



# Understanding pulmonary fibrosis

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Specialty Pharmacy

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# What you need to know about pulmonary fibrosis

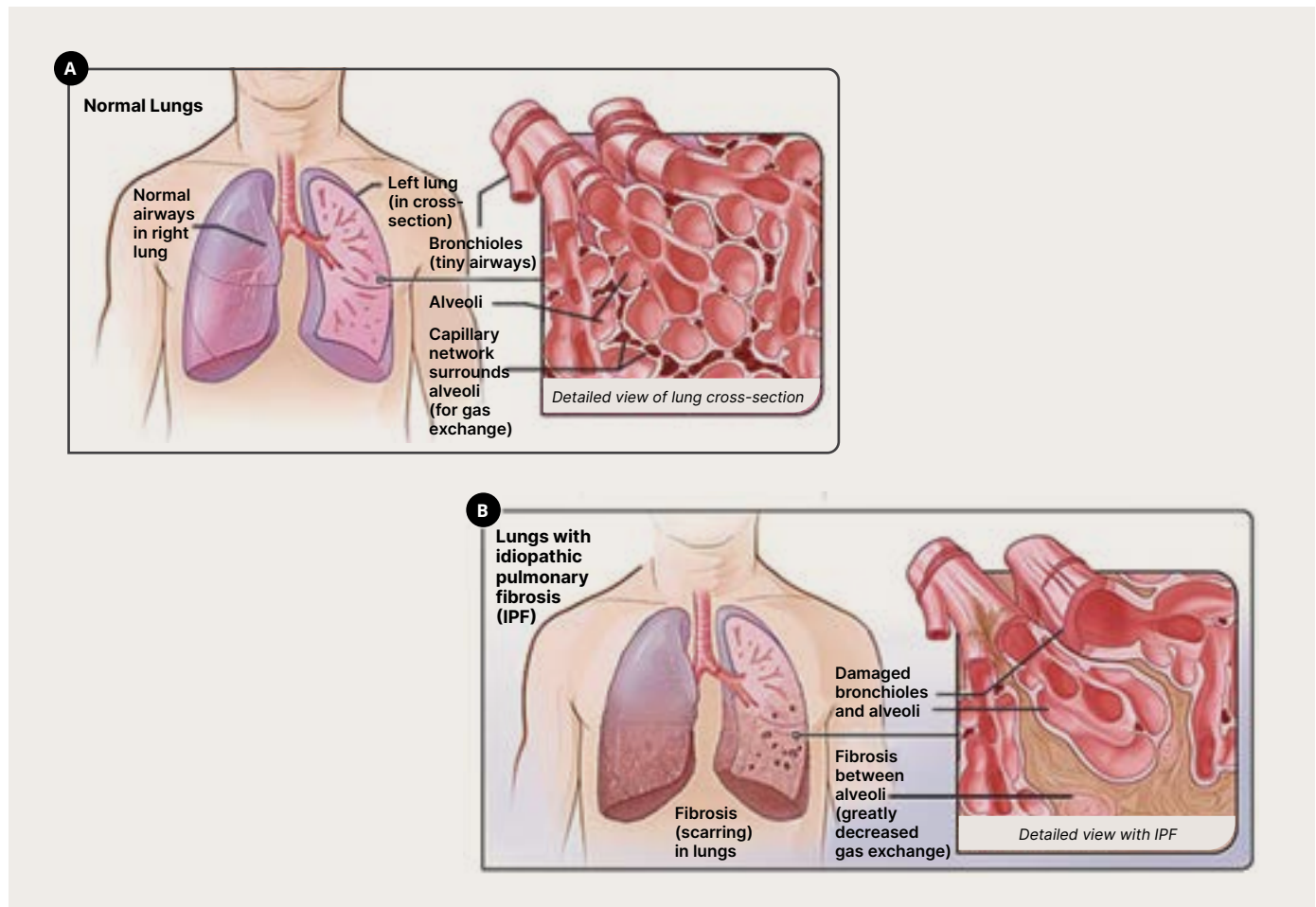
Learning how to manage pulmonary fibrosis (PF) can seem tough, especially at first. But understanding your diagnosis can help you take control of your health. A good treatment plan and key lifestyle changes can help you control symptoms and improve your overall health. This booklet provides information about PF, what to expect after diagnosis and how to keep up with your treatment to live a full and active life.

## Overview

PF is a long-term disease that affects the lungs. Normally, oxygen can pass easily through the air sacs in the lungs, called alveoli. From there, it moves into the bloodstream.<sup>1</sup>

In PF, the tissue around the alveoli becomes thick and stiff. This causes fibrosis, or scarring. The damage makes it difficult for oxygen to enter the bloodstream. This can result in low oxygen levels. It can make it hard to breathe.<sup>1,2</sup>

**Figure 1. How PF affects lungs and airways**



Source: National Heart, Lung, and Blood Institute; National Institutes of Health; U.S. Department of Health and Human Services

## Causes and risk factors

PF can develop for different reasons, including<sup>1,2</sup>:

- Autoimmune diseases, like rheumatoid arthritis or scleroderma
- Exposure to things in the environment, like mold, bacteria or animal proteins
- Gene mutations
- Medications
- Occupational exposure to materials like asbestos, silica or coal dust
- Radiation treatment to the chest
- Unknown reasons (idiopathic)

There are more than 200 types of PF.<sup>2</sup> The most common form is called idiopathic pulmonary fibrosis (IPF). This is when the cause for PF cannot be found.<sup>3</sup> Some people might be at a higher risk for IPF, including<sup>1,2</sup>:

- Children or siblings of people with IPF
- Males
- People over the age of 50
- Smokers

## Diagnosis

A healthcare team uses many pieces of information to confirm a PF diagnosis, such as<sup>1,2,5</sup>:

- Family history and personal risk factors
- Health exams for PF signs and symptoms, such as blue hands and feet or high-pitched crackling sounds in the lungs
- Tests and procedures, including blood tests, breathing tests, chest imaging and tissue samples

The diagnostic process helps rule out other causes of lung disease. It also helps determine what type of PF you have.<sup>2</sup> This information helps your healthcare provider build a treatment plan that is right for you.

### By the Numbers: IPF in the U.S.<sup>2,4</sup>

Most common between the ages of **50** and **70**

Approximately **35,000** new cases diagnosed each year

Results in as many as **40,000** deaths each year

# Symptoms

The most common PF symptoms are<sup>1,4</sup>:

- Shortness of breath
- Dry cough that won't go away

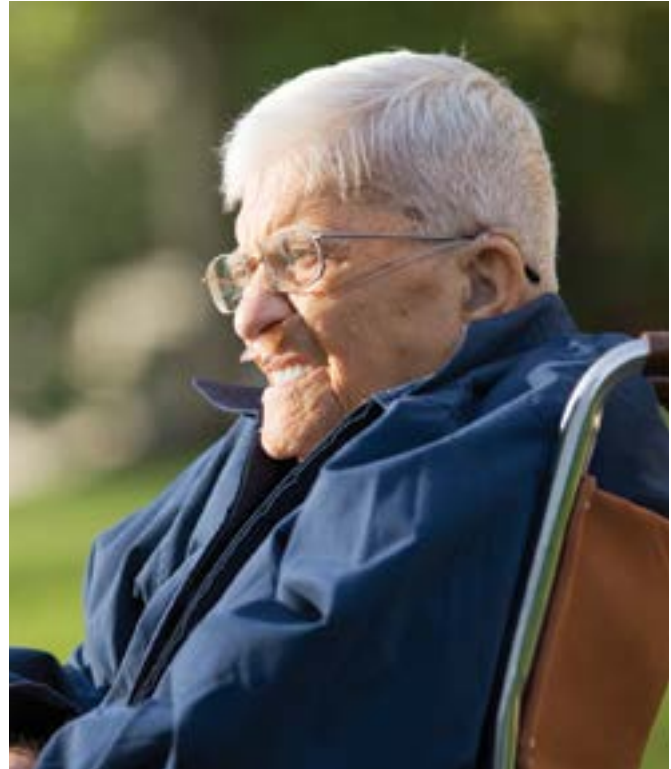
Both of these can get worse over time. For example, feeling breathless while exercising might slowly give way to feeling short of breath during rest. A bothersome cough might turn into long coughing spells that do not get better.<sup>2</sup>

Other signs and symptoms might include<sup>1,4</sup>:

- Aching muscles and joints
- Clubbing, or the widening and rounding of toes and fingertips
- Fatigue
- Weight loss

Each person's experience is different. Some people might have severe symptoms. Some might have just a few mild symptoms. Others might have no symptoms at all. This can change from day to day and over time.<sup>1,2</sup>

Sometimes, symptoms can suddenly get worse. These are called acute exacerbations.<sup>1</sup> These can happen because of a lung infection, heart problems or blood clots in the lungs. Other times, they seem to happen for no clear reason. Treating an acute exacerbation might require oxygen. In some cases, it might require hospital care.<sup>2</sup>



# Complications

PF causes progressive damage to the lungs. For some people, this process is slow. For others, problems might develop more quickly. Over time, PF can lead to other serious lung problems, including<sup>1,2,4,6</sup>:

- Pneumonia
- Pulmonary embolism, or blood clots in the lungs
- Pulmonary hypertension, or high blood pressure in the blood vessels of the lungs
- Lung cancer
- Respiratory failure

People with PF might also be more likely to have other health issues, including<sup>6</sup>:

- Gastroesophageal reflux disease (GERD)
- Chronic obstructive pulmonary disease (COPD)
- Diabetes
- Heart disease
- Sleep apnea



# Living with PF

There is no cure for PF. However, certain therapies can help people manage PF and improve their quality of life.<sup>1</sup> The goals of PF treatment include<sup>1,2</sup>:

- Controlling symptoms
- Encouraging physical activity
- Improving breathing ability
- Raising oxygen levels
- Slowing down lung damage

Treatment needs can vary from person to person. Your healthcare provider will build a plan that is right for you. Your prescribed treatment might include medication, oxygen therapy and pulmonary rehabilitation.<sup>1,2</sup>

## Medications

Medication therapy can help slow the progression of PF. It can also treat symptoms and control other health issues that can make PF worse. The following table lists common types of medications used to treat PF.

**Table 1. Medications for PF<sup>1,2,7</sup>**

Medication type	Action
Antacid, proton pump inhibitor, H2 blocker	Treats GERD to prevent stomach acid from entering and irritating the lungs
Anti-fibrotic	Slows down lung damage and decline in lung function
Anti-inflammatory	Reduces swelling in the lungs
Cough suppressant	Treats chronic cough
Immunosuppressant	Prevents swelling by lowering the body's immune response

There is no single treatment that is right for everyone with PF. The progression and symptoms of PF can differ from person to person. Your healthcare provider will prescribe drug therapy that is tailored to you.

## Oxygen therapy

Your healthcare provider might prescribe oxygen therapy if your oxygen levels get too low — about 88% or lower.<sup>2</sup> Oxygen therapy can bring your levels back up. This can make it easier to breathe. It can also help you feel less tired. This might make it easier to stay active.<sup>2</sup>

Your prescription will tell you how much oxygen you need and when you should use it. This is different for each person. Ask your healthcare provider about your target oxygen level. You might need to monitor your oxygen level from time to time using a fingertip device called a pulse oximeter. This can help you see if the therapy is working.<sup>8</sup>

Most patients have one oxygen system at home and another portable system that travels with them.<sup>8</sup> Your care team will help you find the right type of oxygen system. They can also help you learn how to use and maintain the equipment.

It might take a little while to get used to oxygen therapy. Some people find it awkward and frustrating, especially at first. But oxygen therapy can get easier with time. With guidance from your care team, oxygen therapy can keep you moving and improve your quality of life.

## Oxygen away from home

Your PF therapies should be part of your daily routine. This means you will need to keep up with treatments like oxygen therapy even when you are traveling. There are many things to consider<sup>9,10</sup>:

- Tell your care team about your travel plans well in advance. They can assess if it is safe for you to travel.
- You might need prescription oxygen, depending on where you are going and how you will get there. Flying or traveling to higher elevations will require more oxygen.
- Find out ahead of time if your airline, train or cruise requires paperwork for using oxygen during a trip. Learn what type of equipment you can use on board.
- No matter how or where you travel, be sure to pack a reliable power source and any accessories you need.

Like other PF therapies, PR can help you feel better and do more. But completing a PR program is just a start. Your care team can help you take what you learn in PR and make it part of your daily life.

## Staying on track with treatment

Following your prescribed treatment is crucial. It can slow down the damage to your lungs and help you feel better. No matter which drugs and therapies are part of your treatment, you should keep up with them exactly as your healthcare provider prescribes. This means staying diligent about physical therapy or breathing exercises. It also means taking medications at the right times and the correct doses.

Some days, it might feel hard to stay on track. Other times, you might feel like nothing is working. Do not stop treatment on your own. Tell your care team your concerns. They can help you adjust your treatment as needed so it works best for you.

## Pulmonary rehabilitation

A pulmonary rehabilitation (PR) program is designed to help you breathe more easily. It can include breathing techniques, exercise and health education. These things can improve your quality of life and help you stay active.<sup>2,11</sup>

You might work with different types of healthcare providers, including dietitians, physical therapists, respiratory therapists and social workers. They will build a program based on your needs. This might include<sup>2,11</sup>:

- Breathing exercises
- Counseling and support for anxiety, depression or stress
- Exercise training
- Nutritional counseling
- Physical therapy



# Lifestyle changes

Along with a prescribed treatment plan, certain lifestyle changes can help you manage PF. You might learn some of these wellness strategies through your PR program. These healthy habits can help slow down the disease process. They can also control symptoms and prevent complications. Protecting your lungs, staying active, eating well and finding emotional support are all important for managing PF.<sup>12</sup>

## Protecting your lungs

PF causes inflammation and scarring in the lungs. This makes it crucial to protect your lungs from any further damage. You can do this in many ways<sup>1,2,10,12</sup>:

- If you smoke, quit. Talk to your care team if you need help with this.
- Avoid inhaling substances that can irritate your lungs. This includes things like dust, chemicals or secondhand smoke.
- Lower your risk for respiratory infections. Wash your hands often or use hand sanitizer frequently. Avoid large crowds. Steer clear of others who are sick.
- Ask your healthcare provider about vaccinations against flu, pneumonia and COVID-19.
- If you develop a cold, the flu or other respiratory illness, call your healthcare provider right away for guidance.

## Staying active

Exercise can improve overall health. For those with PF, staying active can help boost energy, improve lung function and strengthen muscles. It can also relieve stress and improve sleep.<sup>1,13</sup> Your prescribed PR program will include plans for exercise training. Activities might include<sup>13</sup>:

- Biking
- Walking
- Stretching
- Weight training

Exercise plans are tailored to each person. Your care team will help you choose the right types of activity and plan how often to exercise. Beginners might start with two to four days of exercise a week. Others who have previously been active might work toward four to seven workouts per week.<sup>14</sup>

It might feel hard to get moving at first, especially if you are feeling short of breath. Your care team can help you set realistic goals for staying active. They can also teach you how to monitor and support your breathing during exercise. Some people might take medicine to open the airways. Others might adjust their oxygen therapy.<sup>1,13,14</sup>

Along with exercise training, consistent everyday movement is important. Try to get up and around to take care of daily tasks, even if you feel tired or breathless. Challenge yourself to keep moving. Make time to rest when needed. With regular movement, some daily activities might start to feel easier.<sup>1,13</sup>

### Staying active safely

It's important to stay safe during workouts. Your PR team might suggest a few precautions<sup>14</sup>:

- Follow your prescribed exercise routines. Use supplemental oxygen as directed.
- Learn how to track your vital signs. This might include your heart rate, oxygen level or breathing. Do this before, during and after your workouts.
- Dress comfortably. Wear supportive shoes that fit well.
- Keep your activity space well-lit, with enough room to move around.
- Have a workout buddy or someone who can stay nearby in case you need help.



# Eating well

Along with staying active, a healthful diet is a key part of your care plan. Eating well can give you energy. It can also help you stay at the right weight. This is important for managing PF.

Being overweight can put pressure on your lungs. This can make it harder to breathe. Being underweight can leave you with less energy. This can weaken the muscles you use to breathe.<sup>15</sup>

A healthy weight is usually based on your body mass index (BMI). This is a measure of your body fat based on your height and weight.<sup>15</sup> You can talk with your healthcare provider about your BMI and your goal weight.

A dietitian can show you how to get the calories and nutrients needed to support your health. Your PR plan will likely include nutritional counseling. There is no special diet for PF. In general, a balanced, heart-healthy diet is a good choice<sup>1,12,15,16</sup>:

- Eat plenty of fresh vegetables, fruits and whole grains like plain oatmeal or brown rice.
- Opt for low or nonfat dairy and lean proteins, like fish, chicken, eggs and beans.
- Choose heart-healthy oils and foods high in healthy fats, like nuts, fish and avocados.
- Limit sodium, added sugars, and saturated and trans fats. These are often found in ready-made foods.
- If you have GERD or other reflux issues, avoid acidic foods, like oranges, coffee or tomatoes.

It is important to choose enough of the right foods. It is also crucial to choose the right meal times and sizes<sup>1,12,15</sup>:

- A very full stomach can make it hard to breathe. Try eating smaller meals throughout the day to avoid discomfort.
- Acid reflux and GERD can trigger PF symptoms or make them worse. Avoid meals close to bedtime to help prevent reflux.



## Emotional health

Some days, living with PF can be hard. You might feel worried, sad or mad. This is normal, and you are not alone. These feelings are common for people with long-term conditions like PF.<sup>12,17</sup> You can find healthy ways to cope<sup>1,12,17</sup>:

- Exercise, eat well and get enough sleep to help boost your mood.
- Find supportive family members and friends. They can help with tasks, motivate you through lifestyle changes and give moral support at medical appointments.
- Join a PF support group. You can make new friends and learn from others who share your experience.
- Practice relaxation techniques. These might include breathing exercises or meditation.
- Talk to a counselor or therapist. They can help you work through your emotions. If needed, they might also recommend medication that could help improve your mood.

Living with PF can be challenging. You do not have to work through everything on your own. Your care team and others close to you can provide the support you need to feel well.

## Lung transplantation

For some people with PF, standard treatments and therapies are not enough. In these cases, lung transplantation is an important option. In 2019, people with IPF accounted for about 35% of lung transplants in the United States.<sup>2</sup>

Transplantation replaces damaged lungs with healthy ones from a donor. It can improve a person's quality of life. After transplantation, most people have no trouble breathing, no chronic cough and no need for oxygen therapy.<sup>2</sup>

Lung transplantation is not an option for everyone with PF. It can carry many risks. The process takes careful preparation, waiting and lifelong care. Thinking about such a major treatment might feel scary. Learning more about the process can be useful. It might help to talk openly with your care team about this option long before it might ever be needed.<sup>18</sup>

## Ongoing care

Work with your care team to manage your PF treatment. Regular checkups will help you and your care team keep track of your condition and your overall health. You can talk about how well your treatment regimen and lifestyle changes are working. Your healthcare provider can also adjust your therapies as needed.

We provide this information because  
the more you know about PF,  
the better you'll be able to manage it.

Additionally, the Walgreens Specialty360 Therapy  
Team is here to support you with **dependable,**  
**personalized** service to help manage your  
medication side effects and stay on track with  
your prescribed therapy.

We look forward to being a member of your  
healthcare team and helping you get the best  
results from your treatment.

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# Resources

You might find it helpful to contact these organizations for additional support and resources.\*

## **Pulmonary Fibrosis Foundation**

www.pulmonaryfibrosis.org  
844-TalkPFF (844-825-5733)  
Facebook: www.facebook.com/PFFORG  
Twitter: @PFFORG

The Pulmonary Fibrosis Foundation is a nonprofit patient advocacy organization for the PF community. Its website features educational materials for patients, families and caregivers as well as clinical resources for healthcare providers and researchers. The PFF supports a number of policy and advocacy initiatives, funds ongoing research efforts and maintains the PFF Care Center Network of clinicians and support groups. The PFF also responds to patient inquiries and provides support to the PF community via the PFF Help Center, at 844-TalkPFF (844-825-5733), help@pulmonaryfibrosis.org and www.pulmonaryfibrosis.org/patients-caregivers/medical-and-support-resources/pff-help-center.

## **American Lung Association**

www.lung.org  
800-LUNGUSA (800-586-4872)  
Facebook: www.facebook.com/lungusa  
Twitter: @LungAssociation

The American Lung Association is a national nonprofit organization dedicated to saving lives by improving lung health and preventing lung disease. Its website features research updates, policy and advocacy initiatives and patient information about lung health and diseases, including PF.

## **National Heart, Lung, and Blood Institute**

www.nhlbi.nih.gov  
877-NHLBI4U (877-645-2448)  
Facebook: www.facebook.com/NHLBI  
Twitter: @nih\_nhlbi

The National Heart, Lung, and Blood Institute at the National Institutes of Health oversees the research, training and education for the prevention and treatment of heart, lung and blood disorders. Its website features educational resources for patients and clinicians, updates about awareness efforts and information about clinical trials for PF and a number of other diseases and conditions.

\*The referenced organizations are provided for informational purposes only. They are not affiliated with, and have not provided funding to, Walgreens Specialty Pharmacy for this booklet. Walgreens Specialty Pharmacy does not endorse or recommend any specific organization.





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