Regional IPF Management

Experts Weigh In

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Disease State Overview

Idiopathic pulmonary fibrosis (IPF) is a chronic, progressive, irreversible, restrictive lung disease that culminates in respiratory failure and death.[1] It is recognizable by the progressive decline of lung function in adults over the course of 2 to 5 years, with increasing symptoms over time.[1-3] The median 5-year survival time for IPF, 3.8 years, is worse than that of many cancers and is the worst among idiopathic interstitial pneumonias (Figure 1).[2,4–6] The 5-year survival rate for IPF is approximately 30%.[7,8]

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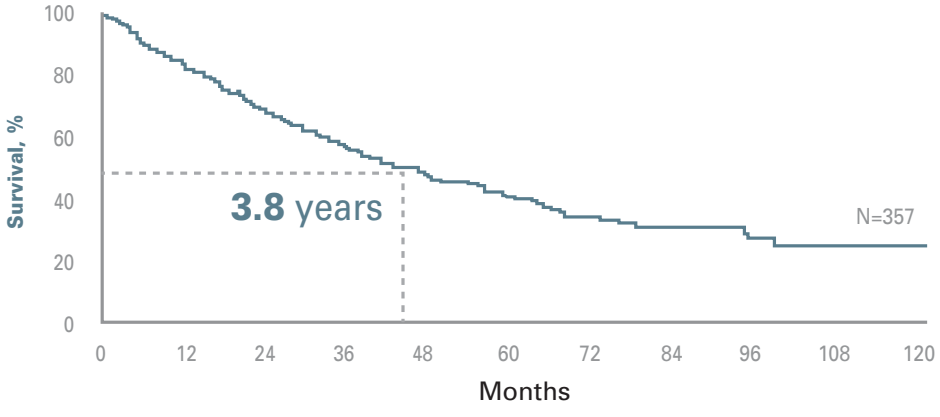
The incidence of IPF in adults age 65 years and older in the United States is about 42,000 cases a year, and the prevalence has been increasing steadily from 202.2 cases per 100,000 people in 2001 to 494.5 cases per 100,000 people in 2011.[2,9]

Figure 1. Median Survival Time for Patients with IPF[4]



▸ Median survival time is 3.8 years[4]

Survival of patients with IPF from time of initial pulmonary function test (lung transplantation recipients excluded). Adapted from Nathan et al (2011).

The cause of IPF is unknown, but there are several risk factors that may contribute to IPF, including smoking, environmental pollutants, viral infections, gastroesophageal reflux disease, and possible genetic factors, such as gene polymorphisms or deletions.[3,10]

Understanding IPF in Texas:

- IPF is more prevalent in white men age 50 years and older, and smoking is one of the biggest risk factors for IPF

State	Percent White[11]	Percent Men Age 50 Years and Older[12]	Percent Smokers[13]
Texas	75	27	15

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"It is important to counsel and empower the patient. Just because we have no way of predicting the course of the disease right now doesn't mean we shouldn't do anything, because once lung function is lost, we can't get it back. It's important to tell patients who are early in the disease and asymptomatic that they will progress eventually and that we should be thinking about management options such as treatment, oxygen use, pulmonary rehab, or palliative care."

Dr Nambiar

"I show my patients on a graph how some do in terms of being initially asymptomatic to symptomatic and how unpredictable IPF is. I highlight that it is very difficult to identify where they are going to be at a given point in time. It is always better to manage the disease with options like pharmacological treatment, supplemental oxygen, pulmonary rehab, or emotional support even though they may not have symptoms initially. It is important to understand that we cannot predict who's going to live longer or shorter."

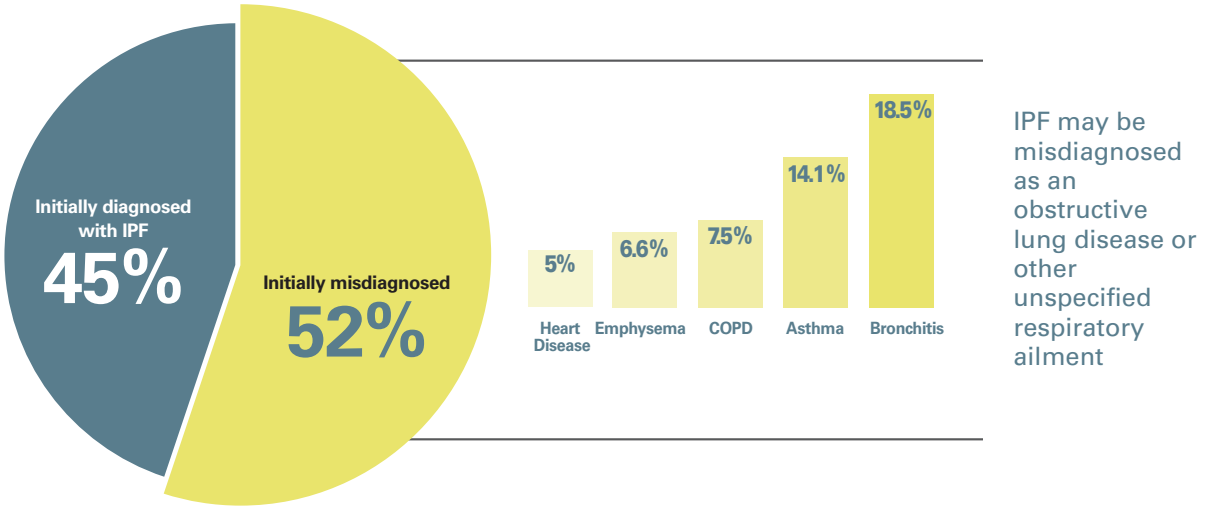
Dr Patel

Prompt Diagnosis is Key for IPF

Because the damage IPF causes is irreversible, prompt and accurate diagnosis is critical for earlier initiation of intervention.[1]

Figure 2. Misdiagnosis of IPF is Common[14]

2004 Coalition for Pulmonary Fibrosis Basic Research Questionnaire of Patients with IPF (N=1448)



In addition, a patient's symptoms do not always reflect the underlying disease progression.[15] One survey demonstrated that more than half of patients experienced at least a 1-year delay between onset of symptoms and diagnosis, and more than a third of patients reported seeing three or more physicians before their IPF diagnosis was established.[14] This gap between the first appearance of symptoms and diagnosis may be a contributor to the short prognostic time in IPF.

A timely diagnosis of IPF is the first step to early management, which is important because loss of lung function is irreversible.[1] During a physical examination of a patient, it is important to suspect IPF in any patient who is 50 years of age or older and has any of the following signs and symptoms[3,16]:

- Inspiratory, bibasilar "Velcro-like" crackles
- A chronic cough
- Dyspnea on exertion
- Fatigue
- Digital clubbing

Refer to the 2011 American Thoracic Society (ATS) guidelines for recommendations on a diagnostic algorithm for IPF.[3]

Pulmonary Function Tests and Oxygen Saturation

A patient's oxygen saturation levels and how they perform on pulmonary function tests are important criteria to consider when diagnosing IPF. Table 1 shows the most common and useful tests. A physician should consider IPF as a diagnosis if a patient shows a reduced total lung capacity (TLC) and/or forced vital capacity (FVC), a forced expiratory volume in 1 second (FEV_1)/FVC ratio that is consistent with a restrictive pattern, a reduced diffusing capacity of the lungs for carbon monoxide ($D_{L_{CO}}$), and low oxygen saturation with exertion (6-minute walk test) or at rest.[17,18]

Table 1. Pulmonary Function Tests

Lung Volume	Flow	Gas Exchange
• TLC	• FEV_1 • FVC • FEV_1/FVC ratio	• $D_{L_{CO}}$
Oxygen Saturation		
• With exertion (6-minute walk test)	• At rest	

Another important tool for diagnosing IPF is high-resolution computed tomography (HRCT). A pattern of usual interstitial pneumonia (UIP) is indicative of IPF when seen on an HRCT. Signs of definitive UIP include[3,19]:

- Subpleural, basal predominance
- Reticular abnormality
- Honeycombing with or without traction bronchiectasis
- Absence of features listed as inconsistent with UIP pattern

For more detail on what to see on HRCT for IPF, refer to the 2011 ATS guidelines.[3]

Importance of Early Management in IPF

Initiating a management plan at diagnosis is important because loss of lung function is irreversible and cannot be restored.[1,7] The timeline for damage is unpredictable,[15] so the goal of management is to conserve lung function for as long as possible.[1] Despite appropriate management, patients will continue to have declines in FVC as a natural progression of the disease.[1,3]

Considerations for Managing IPF

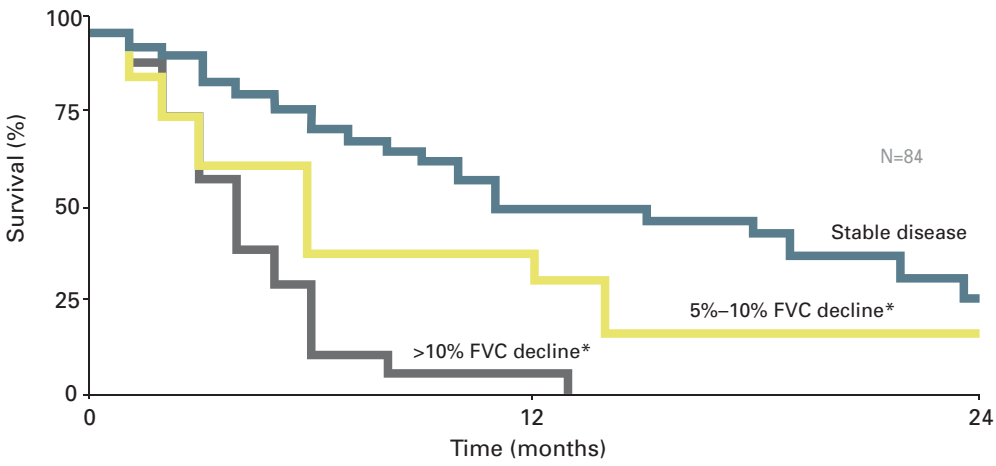
Over time, IPF causes irreversible decline in lung function.[4] Early in the disease, the lung fibrosis is heterogeneous, with fibrotic tissues adjacent to normal lung tissue. However, as the disease progresses and the fibrosis spreads, lung function becomes severely compromised.[7]

Because there is no cure for IPF, slowing the decline of lung function is a crucial goal of IPF management; however, the rate of decline can vary greatly from patient to patient as well as within the same patient.[3,4]

A vital goal for physicians to consider when managing a patient with IPF is the importance of slowing the progression of the disease.

The ATS, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Association guidelines recommend measuring FVC and $D_{L_{CO}}$ during routine monitoring at least every 3 to 6 months, with flexible monitoring for patients who have progressive dyspnea or symptoms that suggest a more rapidly progressing disease (Figure 3; Table 2).[3]

Figure 3. The Relationship Between FVC Decline and 2-Year Progression-Free Survival[20]



A 5%–10% or >10% decline in FVC is associated with worse prognosis than more stable disease (P<0.005)

*Serial change at 6 months in patients with IPF.
Adapted from Zappala et al (2010).

Pulmonologists should be aware of the guidelines and practices for earlier diagnosis and management of IPF.

Table 2. Monitoring the Clinical Course of IPF[3,21]

How to monitor: Follow-up process, most relevant tests to be conducted, frequency of visits	• Every 3-6 months • Monitor changes in symptoms, comorbidities • Need for oxygen supplementation

"In our practice, the patient's management is guided by a few parameters. First, we monitor the pulmonary function tests frequently to see if there is any deterioration. Second, we look at the clinical deterioration with the 6-minute walk test and the distance a patient can tolerate. Third is the oxygen saturation: We want to make sure that the oxygen assessment is done regularly, especially when therapy is initiated. And fourth is the CT scan of the chest to see if there's any progression of the disease."

Dr Surani

"Symptoms are not necessarily an accurate measure of disease progression. We need to dig deeper in patients' history. If they have decreased their activity because of symptoms, they are not going to report dyspnea on exertion. Symptoms, along with the imaging or PFTs, can be helpful in assessing disease progression, particularly when the walk distance has gone down."

Dr Patel

Management Strategies for IPF

The goal of IPF management is to conserve lung function for as long as possible.[1] A management plan should be initiated for all patients diagnosed with IPF, even if their symptoms appear to be stable.[15,22] It is important to keep in mind that progression of fibrosis may not present as worsening of symptoms.[15] Table 3 lists some management strategies to consider.

Table 3. Management Strategies

- The ATS Guidelines contain additional information about the following management strategies:

Pharmacologic strategies[23]:

- FDA-approved IPF-specific therapy[23,24]
- Management of comorbidities[15,24]
- Symptom management[24]
- Clinical trials enrollment[24]

Nonpharmacologic strategies[3]:

- Supplemental oxygen
- Pulmonary rehabilitation
- Disease education
- Psychosocial support
- Evaluation for lung transplant
- Palliative care

It is important to follow patients with IPF frequently and regularly and monitor disease progression.[3]

Continued Management of IPF

It is important to follow patients with IPF frequently and regularly and monitor disease progression.[3] Symptoms that initially appear stable can progress at any time, and progression is both unpredictable and unavoidable.[15] The 2011 Clinical Practice Guidelines recommend monitoring pulmonary function tests, changes in symptoms, and changes in oxygen requirements every 3 to 6 months.[3]

"When it comes to comorbidities, I think what's most important is identifying them. So we screen patients for comorbidities, not just at the initial contact with us, but even at their follow-up visits, and we've got protocols to help determine if a patient's decline is due to one of their comorbidities or progression of IPF."

Dr Isiguzo

"When we talk to patients, we must acknowledge that we're dealing with a disease that's as bad as common cancers. It's very important to be realistic but still provide management options. We can try to slow down their disease, but the disease will still progress. Educating and empowering patients to be aware of that information is important, but it is also important that they realize that, even though the disease does progress, we're not going to give up on managing them."

Dr Nambiar

"PFTs are a key measure to assess IPF. Others would be the 6-minute walk test, dyspnea on exertion, or a declining oxygen level. These parameters help us guide a patient's management."

Dr Surani

"My number one management goal is to try and slow down the disease progression, because that is all that I am equipped to do with what is available to me, aside from a lung transplant. My other goal is to provide emotional and psychological support to give the patient the best quality of life possible."

Dr Isiguzo

Communicating with Patients About Their IPF and Management Plan

When patients present with breathlessness, crackles, or cough, it is important for physicians to account for clinical history and demographics of the patient to determine whether a chest x-ray or HRCT scan is appropriate.

Because IPF is unfamiliar to most patients, receiving a diagnosis of IPF can leave them confused and overwhelmed, so a carefully considered communication plan is needed. In addition, because progression of IPF can be quick and without warning, some patients may have little time to adjust to their diagnosis.[25] It is important that physicians communicate with patients about their perceptions of their prognosis and how to manage the disease. Physicians can discuss available management options and encourage patients to start an appropriate management plan.[3]

Some points to consider when communicating with patients include[23,25,26]:

- Acknowledging the impact of IPF on both the patient and his or her caregiver
- Helping the patient understand the unpredictability of the disease and that every patient's disease is different
- Educating the patient on management options
- Helping patients understand the potential benefits of continuing treatment and managing their comorbidities

"Communication builds a relationship with the patients. The better the relationship you have with your patients, the more compliant they may be. It's important to talk to patients about the unpredictability of IPF. Because IPF is terminal, we make sure patients know they should come see us as soon as possible when they feel they're declining in function. It's also important to let patients vent and tell us what and how they feel. Otherwise, we fail in communication. It's very important to have some empathy and help manage the disease in whatever way we can to support these patients."

Dr Surani

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