Progressive Pulmonary Fibrosis and Progressive Fibrotic Interstitial Lung Disease

Pulmonary Fibrosis Foundation™

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What is progressive pulmonary fibrosis?

Interstitial lung diseases (ILD) are a large category of diseases that have either fibrosis, inflammation, or both, in the walls of the air sacs in the lung. There are over 200 specific diagnoses that are categorized as ILDs. Fibrosis, or tissue scarring, can be present in many types of ILDs, and when the fibrosis worsens over time, this is referred to as "progressive pulmonary fibrosis" (PPF) and may also be termed "progressive fibrotic ILD" (PF-ILD).

How is PPF diagnosed?

PPF is not a specific diagnosis, rather a disease behavior that can occur in many types of ILD. When a patient has ILD, their doctor or other healthcare provider will monitor for signs of progression, with clinical exam, pulmonary function testing (PFT), and imaging with a high-resolution CT scan (also known as CAT scan), which gives detailed images of the lung. PPF is defined by worsening respiratory symptoms combined with worsening PFT results or increased signs of fibrosis on CT scan over time. Healthy lung tissue looks nearly black on a CT scan while scar tissue and inflammation appear grey or white, indicating damage to the lung tissue.

What are the symptoms of PPF?

Early in the disease, many people with ILD have minimal or mild symptoms, including shortness of breath or cough. Some people may experience no symptoms early in the disease. As the fibrosis progresses, breathlessness during exercise and daily activities become common. Not everyone may exercise but everyone has daily activities to complete. Fatigue, depression, and anxiety are also commonly experienced by people living with PPF.

How is PPF treated?

The treatment of PPF depends on the specific underlying ILD diagnosis. Pirfenidone and nintedanib have been studied in certain ILDs where PPF has developed, and may be effective to slow disease progression.

Lung transplantation is an appropriate treatment for some people who have developed PPF. Early evaluation for lung transplant is important because the process involves a series of appointments to provide the patient with information about transplantation, and to determine if they are an appropriate candidate.

Pulmonary rehabilitation, supplemental oxygen, smoking cessation, and routine vaccinations (such as influenza and pneumonia vaccination) are important parts of living with a PPF. You can learn more about supplemental oxygen at: **pulmonaryfibrosis.org/oxygen**. You can learn more about pulmonary rehabilitation at **pulmonaryfibrosis.org/pulmonaryrehab**.

What causes PPF?

ILDs of all kinds can result in progressive tissue scarring. There are many types of ILD that have known causes, such as chronic hypersensitivity pneumonitis, connective tissue disease-related ILD, occupational diseases (pneumoconioses), and drug-induced ILD. There are also many types of ILD that do not have a known cause. Idiopathic pulmonary fibrosis (IPF) is the most common form of ILD and has no known cause.

What is my prognosis?

PPF is defined by progression, which means that fibrosis builds up over time, gradually causing worsening breathlessness and the need for increasing amounts of oxygen. Eventually, lung failure (medically called "respiratory failure") can develop, which is a life-threatening condition. It is important for you to know that there is no way to predict how long someone who develops PPF will live. Monitoring your ILD and detecting PPF early is important. Some patients will progress more slowly, and others will develop respiratory failure more quickly, with some becoming very ill within months or just a few years after their diagnosis. No one can predict exactly how long you will live with PPF. Everyone is different. Your doctor can give you more detailed information about your prognosis.

Are there experimental therapies available?

Many pulmonary fibrosis research studies are underway and enrolling participants. Clinical trials take different forms and are not always in the form of a "treatment." You can learn more and search for research studies closest to you on our PFF Clinical Trial Finder: **trials.pulmonaryfibrosis.org**. Before enrolling in a clinical trial, discuss your participation with your doctor.

Sources

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