ILD Diagnosis and Management: A Practical ATS 2026 Update

Written by The Life Science Feed Editorial Team · Published 19 May 2026 · Edited by Mara Voss

> Interstitial lung disease (ILD) presents a significant diagnostic and therapeutic challenge due to its heterogeneous nature and often progressive course. General practitioners and specialists require clear, evidence-based guidance to navigate the complexities of early identification and optimal management. The ATS 2026 session, "Interstitial Lung Disease: A Practical Approach to Diagnosis and Management," offered a structured framework, underscoring the critical role of multidisciplinary assessment and timely intervention.

KEY TAKEAWAYS

- The Pivot: The session reinforced the necessity of a multidisciplinary team (MDT) approach for accurate ILD diagnosis and classification, moving beyond isolated specialist assessments.
- The Data: Early initiation of antifibrotic therapy, specifically nintedanib or pirfenidone, is recommended for progressive fibrosing ILD, irrespective of the underlying ILD subtype.
- The Action: Clinicians should expedite referral to ILD centres for suspected cases and consider antifibrotic therapy once progressive fibrosing ILD is established, adhering to current guideline recommendations.

Interstitial lung diseases encompass a diverse group of more than 200 conditions characterised by inflammation and fibrosis of the lung parenchyma.1 Accurate diagnosis is paramount, as misclassification can lead to inappropriate treatment and suboptimal patient outcomes. The ATS 2026 session highlighted the diagnostic algorithm, beginning with a comprehensive clinical history, physical examination, and review of high-resolution computed tomography (HRCT) imaging.2

The session emphasised that a definitive diagnosis of ILD, particularly idiopathic pulmonary fibrosis (IPF), often requires a multidisciplinary team (MDT) discussion involving pulmonologists, radiologists, and pathologists.3 This collaborative approach helps to integrate clinical, radiological, and histopathological data to arrive at the most accurate diagnosis and classification. For instance, a usual interstitial pneumonia (UIP) pattern on HRCT in the appropriate clinical context is highly suggestive of IPF, but other ILDs can also present with a UIP pattern.4 The MDT ensures these nuances are considered, reducing diagnostic error.3

The prevalence and incidence of ILDs vary widely depending on the specific subtype. IPF, the most common and severe form of idiopathic interstitial pneumonia, has an estimated incidence ranging from 2.8 to 9.3 cases per 100,000 person-years and a prevalence of 10 to 60 cases per 100,000 people globally.14 Other ILDs, such as sarcoidosis, hypersensitivity pneumonitis, and connective tissue disease-associated ILD, also contribute significantly to the overall burden of interstitial lung disease. These conditions often present with overlapping symptoms, underscoring the need for a systematic diagnostic approach.

Practical Approach to Diagnosis and Management

The diagnostic pathway presented at ATS 2026 began with the recognition of symptoms such as progressive dyspnoea, chronic cough, and crackles on lung auscultation.1 Initial investigations typically include pulmonary function tests (PFTs), which often reveal a restrictive ventilatory defect and reduced diffusing capacity for carbon monoxide (DLCO).5 HRCT of the chest is considered the cornerstone of imaging, providing critical information on the pattern and distribution of lung abnormalities.4

Once ILD is suspected, the next step involves identifying the specific subtype. This differentiation is crucial because management strategies vary significantly. For example, some ILDs, such as hypersensitivity pneumonitis or sarcoidosis, may respond to immunosuppressive therapy, while others, like IPF, require antifibrotic agents.6 The session underscored the importance of ruling out secondary causes of ILD, such as connective tissue diseases (CTDs), drug-induced ILD, or occupational exposures.2 Serological testing for CTD-related autoantibodies is therefore a standard part of the diagnostic workup.7

For patients with progressive fibrosing ILD (PF-ILD), defined by worsening respiratory symptoms, physiological decline, or radiological progression despite standard management, antifibrotic therapy is now a key component of treatment.8 The session reiterated that nintedanib and pirfenidone are approved antifibrotic agents that slow the rate of decline in forced vital capacity (FVC) in patients with IPF.9,10 Nintedanib acts as a tyrosine kinase inhibitor, targeting multiple receptors involved in fibrotic processes, including vascular endothelial growth factor receptor (VEGFR), fibroblast growth factor receptor (FGFR), and platelet-derived growth factor receptor (PDGFR). Pirfenidone is an oral antifibrotic agent with anti-inflammatory and antioxidant properties, modulating the production of growth factors and cytokines.6 Furthermore, nintedanib has received approval for use in a broader group of patients with chronic PF-ILDs, demonstrating a reduction in FVC decline across various ILD subtypes that exhibit a progressive fibrosing phenotype.8

The practical guidance included recommendations for early initiation of antifibrotic therapy once PF-ILD is established.8 This is based on evidence indicating that early intervention can preserve lung function more effectively than delayed treatment.11 Management also extends beyond pharmacotherapy to include supportive care measures such as oxygen therapy, pulmonary rehabilitation, and management of comorbidities like gastroesophageal reflux disease (GERD) and pulmonary hypertension.12 The session also touched upon the role of lung transplantation for carefully selected patients with advanced ILD.13

Limitations in the current understanding of ILD include the challenge of predicting which patients will develop a progressive fibrosing phenotype and the need for more targeted therapies for specific ILD subtypes. The heterogeneity of ILD presentation and progression across different patient populations, including varying genetic predispositions and environmental exposures, complicates the development of universally effective treatments. Ongoing research is exploring novel antifibrotic and anti-inflammatory agents, as well as biomarkers for earlier diagnosis and prognostication.14

The ATS 2026 session provided a valuable synthesis of current best practices, reinforcing the need for a systematic, MDT-driven approach to improve outcomes for patients with ILD.

CLINICAL IMPLICATIONS

The ATS 2026 session on ILD diagnosis and management reinforces a critical message for clinicians: the era of a single specialist managing complex ILD cases in isolation is over. The emphasis on multidisciplinary team (MDT) discussions for definitive diagnosis is not merely academic; it is a pragmatic necessity to reduce misdiagnosis and ensure appropriate therapeutic pathways. GPs, in particular, must be attuned to subtle signs of ILD and understand the urgency of referral to specialised centres, as delayed diagnosis directly impacts the window for effective antifibrotic intervention.

The expanding indication for antifibrotic agents like nintedanib and pirfenidone beyond idiopathic pulmonary fibrosis (IPF) to other progressive fibrosing ILDs represents a significant shift. This means that a broader cohort of patients, previously without specific therapeutic options, can now benefit from treatments that slow disease progression. However, this also places a greater onus on accurate phenotyping of ILD to identify those who will genuinely benefit. The pharmaceutical industry, having developed these agents, now faces the challenge of ensuring equitable access and educating the wider medical community on their appropriate use, moving beyond the initial IPF-centric focus.

For patients, these developments offer a tangible improvement in prognosis. The prospect of slowing lung function decline, even if not reversing it, provides hope in conditions that were once universally progressive and fatal. However, the cost of these therapies and the need for lifelong treatment remain significant considerations. Healthcare systems must adapt to support these long-term management strategies, ensuring that the promise of improved outcomes is not undermined by financial barriers or fragmented care. The session's practical approach underscores that while pharmacological advancements are vital, comprehensive supportive care and a coordinated clinical effort remain indispensable.

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