

# PULMONARY FIBROSIS

## INNOVATION CHALLENGE

THREE LAKES  
FOUNDATION



### **Background: Pulmonary Fibrosis**

#### **What is PF?**

Pulmonary fibrosis (PF), a group of diseases within the broader family of interstitial lung diseases (ILD), is characterized by progressive scarring of the lungs. There are about 200 different forms of PF, the most common being idiopathic pulmonary fibrosis, or IPF. For some forms of PF, doctors are able to identify the cause; IPF has no identifiable cause.

#### **What is the incidence?**

The worldwide incidence of PF is climbing. Whether more patients are presenting with PF or more doctors are detecting is unclear.

- Worldwide, the number of people estimated to be living with PF tops 3 million.
- In the U.S. alone, there are more than 200,000 people with PF. Another 50,000 are diagnosed each year.

#### **How serious is it?**

Every year in the U.S., more people die from pulmonary fibrosis than from breast cancer. The most dire diagnosis is IPF, which has an average lifespan following diagnosis of 3-5 years.

#### **Who are the patients?**

Generally, this disease affects slightly more men than women, and the average age of someone with IPF is over 60 — but younger people get PF, too.

PF can develop in patients who:

- Have rheumatoid arthritis, scleroderma, sarcoidosis or Sjogren's disease
- Have been treated with certain medicines or other therapies
- May be sensitive to certain environmental triggers that can harm the lungs

### **What are the challenges to diagnosing PF?**

Because PF symptoms mimic those of other, much more common illnesses, the average path to an accurate diagnosis is delayed and leads to frustration and anxiety. Patients don't recognize the seriousness of their symptoms and may not act quickly. Doctors must rule out common causes first, meaning that patients sometimes don't get referred to specialists as early as they should. Evaluating CT scans in PF is challenging. The communication between radiologists and clinicians regarding incidental findings isn't always straightforward. An accurate diagnosis is facilitated by a multi-disciplinary discussion between pulmonologist, radiologists, rheumatologist and pathologists, but isn't available in all medical centers. About 1/3 of initial IPF diagnoses are incorrect. Because of these and other reasons, many patients go undiagnosed for about 18-24 months following initial symptoms.

### **What are the challenges to treating PF?**

Currently, the definitive treatment for pulmonary fibrosis is a lung transplant. But, not everyone qualifies for the surgery, recovery is difficult, and there is a very limited supply of lungs. Some forms of PF have had available therapies on the market for years. Two therapies for IPF were approved in the USA in 2014. Since then, many new drugs have entered the pipeline. In addition to medicine, a proper diet, exercise (including pulmonary rehab), infection avoidance, removal of environmental triggers, social-emotional support and supplemental oxygen therapy are important for symptom relief.

Most patient care takes place outside of the clinic; patients participate in the management of their disease. Depression and anxiety, along with challenging physical symptoms, make this difficult. Some therapies include significant side effects. Information and guidance about the disease, nutrition, infection control, mental health, and managing symptoms can be hard to find. In the U.S., there are not nearly enough pulmonary rehabilitation experts and facilities to meet patient demand. The many obstacles surrounding prescribing, receiving, and using oxygen equipment are daunting. Finally, traveling both locally and across distances is increasingly problematic as supplemental oxygen needs grow.

### **Where are the opportunities for innovation?**

Pulmonary fibrosis diagnosis and care is still a fairly new frontier with many opportunities to provide methods for patients and caregivers to improve their quality of life. As new therapies that slow the progression of IPF extend patient lifespans, the need for long-term oxygen therapy, pulmonary rehab and additional treatments increases. Computer-based algorithms, machine learning, better understanding of mental health, the internet, telehealth, relevant innovations from other diseases, research consortiums and new interest from major industry players bring endless possibilities to entrepreneurs and seasoned companies alike.