Pulmonary Fibrosis FOUNDATION

pulmonaryfibrosis.org

ABOUT PULMONARY FIBROSIS

Pulmonary fibrosis (PF) is a family of more than 200 different lung diseases that all look very much alike. The PF family of lung diseases falls into an even larger group of diseases called “interstitial lung diseases.” The word “pulmonary” means “lung” and “fibrosis” means scar tissue – so in its simplest sense, pulmonary fibrosis means scarring in the lungs.

In PF, scar tissue builds up in the walls of the air sacs of the lungs, and eventually the scar tissue makes it hard for oxygen to get into the blood. Low oxygen levels (and the stiff scar tissue itself) can cause shortness of breath, particularly when walking and exercising.

Pulmonary fibrosis is a progressive disease, which means it tends to worsen over time. However, every individual diagnosed with pulmonary fibrosis has a unique experience with the disease and there is no “standard” or expected clinical course. Some people with PF remain stable for extended periods of time; others may experience a rapid progression of symptoms; while others may experience a stepwise deterioration over time, fluctuating between periods of stability and worsening symptoms. Therefore, PF treatment strategies are highly individualized, based on a person’s medical history. With no known cure, the disease is often fatal within three to five years of diagnosis. The good news for people living with PF is that there are treatments designed to specifically manage the symptoms of the disease and researchers are studying new ways to halt its progression.

SYMPTOMS OF PULMONARY FIBROSIS

The most common symptoms of PF are cough and shortness of breath. Symptoms may be mild or even absent early in the disease process. As the lungs develop more scar tissue, symptoms worsen. Shortness of breath initially occurs with exercise, but as the disease progresses it is not uncommon to become breathless during everyday activities, such as showering, getting dressed, speaking on the phone, or even eating.

Other common symptoms of pulmonary fibrosis include:

- Chronic dry, hacking cough
- Fatigue and weakness
- Discomfort in the chest
- Loss of appetite
- Unexplained weight loss

CAUSES OF PULMONARY FIBROSIS

There are a number of potential causes of pulmonary fibrosis, including exposure to airborne toxins, such as asbestos or mold; undergoing radiation treatment; and taking some specific medicines, including those that treat cancer, heart disease and bacterial infections. Genetics are also thought to play a role in PF. Sometimes doctors are able to identify a specific cause of the disease, but it is also common to end up without an answer, despite completing a large number of medical tests. When a cause is not found, or the disease occurs spontaneously, it can be called “idiopathic.” One recent study estimates idiopathic pulmonary fibrosis (IPF) affects 1 out of 200 adults over the age of 65 in the US. Approximately 50,000 new cases are diagnosed each year and as many as 40,000 Americans die from IPF each year.

TREATING PULMONARY FIBROSIS

Many people think there are no treatments for PF. In the past, this may have been true, but doctors do have a number of ways to treat the symptoms of PF, and help keep those with the disease active and healthy.

These treatments include:

- Supplemental oxygen
- Pulmonary rehabilitation
- Lung transplantation
- Preventive measures, such as flu and pneumonia vaccines
- Optimizing weight and exercise
- Medications to manage symptoms

230 East Ohio Street, Suite 500, Chicago, Illinois 60611 844.TalkPFF (844.825.5733) pcc@pulmonaryfibrosis.org info@pulmonaryfibrosis.org

Pulmonary Fibrosis
FOUNDATION
pulmonaryfibrosis.org

Some patients may benefit from disease-specific therapy for IPF:

- Nintedanib is an anti-fibrotic drug that is approved to treat IPF in the United States. In clinical trials, nintedanib has been shown to slow the decline in lung function in mild-to-moderate IPF.
- Pirfenidone is an anti-fibrotic and anti-inflammatory drug that is approved to treat patients in the US, EU, Canada and Asia. In clinical trials, pirfenidone has been shown to slow the progression of mild-to-moderate IPF.
- Prednisone is an anti-inflammatory therapy that can help some people with inflammation in the lungs.

ABOUT THE PULMONARY FIBROSIS FOUNDATION

The Pulmonary Fibrosis Foundation mobilizes people and resources to provide access to high quality care and leads research for a cure so people with pulmonary fibrosis will live longer, healthier lives. The PFF collaborates with physicians, organizations, patients, and caregivers worldwide. The Pulmonary Fibrosis Foundation has a four-star rating from Charity Navigator and is a Better Business Bureau accredited charity. The *PFF Summit 2017*, its fourth biennial international health care conference, will be held November 9-11, 2017 in Nashville, Tennessee. For more information, visit pulmonaryfibrosis.org or call 844.TalkPFF (844.825.5733) or +1 312.587.9272 from outside of the US.