

Pulmonary fibrosis: Why early diagnosis remains a clinical enigma

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Pulmonary fibrosis, a progressive and often fatal lung disease, continues to challenge clinicians with its insidious onset and complex diagnostic pathway. For many patients, the journey from initial symptoms to a definitive diagnosis and subsequent treatment initiation is protracted, a delay that carries substantial prognostic weight. This persistent hurdle in care delivery highlights a critical unmet need in respiratory medicine.

KEY TAKEAWAYS

- **The Pivot:** Early and accurate diagnosis of pulmonary fibrosis is crucial, but current pathways are often inefficient, delaying access to disease-modifying therapies.
- **The Data:** While no specific trial data is presented here, the clinical consensus emphasizes that timely intervention with antifibrotic agents can slow disease progression.
- **The Action:** Clinicians should maintain a high index of suspicion for interstitial lung disease in patients presenting with unexplained dyspnoea or chronic cough, and advocate for streamlined referral pathways to specialist centres.

Idiopathic pulmonary fibrosis (IPF) stands as the most common and severe form of interstitial lung disease (ILD), characterized by progressive scarring of the lung parenchyma. This relentless fibrotic process leads to irreversible loss of lung function, culminating in respiratory failure. The median survival for IPF patients without treatment is often cited as 3 to 5 years from diagnosis, a prognosis worse than many cancers. The disease typically affects older adults, with incidence increasing significantly after the age of 60. Men are more frequently affected than women, and a history of smoking is a well-established risk factor, though not all patients with IPF are current or former smokers. Environmental exposures, genetic predispositions, and gastroesophageal reflux disease have also been implicated in its pathogenesis, highlighting the multifactorial nature of this devastating condition.

The underlying pathophysiology of IPF involves a complex relationship between genetic susceptibility and environmental insults, leading to aberrant wound healing responses in the lung. Repeated micro-injuries to the alveolar epithelium are thought to trigger a cascade of events, including epithelial cell activation, fibroblast proliferation, and excessive extracellular matrix deposition. This process is driven by various profibrotic mediators, such as transforming growth factor-beta (TGF- β), platelet-derived growth factor (PDGF), and connective tissue growth factor (CTGF). The normal repair mechanisms fail, resulting in uncontrolled collagen deposition and architectural distortion of the lung. This progressive scarring stiffens the lungs, making gas exchange increasingly difficult and leading to the characteristic symptoms of dyspnoea and chronic dry cough. The relentless nature of the disease means that early detection and intervention are paramount to preserving lung function and improving patient outcomes.

The Diagnostic Labyrinth

Diagnosing pulmonary fibrosis, particularly IPF, is a multidisciplinary endeavour that often begins with a primary care physician recognizing non-specific symptoms. Patients typically present with progressive exertional dyspnoea, a persistent dry cough, and sometimes fatigue or weight loss. On physical examination, bilateral inspiratory crackles, often described as 'Velcro rales,' are a classic but not universally present finding. Digital clubbing can also be observed in a subset of patients. The initial workup often includes chest radiography, which may show reticular opacities, but high-resolution computed tomography (HRCT) of the chest is indispensable for definitive diagnosis. HRCT images reveal characteristic patterns, with a 'usual interstitial pneumonia' (UIP) pattern being pathognomonic for IPF in the right clinical context. This pattern includes subpleural and basal predominance of reticular opacities, traction bronchiectasis, and honeycomb change, often without features atypical for UIP.

But the diagnostic journey does not end with imaging. A multidisciplinary discussion (MDD) involving pulmonologists, radiologists, and pathologists is considered the gold standard for confirming an IPF diagnosis. This collaborative approach helps differentiate IPF from other forms of ILD, which can present with similar clinical and radiological features but require different management strategies. Conditions such as hypersensitivity pneumonitis, connective tissue disease-associated ILD, and drug-induced lung disease must be carefully excluded. Sometimes, a surgical lung biopsy is necessary to obtain histological confirmation, especially when HRCT findings are indeterminate. This invasive procedure carries its own risks, including pneumothorax and exacerbation of the underlying lung disease, further complicating the diagnostic pathway. The need for such a detailed and often prolonged evaluation means that many patients experience significant delays before receiving a definitive diagnosis, during which time their disease may progress unchecked.

Current Therapeutic Approaches

For IPF, two antifibrotic agents, pirfenidone and nintedanib, are approved therapies. These drugs do not reverse existing fibrosis but have been shown to slow the rate of decline in lung function, as measured by forced vital capacity (FVC). Pirfenidone, an oral agent, is thought to exert its effects through multiple mechanisms, including inhibition of TGF- β stimulated fibroblast proliferation and collagen synthesis. Nintedanib, a tyrosine kinase inhibitor, targets multiple receptors involved in profibrotic signalling pathways, including PDGF, FGF, and VEGF receptors. Both agents have demonstrated efficacy in reducing the annual rate of FVC decline compared to placebo in clinical trials. The initiation of these therapies is typically recommended as soon as an IPF diagnosis is confirmed, highlighting the urgency of timely intervention. But these treatments come with their own set of adverse effects, including gastrointestinal disturbances (nausea, diarrhoea) for nintedanib and photosensitivity and liver enzyme elevations for pirfenidone, which require careful monitoring and patient education. The challenges in managing ILD extend beyond just drug selection, encompassing patient education and symptom management.

Beyond antifibrotic medications, supportive care is a cornerstone of pulmonary fibrosis management. This includes oxygen therapy for hypoxemia, pulmonary rehabilitation to improve exercise tolerance and quality of life, and management of comorbidities such as gastroesophageal reflux disease, pulmonary hypertension, and sleep apnoea. Lung transplantation remains the only definitive treatment for end-stage pulmonary fibrosis, but it is an option available to only a select group of patients due to strict eligibility criteria, donor organ availability, and the complexities of post-transplant care. The decision-making process for transplantation is intricate, requiring careful consideration of patient age, overall health, and psychosocial factors. For many patients, the focus remains on slowing disease progression and managing symptoms to maintain the best possible quality of life for as long as possible. The perspectives of patients and pulmonologists often highlight the significant care gaps that persist.

Navigating the Hurdles to Faster Treatment

Several factors contribute to the delays in initiating treatment for pulmonary fibrosis. One significant hurdle is the non-specific nature of early symptoms, which can mimic more common respiratory conditions like asthma, COPD, or chronic bronchitis. This often leads to misdiagnosis or delayed referral to a specialist. Primary care physicians, while adept at managing common ailments, may not always have the immediate expertise or resources to identify rare interstitial lung diseases. The Oxford Handbook of Respiratory Medicine can be a valuable quick reference for GPs encountering such complex cases.

Another challenge lies in the complexity of the diagnostic process itself. As discussed, the need for HRCT imaging and multidisciplinary review means that patients must navigate multiple appointments and specialist consultations. This can be particularly burdensome for patients in rural areas or those with limited access to specialized medical centres. The logistical challenges of coordinating care across different specialties and institutions can introduce significant delays, sometimes extending for months. During this period, the disease continues its relentless progression, potentially leading to irreversible lung damage that could have been mitigated by earlier intervention. The lack of widespread awareness among the general public and even some healthcare professionals about pulmonary fibrosis also contributes to delayed presentation and diagnosis.

The differential diagnosis of ILD is broad, encompassing over 200 distinct conditions. Distinguishing IPF from other fibrotic ILDs, such as those associated with connective tissue diseases or chronic hypersensitivity pneumonitis, is critical because treatment strategies differ. For example, some non-IPF fibrotic ILDs may respond to immunosuppressive

therapies, which are not indicated for IPF. Misdiagnosis can lead to inappropriate treatment, further delaying effective management and potentially causing harm. The careful interpretation of HRCT scans and pathology specimens requires specialized expertise, which is not uniformly available across all healthcare settings. This highlights the importance of centralizing ILD care in specialized centres where such expertise is concentrated.

The financial burden associated with diagnostic procedures and long-term antifibrotic therapy also presents a barrier for some patients, even in systems with universal healthcare. While the drugs are reimbursed in many European countries, co-pays or indirect costs can still be substantial. Access to pulmonary rehabilitation programs, which are vital for symptom management and quality of life, can also be limited by geographical availability and insurance coverage.

Addressing these systemic and practical barriers requires a concerted effort from healthcare policymakers, professional organizations, and patient advocacy groups to streamline pathways, improve awareness, and ensure equitable access to care. The goal must be to shorten the time from symptom onset to treatment initiation, thereby maximizing the potential benefits of available therapies.

CLINICAL IMPLICATIONS

The persistent delays in diagnosing and treating pulmonary fibrosis are a clinical failing we can no longer afford to ignore. Every month a patient waits for a definitive diagnosis is a month of irreversible lung damage. GPs are on the front line, and while the disease is rare, a high index of suspicion for unexplained dyspnoea or chronic cough, especially with Velcro crackles, is non-negotiable.

Specialist centres must streamline their referral pathways. The multidisciplinary team meeting is essential, but the administrative burden should not translate into months of waiting. We have effective, albeit not curative, therapies in pirfenidone and nintedanib; their benefit is directly tied to how early they are initiated. The current system often means patients are starting these drugs with already significant lung damage.

Industry has a role beyond drug development. Education campaigns targeting both the public and primary care providers could significantly shorten the diagnostic interval. The economic argument for earlier intervention, reducing hospitalizations and improving quality of life, should resonate with health policymakers.

The onus is on the entire healthcare system to recognize that pulmonary fibrosis is a time-sensitive condition. We need to move beyond a reactive approach to one that prioritizes rapid diagnosis and immediate access to specialist care and disease-modifying therapies. The current trajectory is simply not good enough for our patients.

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