Idiopathic Pulmonary Fibrosis

Pulmonary Fibrosis Foundation™

pulmonaryfibrosis.org

What is idiopathic pulmonary fibrosis?

There are more than 200 types of interstitial lung diseases (ILD), which are characterized by varied amounts of inflammation, scarring, or both, that damage the ability of the lung to absorb oxygen from the air. Pulmonary fibrosis (PF), means scarring of the lung, and can be seen in many types of ILD. More than 250,000 Americans are living with PF and ILD. Difficult to diagnose, PF and ILD can be debilitating and, in some cases, incurable. Causes of PF and ILD include the use of certain medications, radiation to the chest, autoimmune diseases, environmental, and occupational exposures. Idiopathic pulmonary fibrosis, which has no known cause, is one of the most common forms of PF. The prevalence of PF and ILD is on the rise with more than 50,000 new cases diagnosed annually.

What are the symptoms of IPF?

Early in the disease, most people with IPF will have no symptoms or might have a bothersome cough. As the disease progresses, breathlessness during exercise and daily activities becomes common. Fatigue, depression, and anxiety are also commonly experienced by people living with IPF.

What causes IPF?

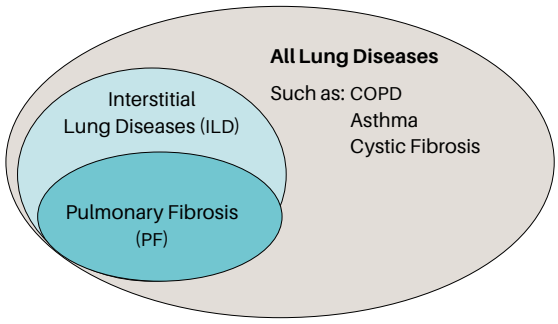
Although the word "idiopathic" means "of unknown cause," we have learned a lot about the causes of IPF. Former and current smokers are more likely to develop IPF than those who have never smoked. A family history of pulmonary fibrosis is also a risk factor, as are certain genes, such as MUC5B, TERT, TERC, DKC1, RTEL1, AKAP13, DSP, FAM13A, DPP9, and TOLLIP. Some evidence suggests that gastroesophageal reflux (acid reflux, or heartburn), certain viral infections, air pollution, and some exposures in the work place may be risk factors for IPF.

What makes it different from other forms of PF?

It is important first to understand that IPF falls into a large category of diseases called "interstitial lung diseases" or "ILD." There are over 200 types of ILD. Most ILDs have either fibrosis or inflammation (or both) in the walls of the air sacs. "Pulmonary fibrosis" (PF) is used to refer to any type of ILD in which fibrosis is present, and since most forms of ILD include fibrosis, most forms of ILD are also forms of PF. IPF is one type of PF. There are many others, some of

Table X

DISEASE	ABBREVIATON
Idiopathic pulmonary fibrosis	IPF
Idiopathic non-specific interstitial pneumonia	Idiopathic NSIP
Respiratory-bronchiolitis-associated ILD	RB-ILD
Desquamative interstitial pneumonia	DIP
Cryptogenic organizing pneumonia	COP or BOOP
Sarcoidosis	Sarcoid



TYPE OF PF	CLUES THAT DOCTORS USE
Drug-Induced	Prior or current use of amiodarone, nitrofurantoin, chemotherapy, methotrexate, or other drugs known to affect the lungs
Radiation-induced	Prior or current radiation treatment to the chest
Environmental (called hypersensitivity pneumonitis)	Exposure to mold, animals, or other triggers
Autoimmune (called connective tissue disease-related)	Joint inflammation, skin changes (particularly on the fingers and face), dry eyes or mouth, abnormal blood tests
Occupational (called pneumoconiosis)	Prior or current exposure to dusts, fibers, (called pneumoconiosis) fumes, or vapors that can cause PF (such as asbestos, coal, silica, and others)

which have known causes, such as chronic hypersensitivity pneumonitis, connective tissue disease-related ILD, occupational diseases (Pneumoconioses), and drug-induced ILD. In addition to IPF, there are many ILDs of unknown cause – meaning that there are many "idiopathic" ILDs other than IPF. Some are listed in Table X (previous page).

How is IPF diagnosed?

When a doctor or other healthcare provider suspects that a patient has ILD, they will perform tests that might include pulmonary function tests, a chest x-ray, blood work, and a high-resolution CT scan. Pulmonary function tests measure how much air the lungs can hold. The more scar tissue the lungs have, the less air they will hold. A doctor will also use a special kind of X-ray of the chest, called a high-resolution computed tomography (HRCT) scan, which gives detailed images of the lung. Healthy lung tissue looks nearly black on a CT scan (also known as CAT scan). Scar tissue and inflammation both appear grey or white. In many cases, a diagnosis of IPF can be made from these tests. In some cases, a lung biopsy may need to be performed.

My doctor said my CT scan (or biopsy) showed "UIP." What is that?

The term "usual interstitial pneumonia" or "UIP" refers to the visual pattern of scarring on a CT scan (image) or in lung tissue obtained from a lung biopsy. Although each person's situation is different, it is common to make a diagnosis of IPF when UIP is present on a CAT scan or on a biopsy, when no cause can be identified.

How is IPF treated?

Pirfenidone (Esbriet®, Pirfenex®, Pirespa®) and Nintedanib (OFEV®) are two drugs approved to treat IPF in many countries around the world. These drugs help to slow the progression of IPF.

Lung transplantation is an appropriate treatment for some people living with IPF. 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, routine vaccinations (such as influenza and pneumonia vaccination) are important parts of living with IPF.

You can learn more about supplemental oxygen at **pulmonaryfibrosis.org/oxygen.**

You can learn more about pulmonary rehabilitation at **pulmonaryfibrosis.org/pulmonaryrehab.**

What is my prognosis?

IPF is a progressive disease, 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 with IPF will live. You may have heard that the average survival of people living with IPF is only "3 to 5 years." This is an outdated statistic. With earlier diagnosis and better treatments available, many people live much longer than 3 to 5 years. Others will develop respiratory failure sooner than three years, 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 IPF. Everyone is different. Your doctor can give you more detailed information about your prognosis.

Are there experimental therapies available?

You can search for research studies closest to you on our PFF Clinical Trial Finder at **trials.pulmonaryfibrosis.org**.

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