understanding pulmonary fibrosis

[ENGLISH]

Pulmonary Fibrosis Foundation
What is pulmonary fibrosis?¹

Pulmonary fibrosis (PF) is a condition in which lung tissue becomes scarred, thickened, and stiff. The scarring is called fibrosis and makes it hard for a person to breathe.

How does pulmonary fibrosis affect the lungs?¹

The air sacs and blood vessels in the lungs work to deliver oxygen to the body.

In people with PF, the tissue inside and between the air sacs becomes scarred and stiff. The scarring and stiffness make it hard for oxygen to pass through the walls of the air sacs into the bloodstream.

As a result, the brain, heart, and other vital organs may not get the oxygen they need to work properly.

What causes pulmonary fibrosis?

PF can be caused by many things, some of the known causes of PF include:

Job-related and environmental factors. Being exposed to toxins and pollutants over the long term can damage the lungs. These toxins and pollutants may include silica dust, asbestos fibers, grain dust, and some bird and animal droppings.²,³,⁴

Radiation treatments. Radiation treatment for lung or breast cancer may also damage the lungs in some people.³

Certain medicines. Some chemotherapy medicines to treat cancer, some heart medicines, and some antibiotics are known to cause lung damage.¹

Certain medical conditions. Lung tissue can also be damaged by conditions such as tuberculosis, pneumonia, lupus, rheumatoid arthritis, sarcoidosis, and scleroderma.³,⁵,⁶,⁷,⁸

Genetics. Genes can play a role in the development of PF.²

When there is no known cause for PF, the disease is called idiopathic pulmonary fibrosis, or IPF.¹

For more information about the causes of PF, visit www.pulmonaryfibrosis.org/causesofpf.

What is IPF?

IPF is a specific form of PF; it is also reported to be the most common type of PF.²

Genetics may play a role since IPF may occur in several members of a family.²

Some researchers have also suggested that gastroesophageal reflux disease (GERD) may have a role in the development of IPF.²

Who gets idiopathic pulmonary fibrosis?

If you have been diagnosed with IPF, you are not alone.

As many as 132,000 people in the United States have idiopathic pulmonary fibrosis.⁹

IPF is most common in people 50 years or older.²
How does idiopathic pulmonary fibrosis make you feel?

IPF symptoms develop over time. The most common symptoms of IPF are:

- Shortness of breath
- A dry, hacking cough

Other symptoms that may occur over time include:

- Fast, shallow breathing
- Fatigue and weakness
- Aching muscles and joints
- Weight loss that cannot be explained
- Clubbing (widening and rounding) of the tips of the fingers and toes

What can happen to your body over time when you have idiopathic pulmonary fibrosis?

IPF can progress slowly or quickly. It varies from person to person and is hard to predict.

In some people, IPF remains stable for years. In others, symptoms get worse over time and it becomes harder to breathe even when you are not active.

IPF can also lead to other medical problems. As the disease gets worse, it can lead to respiratory (lung) failure, infections, high blood pressure in the lungs (pulmonary hypertension), and heart failure.

It is important for people living with a form of IPF to work closely with their health care providers to find ways to try and preserve their overall health.
How are PF and IPF treated?

The treatments used for various forms of PF will depend on its cause. Specifically, for IPF, there are US Food and Drug Administration (FDA) approved medications that can be used to help patients affected by the disease. These medications are taken orally.

Additionally, some treatments may help relieve the symptoms of PF, and may help maintain your activity.

These treatments include:

**Oxygen therapy.** Oxygen may be prescribed if the level of oxygen in your blood gets too low. Using oxygen can help make breathing easier and help you try to stay active.

It can also prevent or reduce problems from low oxygen levels in your blood, reduce the blood pressure in your heart, improve your sleep, and help you feel better.11

**Pulmonary rehabilitation.** Pulmonary rehabilitation programs help people with PF. They focus on physical exercise and breathing techniques. They also provide nutritional counseling and emotional support.12,13

**Lung transplantation.** For some people with PF, lung transplantation may be an option.14

Another option to consider is to enroll in a clinical trial. Your health care provider will talk with you about your options and which steps might be right for you.

Who treats pulmonary fibrosis?

Finding a health care provider who has experience treating people with PF is important. Pulmonologists (lung and breathing specialists) familiar with PF can often be found at major medical centers.

To find a medical center near you with expertise in treating PF, contact the PFF Patient Communication Center at 844.825.5733 or +1 312.587.9272 (from outside the US) or visit www.pulmonaryfibrosis.org.

Can you benefit from participating in a clinical trial?

A clinical trial is a research study to learn more about investigational treatments or new ways to use existing treatments. Some clinical trials are performed to see if a new medicine is safe and effective for people to use. Other clinical trials compare existing treatments to find out which one is better.15

The FDA and other regulatory agencies such as the European Medicines Agency (EMA), have regulations and guidelines to make sure people are provided with reliable information to help them decide whether working with their doctor to join a clinical trial is an appropriate option for them.16,17 Please be sure to talk to your provider about all of your options.

Where can you find support?

**IN-PERSON SUPPORT GROUPS**

Participating in a support group may help individuals with PF and family members better manage the challenges of living with PF.3

To find a support group near you, visit www.pulmonaryfibrosis.org/life-with-pf/support-groups.

If there is no support group in your area and you would like to start one, contact the PFF Patient Communication Center at 844.TalkPFF (844.825.5733) or +1 312.587.9272 (from outside the US) or visit www.pulmonaryfibrosis.org/life-with-pf/support-groups/support-group-leader-network.

**ONLINE SUPPORT GROUPS**

**Inspire.** These online support groups are for people with PF and their caregivers. Learn more about the Pulmonary Fibrosis Foundation’s Inspire community at www.inspire.com/partners/pulmonary-fibrosisfoundation.

**RareConnect.** This international online community is designed to connect people with IPF who speak different languages. Learn more about RareConnect at www.rareconnect.org/en/community/idiopathicpulmonary-fibrosis.
How can you live better with pulmonary fibrosis?

Living with PF is not always easy. But there are some things that you can do to help manage your condition and try to maintain your quality of life.\(^{18,19}\)

**TAKE AN ACTIVE ROLE WITH YOUR CARE TEAM**

Your care team includes your providers, your supporters, and YOU. Being actively involved in your treatment is important for individuals with PF.

Work closely with your health care providers and others on your care team.

Call your health care provider if you do not feel well or notice any unusual symptoms.

Take all of your medications exactly as directed by your health care provider.

Follow your health care provider’s dietary and exercise recommendations.

Get your vaccinations. Respiratory (lung) infections like the flu can make your PF symptoms worse.\(^{20}\) It is important to talk with your doctor about getting a flu shot every year and be up to date with your pneumonia vaccine.

Keep all of your medical appointments.

Speak up for yourself. Talk with your providers about your questions or concerns.

Be prepared for your medical visits. Prepare a list of questions to take to your appointments. Visit www.pulmonaryfibrosis.org/yourappointment for a list of questions.

Take notes at your appointments. Or have a friend or family member come with you to take notes.

Get support and stay informed. Go to educational meetings held by your local medical center, join a support group, and connect with the Pulmonary Fibrosis Foundation.

**CONSIDER MAKING SOME LIFESTYLE CHANGES**

Stay active and keep in shape.

Eat well and maintain a healthy body weight. A dietician can help you create a healthy eating plan that works for you.

Get plenty of rest.

Stop tobacco use and avoid secondhand smoke.

Learn and practice relaxation techniques to help manage stress.

Join a support group. Family members and caregivers can join too.

Try to keep a positive attitude. If you feel depressed or anxious, be sure to tell your provider or another member of your care team.

**SUPPORT FOR YOU FROM THE PULMONARY FIBROSIS FOUNDATION**

The Pulmonary Fibrosis Foundation (PFF) provides support for the entire PF community. Our signature programs include:

- PFF Patient Communication Center
- Disease Education Materials
- International network of support groups and online communities
- PFF Care Center Network and the PFF Patient Registry
- PFF Summit
- PFF Ambassador Program

To learn more about the Pulmonary Fibrosis Foundation contact the PFF Patient Communication Center at 844.TalkPFF (844.825.5733) or pcc@pulmonaryfibrosis.org.