Do you recognize the signs of ILDs?

Time is of the essence in diagnosing interstitial lung diseases (ILDs) like pulmonary fibrosis, but the signs can be difficult to recognize. Delays in diagnosis, ranging from 1 year to more than 5 years, can have devastating effects on patient outcomes.

With ILDs affecting 400,000 people in the US, the likelihood of encountering a patient with an ILD is rare—meaning ILDs are often overlooked as a potential diagnosis.

Hoyer et al reported 41% of patients in their study had at least one alternate diagnosis before IPF diagnosis—most commonly Cardiac disease and Obstructive lung disease.


UBIQUITOUS SYMPTOMS—
WHY ILDS ARE DIFFICULT TO DIAGNOSE

- Patients may be asymptomatic in early stages.
- Symptoms mirror other common conditions.
- ILDs are relatively rare.

“While interstitial lung diseases do not affect a substantial amount of the population, those touched by the disease are impacted tremendously. Any delay in receiving a diagnosis is time that could be dedicated to finding a treatment therapy that can improve their quality of life.”

—Andrew H. Limper, MD, FCCP, Pulmonologist
More Than A Cough™

SHORTENING TIME TO DIAGNOSIS

There is no cure for ILDs, but early diagnosis and treatment can slow progression and lessen the severity of symptoms, allowing patients to live longer with a higher quality of life. Your expertise is essential.

Know risk factors beyond dyspnea and cough:
1. Occupational or environmental exposures
2. Autoimmune disease
3. Sleep disorders
4. Weight loss
5. IPF comorbidities
6. Familial/genetic association

Leverage the right tests to probe for risk factors; understand implications of the findings.

Know when to extend the care team, bridging specialties through referrals or ILD centers to provide additional options for treatment.

Maximize patient quality of life through oxygen therapy, pulmonary rehabilitation, mental health assessment, medication, and nutrition counseling.

Ready to learn more? Scan the QR code to access our web page with links to resources designed to help you advance ILD patient care.

State of Practice: Factors Driving Diagnostic Delays in Idiopathic Pulmonary Fibrosis

Idiopathic pulmonary fibrosis (IPF) is a progressive fibrosing interstitial lung disease (PF-ILD) characterized by a decline in lung function, worsening of symptoms and health-related quality of life, and early mortality. The median survival of patients with IPF is generally reported to be between 3 to 5 years from diagnosis. This range, however, is based on older observations that may not reflect current best practices and newer therapies.

DELAYS IN DIAGNOSIS ARE COMMON IN IPF

Time is of the essence in diagnosing IPF; however, the literature suggests that delays in establishing an IPF diagnosis are common, generally ranging from 1 year to more than 5 years. The non-specificity of IPF symptoms, especially in early disease, and the fact that it is a rare disease most likely contribute to the delay in diagnosis, especially among primary care providers (PCPs) who may be inclined to follow assessment pathways for more common cardiac or respiratory conditions.

Time lost is lung lost in IPF. Delays in diagnosis can result in irreversible decline in lung function and may eliminate some therapeutic options. Reducing the time to diagnosis has the potential to dramatically impact patients’ lives and clinical outcomes, including allowing earlier initiation of medications that slow disease progression, provision of supportive care (eg, pulmonary rehabilitation), palliative care, treatment of comorbidities, and referral for lung transplant evaluation or enrollment in clinical trials.

CHEST and Three Lakes Foundation, a philanthropic organization dedicated to increasing awareness and finding solutions for the pulmonary fibrosis community, partnered to explore factors that drive delays in the diagnosis of IPF and identify educational interventions that could mitigate these delays.

PCP AND PULMONOLOGIST SURVEYS

CHEST Analytics conducted two online surveys: one among PCPs (n=156 family medicine and n=150 general internal medicine); and one among general pulmonologists (n=100) working in a community-based nonacademic settings.

The objectives of this research were to:
1. Understand how providers approach the evaluation of patients who present with nonspecific symptoms consistent with IPF, as well as how they evaluate patients with newly diagnosed interstitial lung disease who are suspected of having IPF.
2. Assess provider knowledge and attitudes regarding the evaluation and management of IPF.
3. Identify motivators and barriers to use of specialized resources available to assist in the diagnosis and management of patients with IPF.

PCP respondents were recruited from syndicated commercial research panels, and pulmonologist respondents were recruited from CHEST Analytics database. Respondents were paid a market rate incentive for their time and effort in taking the survey. Data were collected during April 11-May 16, 2022.

Bridging Specialties™: Timely Diagnosis for ILD Patients is a collaborative initiative between the American College of Chest Physicians (CHEST) and Three Lakes Foundation that brings together pulmonologists and primary care physicians to define a clearer clinician-guided approach to diagnosis for ILDs like PF and IPF. The findings above were drawn from a clinician survey that was conducted as a part of this initiative. Read a summary of the findings on the following pages, or scan the QR code to connect to the Clinical Perspectives white paper containing the full survey results.
KEY SURVEY FINDINGS

PCPs
Survey results point to gaps in knowledge and clinical behavior that contribute to significant delays in diagnosis, limitation of treatment options, and worsening quality of life.

In response to reviewing a sample case (woman, age 55, presenting with the following nonspecific symptoms: shortness of breath during moderate exertion, cough, exhaustion, and difficulty sleeping), PCPs indicate the following clinical behaviors:

- Almost half (46%) evaluate/refer patients for cardiac issues as the leading differential diagnosis for this patient.
- A substantial minority (39%) say they would proceed with modification of current medications over evaluating symptoms for this patient if she is using inhaled therapy or has a respiratory diagnosis.
- Only a minority of respondents say that spirometry (47%), full PFT (40%) diffusion capacity (11%) would be part of their initial workup.
- Reported items most likely to be included when taking a patient's history frequently do not include early symptomatic clues of IPF (presence of prescription-drug induced lung disease, various household exposures, review of autoimmune conditions in presence of joint disease).
- Less than half of PCP respondents (46%) would refer patient for pulmonary consult if initial results showed pulmonary findings; 54% would retain the patient for initial testing or attempt treatment.
- Only 62% would order HRCT scan if chest radiograph is unremarkable but shows lower lobe opacity; and only 50% would order if examination reveals crackling upon inspiration.

Pulmonologists
While pulmonologists generate strong knowledge scores regarding guidelines for IPF diagnosis, reported use of anti-fibrotics in confirmed patients is far from universal. Most (65%) say they routinely prescribed anti-fibrotics only after the patient shows progression.

POTENTIAL OPPORTUNITIES TO REDUCE TIME TO IPF DIAGNOSIS

A key takeaway from both the PCP and pulmonology surveys is that no one factor or set of factors drives delays in diagnosis of IPF; thus, education needs to be multifaceted and targeted to those factors that have the potential to most significantly impact clinician behavior. The full report outlines areas where educational interventions have the potential to reduce delays in diagnosing IPF and identifies opportunities to create bridges between the PCP and pulmonology communities. You can learn more at Clinical Perspectives: State of Practice: Factors Driving Diagnostic Delays in Idiopathic Pulmonary Fibrosis.

References