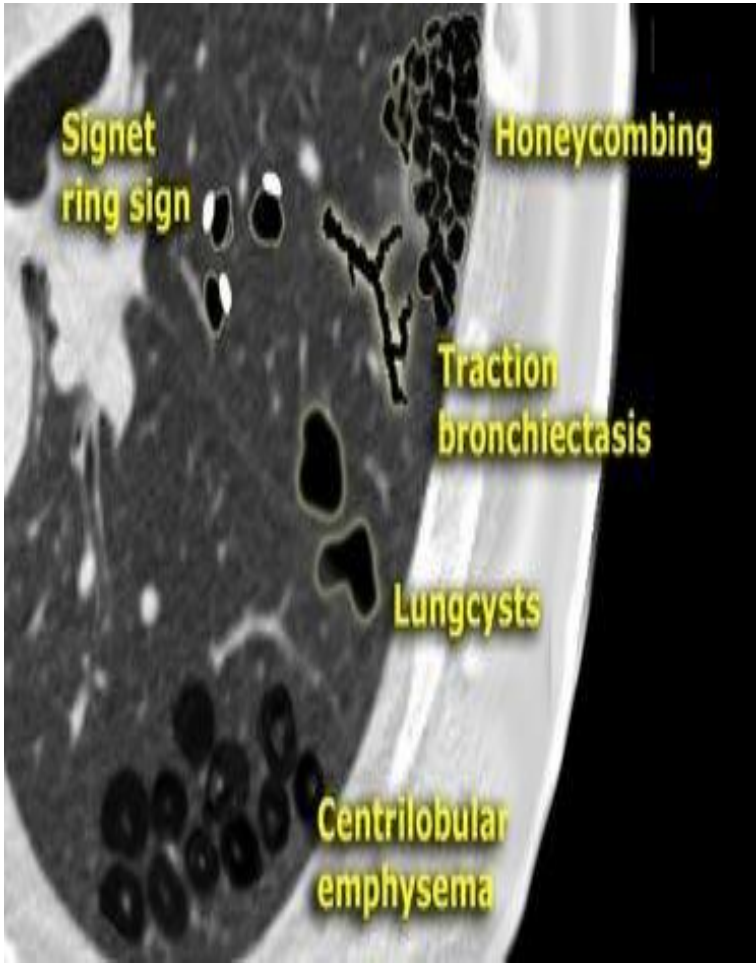
Reticulonodular

Interlobular septal thickening

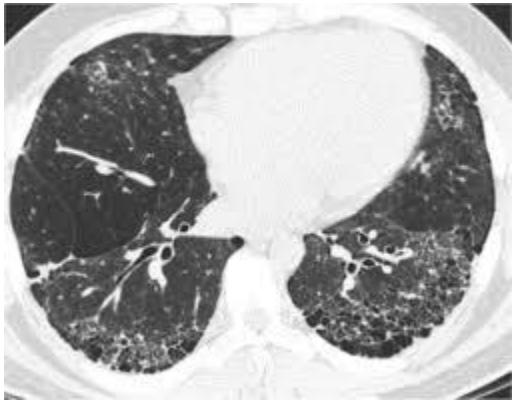


HRCT OF IPF




Signet
ring sign
Honeycombing
Traction
bronchiectasis
Lungcysts
Centrilobular
emphysema

Honeycombing
IPF

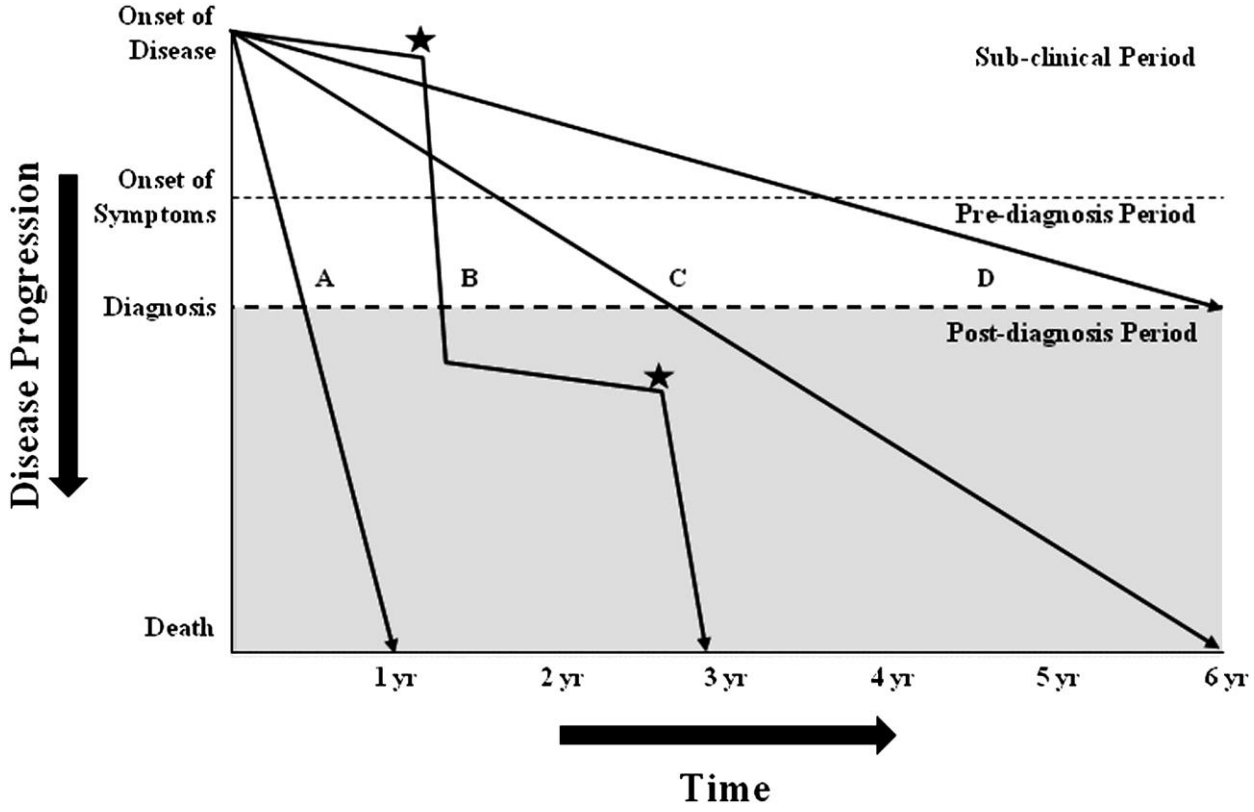


High resolution computed tomography patterns and UIP diagnosis

UIP	Probable UIP	Indeterminate for UIP	Alternative diagnosis
▪ Subpleural and basal predominant; distribution is often heterogeneous* ▪ Honeycombing with or without peripheral traction bronchiectasis or bronchiolectasis¶	▪ Subpleural and basal predominant; distribution is often heterogeneous ▪ Reticular pattern with peripheral traction bronchiectasis or bronchiolectasis ▪ May have mild GGO	▪ Subpleural and basal predominant ▪ Subtle reticulation; may have mild GGO or distortion ("early UIP pattern") ▪ CT features and/or distribution of lung fibrosis that do not suggest any specific etiology ("truly indeterminate for UIP")	▪ Findings suggestive of another diagnosis, including: • CT features: ○ Cysts ○ Marked mosaic attenuation ○ Predominant GGO ○ Profuse micronodules ○ Centrilobular nodules ○ Nodules ○ Consolidation • Predominant distribution: ○ Peribronchovascular ○ Perilymphatic ○ Upper or mid-lung • Other: ○ Pleural plaques (consider asbestosis) ○ Dilated esophagus (consider CTD) ○ Distal clavicular erosions (consider RA) ○ Extensive lymph node enlargement (consider other etiologies) ○ Pleural effusions, pleural thickening (consider CTD/drugs)

UIP: usual interstitial pneumonia; GGO: ground-glass opacities; CT: computed tomography; CTD: connective tissue disease; RA: rheumatoid arthritis.
* Variants of distribution: occasionally diffuse, may be asymmetrical.
¶ Superimposed CT features: mild GGO, reticular pattern, pulmonary ossification.

IPF(Idiopathic pulmonary fibrosis)
Histologically : UIP(usual interstitial pneumonia)
Most common form of ILD .



Published in: Brett Ley; Harold R. Collard; Talmadge E. King Jr.; *Am J Respir Crit Care Med* 2011 183431-440.

ATS/ERS definition of IPF

- A type of chronic fibrosing interstitial pneumonia.
- Unknown etiology limited to the lungs.
- Associated with a histologic pattern of usual interstitial pneumonia (UIP).
- ATS/ERS. *Am J Respir Crit Care Med*. 2002;165:277-304.

How can ILD be investigated?

- High resolution CT chest
  - Some causes have specific appearances
- Pulmonary function tests
- Lung Biopsy
- Blood tests
- MDT

…the choice of tests depends on the clinical presentation

Multidisciplinary meeting

Idiopathic Interstitial Pneumonia

What Is the Effect of a Multidisciplinary Approach to Diagnosis?

Kevin R. Flaherty, Talmadge E. King, Jr., Ganesh Raghu, Joseph P. Lynch III, Thomas V. Colby,

Nonpharmacologic Management

- Smoking cessation .
- Influenza, pneumococcal, and other age-appropriate vaccines should be administered.
- Supplemental Oxygen.
- Pulmonary Rehabilitation .
- Lung Transplantation .
- Lung Cancer identification.

Pharmacologic Management

Pirfenidone .

Nintedanib .

The NEW ENGLAND JOURNAL of MEDICINE

ESTABLISHED IN 1812 MAY 29, 2014 VOL. 370 NO. 22

Efficacy and Safety of Nintedanib in Idiopathic Pulmonary Fibrosis

Luca Richeldi, M.D., Ph.D., Roland M. du Bois, M.D., Ganesh Raghu, M.D., Arata Azuma, M.D., Ph.D., Kevin K. Brown, M.D., Ulrich Costabel, M.D., Vincent Cottin, M.D., Ph.D., Kevin R. Flaherty, M.D., David M. Hansell, M.D., Yoshikazu Inoue, M.D., Ph.D., Dong Soon Kim, M.D., Martin Kolb, M.D., Ph.D., Andrew G. Nicholson, D.M., Paul W. Noble, M.D., Moisés Selman, M.D., Hiroyuki Taniguchi, M.D., Ph.D., Michèle Brun, M.Sc., Florence Le Maulf, M.Sc., Mannaïg Girard, M.Sc., Susanne Stowasser, M.D., Rozsa Schlenker-Herceg, M.D., Bernd Disse, M.D., Ph.D., and Harold R. Collard, M.D., for the INPULSIS Trial Investigators*

ORIGINAL ARTICLE
INTERSTITIAL LUNG DISEASE

Pirfenidone for idiopathic pulmonary fibrosis: analysis of pooled data from three multinational phase 3 trials

Paul W. Noble[1], Carlo Albera[2], Williamson Z. Bradford[3], Ulrich Costabel[4], Roland M. du Bois[5], Elizabeth A. Fagan[3], Robert S. Fishman[3], Ian Glaspole[6], Marilyn K. Glassberg[7], Lisa Lancaster[8], David J. Lederer[9], Jonathan A. Leff[3], Steven D. Nathan[10], Carlos A. Pereira[11], Jeffrey J. Swigris[12], Dominique Valeyre[13] and Talmadge E. King Jr[14]

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Nintedanib for Systemic Sclerosis–Associated Interstitial Lung Disease

Oliver Distler, M.D., Kristin B. Highland, M.D., Martina Gahlemann, M.D., Arata Azuma, M.D., Aryeh Fischer, M.D., Maureen D. Mayes, M.D., Ganesh Raghu, M.D., Wiebke Sauter, Ph.D., Mannaig Girard, M.Sc., Margarida Alves, M.D., Emmanuelle Clerisme-Beaty, M.D., Susanne Stowasser, M.D., Kay Tetzlaff, M.D., Masataka Kuwana, M.D., and Toby M. Maher, M.D., for the SENSCIS Trial Investigators*

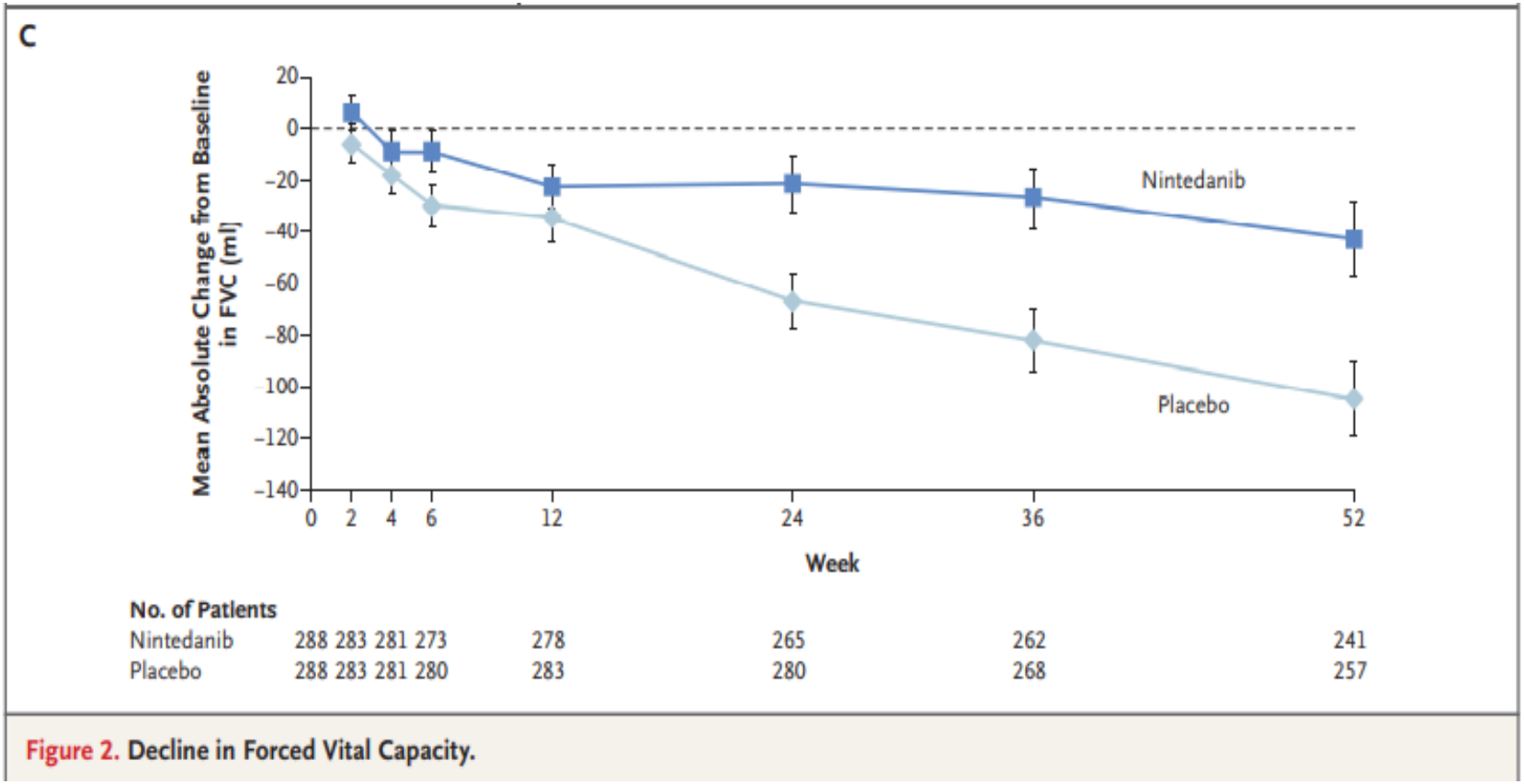


Figure 2. Decline in Forced Vital Capacity.

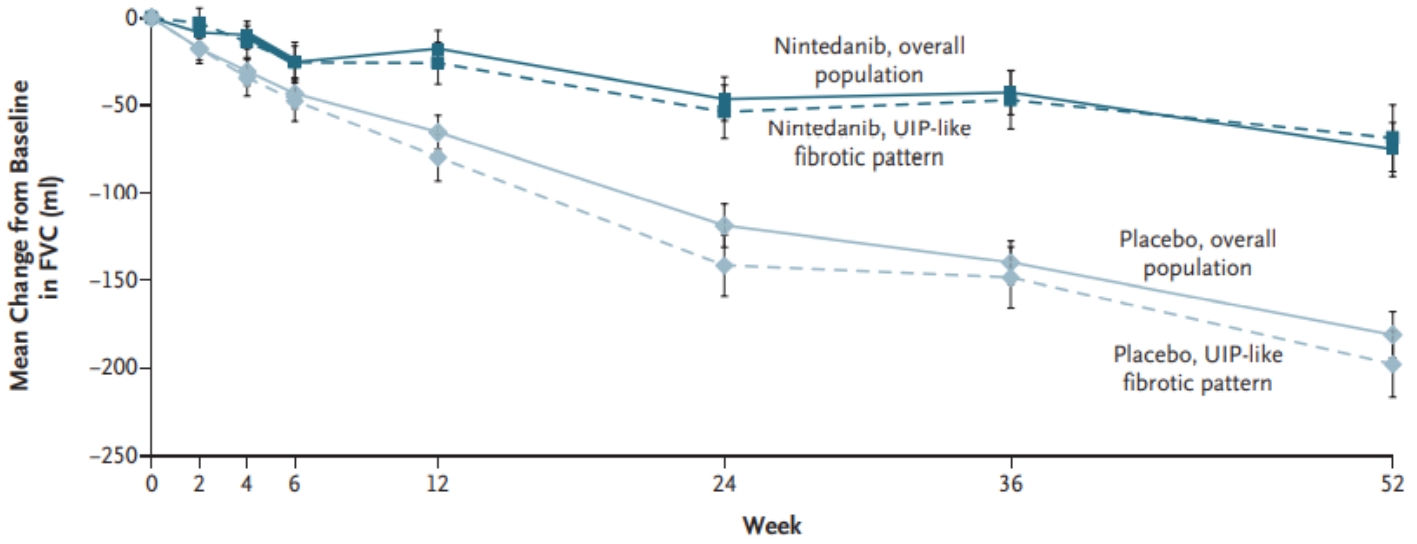
Oliver distler,et al, Safety and Efficacy of Nintedanib in ILD associated with systemic sclerosis. N Engl J Med 2019;380:2518-28

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Nintedanib in Progressive Fibrosing Interstitial Lung Diseases

K.R. Flaherty, A.U. Wells, V. Cottin, A. Devaraj, S.L.F. Walsh, Y. Inoue, L. Richeldi, M. Kolb, K. Tetzlaff, S. Stowasser, C. Coeck, E. Clerisme-Beaty, B. Rosenstock, M. Quaresma, T. Haeufel, R.-G. Goeldner, R. Schlenker-Herceg, and K.K. Brown, for the INBUILD Trial Investigators*



No. of Patients								
Overall population								
Nintedanib	332	326	320	322	314	298	285	265
Placebo	331	325	326	325	320	311	296	274
Patients with UIP-like fibrotic pattern								
Nintedanib	206	203	200	199	193	180	171	160
Placebo	206	202	202	201	197	190	176	162

Figure 2. Decline from Baseline in Forced Vital Capacity (FVC).

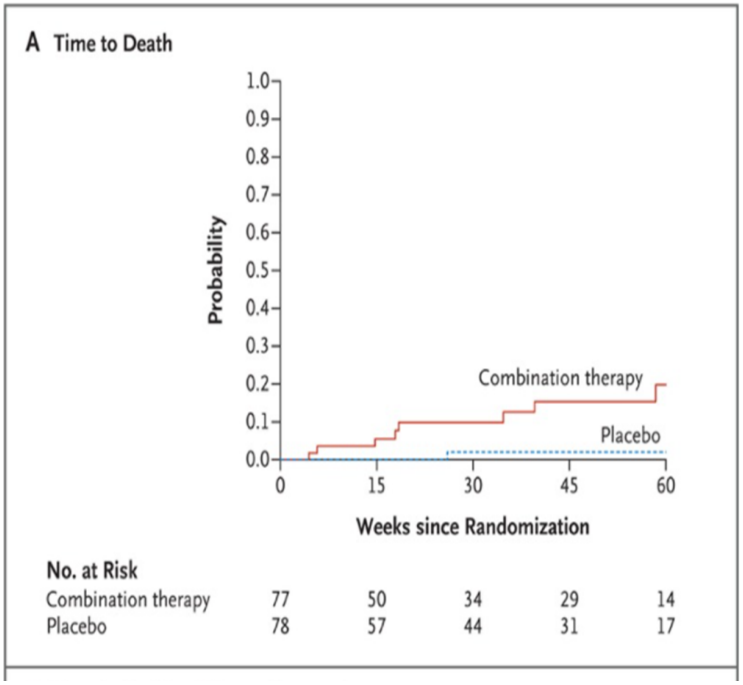
2019;381:1718-27

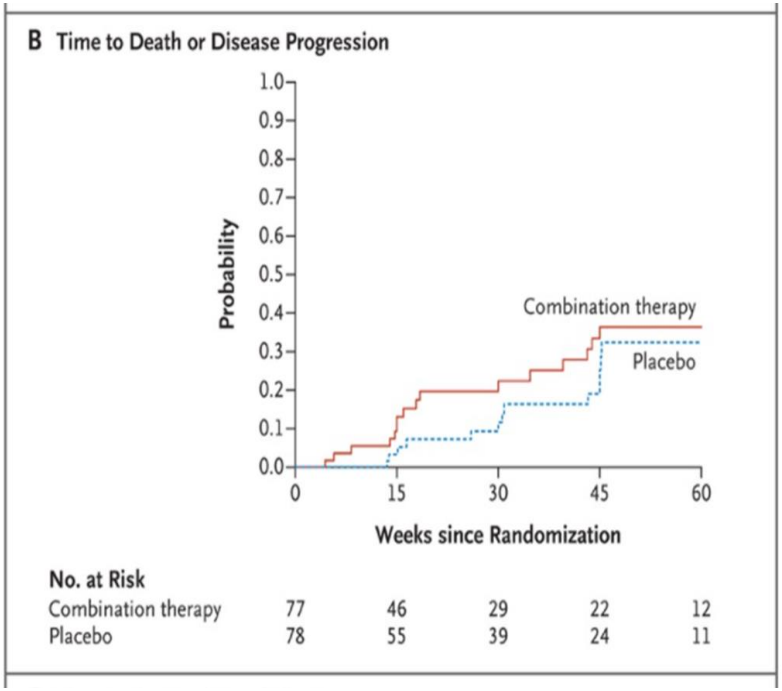
The NEW ENGLAND JOURNAL of MEDICINE

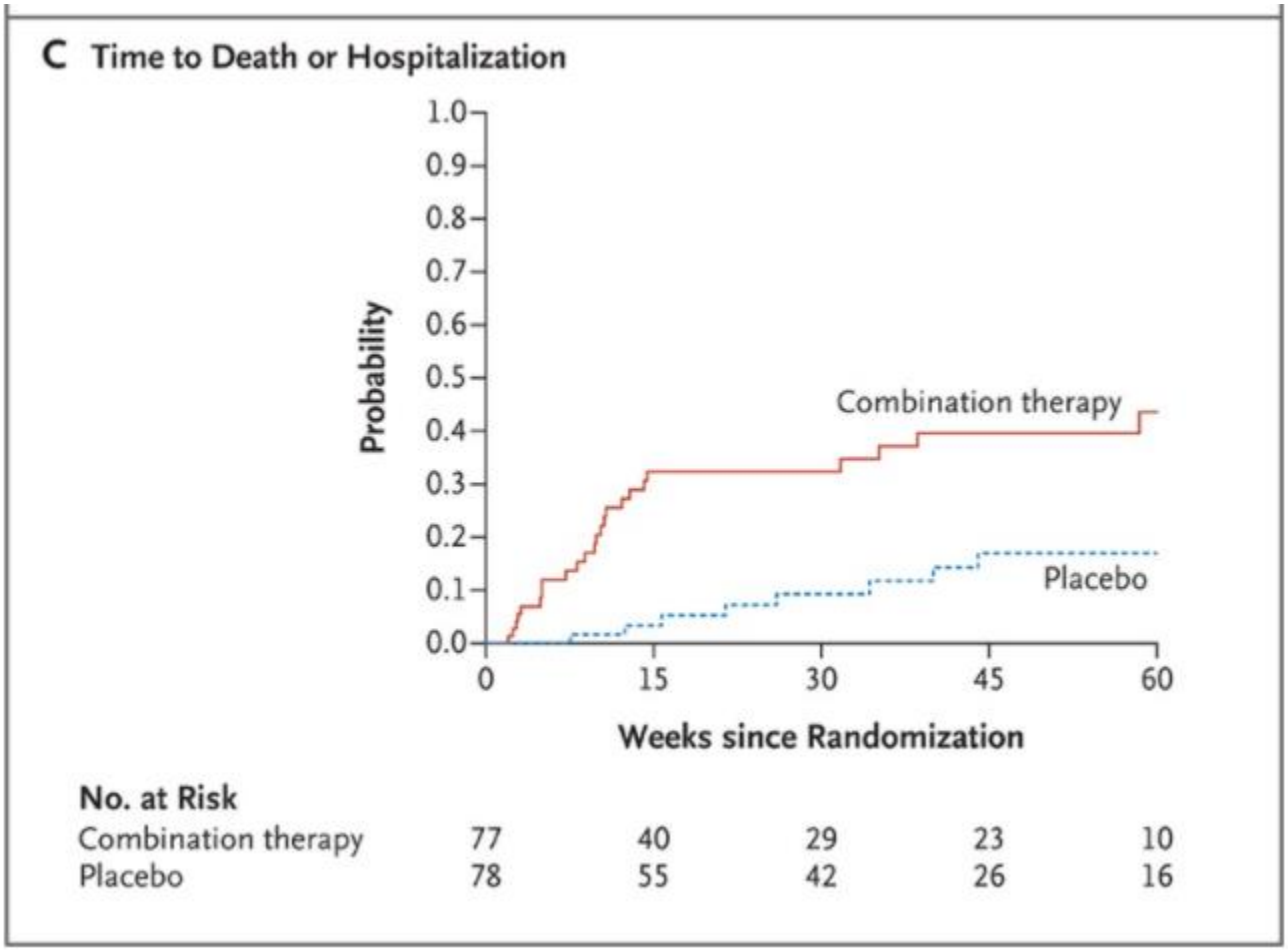
ORIGINAL ARTICLE

Prednisone, Azathioprine, and N-Acetylcysteine for Pulmonary Fibrosis

The Idiopathic Pulmonary Fibrosis Clinical Research Network*





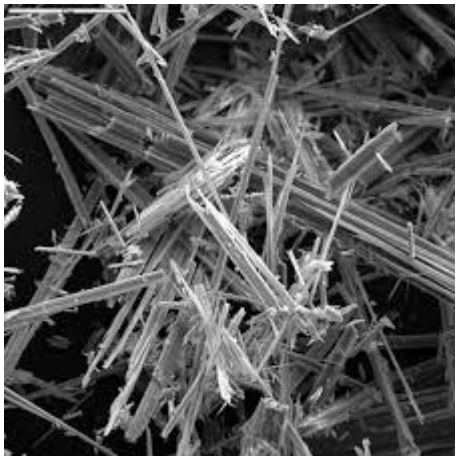


Prednisone, azathioprine, and N-acetylcysteine for pulmonary fibrosis.Raghu G, Anstrom KJ, King TE Jr, Lasky JA, Martinez FJ SON Engl J Med. 2012;366(21):1968.

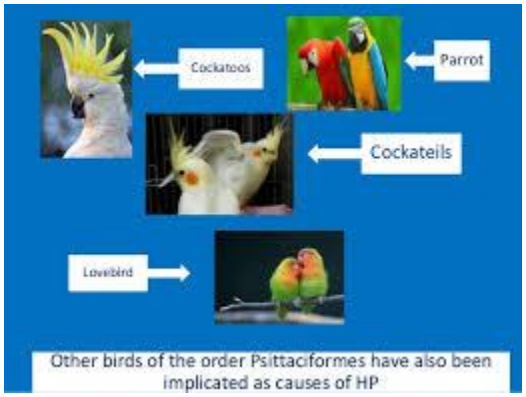
Other ILDS


DANGER
ASBESTOS
CANCER AND LUNG DISEASE HAZARD
KEEP OUT
AUTHORIZED PERSONNEL ONLY
RESPIRATORS AND PROTECTIVE CLOTHING ARE REQUIRED IN THIS AREA.


14.2% Construction
7% Exposed at Home
4.3% Industry Not Reported
61.4% Industrial
3.7% Elementary & Secondary
1.7% Railroads
2.4% Government
1.7% Hospital
1.9% Agricultural
1.9% Electric Light & Power



Hypersensitivity pneumonitis (HP)



Velcro crackles
Fine end inspiratory

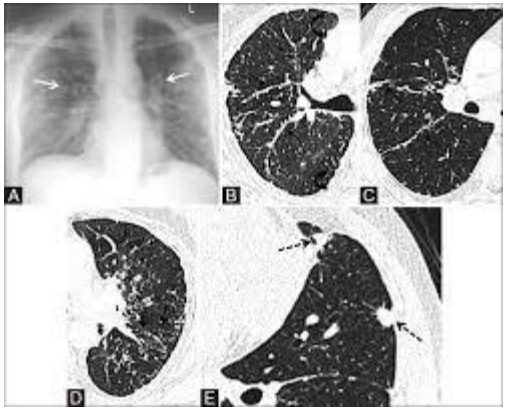
Connective tissue related ILD

1. Systemic lupus erythematosus
2 ,4,5 Scleroderma
3.Rheumatoid arthritis .

Sarcoidosis

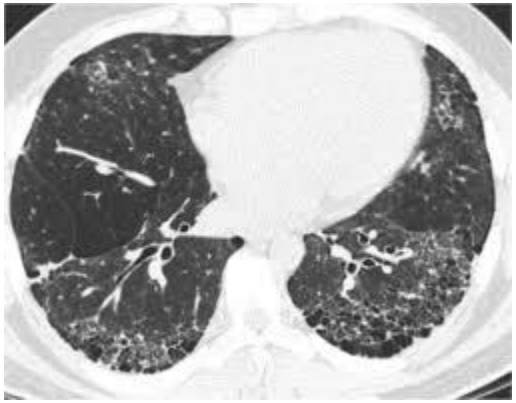
Dermatomyositis/Polymyositis

Sarcoidosis

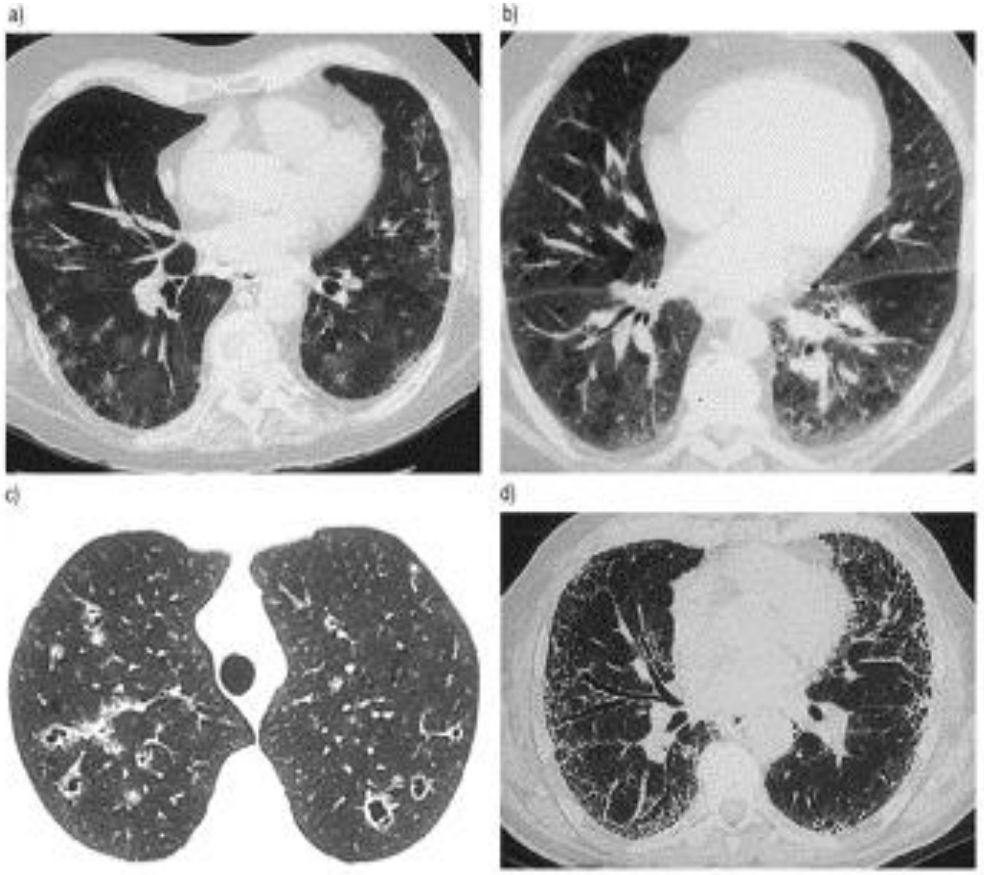


Asbestosis

Honeycombing
IPF



Smoking related interstitial lung disease



Drug induced
Radiation induced

Thank you