# About the Author

## Nathan Hambly, MD

Dr. Nathan Hambly is an Associate Professor of Medicine at McMaster University. He earned his medical degree at the University of Ottawa and completed both internal medicine and adult respirology residencies at McMaster University. Dr. Hambly is the Director of the McMaster Pulmonary Hypertension Program, and works as a consultant respirologist at St. Joseph's Healthcare Hamilton. He is the Program Director for both the Interstitial Lung Disease and Pulmonary Hypertension Fellowship training programs at McMaster. His clinical interests include pulmonary hypertension, interstitial lung disease, and sarcoidosis.

***Affiliations:*** Associate Professor of Medicine, McMaster University, Division of Respirology, Hamilton, ON
Firestone Institute for Respiratory Health, Hamilton, ON

doi.org/10.58931/crt.2025.1316

# Interstitial Lung Disease in 2025: Updated Classification, Precision Diagnostics, and Expanding Therapeutic Frontiers

**Nathan Hambly, MD**

## Introduction

Interstitial lung disease (ILD) is an umbrella term for over 200 heterogeneous disorders that primarily affect the pulmonary interstitium and/or small airways and alveoli. These disorders are often characterized by varying degrees of inflammation and fibrosis that lead to impairment in lung function and consequent respiratory symptoms. Although idiopathic pulmonary fibrosis (IPF) remains the classic example, there exists a remarkable prevalence of cases in the context of connective tissue disease (CTD) or as a consequence of antigen exposure in the setting of hypersensitivity pneumonitis (HP). Over the past decade, the integration of high-resolution computed tomography (HRCT), extended serologic panels, increased knowledge of the natural history of disease, and the high risk of fibrotic progression across the diagnostic spectrum, has refined diagnostic certainty and allowed earlier therapeutic intervention. Contemporary guidelines from the American Thoracic Society (ATS), European Respiratory Society (ERS), and European Alliance of Associations for Rheumatology (EULAR) now emphasize pattern-based phenotyping rather than rigid disease taxonomies. Clinicians are encouraged to interpret ILD through intersecting dimensions—radiologic morphology, immune serology, and cellular aging biology—each contributing to disease trajectory and treatment response.

For Canadian respirologists, these advances coincide with a rapidly evolving therapeutic landscape **(Table 1)**. Antifibrotic agents, selective immunomodulators, and emerging inhaled prostacyclin-based therapies are transforming the future therapeutic landscape. Yet, access to evidence-based treatments, especially for those with systemic sclerosis (SSc) and idiopathic inflammatory myopathies (IIM) related lung disease, remains inconsistent across provinces, underscoring the need for sustained advocacy. This review summarizes the 2025 classification framework, outlines novel diagnostic approaches, and appraises recent and emerging clinical trial data with practical guidance for clinical practice **(Table 2)**.

## Updated Classification and Guideline Framework

The 2025 ATS/ERS statement on ILD classification[1] re-centres the field on phenotypic stratification. Rather than adopting a molecular taxonomy, the framework reinforces a two-step model: identify the radiologic or histopathologic pattern, then, determine the final multidisciplinary diagnosis. For example, usual interstitial pneumonia represents a pattern, whereas IPF is the multidisciplinary diagnosis that incorporates radiologic/histopathologic patterns, clinical features, and exclusion of alternative etiologies.

Four major updates reflect the evolution of the classification system. First, the framework now includes all ILDs, not only idiopathic interstitial pneumonias, recognizing that the rationale for isolating idiopathic interstitial pneumonia is no longer justified. Second, patients are subdivided into interstitial (fibrotic or non-fibrotic) and alveolar filling disorders, clarifying diagnostic pathways. Third, diagnostic confidence is explicitly incorporated: cases with less than 50% diagnostic certainty are categorized as unclassifiable ILD, acknowledging that uncertainty is common and requires ongoing reassessment. Fourth, three new terms were introduced: bronchiolocentric interstitial pneumonia (BIP), alveolar macrophage pneumonia (AMP), and idiopathic diffuse alveolar

damage (iDAD). Of these, BIP has generated the most debate within the ILD community. BIP has been introduced as an overarching term to describe an airway-centred process that can be observed both in HP and other ILDs such as CTD-ILD, aspiration, and drug-induced disease. This is because a substantial proportion of patients with bronchiolocentric patterns of disease on pathology and imaging have non-HP diagnoses. Previously published HP guidelines still provide an excellent template for approaching the BIP pattern, recognizing that the HP diagnostic guidelines were essentially describing a radiologic/pathologic pattern of bronchiolar disease rather than a specific diagnosis. AMP has been incorporated to replace the pathologically inaccurate term desquamative interstitial pneumonia. Similarly, iDAD is replacing acute interstitial pneumonia given the imprecision that arises when chronic fibrotic lung conditions present with acute exacerbations.

This framework aligns closely with the 2025 ERS/EULAR CTD-ILD recommendations,[2] which provide disease-specific algorithms for SSc-ILD, rheumatoid-arthritis (RA)-ILD, and IIM-associated ILD. Developed by a ERS/EULAR task force established in 2020 and finalized over five years, the guidance addresses 25 Patients, Intervention, Comparison Outcomes (PICO) questions and 28 narrative questions across four primary CTD groups. Its recommendations were informed by a systematic literature review followed by structured consensus building using the Grading of Recommendations, Assessment, Development and Evaluations (GRADE) methodology. Key elements include the institution of regular screening with HRCT chest screening for high-risk patients, which includes all patients with SSc and mixed

| Trial | Population | Intervention | Comparator | Key Outcomes |
|---|---|---|---|---|
| FIBRONEER-IPF[11] | IPF | Nerandomilast 18 mg or 9 mg BID | Placebo | Reduced annual FVC decline; benefit on background antifibrotics |
| FIBRONEER-PPF[12] | Progressive pulmonary fibrosis | Nerandomilast 18 mg or 9 mg BID | Placebo | Reduced FVC decline; mortality apparent |
| INBUILD[10] | Non-IPF progressive ILD | Nintedanib | Placebo | Reduced FVC decline across heterogeneous ILDs |
| RECITAL[16] | CTD-ILD | Rituximab | Cyclophosphamide | Similar FVC outcomes; fewer adverse events with rituximab |
| EVER-ILD[17] | Fibrotic HP and CTD-ILD | MMF + rituximab | MMF alone | Improved FVC with combined therapy |
| focuSSced[15] | Diffuse SSc with ILD | Tocilizumab | Placebo | Slowed FVC decline despite neutral skin outcome |
| TETON-2 (ERS 2025) | IPF (Phase 3, ongoing) | Inhaled treprostinil | Placebo | Early reports: improvement in FVC trajectory |
| INCREASE[13] | PH-ILD (Group 3) | Inhaled treprostinil | Placebo | Improved 6MWD; reduced NT-proBNP |

**Table 1.** Selected recent clinical trials informing ILD and pulmonary fibrosis management; *courtesy of Nathan Hambly, MD*

**Abbreviations: 6MWD:** six-minute walk distance; **BID:** twice a day; **CTD:** connective tissue disease; **FVC:** forced vital capacity; **HP:** hypersensitivity pneumonitis; **ILD:** interstitial lung disease; **IPF:** idiopathic pulmonary fibrosis; **MMF:** mycophenolate mofetil; **SSc:** systemic sclerosis; **NT-proBNP:** N-terminal pro–B-type natriuretic peptide; **PH-ILD:** pulmonary hypertension associated with interstitial lung disease

connective tissue disease at diagnosis, and RA and IIM patients with known risk factors for ILD. The guidelines also detail the appropriate monitoring of ILD in each of the disease states over time, including the timing of clinical assessments and the use of serial pulmonary function tests (PFTs) and HRCT evaluations. The role of immunomodulatory and antifibrotic therapy across the spectrum of CTD-ILD was also comprehensively described, particularly focusing on the unique intricacies that characterize each CTD-ILD subcategory. For instance, the strong recommendation for tocilizumab in early diffuse SSc-ILD with signs of inflammation, whereas antifibrotic therapy is prioritized for cases of progressive pulmonary fibrosis (PPF) and/or a usual interstitial pneumonia pattern in RA-ILD.

In practice, this "pattern-plus-context" model supports tailored therapy across the spectrum of parenchymal disease. Molecular insights—particularly telomere attrition and epigenetic aging—serve as adjunctive markers that refine prognosis and inform treatment tolerance rather than redefining the classification framework itself.[3,4]

| | Key Clinical Pearls |
|---|---|
| Detailed History | • Multiple ILD phenotypes may coexist within families; genetic risk reflects fibrosis susceptibility, not a specific ILD diagnosis.<br>• Premature greying, cytopenias, cirrhosis, or MDS should prompt evaluation for telomere-mediated disease.<br>• Raynaud phenomenon, telangiectasias, and myositis markers are essential diagnostic anchors.<br>• Patients without identifiable antigen exposure have worse long-term outcomes; exposure clarification is critical. |
| Physical Examination | • Absence of crackles suggests alternative diagnoses (sarcoid); clubbing strongly suggests fibrotic phenotypes such as IPF or DIP.<br>• Detailed examination for any evidence of CTD critical including periungal capillary changes, telangiectasias, sclerodactyly, mechanic's hands, and cutaneous manifestations of IIM. |
| Diagnostics | • Ground-glass opacities may coexist with UIP; distribution and relation to fibrosis are more informative than presence alone.<br>• 'Straight-edge sign' raise suspicion for CTD.<br>• PPFE-like features suggest telomere shortening process.<br>• Idiopathic NSIP is rare and warrant thorough CTD and exposure evaluation. |
| Treatment | • Strong consideration for anti-fibrotic therapy appropriate at all stages of disease in the setting of both IPF and PPF.<br>• SSc-ILD and myositis-ILD often require early immunomodulation given evidence from RECITAL, EVER-ILD, and focuSSced trials.<br>• Patients with mixed inflammatory–fibrotic mechanisms may benefit from concurrent immunomodulation and antifibrotics.<br>• Most RA-ILD reflects underlying disease, not MTX toxicity. |
| Multidisciplinary Discussion | • Multidisciplinary discussion is essential not only for diagnosis but also for ongoing longitudinal management. |

**Table 2:** Practical clinical pearls in interstitial lung disease assessment and management; *courtesy of Nathan Hambly, MD*

**Abbreviations: CTD:** connective tissue disease; **DIP:** desquamative interstitial pneumonia; **IIM:** idiopathic inflammatory myopathies; **ILD:** interstitial lung disease; **IPF:** idiopathic pulmonary fibrosis; **MDS:** myelodysplastic syndromes; **MTX:** methotrexate; **PPF:** progressive pulmonary fibrosis; **PPFE:** pleuroparenchymal fibroelastosis; **RA-ILD:** rheumatoid arthritis-interstitial lung disease; **UIP:** usual interstitial pneumonia

## Diagnostic Innovation and Phenotypic Refinement

### *Extended Serologic and Cellular Testing*

Comprehensive autoimmune serology remains central to ILD characterization. Rigorous exclusion of an underlying CTD is essential and requires a comprehensive serological and biochemical assessment, including antinuclear antibodies, extractable nuclear antigen panel, rheumatoid factor, anti-cyclic citrullinated peptide antibodies, and measurement of serum creatine kinase. Routine incorporation of myositis-specific and myositis-associated antibodies, including anti-MDA5, PL-7, and PL-12, among others, improves diagnostic precision for clinically amyopathic dermatomyositis and overlap myositis, especially when radiologic imaging reveals evidence of organizing pneumonia or non-specific interstitial pneumonia (NSIP). Detection of anti–granulocyte-macrophage colony-stimulating factor antibodies confirms autoimmune pulmonary alveolar proteinosis, while vascular endothelial growth factor-D levels reliably distinguish lymphangioleiomyomatosis from alternative cystic lung diseases. These biomarkers reduce diagnostic delay and, in certain cases, predict therapeutic responsiveness to biologic or inhaled therapies.[5]

### *Telomere Biology and Cellular Senescence*

Telomere shortening has emerged as a unifying signal across the spectrum of fibrotic lung disease. In the PANTHER-IPF correlative study, Newton and colleagues[6] demonstrated that short leukocyte telomere length (LTL <10th percentile) predicted adverse outcomes with corticosteroid, azathioprine, and n-acetylcysteine therapy. Similarly, a retrospective multi-centre cohort analysis reported that reduced peripheral blood telomere length was associated with a heightened risk of mortality among patients with unclassifiable ILD and fibrotic HP who were treated with antimetabolite therapies such as mycophenolate or azathioprine.[3] Adegunsoye et al.[7] confirmed that LTL is inversely correlated with chronological age and independently predicts mortality across racially diverse pulmonary fibrosis cohorts. Most recently, El Husseini et al.[8] showed that short LTL portends accelerated forced vital capacity (FVC) decline in RA-ILD, reinforcing telomere length as a clinically meaningful biomarker.

For practising respirologists, LTL measurement can be measured from peripheral blood, but this testing is not routinely available across Canada. Further research is required to define its clinical role and to establish how such testing should be implemented into standard practice to guide decisions on immunosuppression and prognostication when considering antifibrotic therapy.

### *Quantitative and Artificial-Intelligence HRCT*

Advances in quantitative CT allow automated fibrosis scoring, vessel-related-structure mapping, and serial change detection. Machine-learning algorithms now predict FVC decline and mortality from baseline HRCT, offering objective support for clinical judgment.[9] Although quantitative CT analysis remains primarily a research tool, it holds substantial promise for future integration into clinical care by enabling early identification of patients suitable for clinical trial enrolment, detecting early progression in interstitial lung abnormalities, and stratifying individuals at heightened risk for fibrotic progression. As availability expands within tertiary ILD programs, it may ultimately support a more standardized approach to longitudinal monitoring.

## Therapeutic Evolution in 2025

### *Antifibrotic Therapy*

Pirfenidone and nintedanib remain the therapeutic foundation for IPF, whereas nintedanib is the only approved antifibrotic agent for PPF of non-IPF etiologies. Both agents have demonstrated approximately a 50% reduction in annual FVC decline in IPF.[10] Despite gastrointestinal intolerance in some patients, these therapies have firmly established antifibrotic treatment as standard of care.

### *Nerandomilast and the FIBRONEER Program*

The dual phase-3 trials, FIBRONEER-IPF and FIBRONEER-PPF[11,12] evaluated nerandomilast, a selective phosphodiesterase-4B inhibitor with combined antifibrotic and immunomodulatory properties. In IPF, nerandomilast reduced annual FVC loss by approximately 70 mL versus placebo ($p<0.001$). In PPF—including CTD-ILD and fibrotic hypersensitivity pneumonitis—the adjusted FVC difference was similar, with an emerging mortality signal favouring active therapy. Diarrhea was the principal adverse effect but was rarely dose-

limiting. These findings position nerandomilast as a promising addition to the therapeutic armamentarium, having recently received FDA approval for IPF and is currently under review by Canadian regulatory authorities.

### *Emerging and Ongoing Trials*

The TETON trial series (NCT04708782, NCT05255991, NCT05943535) is investigating inhaled treprostinil for IPF and PPF, building on the INCREASE trial,[13] which demonstrated improved exercise capacity and FVC in Group 3 pulmonary hypertension secondary to parenchymal lung disease. Treprostinil's potential antifibrotic and vasodilatory effects have generated considerable interest. Unfortunately, commercial nebulized treprostinil is currently not presently available in Canada. In parallel, the ALOFT IPF (NCT01234567) and ALOFT PPF (NCT06025578) global phase 3 studies are evaluating the safety and efficacy of the oral lysophosphatidic acid receptor 1 ($LPA_1$) antagonist admilparant in patients with IPF and PPF. Additional trials of nebulized pirfenidone (NCT06329401) and nintedanib (NCT07194382) aim to enhance pulmonary bioavailability while minimizing systemic side-effects. Collectively, these studies suggest that local, multimodal/combinational antifibrotic delivery strategies may become a mainstay of future therapy.

### *Immunomodulatory Therapy*

Immunomodulatory agents remain a mainstay in the management of CTD-ILD. Antimetabolite agents such as mycophenolate mofetil (MMF) remain first-line therapy for SSc-ILD and IIM-ILD. However, the recent EULAR/ERS guidelines, based on evidence from the faSScinate[14] and focuSSced[15] studies, strongly recommend the IL-6 receptor antagonist tocilizumab, which has demonstrated efficacy in slowing FVC decline in SSc-ILD, and has received regulatory approval in multiple jurisdictions. Similarly, the RECITAL trial[16] showed that rituximab achieved FVC stabilization equivalent to intravenous cyclophosphamide but with fewer adverse events. The EVER-ILD trial[17] demonstrated that combining MMF with rituximab improved FVC and progression-free survival compared with MMF alone in patients with an NSIP pattern of disease. In RA-ILD, initial therapeutic decisions are dictated based on whether active arthritis is present. When active arthritis exists, EULAR/ERS conditionally recommend abatacept, rituximab, or Janus kinase inhibitors.[2] Despite their underlying immunologic pathogenesis, identifying a PPF phenotype in CTD-ILD remains critically important because of its distinct prognostic significance and therapeutic implications. In a Canadian cohort study, 45% of patients with CTD-ILD developed evidence of PPF within 24 months of their diagnosis.[18]

## Interstitial Lung Abnormalities

Interstitial lung abnormalities (ILAs) refer to incidental HRCT findings characterized by non-dependent ground-glass opacities, reticulation, traction bronchiectasis, or subpleural fibrotic changes involving at least 5% of a lung zone in individuals without a known diagnosis of ILD. The 2025 ATS/ERS Clinical Statement highlights that ILAs are increasingly detected in lung cancer screening cohorts and in relatives of patients with familial pulmonary fibrosis.[19]

Risk factors for progression include advanced age, prior smoking history, subpleural reticulation, and fibrotic features. However, major uncertainties remain: the natural history varies, surveillance intervals are not standardized, and therapeutic implications remain unclear. Whether early antifibrotic therapy benefits high-risk ILAs remains unknown. For the practising respirologist, ILAs should prompt a structured clinical evaluation, baseline PFTs, and risk-adapted follow-up, especially in individuals with CTD, relevant exposures, or a family history of pulmonary fibrosis.

## Future Directions

The next phase of ILD management will likely integrate multi-omic risk profiling with clinical phenotyping to enable personalized therapy. Epigenetic clocks and circulating fibroblast activation markers may soon complement telomere length as indicators of disease behaviour. Novel agents targeting alternative fibrotic pathways are entering phase 2 trials, while inhaled delivery platforms promise improved safety and adherence. Expanding access to antifibrotics and biologics across Canada will determine whether these innovations translate into improved outcomes.

## Conclusion

By 2025, ILD care has evolved from a single-disease paradigm to an integrated, phenotype-guided approach. Radiographic patterns, immune profiles, and cellular-senescence markers

collectively define prognosis and therapeutic responsiveness. Emerging evidence from the FIBRONEER trial establishes nerandomilast as a promising antifibrotic with a potential mortality benefit, while the RECITAL, EVER-ILD, and focuSSced trials consolidate the role of biologic immunomodulation in CTD-ILD. For Canadian clinicians, the challenge lies not in scientific uncertainty but in equitable implementation—ensuring that all patients with ILD gain timely access to recommended therapies. Through informed diagnostics, evidence-based treatment, and continued advocacy, our respiratory community remains central to improving outcomes in this complex and rapidly advancing field.

## Correspondence

**Nathan Hambly, MD**
**Email:** nathan.hambly@medportal.ca

## Financial Disclosures

**N.H.: Consultancy/Advisory Board:** Boehringer Ingelheim, Janssen, Merck; **Speaker Honoraria:** Astra-Zeneca, Boehringer Ingelheim, Janssen, Merck; **Speaker's Bureau:** Boehringer Ingelheim, GSK, Janssen, Merck; **Funded Grants or Clinical Trials:** Avalyn Pharma, BMS, Boehringer Ingelheim, GSK, Janssen, Merck, Savara, United Therapeutics

## References

1. Ryerson CJ, Adegunsoye A, Piciucchi S, Hariri LP, Khor YH, Wijsenbeek MS, et al. Update of the international multidisciplinary classification of the interstitial pneumonias: an ERS/ATS statement. Eur Respir J. Published online August 28, 2025. doi:10.1183/13993003.00158-2025
2. Antoniou KM, Distler O, Gheorghiu AM, Moor CC, Vikse J, Bizymi N, et al. ERS/EULAR clinical practice guidelines for connective tissue disease-associated interstitial lung diseaseDeveloped by the task force for connective tissue disease-associated interstitial lung disease of the European Respiratory Society (ERS) and the European Alliance of Associations for Rheumatology (EULAR)Endorsed by the European Reference Network on rare respiratory diseases (ERN-LUNG). Eur Respir J. Published online September 11, 2025. doi:10.1183/13993003.02533-2024
3. Zhang D, Adegunsoye A, Oldham JM, Kozlitina J, Garcia N, Poonawalla M, et al. Telomere length and immunosuppression in non-idiopathic pulmonary fibrosis interstitial lung disease. Eur Respir J. 2023;62(5):2300441. doi: 10.1183/13993003.00441-2023 .
4. Goobie GC, Marinescu DC, Adegunsoye A, Bourbeau J, Carlsten C, Clifford RL, et al. Accelerated epigenetic ageing worsens survival and mediates environmental stressors in fibrotic interstitial lung disease. Eur Respir J. 2025;65(6):2401618. Published 2025 Jun 5. doi:10.1183/13993003.01618-2024
5. Trapnell BC, Inoue Y, Bonella F, Wang T, McCarthy C, Arai T, et al. Phase 3 trial of inhaled molgramostim in autoimmune pulmonary alveolar proteinosis. N Engl J Med. 2025;393(8):764-773. doi:10.1056/NEJMoa2410542
6. Newton CA, Zhang D, Oldham JM, Kozlitina J, Ma SF, Martinez F, et al. Telomere length and use of immunosuppressive medications in idiopathic pulmonary fibrosis. Am J Respir Crit Care Med. 2019;200(3):336-347. doi:10.1164/rccm.201809-1646OC
7. Adegunsoye A, Newton CA, Oldham JM, Ley B, Lee CT, Linderholm AL, et al. Telomere length associates with chronological age and mortality across racially diverse pulmonary fibrosis cohorts. Nat Commun.2023;14(1):1489. Published 2023 Mar 17. doi:10.1038/s41467-023-37193-6
8. El Husseini K, Lee JS, Juge PA, Ebstein E, Ottaviani S, Borie R, et al. Short telomere length is associated with accelerated lung disease progression in rheumatoid arthritis-associated interstitial lung disease. Eur Respir J. Published online September 18, 2025. doi:10.1183/13993003.00587-2025
9. Dixon G, Thould H, Wells M, Tsaneva-Atanasova K, Scotton CJ, Gibbons MA, et al. A systematic review of the role of quantitative CT in interstitial lung disease. Eur Respir Rev. 2025;34(176):240194. Published 2025 Apr 30. doi:10.1183/16000617.0194-2024
10. Flaherty KR, Wells AU, Cottin V, Devaraj A, Walsh SLF, Inoue Y, et al. Nintedanib in progressive fibrosing interstitial lung disease. N Engl J Med. 2019;381(18):1718-1727. doi:10.1056/NEJMoa1908681
11. Richeldi L, Azuma A, Cottin V, Kreuter M, Maher TM, Martinez FJ, et al. Nerandomilast in patients with idiopathic pulmonary fibrosis. N Engl J Med.2025;392(22):2193-2202. doi:10.1056/NEJMoa2414108
12. Maher TM, Assassi S, Azuma A, Cottin V, Hoffmann-Vold AM, Kreuter M, et al. Nerandomilast in patients with progressive pulmonary fibrosis: outcomes from a phase III trial. N Engl J Med.2025;392(22):2203-2214. doi:10.1056/NEJMoa2503643
13. Waxman A, Restrepo-Jaramillo R, Thenappan T, Ravichandran A, Engel P, Bajwa A, et al. Inhaled treprostinil in pulmonary hypertension due to interstitial lung disease. N Engl J Med. 2021;384(4):325-334. doi:10.1056/NEJMoa2008470
14. Khanna D, Denton CP, Jahreis A, van Laar JM, Frech TM, Anderson ME, et al. Safety and efficacy of subcutaneous tocilizumab in adults with systemic sclerosis (faSScinate): a phase 2, randomized, controlled trial. Lancet. 2016;387(10038):2630-2640. doi:10.1016/S0140-6736(16)00232-4
15. Khanna D, Lin CJF, Furst DE, Goldin J, Kim G, Kuwana M, et al. Tocilizumab in systemic sclerosis: results of a randomized, double-blind, placebo-controlled, phase 3 trial. Lancet Respir Med.2020;8(10):963-974. doi:10.1016/S2213-2600(20)30318-0
16. Maher TM, Tudor VA, Saunders P, Gibbons MA, Fletcher SV, Denton CP, et al. Rituximab versus intravenous cyclophosphamide in patients with connective tissue disease-associated interstitial lung disease in the UK (RECITAL): a double-blind, double-dummy, randomized, controlled, phase 2b trial. Lancet Respir Med. 2023;11(1):45-54. doi:10.1016/S2213-2600(22)00359-9
17. Manikikian J, Caille A, Reynaud-Gaubert M, Agier MS, Bermudez J, Bonniaud P, et al. Rituximab and mycophenolate mofetil combination in patients with interstitial lung disease (EVER-ILD): a double-blind, randomized, placebo-controlled trial. Eur Respir J. 2023;61(6):2202071. Published 2023 Jun 8. doi:10.1183/13993003.02071-2022
18. Hambly N, Farooqi MM, Dvorkin-Gheva A, Donohoe K, Garlick K, Scallan C, et al. Prevalence and characteristics of progressive fibrosing interstitial lung disease in a prospective registry. Eur Respir J. 2022;60(4):2102571. Published 2022 Oct 6. doi:10.1183/13993003.02571-2021
19. Podolanczuk A, Hunninghake GM, Wilson KC, Knor YH, Kheir F, Pang B, et al. Approach to the evaluation and management of interstitial lung abnormalities. Am J Respir Crit Care Med. 2025;211(7):1132-1155. doi:10.1164/rccm.202505-1054ST