INNOVATIONS IN MANAGING ILD IN THE VA SETTING:

New Therapies and Guideline Recommendations for Assessment

MED LEARNING GROUP

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Innovations in Managing ILD in the VA Setting: New Therapies and Guideline Recommendations for Assessment

PROGRAM CHAIR
Sonye K. Danoff, MD, PhD
Professor of Medicine
Division of Pulmonary and Critical Care Medicine
Director, Interstitial Lung Disease Program
Associate Director, Myositis Center
Johns Hopkins University School of Medicine
Baltimore, Maryland

FACULTY PRESENTER
Nirav G. Shah, MD, FCCP, ATSF
Professor of Medicine and Clinical Chief
Division of Pulmonary, Critical Care, & Sleep Medicine
Assistant Dean for Medical Education
University of Maryland School of Medicine
Baltimore, Maryland

PROGRAM OVERVIEW
Fibrosing interstitial lung disease (F-ILD) has a higher prevalence among veterans, likely due to the combined impact of aging, history of smoking, and military-related exposures. This activity is designed to help clinicians who treat veterans identify and manage patients with or at risk for developing F-ILD. The program reviews the latest clinical evidence on current and emerging treatments and provides data and resources on ILD specific to the veteran population.

TARGET AUDIENCE
This activity is designed to meet the educational needs of primary care clinicians, pulmonologists, rheumatologists, and radiologists, as well as related nurse practitioners and physician assistants in the VA setting.

LEARNING OBJECTIVES
Upon the completion of this program, attendees should be able to:

- Analyze physical assessment and medical exposure data to determine connection of F-ILD/ PFF signs and symptoms to military exposures and other risk factors
- Identify resources to advocate for patients with F-ILD/PPFs potentially related to military exposures
- Recognize the implications of current, newly approved, and emerging therapies

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Faculty Member	Disclosures
Sonye Danoff, MD	**Salary:** Pulmonary Fibrosis Foundation (paid to Johns Hopkins) **Royalty**: UpToDate **Consulting fee:** Boehringer Ingelheim Pharmaceuticals, AstraZeneca, Avalyn Pharma, Bristol Myers Squibb, CSL Behring, AbbVie **Non-CME/CE Service Fees Received Directly from a Commercial Interest or its Agent:** Boehringer Ingelheim Pharmaceuticals, United Therapeutics, Bristol Myers Squibb, Mediar Therapeutics **Other:** Avalyn Pharma (DSMB), Bristol Myers Squibb (Adjudication Committee)
Nirav G. Shah, MD	Has nothing to disclose.

All relevant financial relationships have been mitigated.

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5 min.	Introduction
25 min.	Didactic Content Shared by a Filmed Faculty Using Slides and Infographic Data Initial Assessment ○ Identifying at-risk patients with persistent, unexplained, non-specific respiratory symptoms ○ History and Physical assessment Connecting the Diagnosis to Military Exposures ○ Military exposures ○ Burn pits, Agent Orange ○ The Pact Act ○ Presumptive conditions ○ Vietnam Veterans Standard, New, and Emerging Therapies ○ Anti-inflammatory/immunosuppressives ○ Antifibrotic agents ○ Emerging therapies
20 min.	Case Discussion
10 min.	Q&A

Innovations in Managing ILD in the VA Setting: New Therapies and Guideline Recommendations

Nirav G Shah, MD, FCCP, ATSF
Professor of Medicine and Clinical Chief
Division of Pulmonary, Critical Care, and Sleep Medicine
Assistant Dean for Medical Education
University of Maryland School of Medicine
Baltimore, Maryland

Disclosures

- **Dr Shah** has nothing to disclose
- During this lecture the use of medications for both US Food and Drug Administration (FDA)-approved and non-FDA-approved indications may be discussed
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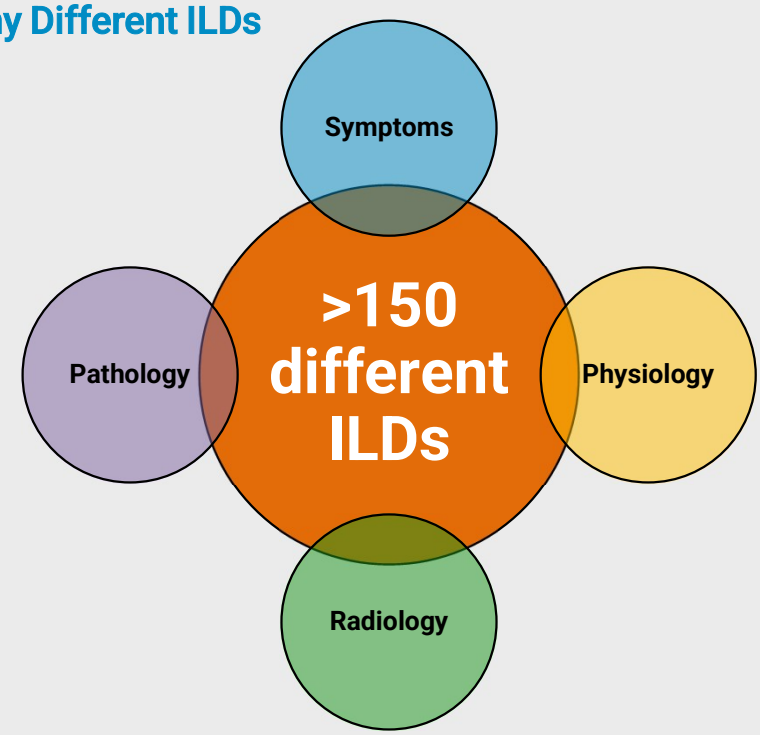
Learning Objectives

- Analyze physical assessment and medical exposure data to determine the connection of fibrosing interstitial lung disease/progressive pulmonary fibrosis (F-ILD/PPF) signs and symptoms to military exposures and other risk factors
- Identify resources to advocate for patients with F-ILD/PPFs potentially related to military exposures
- Recognize the implications of current, newly approved, and emerging therapies

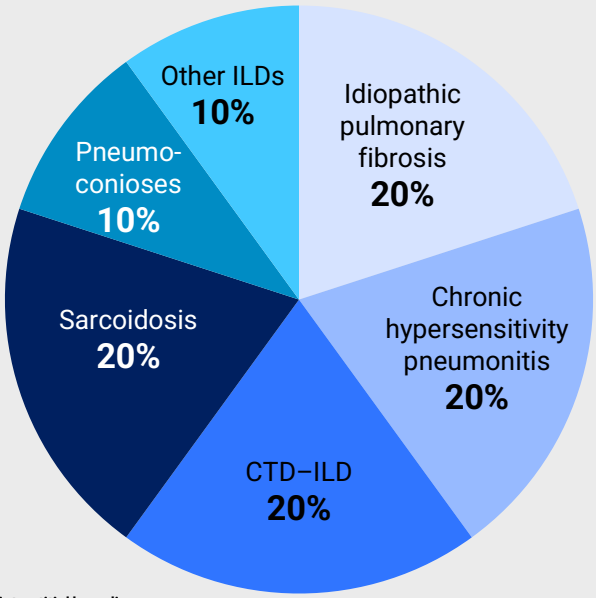
F-ILD/PPF

There Are Many Different ILDs



ILDs = interstitial lung diseases.

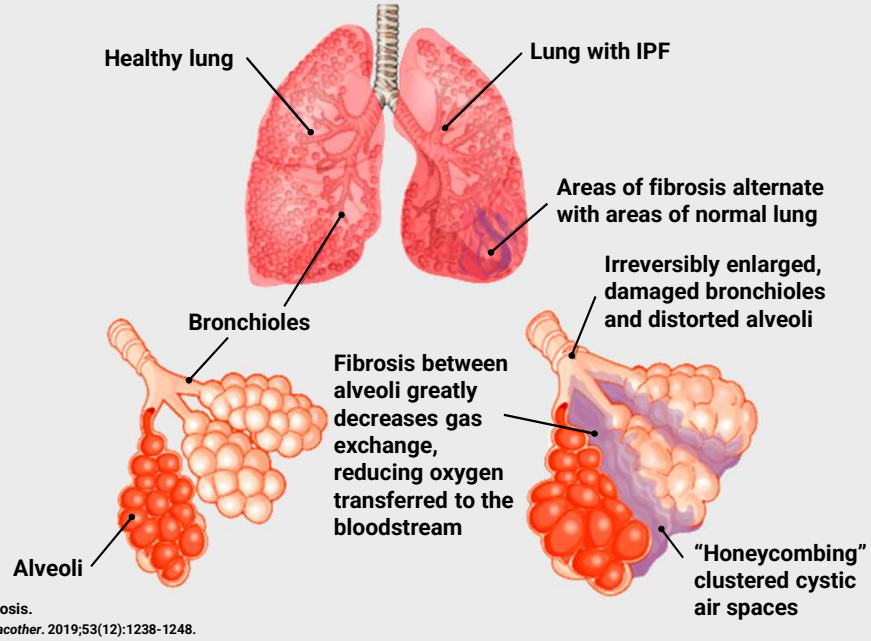
Variability of ILDs



CTD-ILD = connective tissue disease-associated interstitial lung disease.

Lederer DJ, Martinez FJ. *N Engl J Med.* 2018;378:1811-1823.

Fibrosis



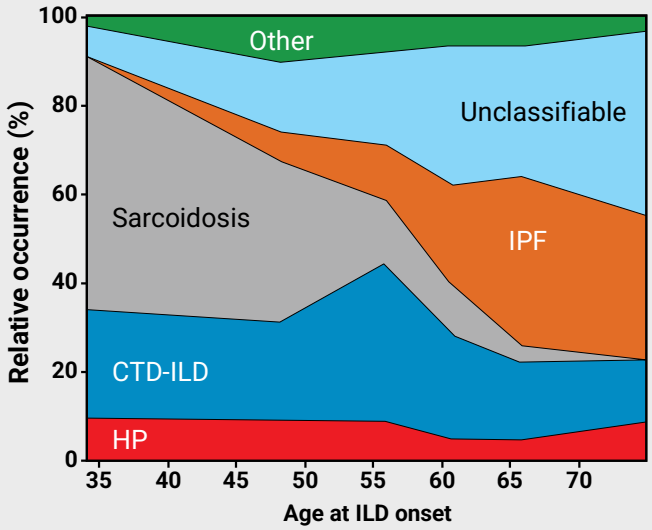
IPF = idiopathic pulmonary fibrosis.

Pleasants R, Tighe RM. *Ann Pharmacother*. 2019;53(12):1238-1248.

ILD Epidemiology

Distribution of ILD by age of onset

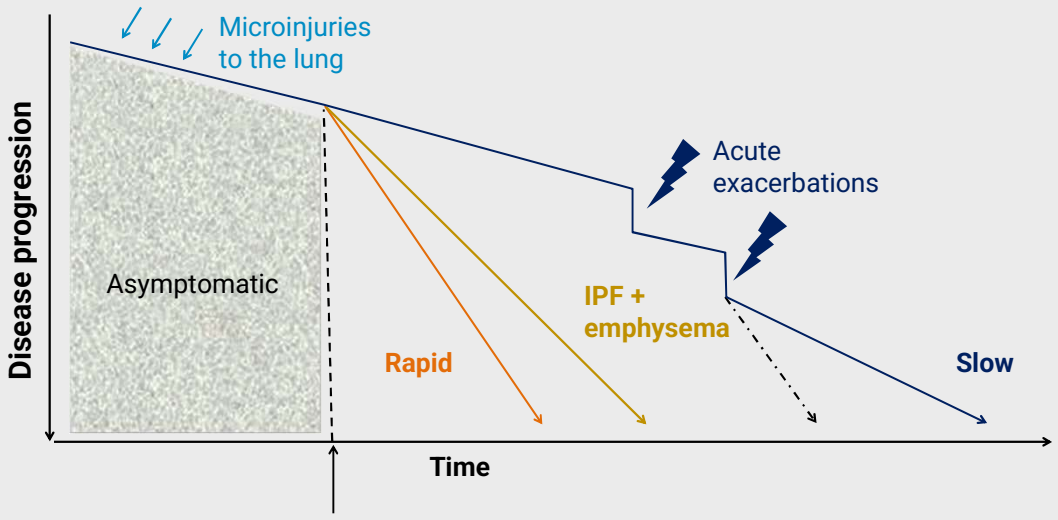
- N = 327 with ILD
- 98% had ILD for 21 years
- Among elderly subjects (>70 years), 43% had "unclassifiable" ILD
- Lack of surgical biopsy in most led to inadequate data to make a definitive diagnosis



HP = hypersensitivity pneumonitis.

Patterson KD. *Chest*. 2017;151(4):838-844.

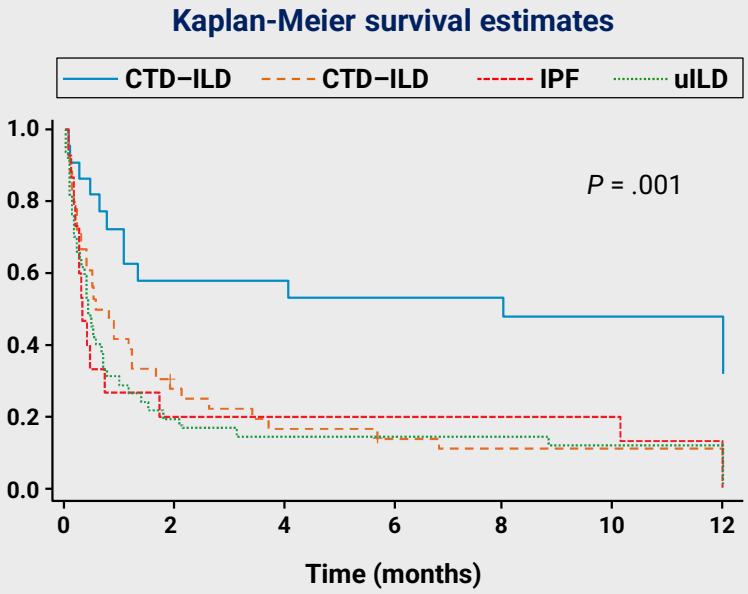
Natural History of IPF



King TE, et al. *Lancet.* 2011;378(9807):1949-1461.

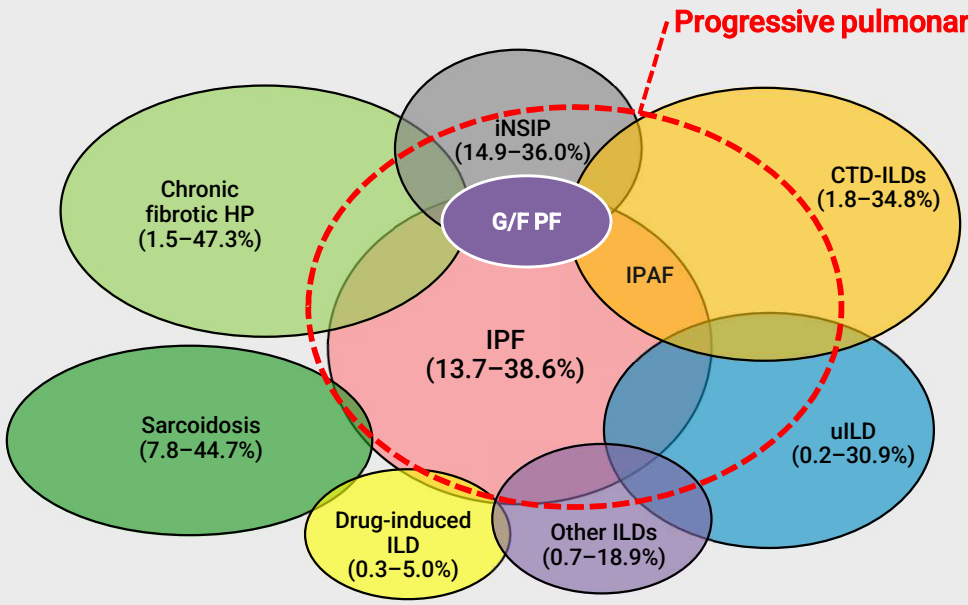
Mortality

- In-hospital mortality rates of 73% to 100% with invasive mechanical ventilation
- CTD-ILD < IPF



uILD = unclassifiable interstitial lung disease.
Gannon WD, et al. *Chest.* 2018;153(6):1387-1395.

Progressive Pulmonary Fibrosis



G/F PF = genetic and/or familial pulmonary fibrosis; iNSIP = idiopathic nonspecific interstitial pneumonia; IPAF = interstitial pneumonia with autoimmune features; uILD = unclassifiable ILD.

Rajan SK, et al. *Eur Resp J*. 2023;61:2103187.

Definition of Progressive Pulmonary Fibrosis (PPF)

In a patient with ILD of known or unknown etiology other than IPF who has radiologic evidence of pulmonary fibrosis, PPF is defined as at least 2 of the following 3 criteria occurring within the past year with no alternative explanation*

1. Worsening respiratory symptoms
2. Physiologic evidence of disease progression (either of the following)
   a) Absolute decline in FVC ≥5% predicted within 1 year of follow-up
   b) Absolute decline in DLCO (corrected for Hb) ≥10% predicted within 1 year of follow-up
3. Radiological evidence of disease progression (1 or more of the following)
   a) Increased extent or severity of traction bronchiectasis and bronchiolectasis
   b) New ground-glass opacity with traction bronchiectasis
   c) New fine reticulation
   d) Increased extent or increased coarseness of reticular abnormality
   e) New or increased honeycombing
   f) Increased lobar volume loss

*Although it is critical to exclude alternative explanations of worsening features for all patients with suspected progression, this is particularly important in patients with worsening respiratory symptoms and/or decline in DL_{CO} given the lower specificity of these features for PPF compared with FVC and chest computed tomography.

DLCO = diffusing capacity for of the lungs for carbon monoxide; FVC = forced vital capacity; Hb = hemoglobin; IPF = idiopathic pulmonary fibrosis.

Raghu G, et al. *Am J Respir Crit Care Med*. 2022;205(9):e18-e472022.

Criteria for Progressive Pulmonary Fibrosis (PPF)

The 3 diagnostic criteria for PPF			
Domain	**0.5-year**	**1-year**	**2-year**
Symptoms	Worsening respiratory symptoms	Worsening respiratory symptoms	Worsening respiratory symptoms
Pulmonary function	Absolute decline in FVC% of over 5%	Absolute decline in predicted FVC% of over 5% or an absolute decline in DLCO% of 10%	Absolute decline in predicted FVC% of over 10%, or an absolute decline in predicted FVC% of 5%–10%
Radiology	—	Increased fibrosis on HRCT	Increased fibrosis on HRCT

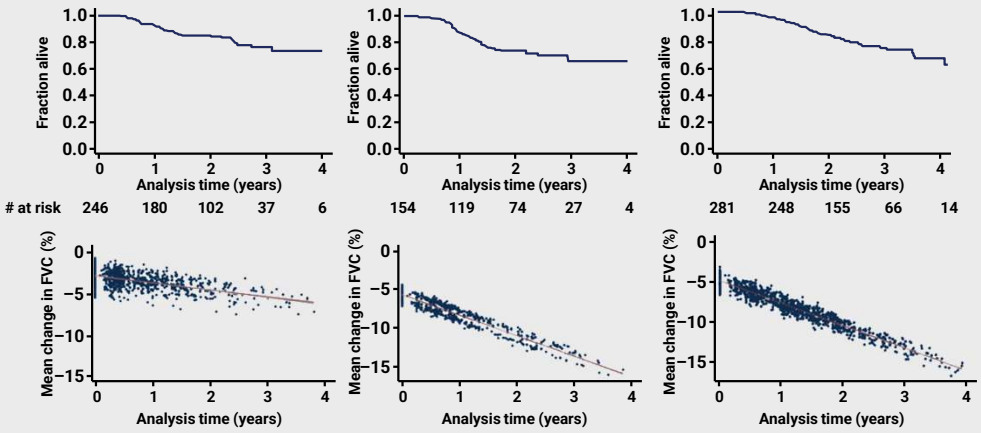
PPF was diagnosed using 3 criteria: (1) The 0.5-year criterion: In the preceding 6 months, an absolute decline in FVC% of over 5% or worsening respiratory symptoms; (2) The 1-year criterion: PPF was defined with at least 2 of the 3 criteria: Worsening respiratory symptoms; in the preceding 12 months, an absolute decline in predicted FVC% of over 5% or an absolute decline in predicted percent DLCO of 10%; increased fibrosis on HRCT scan; (3) The 2-year criterion: In the preceding 24 months, an absolute decline in predicted FVC of over 10%, or an absolute decline in predicted FVC of 5% to 10% with worsening respiratory symptoms or increased fibrosis on HRCT scan, or worsening respiratory symptoms and increased fibrosis on HRCT.

HRCT = high-resolution computed tomography.

Chen T, Zeng C. *J Thorac Dis.* 2024;16:1034-1043.

PPF Is Associated With Poor Survival

- Single center study in China
- Total of 2476 patients diagnosed with non-IPF fibrotic ILD; 246 patients with PPF by 0.5-year standard, 154 patients by 1-year standard, and 281 patients by 2-year standard
- In these 3 groups 58%, 50%, and 49%, respectively, had CTD-ILD



Chen T, Zeng C. *J Thorac Dis.* 2024;16:1034-1043.

History

Occupational/exposure history
- Asbestos, silica, paint/spray-painting, beryllium, environmental services, farm worker, woodworker
- Oily nose drops, laxatives, mold

Hobbies
- Hot tub, musical instruments, gardening, spelunking

Pets
- **Birds**, rabbits, gerbils, guinea pigs, hamsters

Travel

Smoking

Drugs
- Amiodarone, nitrofurantoin, bleomycin, methotrexate, cyclophosphamide, chemotherapy

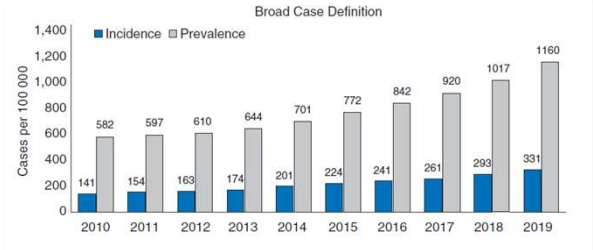
Provided by Dr Nirav Shah.

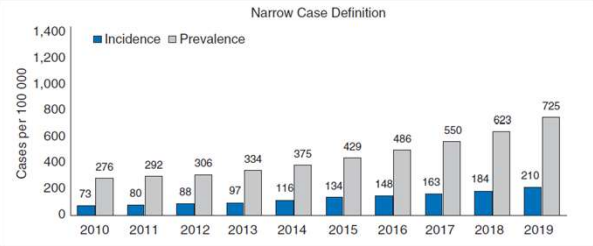
ILDs and Military Exposures

ILD in the US Veteran Population

Epidemiology

- Incidence and prevalence of IPF is increasing among the veteran population
  - 2010–2019 incidence: 141 to 331 cases/100,000 person-years (broad definition[a])
  - 2010–2019 annual prevalence: 582 to 1160 cases/ 100,000 (narrow definition[b])
- 139,116 veterans receiving care from the VHA between 2010 and 2019 were diagnosed with IPF
- Risk factors: Older age, White race, tobacco use, rural residence
  - Difference between males and females in IPF prevalence was small





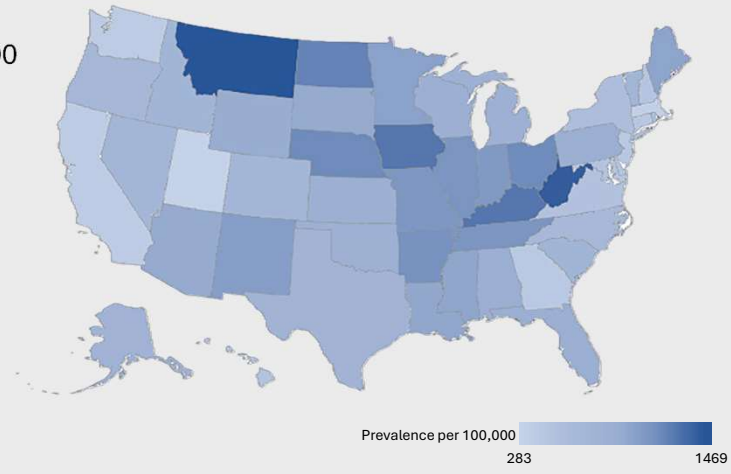
[a]Broad definition = 1 IPF ICD code without evidence of a procedural code for CT scan or lung biopsy.
[b]Narrow definition = 1 IPF ICD code with evidence of a procedural code for CT scan or lung biopsy.
VHA = Veterans Health Administration.
Kaul B, et al. *Ann Am Thorac Soc*. 2022;19:196-203. Tighe RM, et al. *Ann Am Thorac Soc.* 2022;19:161-170.

Geographic Distribution of IPF in US Veteran Population

Epidemiology

- Iowa, Kentucky, Montana, and West Virginia had the highest standardized prevalence rates in 2019
- Highest: Montana—1469 cases/100,000
- Lowest: Utah—430 cases/100,000

Standardized IPF prevalence* among veterans by state in 2019 (narrow definition)



*State prevalence was calculated by identifying the number of unique veterans with IPF in each state divided by the total number of veterans living in the state.
Kaul B, et al. *Ann Am Thorac Soc*. 2022;19:196-203. Tighe RM, et al. *Ann Am Thorac Soc.* 2022;19:161-170.

Airborne Hazards and Burn Pit Exposures

Southwest Asia

- Over the past decades, evidence has emerged that US military personnel deployed to Iraq, Afghanistan, and other sites in Southwest Asia are at increased risk for developing respiratory symptoms and diseases
- These exposures include
  - Open-air burn pits
  - Desert dust, sandstorms
  - Industrial fires and emissions
  - Workplace vapors, dusts, gases, and fumes
  - Jet fuels
  - IED blasts
  - Combat dust and debris
  - Temperature and humidity extremes
  - Vehicular diesel exhaust

IED = improvised explosive device.

National Jewish Health. Military & veterans lung disease (https://www.nationaljewish.org/conditions/veterans-lung-disease). US Dept of Veterans Affairs. Public health: airborne hazards and burn pit exposures (https://www.publichealth.va.gov/exposures/burnpits). URLs accessed 4/20/2026.

PACT Act of 2022

The Sergeant First Class (SFC) Heath Robinson Honoring our Promise to Address Comprehensive Toxics (PACT) Act

- Largest healthcare and benefit expansion in VA history
- Expands and extends eligibility for VA healthcare for veterans with toxic exposures and veterans of the Vietnam, Gulf War, and post-9/11 eras
- Adds 20+ more presumptive conditions for burn pits, Agent Orange, and other toxic exposures
- Adds more presumptive-exposure locations for Agent Orange and radiation
- Requires VA to provide a toxic exposure screening to every veteran enrolled in VA healthcare
- Helps improve research, staff education, and treatment related to toxic exposures

VA = Veteran Affairs.

US Department of Veterans Affairs. The PACT Act and your VA benefits (https://www.va.gov/resources/the-pact-act-and-your-va-benefits/). Accessed April 20,2026.

Presumptive Illnesses and Eligible Locations and Operations

PACT Act

Presumptive illnesses

- Asthma (diagnosed after service)
- Chronic bronchitis, rhinitis, or sinusitis
- Chronic obstructive pulmonary disease (COPD)
- Constrictive or obliterative bronchiolitis
- Emphysema
- Granulomatous disease
- **ILD**
- Pleuritis
- **Pulmonary fibrosis**
- Sarcoidosis

- **On/after September 11, 2001:** Afghanistan, Djibouti, Egypt, Jordan, Lebanon, Syria, Uzbekistan, and Yemen, including airspace above these locations
- **On/after August 2, 1990:** Bahrain, Iraq, Kuwait, Oman, Qatar, Saudi Arabia, Somalia, and United Arab Emirates (UAE), including airspace above these locations
- **Operations:** Operation Enduring Freedom, Operation Freedom's Sentinel, Operation Iraqi Freedom, Operation New Dawn, Operation Inherent Resolve, and Resolute Support Mission

US Department of Veterans Affairs. The PACT Act and your VA benefits (https://www.va.gov/resources/the-pact-act-and-your-va-benefits/). Accessed 4/20/2026.

Agent Orange and Vietnam Veterans

- A 2022 cohort study showed that IPF occurred in 2.2% (20,409) of Vietnam veterans with Agent Orange exposure vs 1.9% (51,086) of Vietnam veterans without exposure (unadjusted odds ratio [OR], 1.14; 95% confidence interval [CI], 1.12–1.16; $P < .001$)[1]
  - Relationship held true after adjusting for known IPF risk factors (age, race, ethnicity, smoking, and rural residence) and accounting for the interaction between Agent Orange x age ($P = .008$) and Agent Orange x smoking ($P = .026$)[1]
- Odds of IPF among Vietnam veterans with presumptive Agent Orange exposure was 8% higher than those with no exposure (adjusted OR, 1.08; 95% CI, 1.06–1.10; $P < .001$)[1]
- ILDs are **NOT** presumptive conditions for Vietnam veterans exposed to Agent Orange[2,3]

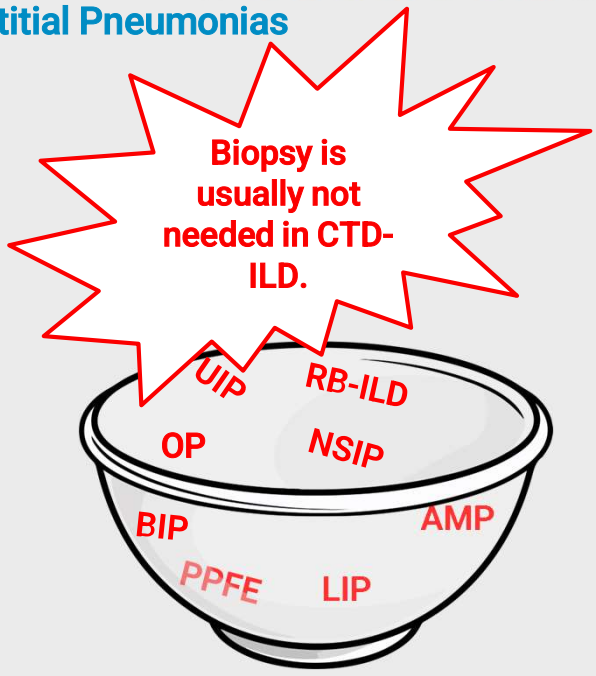
1. Kaul B, et al. *Am J Respir Crit Care Med*. 2022;206:750-757. 2. US Department of Veterans Affairs. Public health: exposure to Agent Orange in Vietnam (https://www.publichealth.va.gov/PUBLICHEALTH/exposures/agentorange/ locations/vietnam.asp). 3. US Department of Veterans Affairs. Public health: veterans' diseases associated with Agent Orange (https://www.publichealth.va.gov/PUBLICHEALTH/exposures/agentorange/conditions/index.asp). 4. US Department of Veterans Affairs. Agent Orange exposure and disability compensation (https://www.va.gov/disability/eligibility/hazardous-materials-exposure/agent-orange/). URLs accessed 4/20/2026.

Clinical and Imaging Features

Revised ATS/ERS Classification of Interstitial Pneumonias

THE ALPHABET SOUP

Interstitial patterns
Usual interstitial pneumonia (UIP)
Nonspecific interstitial pneumonia (NSIP)
Bronchiolocentric interstitial pneumonia (BIP)
Diffuse alveolar damage (DAD)
Pleuroparenchymal fibroelastosis (PPFE)
Lymphoid interstitial pneumonia (LIP)
Alveolar filling patterns
Organizing pneumonia (OP)
Respiratory bronchiolitis-ILD (RB-ILD)
Alveolar macrophage pneumonia (AMP)
Rare alveolar filling disorders
Other
Combined pattern
Unclassifiable pattern



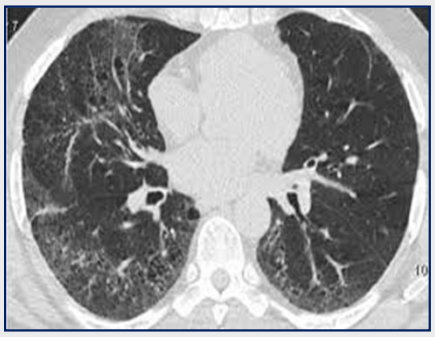
ATS = American Thoracic Society; CTD-ILD = connective tissue disease-associated interstitial lung disease; ERS = European Respiratory Society.

Ryerson CJ, et al. *Eur Respir J*. 2025;2500158.

Interstitial Pneumonias

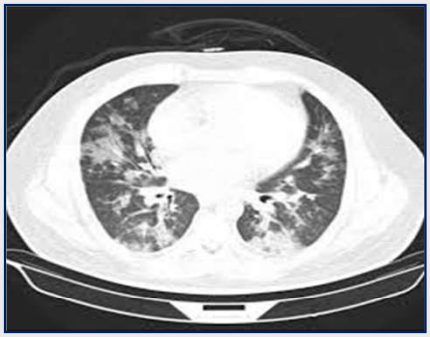
Potentially reversible

NSIP



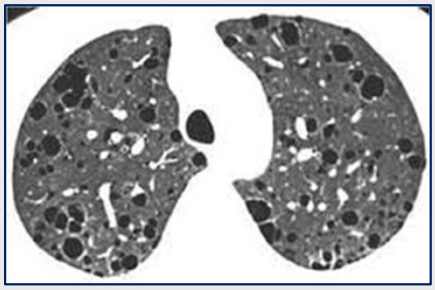
NSIP/OP overlap

OP

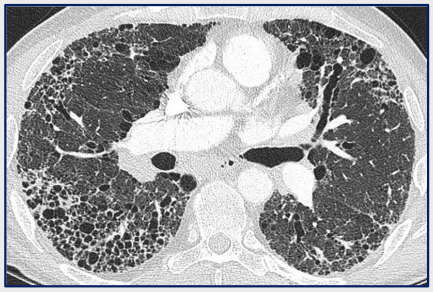


Not reversible

LIP



UIP



LIP = lymphoid interstitial pneumonia; NSIP = nonspecific interstitial pneumonia; OP = organizing pneumonia; UIP = usual interstitial pneumonia.

Clinical Presentation of IPF

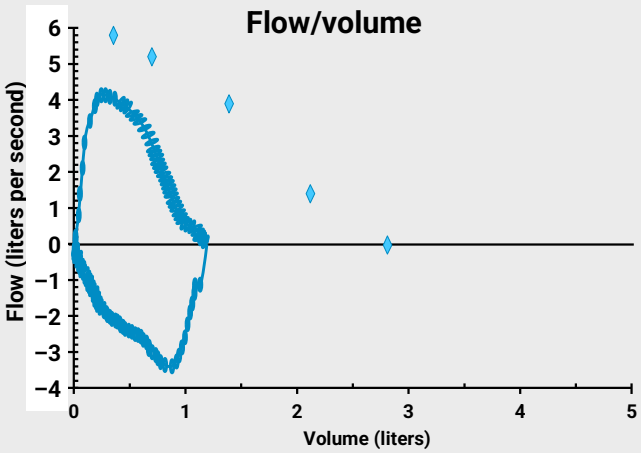
- Chronic exertional dyspnea, dry cough, bibasilar inspiratory crackles, and finger clubbing
- Sixth and seventh decades
- Men > women
- History of cigarette smoking
- Prevalence estimate of between 14.0 and 42.7 per 100,000

Raghu G, et al. *Am J Respir Crit Care Med*. 2018;198(5):e44-e68. Nalysnyk L, et al. *Eur Respir Rev*. 2012;21(126):355-361.

Pulmonary Function Testing for Suspected ILD

- ILD is characterized by restrictive lung physiology
  - FVC <80% of control is abnormal; <50% is severely abnormal
- DLCO is often impaired
- Patients with concurrent emphysema may exhibit normal lung volumes and spirometry but reduced DLCO
- Low baseline FVC, decline in FVC, low DLCO, and decline in 6MWT are associated with decreased survival

Abnormal flow volume indicating restrictive lung physiology

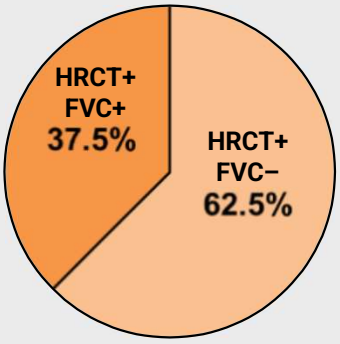


DLCO = diffusing capacity of the lung for carbon monoxide; FVC = forced vital capacity; 6MWT = 6-minute walk test.

Wallace B, et al. *Curr Opin Rheumatol*. 2016;28:236-245.

Pulmonary Function Tests Alone May Miss ILD

- N = 102 patients with systemic sclerosis-associated (SSc) ILD
- 64/102 (63.0%) with significant ILD on HRCT
- 27/102 (26.0%) with FVC <80%



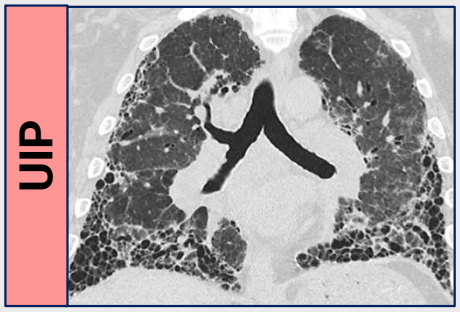
40/64 (62.5%) patients with significant ILD on HRCT had normal FVC

↔ high false-negative rate → high risk of missed diagnoses in clinical practice

Suliman YA, et al. *Arthritis Rheumatol*. 2015;67:3256-3261.

Crackles Can Be Heard Early in IPF

- Bilateral, Velcro™-like crackles can be heard during mid-to-late inspiration, first occurring in the basal areas and then progressing to the upper lungs



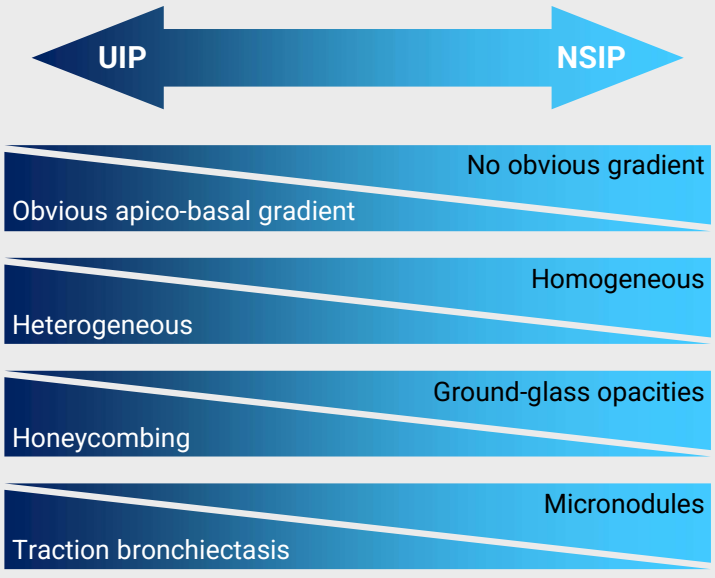
When performing chest auscultation, focus on the bases of both lungs and the midaxillary lines.

Image courtesy of and used with permission from Robert Suh, MD.

UIP = usual interstitial pneumonia.

Sellarés J, et al. *Medicine (Baltimore).* 2016;95(5):e2573. Sgalla G, et al. *BMC Pulm Med.* 2018;18(1):103.

Key Imaging Features for Differentiation Between UIP and NSIP



NSIP = nonspecific interstitial pneumonia; UIP = usual interstitial pneumonia.

Mueller-Mang C, et al. *RadioGraphics*. 2007;27(3):595-615.

Clinical Guideline Updates for HRCT

High-resolution computed tomography scanning patterns

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)

*Variants of distribution: Occasionally diffuse, may be asymmetrical. †Superimposed CT features: Mild GGO, reticular pattern, pulmonary ossification.

CT = computed tomography; CTD = connective tissue disease; GGO = ground-glass opacities; HRCT = high-resolution computed tomography; RA = rheumatoid arthritis; UIP = usual interstitial pneumonia.

Raghu G, et al. *Am J Respir Crit Care Med*. 2018;198(5):e44-e68.

Clinical Guideline Updates for Histology

Histopathology patterns and features

UIP	Probable UIP	Indeterminate for UIP	Alternative diagnosis
▪ Dense fibrosis with architectural distortion (ie, destructive scarring and/or honeycombing) ▪ Predominant subpleural and/or paraseptal distribution of fibrosis ▪ Patchy involvement of lung parenchyma by fibrosis ▪ Fibroblast foci ▪ Absence of features to suggest an alternate diagnosis	▪ Some histologic features from column 1 are present but to an extent that precludes a definite diagnosis of UIP/IPF *and* ▪ Absence of features to suggest an alternative diagnosis *or* ▪ Honeycombing only	▪ Fibrosis with or without architectural distortion, with features favoring either a pattern other than UIP or features favoring UIP secondary to another cause* ▪ Some histologic features from column 1, but with other features suggesting an alternative diagnosis†	▪ Features of other histologic patterns of IIPs (eg, absence of fibroblast foci or loose fibrosis) in all biopsies ▪ Histologic findings indicative of other diseases (eg, hypersensitivity pneumonitis, Langerhans cell histiocytosis, sarcoidosis, LAM)

*Granulomas, hyaline membranes (other than when associated with acute exacerbation of IPF, which may be the presenting manifestation in some patients), prominent airway-centered changes, areas of interstitial inflammation lacking associated fibrosis, marked chronic fibrous pleunitis, organizing pneumonia. Such features may not be overt or easily seen to the untrained eye and often need to be specifically sought. †Features that should raise concerns about the likelihood of an alternative diagnosis include a cellular inflammatory infiltrate away from areas of honeycombing, prominent lymphoid hyperplasia including secondary germinal centers, and a distinctly bronchiolocentric distribution that could include extensive peribronchiolar metaplasia.

IIP = idiopathic interstitial pneumonia; IPF = idiopathic pulmonary fibrosis; LAM = lymphangioleiomyomatosis; UIP = usual interstitial pneumonia.

Raghu G, et al. *Am J Respir Crit Care Med*. 2018;198(5):e44-e68.

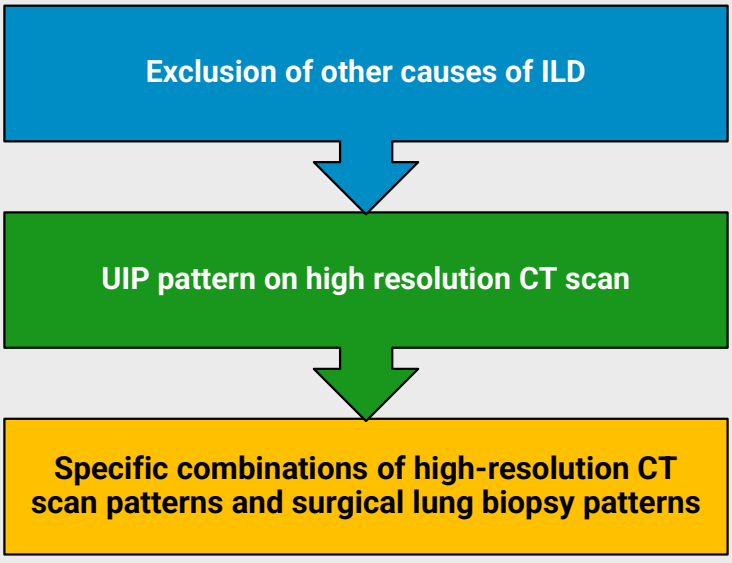
Predicting IPF Mortality

Low risk (0–4 points)
Moderate risk (5–8 points)
High risk (9–13 points)

Predictor	Points
Male sex	2
IPF diagnosis	1
Body mass index (BMI)	
<25	0
26–30	1
31–35	2
>35	3
Mechanical ventilation or ECMO	2
No ambulation	1
SAPS II	
<20	0
21–30	3
>30	4
Total points	13

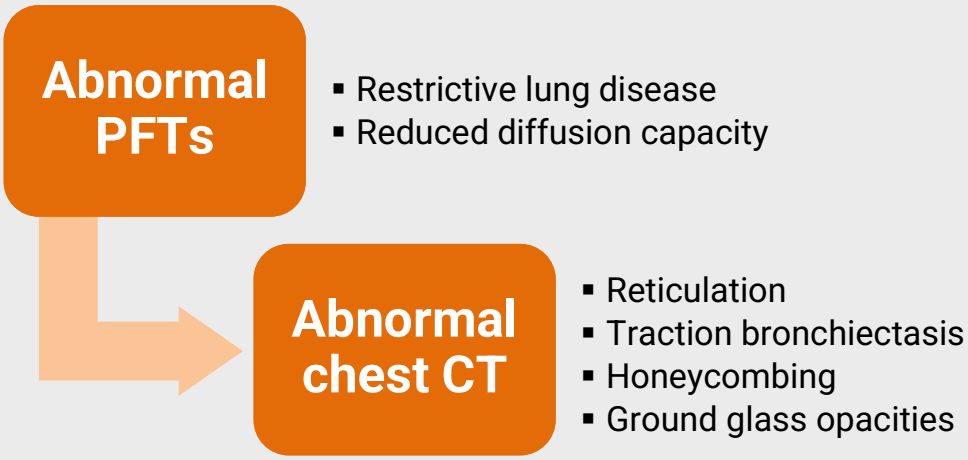
ECMO = extracorporeal membrane oxygenation; SAPS II = Simplified Acute Physiology Score II.
Gannon WD, et al. *Chest*. 2018;153(6):1387-1395.

Criteria to Diagnose IPF



Raghu G, et al. *Am J Respir Crit Care Med*. 2018;198(5):e44-e68.

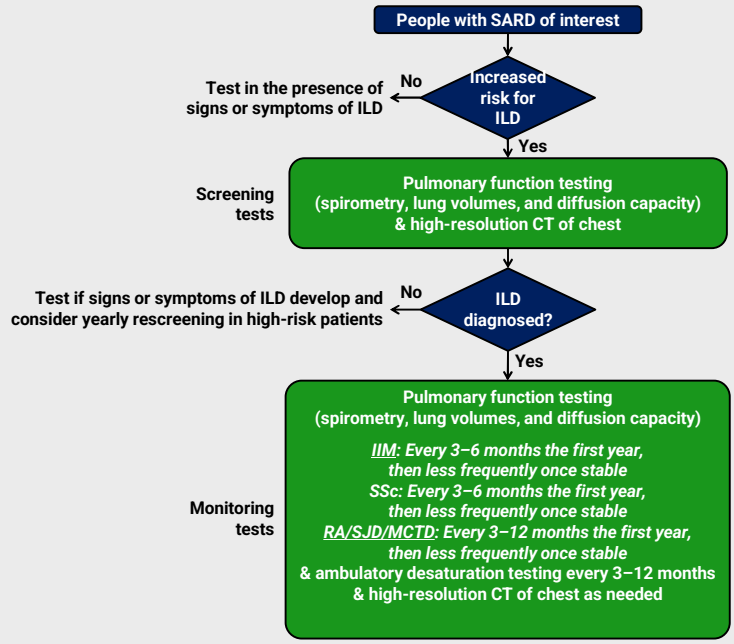
Making a Diagnosis of ILD



Raghu G, et al. *Am J Respir Crit Care Med*. 2018;198(5):e44-e68.

Guideline Recommendations

Screening for ILD: American College of Rheumatology (ACR) Guidelines



■ Conditional recommendation

CT = computed tomography; IIM = idiopathic inflammatory myopathies; ILD = interstitial lung disease; MCTD = mixed connective tissue disease; RA = rheumatoid arthritis; SARD = systemic autoimmune rheumatic disease; SjD = Sjögren's disease; SSc = systemic sclerosis.

Johnson SR, et al. *Arthritis Rheumatol.* 2024;76(8):1201-1213.

ACR/ACCP First-Line Therapy Recommendations for SARD-ILD

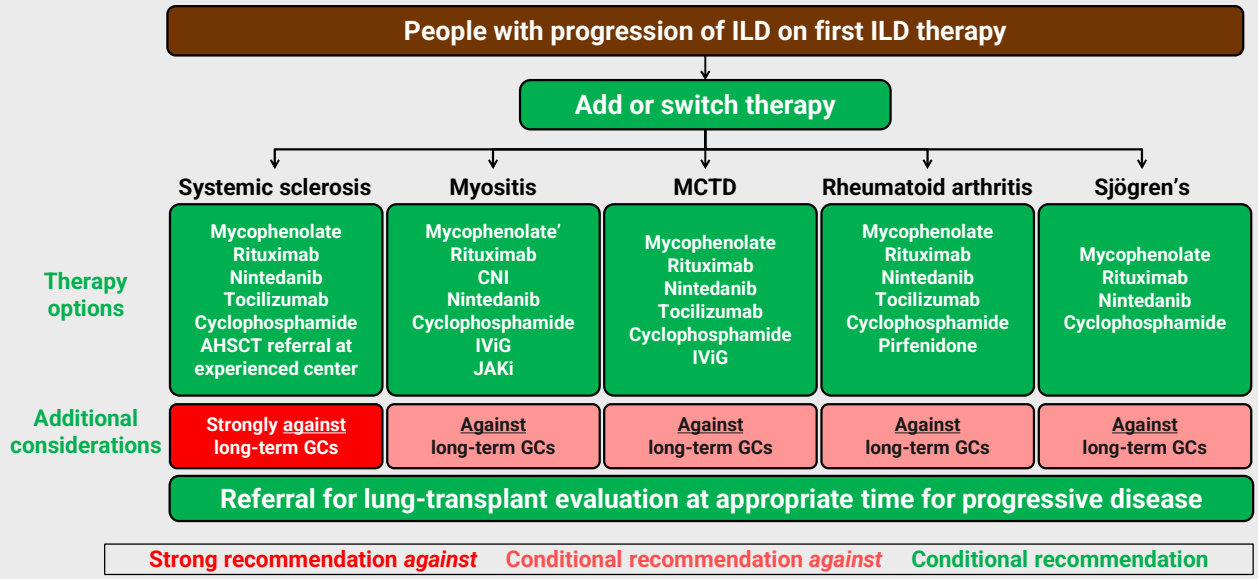
	Systemic sclerosis	Myositis	MCTD	Rheumatoid arthritis	Sjögren's
Preferred First-line ILD therapy	Mycophenolate Tocilizumab Rituximab	Mycophenolate Azathioprine Rituximab CNI	Mycophenolate Azathioprine Rituximab	Mycophenolate Azathioprine Rituximab	Mycophenolate Azathioprine Rituximab
Additional options	Cyclophosphamide Nintedanib Azathioprine	JAKi Cyclophosphamide	Tocilizumab Cyclophosphamide	Cyclophosphamide	Cyclophosphamide
+ Glucocorticoids	Strong recommendation against GCs	Short-term GCs	Short-term GCs	Short-term GCs	Short-term GCs

Strong recommendation *against* | Conditional recommendation

ACCP (also CHEST) = American College of Chest Physicians; ACR = American College of Rheumatology; CNI = calcineurin inhibitor; GC = glucocorticoid; JAKi = Janus kinase inhibitor.

Johnson SR. *Arthritis Rheumatol.* 2024;76:1182-1200.

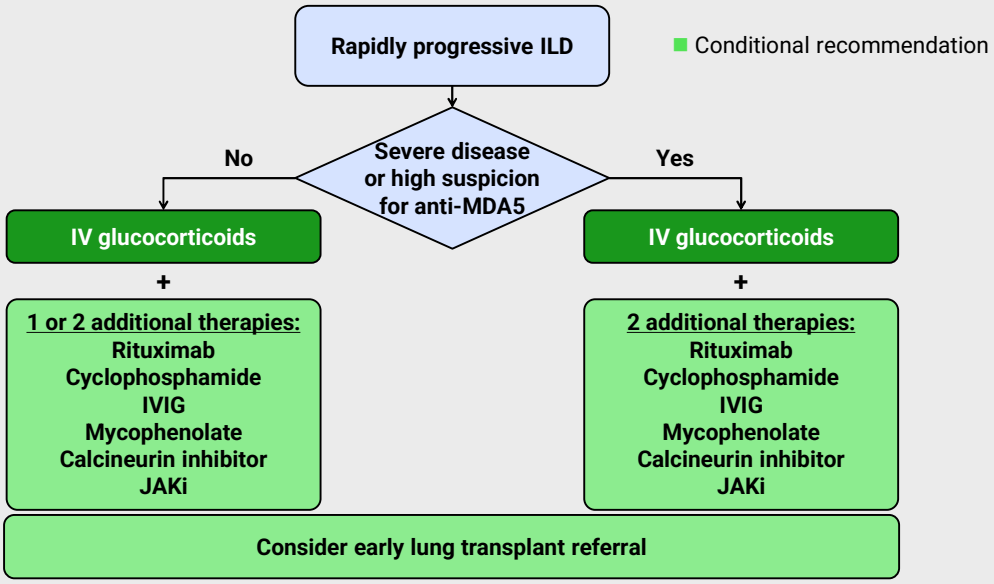
ACR/CHEST Treatment Algorithm for Progression on First SARD-ILD Therapy



AHSCT = autologous hematopoietic stem cell transplant; IVIG = intravenous immunoglobulin.

Johnson SR. *Arthritis Rheumatol.* 2024;76:1182-1200.

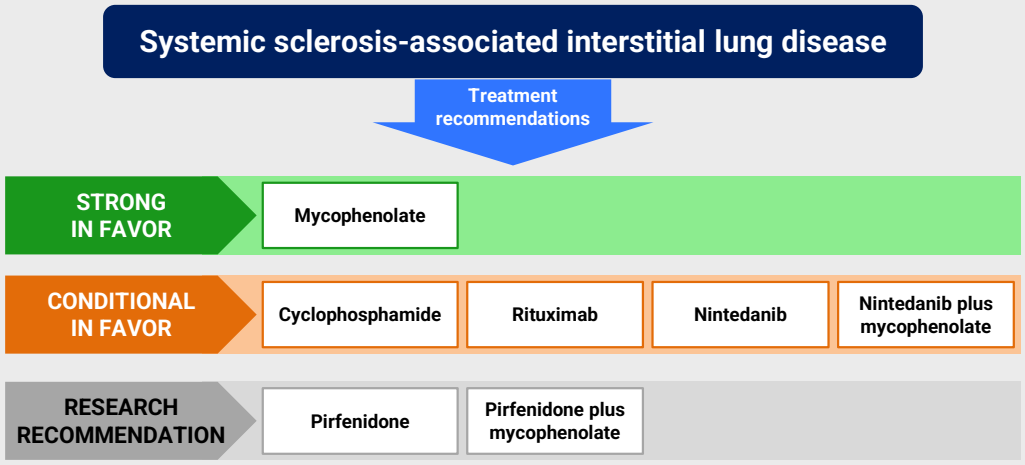
ACR/ACCP Treatment Algorithm for Rapidly Progressive SARD-ILD



Johnson SR, et al. *Arthritis Rheum.* 2024;76(8):1182-1200.

Treatment of Systemic Sclerosis-Associated Interstitial Lung Disease: Evidence-Based Recommendations

An official American Thoracic Society Clinical Practice Guideline



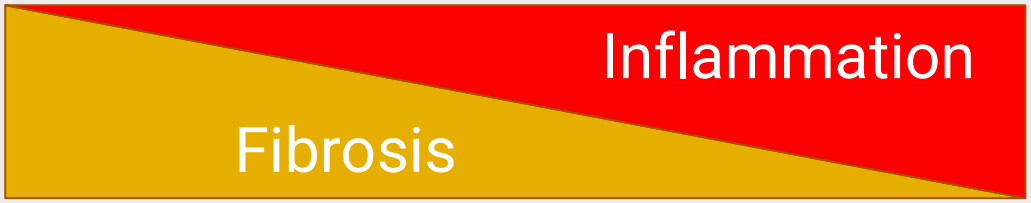
Summary of treatment recommendations for patients with systemic sclerosis-associated interstitial lung disease (SSc-ILD) from the SSc-ILD Guideline Committee

Mycophenolate and cyclophosphamide are not FDA-approved for this indication.

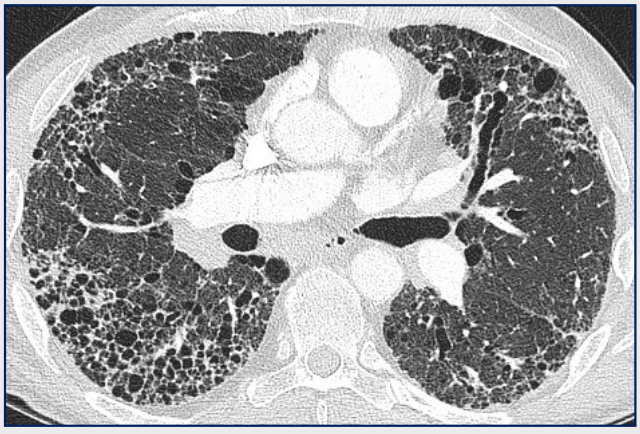
Raghu G, et al. *Am J Respir Crit Care Med.* 2024;209(2):137-152.

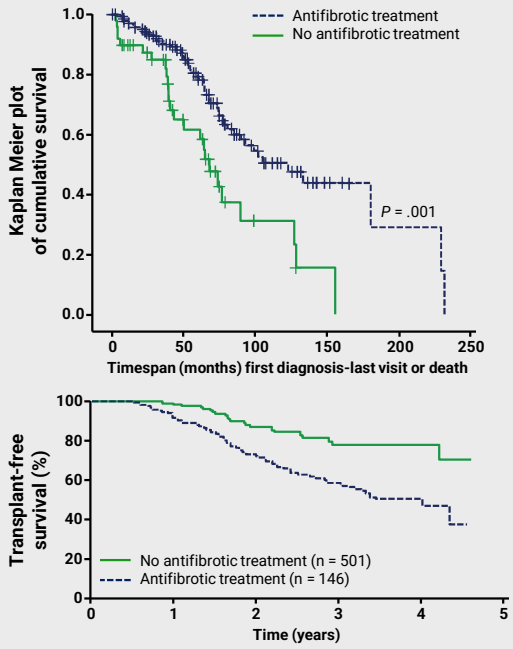
Treatment Review

What Are You Treating?



Benefits of Antifibrotic Therapy in IPF

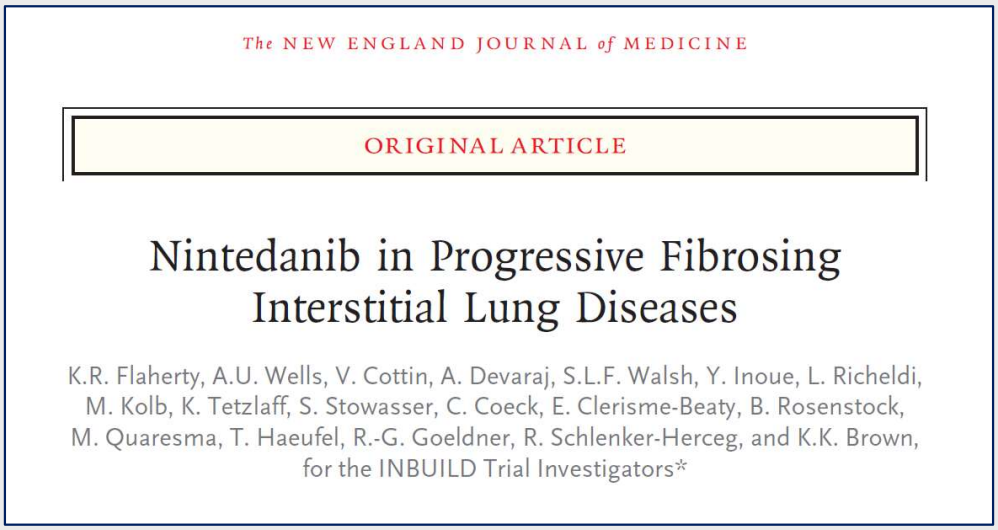




IPF = idiopathic pulmonary fibrosis.

Guenther A, et al. *Respir Res.* 2018;19:141. Richeldi L, et al. *N Engl J Med.* 2014;370:2071-2078. Jo HE, et al. *Eur Respir J.* 2017;49:1601592.

Nintedanib*



The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

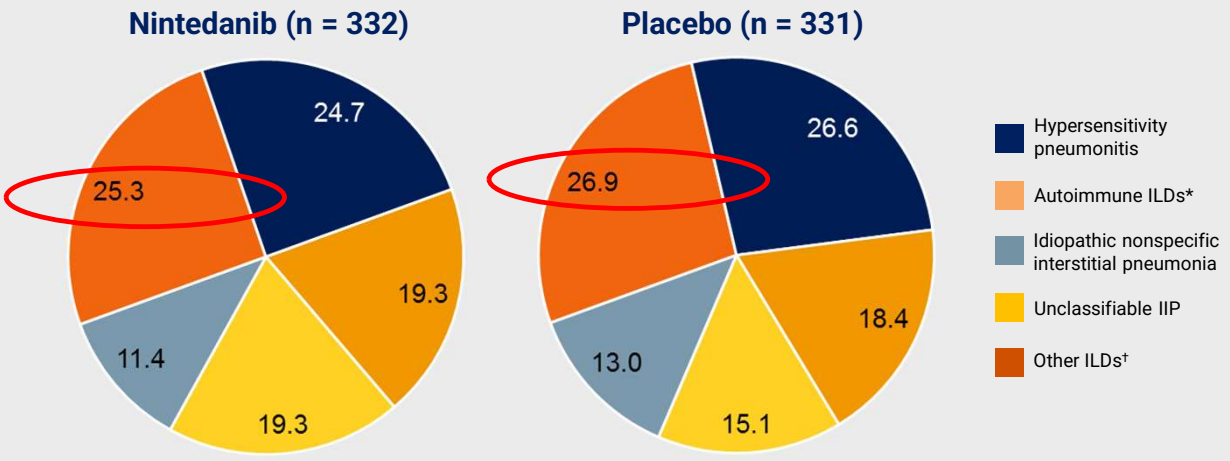
Nintedanib in Progressive Fibrosing Interstitial Lung Diseases

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*

Flaherty KR, et al. *N Engl J Med*. 2019;381(18):1718-1727.

*Nintedanib is FDA-approved to treat adults with IPF, F-ILD, and SSc-ILD.

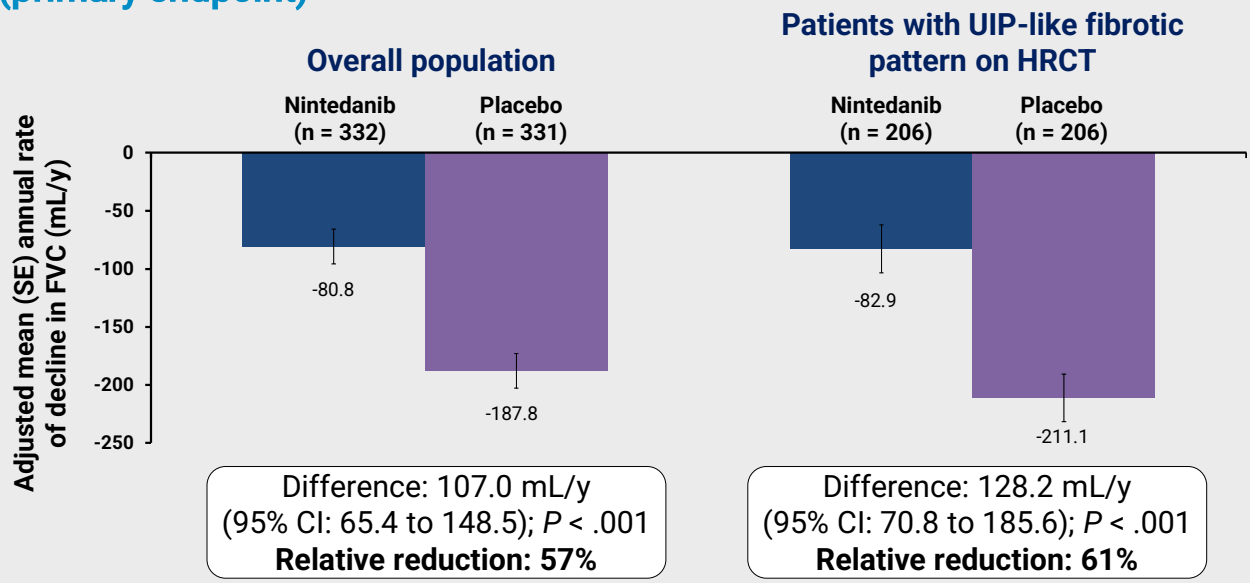
Clinical ILD Diagnoses in Overall Population



*Included rheumatoid arthritis-associated ILD (nintedanib 12.7%; placebo 14.2%), systemic sclerosis-associated ILD (nintedanib 6.9%; placebo 4.8%), and mixed connective tissue disease-ILD (nintedanib 2.1%; placebo 3.6%).

Flaherty KR, et al. *N Engl J Med*. 2019;381(18):1718-1727.

Adjusted Annual Rate of Decline in FVC (mL/y) Over 52 Weeks (primary endpoint)



SE = standard error.

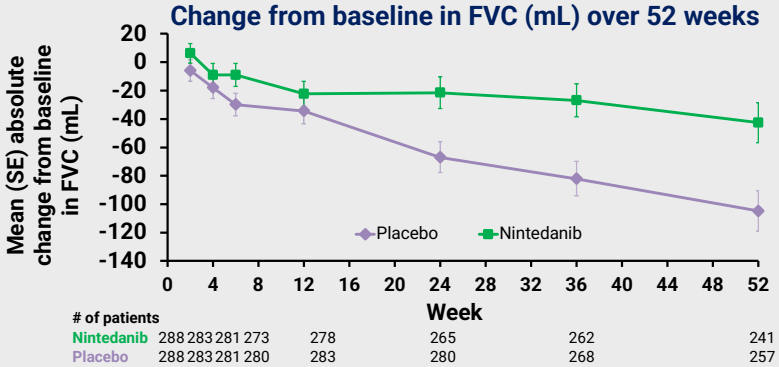
Flaherty KR, et al. *N Engl J Med.* 2019;381(18):1718-1727.


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*

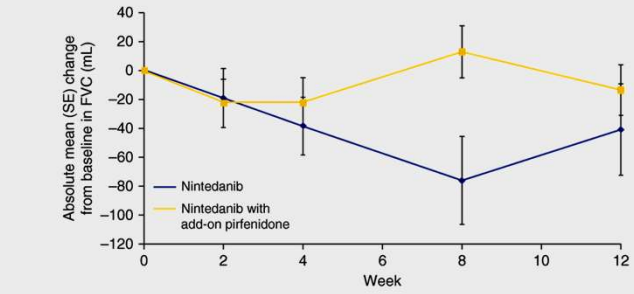


SE = standard error.

Distler O, et al. *N Engl J Med.* 2019;380(26):2518-2528.

The Future of IPF Management: Combination Therapy?

INJOURNEY trial: Pirfenidone + nintedanib in IPF



n					
Nintedanib	51	49	48	45	44
Nintedanib with add-on pirfenidone	53	52	50	50	48

Future drug development

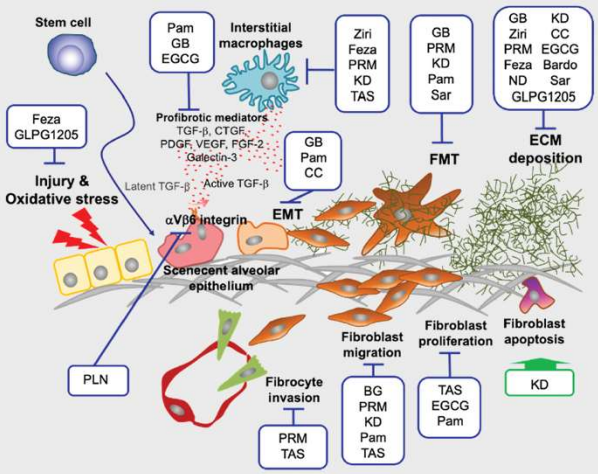


Image courtesy of Dr Toby Maher.

Yanagihara T, et al. *Expert Opin Ther Targets*. 2021;1-10. Vancheri C, et al. *Am J Respir Crit Care Med*. 2018;197(3):356-363.

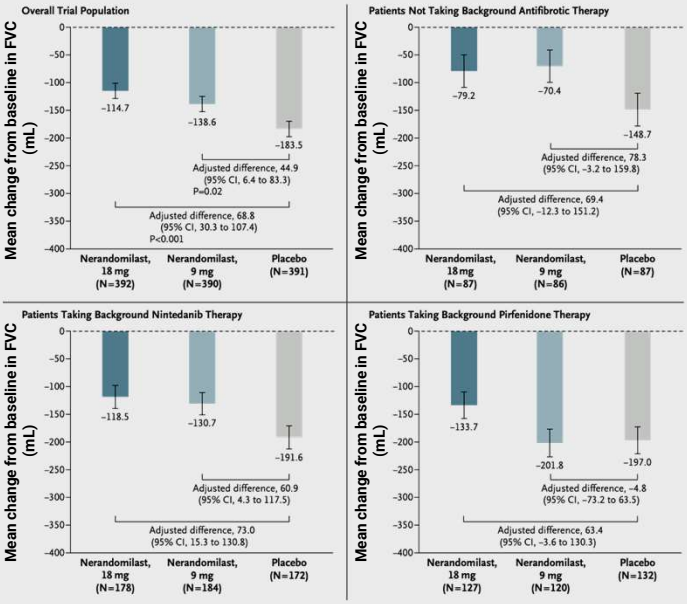
Nerandomilast*

- First molecule in a new class of PDE4B inhibitors
- Approved in adult patients with idiopathic pulmonary fibrosis and progressive pulmonary fibrosis
- FIBRONEER™ program included 2 phase 3 randomized, double-blind, placebo-controlled trials
  - FIBRONEER-IPF: 1177 patients with IPF
  - FIRBONEER-ILD: 1176 patients with PPF
- The primary endpoint was absolute change from baseline in FVC (mL) at Week 52

*Nerandomilast is FDA-approved to treat adults with IPF and PPF.

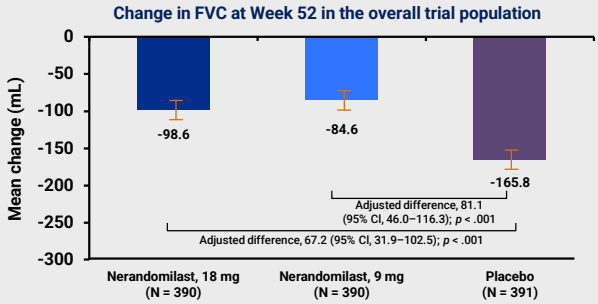
Richeldi L, et al *N Engl J Med*. 2025;392(22):2193-2202.

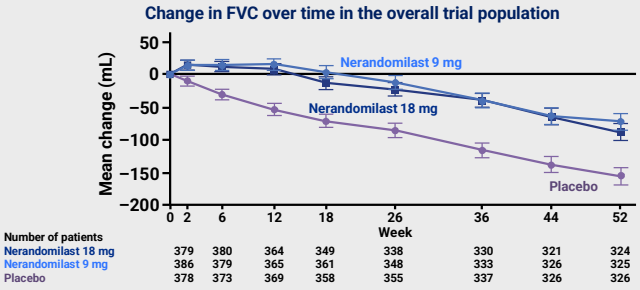
Change from baseline in forced vital capacity at Week 52 (FIBRONEER-IPF)

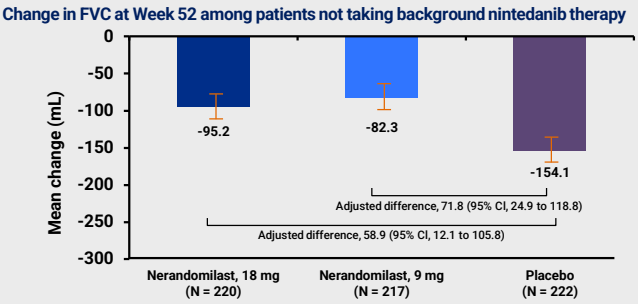


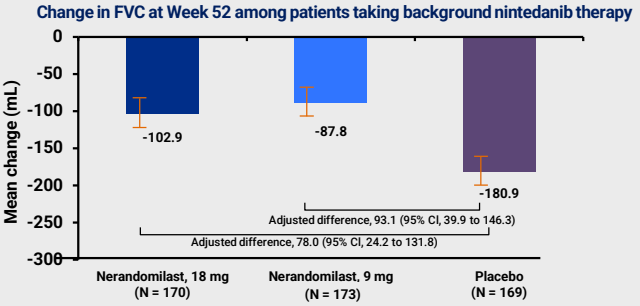
Patients were randomly assigned in a 1:1:1 ratio to receive nerandomilast at a dose of 18 mg twice daily, nerandomilast at a dose of 9 mg twice daily, or placebo twice daily. I bars indicate standard errors.

Fibroneer-ILD: Changes From Baseline to Week 52 in the Forced Vital Capacity (FVC)



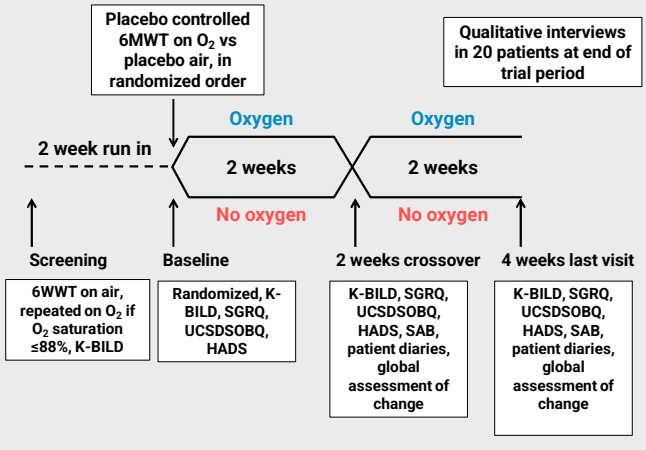


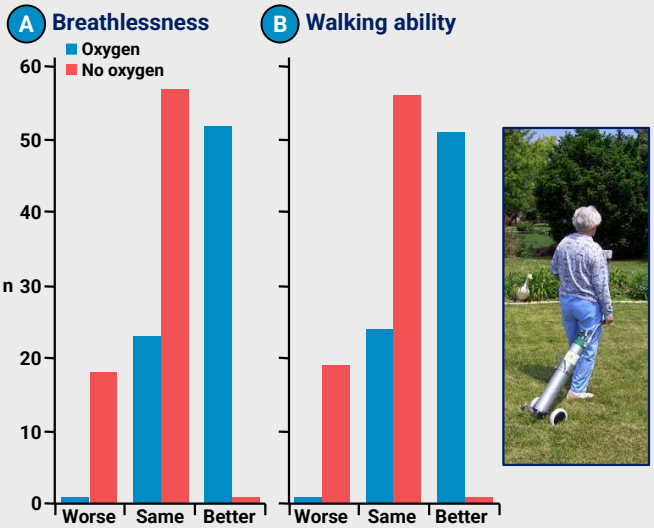




Maher TM, et al. *N Engl J Med.* 2025;392:2203-2214.

Oxygen



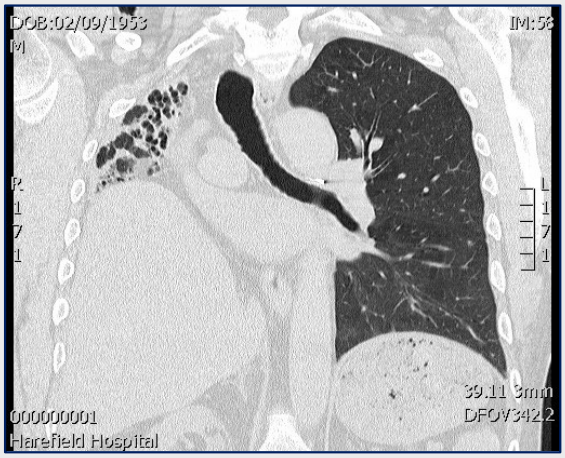


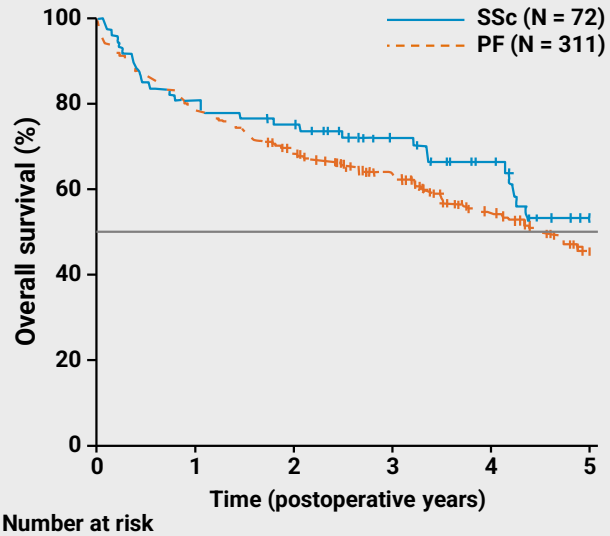
6MWT = 6-minute walk test; HADS = Hospital Anxiety and Depression Scale; K-BILD = King's Brief Interstitial Lung Disease questionnaire; O_2 = oxygen; SAB = SenseWear Pro Armband; SGRQ = St George's Respiratory Questionnaire; UCSDSOBQ = University of California, San Diego Shortness of Breath Questionnaire.

Visca D, et al. *Lancet Resp Med.* 2018;6(10):759-770.

Numbers of patients reporting improved, same, or worse breathlessness (A) and walking ability (B) after 2 weeks on ambulatory oxygen or no treatment.

Lung Transplant





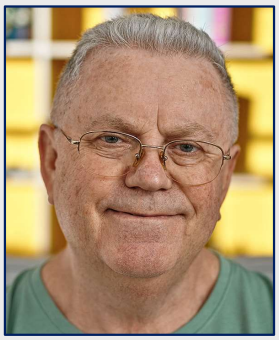
Number at risk						
PF	311	245	207	166	119	82
SSc	72	58	53	39	27	15

PF = pulmonary fibrosis; SSc = systemic sclerosis.

Crespo MM, et al. *Ann Am Thorac Soc.* 2016;13:784-792.

Case Discussions

Case 1: Rick



- A 63-year-old US Army veteran
  - Served in Kuwait 1990 to 1991
- 35-year history of smoking
- 5-year lung cancer survivor
- Treated with radiation and chemotherapy
- Presents with fatigue and shortness of breath when walking the dog with his wife

▪ FVC: 2.91 L (69%)	• TLC: 4.16 L (63%)	▪ Arterial blood gas: 7.41/38/84
▪ FEV1: 2.78 L (84%)	• DLCO: 18.8 mL/min/mmHg (73%)	▪ CMP: WNL
▪ FEV1/FVC: 95		

- Velcro-like crackles heard during late inspiration and honeycombing observed on chest CT

CMP = comprehensive metabolic panel; DLCO = diffusing capacity of the lungs for carbon monoxide; FVC = forced vital capacity; FEV1 = forced expiratory volume at 1 second; TLC = total lung capacity; WNL = within normal limits.

Case 1: Rick Discussion 1

Poll

What would be the initial diagnosis?

a) Chronic obstructive pulmonary disease (COPD)

b) Heart failure

c) F-ILD

d) Asthma

Case 1: Rick Discussion 2

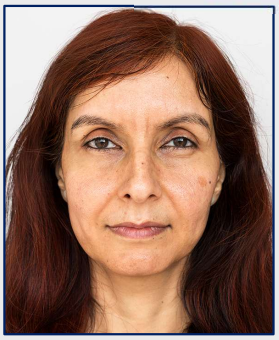
Discussion

- What is the next best step for Rick?

Case 2: Cristina

Discussion

- A 58-year-old woman with diffuse cutaneous systemic sclerosis x 10 years
- ILD diagnosed 8 years ago; on MMF and nintedanib
- Worsening dyspnea on exertion
- HRCT fibrotic NSIP pattern
- FVC 52% predicted, DLCO 25% predicted, FVC/DLCO 2.08
- 6-minute walk distance (6-MWD): 265 meters, O_2 saturation: 83% predicted
- NT-pro BNP 193



What is the next best step for Christina?

MMF = mycophenolate mofetil; NSIP = nonspecific interstitial pneumonia; NT-pro BNP = N-terminal pro-brain natriuretic peptide.

Q & A

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Thank you!

Resource	Address
Alevizos MK, et al. Risk of progression of interstitial pneumonia with autoimmune features to a systemic autoimmune rheumatic disease. *Rheumatology (Oxford).* 2020;59:1233-1240.	https://pubmed.ncbi.nlm.nih.gov/31550371/
Bernstein EJ, et al. Survival of adults with systemic sclerosis following lung transplantation: A nationwide cohort study. *Arthritis Rheumatol.* 2015;67:1314-1322.	https://pubmed.ncbi.nlm.nih.gov/25581250/
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