

# pulmonary fibrosis information guide



**Pulmonary<sup>TM</sup>  
Fibrosis  
Foundation**

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# about this guide

People living with pulmonary fibrosis (PF) and their family members often feel confused, concerned, and overwhelmed by the disease and by the lack of clear and consistent information about what PF is. You may have experienced this yourself. Physicians do not always have the necessary time or resources to explain the details of the disease or to provide the support that people living with PF need.

The purpose of this guide is to help provide an overview of pulmonary fibrosis for patients and their caregivers, and to help provide clear and consistent information about PF, so that you can live a healthier, more normal life.

Please remember that this information is a brief overview and is for educational purposes only. It is not intended to be a substitute for professional medical advice. Always consult personal physicians or health care providers with any questions regarding specific medical conditions.

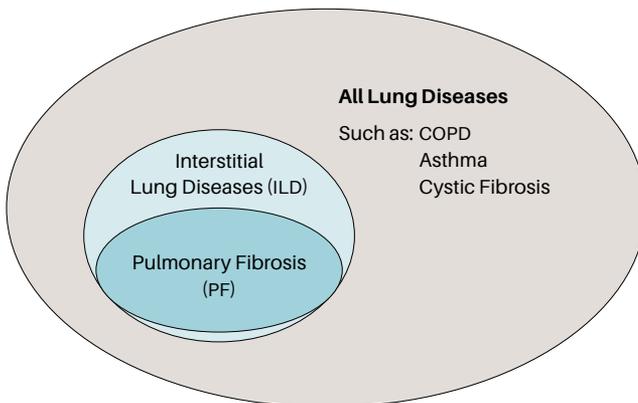
We are here to help. Contact the **PFF Help Center** with any questions or concerns you have about PF and available resources during the course of your care. Our staff can be reached at **844.TalkPFF** (844.825.5733) or [help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org).

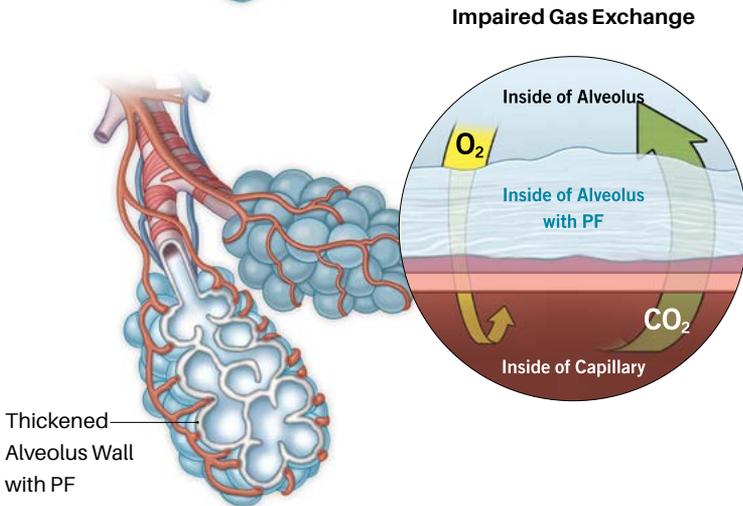
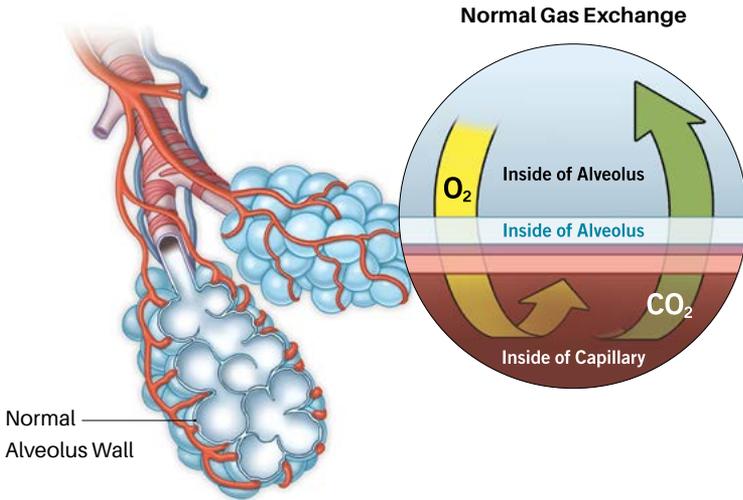
# about pulmonary fibrosis

## What is pulmonary fibrosis?

The word “pulmonary” means lung and the word “fibrosis” means scar tissue—similar to scars that you may have on your skin from an old injury or surgery. So, in its simplest sense, pulmonary fibrosis (PF) means **scarring in the lungs**. Over time, the scar tissue can destroy the normal lung and make it hard for oxygen to get into your blood. Low oxygen levels (and the stiff scar tissue itself) can cause you to feel short of breath, particularly when walking and exercising.

Pulmonary fibrosis isn’t just one disease. It 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 **the interstitial lung diseases** (also known as ILD), which includes all of the diseases that have inflammation and/or scarring in the lung. Some interstitial lung diseases don’t include scar tissue. When an interstitial lung disease does include scar tissue in the lung, we call it pulmonary fibrosis.





No one is certain how many people are affected by PF. One recent study estimated that idiopathic pulmonary fibrosis (or IPF, which is just one of more than 200 types of PF) affects 1 out of 200 adults over the age of 70 in the United States – that translates to more than 200,000 people living with IPF today. Approximately 50,000 new cases are diagnosed each year and as many as 40,000 Americans die from IPF each year.

## I was just diagnosed with pulmonary fibrosis.

### What do I do next?

We suggest that you consider making an appointment with a pulmonologist who has experience caring for patients with PF. A knowledgeable team of PF experts will help make sure you receive an accurate diagnosis and the most up-to-date treatments and management recommendations. To assist you in identifying pulmonologists closer to home and developing expertise in the care of patients with PF, the Pulmonary Fibrosis Foundation established the PFF Care Center Network that includes 68 medical centers throughout the United States. You can find a list of sites within the Network at [pulmonaryfibrosis.org/life-with-pf/find-medical-care](https://pulmonaryfibrosis.org/life-with-pf/find-medical-care).

We also recommend that you consider joining a PF support group. Connecting with other individuals facing the same illness can help you and your family not feel so alone in your journey with pulmonary fibrosis. Support groups can supplement the care you receive from your health care team by providing emotional support and education.

Support groups can help those living with pulmonary fibrosis

- learn about their disease and available treatments;
- feel supported by others who are going through the same thing;
- learn to navigate the health care system more effectively; and
- improve coping skills, among other things.

You can learn more and find a local PF support group at [pulmonaryfibrosis.org/life-with-pf/support-groups](https://pulmonaryfibrosis.org/life-with-pf/support-groups).

Contact the PFF Help Center by calling 844.TalkPFF (844.825.5733) or email [help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org) with any questions or concerns you may have and to connect you with local resources.

## What causes pulmonary fibrosis?

It can be challenging for doctors to figure out what causes PF. Sometimes they are able to identify one or more causes of your disease, which are discussed here. PF of unknown cause is called “idiopathic” and discussed on page 8.

There are five main categories of identifiable causes of pulmonary fibrosis: Medications, radiation, environmental exposures, autoimmune diseases, and occupational exposures. In the United States, environmental and autoimmune causes seem to be the most common types of PF of known cause. This table shows some of the clues that doctors use to identify these known causes of PF.

TYPE OF PULMONARY FIBROSIS	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, fumes, or vapors that can cause PF (such as asbestos, coal, silica, and others)
Idiopathic	When no cause can be identified (see page 8)

Some **MEDICATIONS** can cause PF. Drugs used to treat cancer (chemotherapy), drugs used to treat abnormal heart rhythms (such as amiodarone), drugs used to treat inflammatory conditions (such as methotrexate), and an antibiotic used to treat urinary tract infections (nitrofurantoin) are some of the better known drugs that can cause injury, inflammation, and scarring in the lungs. Numerous other drugs have been implicated as causes of PF in some cases.

**RADIATION** to the chest for lymphoma; Hodgkin's disease; or breast, lung, and other cancers can injure the lung and cause fibrosis.

**ENVIRONMENTAL CAUSES OF PF** are typically called hypersensitivity pneumonitis (HP) or chronic hypersensitivity pneumonitis. HP occurs when the lungs react with inflammation and scarring after breathing in mold spores, bacteria, animal proteins (especially from indoor or caged birds), or other known triggers. No one is certain why some people are so susceptible to developing HP and others are not.

**AUTOIMMUNE DISEASES** are also called connective tissue diseases, collagen vascular diseases, or rheumatologic diseases. "Auto" means self and "immune" refers to your immune system. So if you have an autoimmune disease affecting your lungs, it means that your body's immune system is attacking your lungs. Examples of autoimmune diseases that can cause PF include:

- rheumatoid arthritis;
- scleroderma (also called systemic sclerosis);
- Sjögren's syndrome; and
- polymyositis, dermatomyositis, and antisynthetase syndrome.

**OCCUPATIONAL CAUSES OF PF**, also called pneumoconioses, can develop after significant exposure to a wide variety of inorganic dusts, including asbestos, silica, coal dust, beryllium, and hard metal dusts.

## Are there genetic or inherited forms of pulmonary fibrosis?

Yes, but in most cases, it is not as straightforward as inheriting blue eyes or red hair. There are three different scenarios:

**1) Did the genes I inherited from my parents contribute to PF even if I am the only one in my family with PF?** Yes. The risk of developing most human diseases is influenced by the genes you inherited from your parents. There have been several genes identified that appear to increase the risk of developing PF when an abnormal form of the gene is inherited. There are no official medical guidelines on whether or not genetic testing should be performed if you have PF, although in some cases, your doctor might wish to test you for one or more abnormal forms of these genes.

**2) Can PF run in the family?** Yes. Anywhere from three to 20% of people with PF have another family member with PF. But this doesn't mean that up to 20% of your family members will develop PF. In most cases, the chance that one of your family members will develop PF is very low. We encourage you to talk to your doctor or to a genetic counselor about the chances that one of your family members will develop PF. In certain patients and family members, genetic testing may be appropriate. Additional information is provided in the Genetic Testing in Pulmonary Fibrosis PFF Position Statement at [bit.ly/familialpf](https://bit.ly/familialpf).

**3) Are there specific types of PF where the entire disease is due to one gene?** Yes. There are a few rare genetic forms of PF that can affect both children and adults when just a single gene is inherited in an abnormal form (usually from both parents). Hermansky-Pudlak syndrome is an example of a genetic form of PF that simply requires abnormalities in one gene. People living with Hermansky-Pudlak syndrome often also have very light skin pigmentation (due to low levels of melanin in their skin) and bleeding problems (due to poorly functioning platelets in

their blood). Dyskeratosis congenita is another example of a genetic form of PF that can be accompanied by poorly growing fingernails, changes in skin pigmentation, increased risks of developing problems in the bone marrow, and other conditions.

## My doctor said my disease was “idiopathic.” What does that mean?

Even after an exhaustive evaluation, doctors sometimes end up telling their patients, “We don’t know why you have this.” Often, the word “idiopathic” is used to refer to any disease of unknown cause.

One type of idiopathic lung disease that you may have heard of is **idiopathic pulmonary fibrosis (IPF)**, but there are actually many other forms of ILD that are idiopathic.

Most people find these idiopathic ILDs and their abbreviations to be confusing, and there are too many different kinds of idiopathic ILDs to discuss in this booklet. Here are few of the names and abbreviations you might come across:

Disease	Abbreviation
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

**IDIOPATHIC PULMONARY FIBROSIS (IPF)** is a scarring disease of the lungs of unknown cause. To make a diagnosis of IPF, your doctor will perform a thorough history to try to identify potential exposures or other diseases that might lead to scarring of the lung. If a plausible cause is found, then you do not have IPF. The scarring pattern of IPF is technically called usual interstitial pneumonia (UIP). Your doctor will use detailed X-rays of your lungs called high-resolution computed tomography (HRCT) and sometimes a lung biopsy to look for this pattern. A diagnosis of IPF requires that your doctor cannot find a cause and the presence of a pattern of UIP on either HRCT or a surgical lung biopsy sample. Possible medications used to treat IPF are discussed on pages 13–16.

Although IPF is still considered to be a disease of unknown cause, we do know some factors that increase the risk of getting IPF, including aging (IPF is rare before age 50), cigarette smoking, and having certain genetic predispositions (see page 7).

**“If I had said, ‘I am short of breath and I shouldn’t be,’ I might have gotten a diagnosis faster.”**

PATIENT LIVING WITH PF

# diagnosis & treatment options

## How do doctors recognize and diagnose pulmonary fibrosis?

There are three consequences of PF. Doctors use these consequences to recognize that someone has PF.

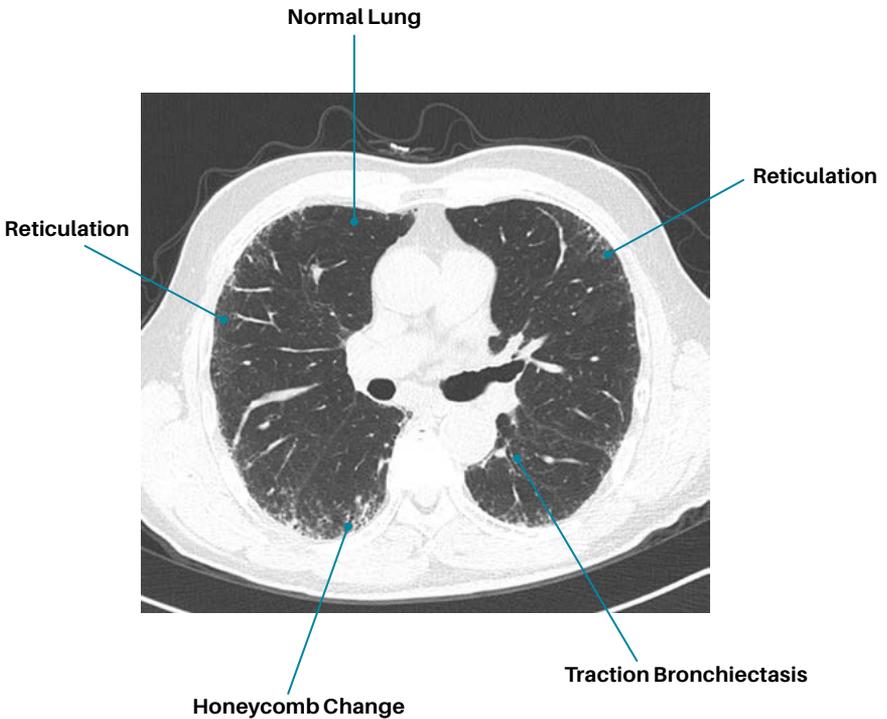
- **STIFF LUNGS** – Scar tissue and inflammation make your lungs stiff. Stiff lungs are hard to stretch, so your breathing muscles have to work extra hard just to pull air in with each breath. Your brain senses this extra work, and it lets you know there’s a problem by triggering a feeling of **breathlessness** (or “shortness” of breath) while exerting yourself. Also, stiff lungs hold less air (they shrink up a bit). Doctors take advantage of this “shrinking” to diagnose and track the disease using breathing tests (**called Pulmonary Function Tests**) that measure how much air your lungs can hold. The more scar tissue your lungs have, the less air they will hold.
- **LOW OXYGEN LEVELS IN THE BLOOD** – Scar tissue blocks the movement of oxygen from the inside of your air sacs into your bloodstream. For many people living with PF, oxygen levels are only reduced a little bit while resting, but their oxygen levels drop quite a bit during exertion. The brain can sense these low oxygen levels, triggering **breathlessness**. Doctors will **check your oxygen levels to see if they drop after walking**, a clue that PF might be present. Doctors also often **prescribe oxygen** to be used through a nasal cannula or a facemask during exertion and sleep for those with PF. As PF progresses, oxygen may be needed 24 hours a day.

- **“CRACKLES”** – Your doctor may have told you that **“crackles” were heard** in your lungs. Crackles (also called “rales”) sound like Velcro being pulled apart. They are heard in many lung diseases because any type of problem affecting the air sacs (such as PF, pneumonia, or a buildup of fluid in the lungs from heart failure) can cause crackles. Some people with PF don’t have crackles, but most do.

Once your doctor recognizes that you might have PF, the next step is to try to diagnose the specific kind of PF you have – there are more than 200 different kinds. Doctors typically start by asking many questions, performing a careful physical examination, and ordering a lot of blood tests. See **What Causes Pulmonary Fibrosis?** on page 5 for more information.

A doctor will also use a special kind of X-ray of the chest, called a high-resolution computed tomography (HRCT) scan, so that they can see what your lung tissue looks like. HRCT scans give a close-up view of the lungs, providing more detail than routine CT scans (also known as CAT scans). Healthy lung tissue looks nearly black on a CT scan. **Scar tissue and inflammation both appear grey or white.** Many forms of PF look similar on a CT scan to the untrained eye (see page 12), but subtle findings on HRCT scans are critically important when trying to identify which type of PF you have. An HRCT protocol recommended by the PFF and PF experts is available on the PFF website.

Sometimes, even after all of the testing is complete, a doctor will still not have an answer and will have to perform a lung biopsy to sort out which of the 200 different types of PF you have. When indicated, a lung biopsy can also help you and your doctor decide which treatments might be helpful.



On CT imaging, lung slices are obtained from the neck through the stomach similar to slicing a loaf of bread. Air is typically black as noted by the air above the chest and throughout most of the lung in the normal patients. With abnormalities such as inflammation or fibrosis in the lung, varying degrees of white opacities are noted in the lung with structural changes such as honeycomb change or traction bronchiectasis.

**Reticulation** - extra lines in the lung

**Honeycomb** - distorted, end stage fibrotic lung

**Ground glass** - nonspecific abnormality that may reflect inflammation, fibrosis, bleeding, fluid in the lung.

## What kinds of treatments are available for pulmonary fibrosis?

Doctors have a number of ways to treat PF, including the use of medications, oxygen therapy, non-medical treatments (such as exercise), and even lung transplantation.

The following information is meant as a general overview of some of the treatments that physicians offer their patients with PF. **This information is not medical advice.** Some of these treatments may be right for some people, but no one treatment is right for everyone. You should speak with your doctor before starting, changing, or stopping any medical treatment. You can also learn more by contacting the PFF Help Center at 844.TalkPFF (844.825.5733) or [help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org).

### 1. OXYGEN THERAPY

Oxygen is prescribed when oxygen levels go below about 88%. Your doctor may choose to measure your oxygen level in different circumstances: while sitting at rest, while walking in the hallway or on a treadmill, and during sleep. If your oxygen level drops below about 88%, your doctor may prescribe oxygen to prevent your oxygen level from dropping further. Many people report that they have less breathlessness and fatigue, and are better able to live an active lifestyle when using oxygen.

Having to use oxygen can be frustrating, inconvenient, and, for many, embarrassing. We recommend that you talk openly with your doctors about your concerns. Many people find it difficult to adjust to using oxygen, particularly when outside the home. Over time, most people find that the benefits of using oxygen greatly outweigh the downside. In a very real sense, using oxygen to

breathe is no different than wearing eyeglasses to see or using a cane to walk – it’s only the judgment of others that seems to make oxygen different from other medical devices. Contrary to what people may think when they see you wearing oxygen, you are doing something important for your health. Those living with PF can jog, work out, and play sports (with permission from their doctors) while using oxygen. Wearing oxygen is a sign of strength, not weakness. More information about supplemental oxygen use and equipment is available in the [PFF Oxygen Basics Booklet](#), available on the PFF website or through the PFF Help Center.

## 2. PULMONARY REHABILITATION

Pulmonary rehabilitation includes exercise training; breathing exercises; anxiety, stress, and depression management; nutritional counseling; education; and more. The goal of pulmonary rehabilitation is to restore your ability to function without extreme breathlessness and **allow you to safely exercise**. It improves both exercise capacity and health-related quality of life for many people living with PF. These programs offer a variety of services and can be inpatient, outpatient, or home/community-based. While pulmonary rehabilitation programs vary, they may comprise a multidisciplinary team of nurses, respiratory therapists, physical therapists, social workers, dietitians, and others.

## 3. DRUG THERAPY

Drug therapy for PF is often specific for the particular type of PF a patient has. In other words, the treatment for one disease may not be the right one for another. It is important to discuss drug therapy with your physician relative to your form of PF. Some common therapies (but not all potential therapies) are listed on the following page.

**Nintedanib (Ofev®):** Nintedanib is an anti-fibrotic drug that is approved in the United States to treat IPF, scleroderma-associated ILD (SSc-ILD), and chronic interstitial lung diseases (ILDs) in which fibrosis continues to progress. In clinical trials, nintedanib has been shown to slow the decline in lung function in SSc-ILD, progressive fibrosing ILD, and mild-to-moderate IPF. It is taken by mouth twice a day. For more information, please visit [ofev.com](https://www.ofev.com).

**Pirfenidone (Esbriet®, Pirfenex®, Pirespa®):** Pirfenidone is an anti-fibrotic and anti-inflammatory drug approved to treat IPF in the US, Europe, Canada, and Asia. In clinical trials, pirfenidone has been shown to slow progression of mild-to-moderate IPF. It is taken by mouth three times a day. For more information, please visit [esbriet.com](https://www.esbriet.com).

**Corticosteroids (prednisone):** Prednisone is used to treat and prevent inflammation by weakening the immune system. While prednisone is not usually used to treat IPF, it is sometimes used to treat inflammation in the lungs of people living with other forms of PF. Since prednisone suppresses the immune system, it can potentially increase the frequency and severity of infections. Prednisone has many other side effects as well.

**Mycophenolate mofetil/mycophenolic acid (CellCept®):** Similar to prednisone, mycophenolate mofetil (MMF) also can treat and prevent inflammation by suppressing the immune system. MMF is sometimes used in combination with prednisone, but is also used alone. MMF is not indicated for the treatment of IPF, but like prednisone can be used for people living with other forms of PF. Some possible side effects include infections, diarrhea, and low blood cell counts. Pregnant women should not take MMF, and women of childbearing age need to use contraception while taking MMF.

**Azathioprine (Imuran®):** Azathioprine is used to suppress the immune system similarly to MMF. Some possible side effects include infections, low blood cell counts, and liver and pancreatic problems.

Several other anti-inflammatory therapies such as methotrexate, cyclophosphamide, cyclosporine, rapamycin (sirolimus), and tacrolimus have been used to treat different forms of interstitial lung disease. As mentioned previously, treatment with these agents is often tailored to specific diseases or forms of ILD and PF.

#### 4. LUNG TRANSPLANTATION

Lung transplantation is discussed on page 18.

#### 5. SYMPTOM MANAGEMENT

Breathlessness and cough are the major symptoms experienced by people living with PF. Patients may also experience fatigue, anxiety, and depression. Please talk to your doctor about these symptoms—there are treatments that might be of help.

##### **Preventing breathlessness**

- Your doctor may recommend that you use enough oxygen to keep your oxygen level over 90% during exertion, as low oxygen concentrations are a common cause of breathlessness in patients with PF. Breathlessness may also occur in the absence of low oxygen concentrations.
- Pulmonary rehabilitation can help you get in shape and learn strategies to minimize breathlessness.
- Doctors sometimes treat severe breathlessness with drugs like morphine when PF is advanced. Palliative care is discussed further below.

##### **Preventing cough**

- There are many causes of cough in addition to PF. Talk to your

doctor to see if post-nasal drip or heartburn (gastroesophageal reflux, or GERD) may be contributing to your cough.

- Low oxygen levels can trigger cough in some people.
- Talk to your doctor to see if there are other treatments that might help your cough.

### **Palliative care**

Palliative care, also referred to as supportive care, is medical treatment focused on relieving and preventing symptoms that are bothersome or distressing. Palliative care also helps you address advanced care planning, which allows you and your loved one to decide your goals of care as your disease progresses. It is important to understand that you can also receive “curative care” at the same time. Curative care refers to participating in research studies, being evaluated for lung transplantation, and considering new medications. This is distinct from hospice care, which is treatment administered in the last months of life (discussed on page 30).

Palliative medicine is usually administered in a team approach, involving physicians, pharmacists, nurses, religious leaders, social workers, psychologists, and other health care professionals working with you. Palliative care providers consider physical, psychosocial, and spiritual factors in their treatment approach. Palliative care is appropriate for anyone experiencing symptoms from their disease.



Adapted from National Consensus Project for Quality Palliative Care, 2004.  
Clinical practice guidelines for quality palliative care

## Is lung transplantation an option for pulmonary fibrosis?

Yes. Lung transplantation is the replacement of one or both lungs with a new lung (or lungs) from a donor. In 2019, about 2,714 people underwent lung transplantation in the United States, and about 35% of those people had IPF. Lung transplantation is an important therapy for some PF patients, but with only 1,000 lungs available per year for those with PF, most people living with the disease will never receive a transplant.

Transplant centers have strict criteria for being a candidate. The evaluation process and waiting time can take months, even years in some cases. Thus, early referral to a lung transplant center is critically important. The first visit is often just a regular medical visit with a doctor and a nurse or nurse practitioner. At that time, the transplant team might decide to order extensive testing, which can include CT scans, heart catheterizations, exercise tests, as well as visits with other members of the transplant team. Talk to your doctor about whether a lung transplant is appropriate for you.

Once placed on the waiting list, about half of people receive their lung transplant within two months (as of 2019). However, there are a number of factors that can influence how long you wait for your transplant.

- Since the availability of donors varies around the country and each transplant center has different criteria for the types of donors they find acceptable, there can be differences in waiting times between different transplant centers. Be sure to ask your transplant team what the average waiting time is.
- People who require replacement of both lungs (called a “double” or “bilateral” lung transplant) often wait longer than those who can receive a “single” lung transplant.
- Children and shorter adults often wait longer for a transplant than those who are taller.

- When you are placed on the waiting list, you receive a “priority score” called the lung allocation score (LAS). This score varies between 0 and 100, with higher scores indicating higher placement on the waiting list. For most people, the LAS is between 35 and 50. On average, as a person becomes sicker, the LAS gets higher and they move up the waiting list. There are many factors that go into calculating the LAS. See the link below for the “LAS calculator,” where you can estimate your LAS.

After recovery from transplant surgery, many people feel much better than they did before transplantation. Most people have no breathlessness, no cough, and require no extra oxygen.

There are also many risks after lung transplantation, including infections, rejection, and medication side effects. While more than one out of four people live more than 10 years after their transplantation, about one out of 10 do not survive the first year. These “average” risks don’t apply to everyone. Talk to your doctors about your risks. Some people have much lower or higher risks.

Here are some links to transplant websites that may be useful:

### **LAS calculator**

<https://optn.transplant.hrsa.gov/resources/allocation-calculators/las-calculator/>

### **Program-specific reports for each transplant center**

<https://www.srtr.org/reports-tools/program-specific-reports/>

### **Organ Procurement and Transplantation Network (OPTN/UNOS)**

<https://optn.transplant.hrsa.gov/>

Please contact the PFF Help Center at 844.TalkPFF (844.825.5733) or email [help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org) for assistance to navigate these reports.

## How will my doctor monitor my PF?

Every doctor will have a different strategy to monitor your PF. Usually, doctors use a combination of the following to determine if your disease is stable or changing:

### 1. SYMPTOMS

One of the most important signs that something has changed in your lungs is a change in the amount of breathlessness you experience while exerting yourself. A new or worsening cough can also be a sign that something in the lungs has changed. If you experience one of these problems, your doctor will first try to determine whether your symptoms are due to progression (new scar tissue in the lungs) or something else (an infection, a blood clot, a heart problem, etc.).

Spirometry		Ref	Pre Meas	Pre % Ref
FVC	Liters	3.45	1.64	48
FEV1	Liters	2.74	1.40	51
FEV1/FVC	%	79	86	
FEF25-75%	L/sec	2.69	2.23	83
FEF25%	L/sec	5.64	4.08	72
FEF50%	L/sec	3.61	5.25	145
FEF75%	L/sec	1.32	0.64	48
PEF	L/sec	6.09	5.25	86
MVV	L/min	95	2.46	59
PIF	L/sec	4.17	2.23	60
PIF50%	L/sec	3.73	7.21	
FET100%	Sec			
Lung Volumes				
VC	Liters	3.45	1.64	48
TLC	Liters	5.37	2.55	47
RV	Liters	2.01	0.91	45
RW/TLC	%	36	36	
FRC PL	Liters	3.04		
FRC N2	Liters	3.04	1.11	37
FRC He	Liters	3.04		
Vtg	Liters			
Diffusion				
DLCO	mL/mmHg/min	26.6	5.9	22
DL Adj	mL/mmHg/min	26.6	5.9	22
VA	Liters		2.32	
DLCO/VA	mL/min/L	5.18	2.55	49
IVC	Liters		1.63	

### 2. PULMONARY FUNCTION TESTS (PFTs)

Doctors routinely order one or more of the following pulmonary function tests to monitor your PF:

- Spirometry (spy-ROM-it-tree)
- Lung volume measurement
- Diffusing capacity (DLCO)

### Pulmonary Function Test (PFT) Report

See the following pages (22 and 23) for a large annotated graphic of this thumbnail test report.

**Spirometry** is performed by taking a deep breath in and then blowing out as hard and fast as you can through a mouthpiece connected to a machine. Spirometry measures how much air you can blow out (called the forced vital capacity or FVC) and how fast the air comes out. For most people with PF, monitoring the FVC can tell your doctor whether your disease is stable or progressing and whether your disease is mild, moderate, or severe. It's easiest to track your FVC expressed as a percentage of normal. Normal is between 80 and 100% of the predicted value (which is based on your age, gender, height, and ethnicity). As more scar tissue develops in your lungs, your FVC will go down.

**Lung volume measurements** can tell how much air your lungs can hold. Just like FVC, the results are expressed as a percentage of normal. The result that is usually tracked is called the total lung capacity (TLC). Normal is between 80 and 100% of the predicted value for age, gender, height, and ethnicity and decreases as more scar tissue develops in the lungs. This test is not performed as frequently as spirometry as it is more costly and time consuming and provides similar information as spirometry.

**Diffusing capacity (DLCO)** measures how easy it is for gases (like oxygen) to move from your lungs into your bloodstream. Scar tissue makes the transfer of oxygen less efficient. Just like FVC and TLC, DLCO is expressed as a percentage of the normal value and goes down as more scar tissue develops in the lungs. Do not be surprised by a DLCO that is much lower than the FVC or TLC—that is expected in people living with PF.

## PULMONARY FUNCTION TEST (PFT) RE-

**Spirometry is a test of your ability to breath out as hard and fast as you can.**

It is the primary test that tells us how healthy your lungs are. Each row below is a different measurement obtained during a Spirometry test. The first three rows are the most important ones. We will only focus on those three.

**FVC stands for "Forced Vital Capacity."**

This is the amount of air that comes out when you take a deep breath and then blow out as hard as you can.

**The FEV1 is the amount of air that came out immediately** (in the first second) when you blow out as hard and fast as you can. This number is often reduced in PF.

**This result is often called the "F.E.V.1.F.V.C. ratio."**

It is simply the FEV1 divided by the FVC and expressed as a percentage. A normal FEV1/FVC is greater than 70% — and most people with PF have an FEV1/FVC that is normal. A reduced FEV1/FVC suggests other kinds of lung diseases, such as asthma or COPD.

**This test literally measures how much air is sitting in your lungs.** We usually pay attention to the "TLC", since it is the best test of how much air your lungs can hold.

**"TLC" stands for Total Lung Capacity.**

It is the amount of air that your lungs can hold when you take the deepest breath possible.

A normal % Predicted value is greater than about 80%.

**A diffusion capacity test measures how easily gases (like oxygen) can move from the inside of the air sacs into the bloodstream.** The most important result in this section is the "DLCO", which is the primary diffusion test result.

### Spirometry ←

→ FVC	Liters
→ FEV1	Liters
→ FEV1/FVC	%
FEF25-75%	L/sec
FEF25%	L/sec
FEF50%	L/sec
FEF75%	L/sec
PEF	L/sec
MVV	L/min
PIF	L/sec
FIF50%	L/sec
FET100%	Sec

### Lung Volumes

→ VC	Liters
→ TLC	Liters
RV	Liters
RV/TLC	%
FRC PL	Liters
FRC N2	Liters
FRC He	Liters
Vtg	Liters

### Diffusion

→ DLCO	mL/mmHg/min
DL Adj	mL/mmHg/min
VA	Liters
DLCO/VA	mLmHg/min/L
IVC	Liters

Ref	Pre Meas	Pre % Ref
3.45	1.64	48
2.74	1.40	51
79	86	
2.69	2.23	83
5.64	4.08	72
3.61	5.25	145
1.32	0.64	48
6.09	5.25	86
95		
4.17	2.46	59
3.73	2.23	60
	7.21	
3.45	1.64	48
5.37	2.55	47
2.01	0.91	45
38	36	
3.04		
3.04	1.11	37
3.04		
26.6	5.9	22
26.6	5.9	22
	2.32	
5.18	2.55	49
	1.63	

“Ref” stands for “Reference. Sometimes this column is also labeled “Predicted.” These are the test results we expect if your lungs are healthy.

This is the actual result of each test. For example, for FVC, we expected this person to be able to blow out 3.45 Liters of air (Reference value), but he or she was only able to blow out 1.64 Liters of air. A low FVC is common in people with PF.

This column is often called “Percent Predicted.” It is calculated as the Measured value divided by the Reference (predicted) value. A normal value in someone with healthy lungs might range from 80% to over 100%.

In this case, the FVC is 48% of the predicted value. Since 48% is lower than the normal range of 80% or more, the FVC in this case is abnormal. A low FVC is common in people living with PF. This is one of the most important test results that doctors pay attention to. As PF worsens, FVC tends to go lower and lower.

In this case the TLC is 47% of the normal value. Most people with PF have a low TLC.

### 3. WALKING TESTS

Doctors use a walking test to see how far you can walk (usually in six minutes) and how low your oxygen levels go when you walk. Sometimes the test is performed when you are using oxygen, and sometimes it is performed without oxygen. Doctors will compare your walking distance and your oxygen levels to your results from previous visits to determine whether your lung disease has progressed. Doctors often also use this test to determine whether or not you need to use supplemental oxygen while exerting yourself in your home and outdoors.

### 4. HRCT SCANS

A high resolution computed tomography (HRCT) scan is a test involving X-rays that lets your doctor see a picture of your lungs. An HRCT scan gives a much clearer picture of your lungs than a regular chest X-ray or even a regular CT scan. An HRCT scan is a necessary test to diagnose PF. When repeated later on, comparing the change between two HRCT scans can tell your doctor if there is more scarring in the lungs. Some doctors perform multiple HRCT scans each year, some once per year, and some only when there is a change in your symptoms, spirometry, or walking test results. An HRCT protocol recommended by the PFF and PF experts in the United States is available on the PFF website at [bit.ly/pffhrct](https://bit.ly/pffhrct).

There are other tests doctors sometimes use to better understand your disease and its impact on your health, including arterial blood gas testing, cardiopulmonary exercise testing, and an echocardiogram. An arterial blood gas directly measures the amount of oxygen and carbon dioxide in your bloodstream and helps your doctor understand how well or poorly your lungs are functioning. Some forms of PF may also affect your heart. To investigate this possibility, your doctor might order an echocardiogram (or ultrasound) of the heart or a cardiopulmonary exercise test.

# clinical trials

## What are clinical trials?

Clinical trials are research studies that explore whether a medical strategy, treatment, or device is safe and effective for humans. These studies also may show which medical approaches work best for certain illnesses or groups of people. Clinical trials produce the best data available for health care decision making. The purpose of clinical trials is research, so the studies follow strict scientific standards. These standards protect patients and help produce reliable study results.

## Should I participate in a clinical trial?

Each potential participant should make an informed and voluntary decision regarding participation in clinical trials. Clinical trials are not intended to act as a treatment for individual patients; they are intended to provide information about a therapy or outcome that may benefit a population of patients. If a patient wants to participate in trials, he or she should have a full understanding of the specifics of participation. Participants should discuss their participation with family and caregivers if they wish, as well as their healthcare providers to ensure complete understanding of the trial.

## Why are clinical trials important?

Research is the pathway to finding new therapies, improving quality of life, and ultimately discovering a cure for pulmonary fibrosis. An important way that patients can help advance research is by joining clinical trials that study the effectiveness of therapies in development. By participating in clinical studies, patients provide crucial data to researchers. New treatments must demonstrate safety and efficacy before receiving approval from the FDA. Additional information

about clinical trials and their importance is available on the PFF website at [pulmonaryfibrosis.org/clinicaltrials](https://pulmonaryfibrosis.org/clinicaltrials).

## Where can I find a clinical trial?

The PFF has developed tools to make searching for and learning about clinical trials easier for patients and their families.

### PFF CLINICAL TRIAL FINDER

The PFF Clinical Trial Finder is a tool designed to help people living with pulmonary fibrosis, their caregivers, and loved ones to learn about clinical research opportunities closest to them. This searchable platform filters the type of trial, patient characteristics, and proximity to home thereby accelerating enrollment and ultimately the development of new treatment options for patients. If users are interested in learning more about any particular study, they should contact the listed site coordinator via email or phone.

The PFF Clinical Trial Finder obtains information directly from ClinicalTrials.gov, a service of the National Institutes of Health, which provides details on publicly and privately supported clinical trials. We strongly recommend that patients consult with their healthcare provider about the trials that may interest them and refer to our terms of service. Get started at [trials.pulmonaryfibrosis.org](https://trials.pulmonaryfibrosis.org).

### PF DRUG DEVELOPMENT PIPELINE

The PFF has launched a PF Drug Development Pipeline tool to educate the PF community about drug and device development in relevant areas of interstitial lung disease, including IPF, HP, RA-ILD, SSc-ILD, Sarcoid-ILD, chronic cough and lung transplant.

The PF Drug Development Pipeline includes interventions, such as novel drugs or devices, that are in development or have been approved for the market in the United States. This tool allows

viewers to filter trials by Study Phase, Disease Condition, Intervention Type, and Funder Type so that they can find information that is relevant to them. All drugs with active clinical trials link to study-specific information in the PFF Clinical Trial Finder on the PFF website. Additional features of the Pipeline tool include a Drug Development Pipeline Glossary, a User Feedback Form, and links to pertinent news and announcements about drug development and corresponding trials. Access the PF Drug Development Pipeline at [pulmonaryfibrosis.org/clinicaltrials](https://pulmonaryfibrosis.org/clinicaltrials).

# maintaining your health

## What is it like to live with pulmonary fibrosis?

First of all, you should know that **each person's experience living with PF is different**. There is no "usual" experience. Some people have severe symptoms, and some have none at all. Some people live an almost normal life for many years, but others experience severe symptoms on a daily basis. The following discussion focuses on the common experiences that many, but not all, people living with PF report.

Early on, people with PF may have no symptoms at all. The most common symptoms are **breathlessness during exertion**, bothersome **cough**, and **fatigue** (tiredness). As the lungs develop more scar tissue, symptoms worsen. Shortness of breath initially occurs with exercise, but as the disease progresses patients may become breathless while taking part in everyday activities, such as showering, getting dressed, speaking on the phone, or even eating.

People living with PF share many of the same challenges faced by

people living with other chronic diseases, such as

- dealing with side effects from medications and other therapies;
- distress resulting from the symptoms of your disease;
- frustration resulting from the limits your disease places on your body;
- inconvenience and cost of medical tests and frequent doctor visits, sometimes far from home; and
- worrying about the “three big Fs”—your family, your finances, and your future.

In addition, people living with PF often need to use oxygen therapy, a burdensome but important treatment that may add inconvenience and frustration to your life.

Over time, most people with PF experience **progression** of their disease. When doctors use the term progression, they mean that there is more scar tissue in your lungs, which usually shows up on breathing tests (lower numbers or percentages on spirometry and diffusing tests). More scar tissue can make you feel more breathless than before. Often progression also means that you might need to start using oxygen, use more oxygen than before, or use oxygen for longer and longer periods during the day or night.

It is important for you to understand that everyone seems to experience his or her own pattern of progression. Remember, there is no “usual” experience. Some people remain stable for years before progression begins. Others seem to have rapid progression starting from the time they receive their diagnosis. Most people fall in between these two extremes, with periods of stability alternating with periods of progression. Some people will develop more rapid progression over days or weeks—this is called an “acute exacerbation.” This type of progression is a little bit different and will be covered in the “Acute Exacerbation” section on page 30.

**“I would advise others to stay as healthy as possible. Get into a regimen of working out with oxygen, supervised if you can, to try to ward off advancement. It’s so individualized, you never know – it may never progress, or it may progress fast.”**

PATIENT LIVING WITH PF

## **What is the life expectancy for someone living with PF?**

Pulmonary fibrosis, particularly idiopathic pulmonary fibrosis, is a serious, life-limiting illness. It is very important to understand what type of PF you have as different forms of PF affect patients in different ways. It is difficult to predict how long you will live. An important goal is to learn to live with this diagnosis despite its serious nature.

No one can tell you how long you will live with PF. You may have read or been told that, on average, people with IPF live three, four, or five years, but this statistic only applies to people diagnosed with idiopathic pulmonary fibrosis, and the statistics for the many other kinds of PF can be different and are often better. Also, there is a lot of variation in how long people with PF live. If four years were the average, that would mean that half of people living with PF live longer than four years. Some live much longer.

Your doctors may be able to give you more information about their expectations (another way of saying “on average”) of what’s to come for you. For example, people with very low breathing tests, rapidly progressing disease, and those who require a lot of oxygen do more poorly than others.

If your doctor believes you are at risk to die in the next six months, they may recommend that you receive hospice care, which is intended to help people who are dying have peace, comfort, and dignity. People receiving hospice care receive treatments to control pain and other symptoms. Hospice care also provides support to families. Care may be provided at a hospice center, but can also be done in nursing facilities, hospitals, or often at home. You can learn more at:

- [caringinfo.org](http://caringinfo.org)
- <http://nia.nih.gov/health/end-of-life>
- [prepareforyourcare.org](http://prepareforyourcare.org)

## I heard that some people with pulmonary fibrosis develop an “acute exacerbation.” What is that?

Some people with PF can develop a sudden worsening of their condition over the course of days or weeks, including increasing breathlessness and cough, the need for more oxygen, and consideration of hospitalization. Sometimes there is an obvious explanation, such as pneumonia (a lung infection), heart problems, or blood clots in the lung (a pulmonary embolism). But in many cases, things are worse without an alternative explanation and represent rapid worsening of the underlying pulmonary fibrosis — this is called an “acute exacerbation.”

Doctors can recognize an acute exacerbation based on your symptoms, oxygen levels, CT scan results, and other tests. Usually doctors will find that your lungs have “ground-glass” on your CT scan, which means that some parts of your lungs appear grey or hazy. This ground-glass effect can result from infection, fluid build up, inflammation, or microscopic injury to the lungs.

An acute exacerbation can be a serious event, and for many it can be fatal within days or weeks. When an acute exacerbation is severe and

the lungs are failing to put enough oxygen into the bloodstream, doctors often bring up the possibility of being treated with a mechanical ventilator (a “breathing machine”) attached to a plastic tube that is placed through your mouth into your windpipe. Most people with a PF exacerbation treated with a mechanical ventilator do not survive very long. It is very important that you discuss this possibility with your doctors before this kind of treatment is needed, so that you can make your wishes known regarding this kind of invasive therapy. While this is a sensitive topic, it is important to discuss with your decision maker to ensure your wishes are honored as your health declines.

## for the caregiver

**I am a caregiver for someone with pulmonary fibrosis.**

### **What do I need to know?**

Whether you are spouse, partner, child, or friend, caring for someone through a serious illness is a challenge. There are only two rules.

- 1 Do what needs to be done each day to take care of your loved one.
- 2 Take care of yourself. The day-to-day life of a caregiver can be physically and emotionally draining.

Here are some suggestions that may help you be an effective caregiver.

### **Be actively involved in your loved one’s medical treatment.**

- The diagnostic and treatment journey for patients with PF can be confusing and difficult. By participating and helping them in their journey, you may alleviate anxiety and frustration for both you and your loved one.

- Bring a list of questions that you want to ask your medical team.
- Go to doctor appointments and write everything down.
- Be prepared to advocate for the patient if you feel it is necessary.

**Know which medications your loved one takes.**

- Keep a current list of all medications and supplements.
- Print out copies of the current medication list to hand to medical staff to copy for their records.

**Know how to operate any medical equipment used by your loved one.**

- A handheld pulse oximeter is an inexpensive device that measures oxygen levels on the tip of the finger.
- Many PF patients need supplemental oxygen. Know how to operate the oxygen concentrator and tanks in case of an emergency.
- Plan for adequate oxygen when away from home.

**“It has been worrying for me. I think it’s just not knowing what the future holds. But I would tell someone else to just really try to stay positive and enjoy every day while you can.”**

PF CAREGIVER

**Let your loved one do everything he or she can for as long as possible.**

Don’t take away a PF patient’s independence any sooner than necessary, but also be prepared to take over responsibility for things your loved one may no longer be able to do now or in the future, such as

- driving;
- ordering and dispensing medications;
- paying bills, managing money, and filing tax returns; or
- buying groceries and cooking meals.

Help your loved one maintain a healthy diet and exercise program.

Learn about Advanced Directives (Living Wills & Medical Power of Attorney), Physician Orders for Life-Sustaining Therapy (POLST), and Palliative Care options, even long before they seem to be needed.

- These documents should be kept in a place where they are easy for you to find and also on file with your local hospital.
- It's important to select a person to be your decision maker when you are no longer able to make decisions, and make that person aware of your wishes and those of your loved one with PF so that he or she can honor those wishes.
- Learn more at
  - [polst.org](http://polst.org)
  - [caringinfo.org](http://caringinfo.org)

**“We look out for each other; we do for each other. Some days I do all the cooking and some days he will decide he wants to cook. Of course, he has to use oxygen, and I worry about the tubing, but he makes sure it’s out his way.”**

PF CAREGIVER

### **Help prevent respiratory infections.**

- Everyone older than six months of age in the home needs a flu vaccine every year, including you.
- Adults over age 65 need to be vaccinated against pneumococcal pneumonia. Younger adults with certain health conditions may also need to be vaccinated against pneumococcal pneumonia.
- Use hand sanitizer frequently.
- Notify your loved one’s primary care provider immediately at the

onset of a cold, cough, or flu.

- Keep your loved one away from large crowds.
- **Know when to say NO** to visits from family or friends; to sitting in a hospital emergency waiting room with sick people; or anything else you feel is unsafe for your patient. You are the best judge of what is OK or not.

**Take care of yourself.**

- **Be willing to ask for and accept help.**
- Keep up with your own medical appointments.
- Find time for yourself. Get your hair cut, schedule a massage, go to a movie, or have lunch with friends.
- Find someone you can talk to when things are hard.

# about the foundation

## The Pulmonary Fibrosis Foundation (PFF)

is proud to serve as the leading patient advocacy organization of pulmonary fibrosis and is committed to funding research to find effective therapies, and hopefully one day, a cure. By actively engaging the PF community, the PFF has developed essential programs available to those living and working with pulmonary fibrosis.

The PFF is focused on our mission to mobilize people and resources to provide access to high quality care and lead research for a cure so people with pulmonary fibrosis will live longer, healthier lives.

**“As the leading advocate for the pulmonary fibrosis community, we are dedicated to advancing the care of people living with this disease.”**

**GREGORY COSGROVE, MD / PFF CHIEF MEDICAL OFFICER**

Some of our signature programs include

- **PFF CARE CENTER NETWORK:** Improves the clinical care of those living with this disease via an expanding group of medical centers nationwide that have proven experience and expertise in treating patients with fibrotic lung diseases
- **PFF REGISTRY:** Collects accurate clinical data on patients with diverse forms of PF in order to understand the current care, treatment, and impact upon the quality of life for those living with PF
- **PFF HELP CENTER:** Provides patients, caregivers, and health care providers with up-to-date medical information; communicates the availability of support services; and provides information about essential resources
- **PFF AMBASSADOR PROGRAM:** Encourages and empowers patients, caregivers, and health care professionals to become spokespeople for the PF community on behalf of the PFF
- **PFF SUPPORT GROUP LEADER NETWORK:** Connects more than 150 local PF support groups and provides a forum for PF support group leaders to connect, exchange ideas, learn from one another, and discuss best practices for their meetings
- **PFF SUMMIT:** Facilitates an environment where world-renowned experts come together at our biennial health care conference to exchange ideas and information with fellow physicians, researchers, patients, caregivers, allied health professionals, and industry representatives

## Learn more about pulmonary fibrosis

The PFF is committed to providing quality disease education to the PF community. We provide patients, caregivers, family members, and health care providers with the resources necessary to

more fully understand PF, and provide patients with the tools necessary to live with PF and improve their quality of life.

- The **PFF DISEASE EDUCATION WEBINAR SERIES** enables the PF community to easily learn about the disease from leading PF specialists. You can view the online archive on the PFF website ([pulmonaryfibrosis.org/webinars](https://pulmonaryfibrosis.org/webinars)).
- **PFF EDUCATION MATERIALS** assist patients, caregivers, family members, and health care providers to learn more about PF. You can download materials from [pulmonaryfibrosis.org](https://pulmonaryfibrosis.org) or request hard copies through the PFF Help Center at 844.TalkPFF (844.825.5733) or [help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org).

## Get involved with the PFF

Your actions have the power to make an impact and create hope for pulmonary fibrosis patients and caregivers everywhere. You can get involved in a variety of ways.

- **FUNDRAISE FOR TEAM PFF:** Host an event that is personally meaningful to you. Organize an online fundraiser, hold a bake sale, or host a golf tournament. Take action in the fight against PF today!
- **PULMONARY FIBROSIS AWARENESS MONTH:** Let the world know September is Pulmonary Fibrosis Awareness Month! Together we can make a difference in spreading disease awareness, providing enhanced patient support, increasing essential research funding, and ultimately finding a cure.
- **VOLUNTEER FOR THE PFF:** Join a network of leaders, advocates, and fundraisers dedicated to helping the PFF fulfill its mission.

Visit [pulmonaryfibrosis.org](https://pulmonaryfibrosis.org) to learn more.

# glossary

**Acute exacerbation:** An episode of rapid worsening of a pulmonary (relating to lungs) condition

**Alveoli:** Tiny air sacs in the lungs where carbon dioxide leaves the bloodstream and oxygen enters the bloodstream

**Bronchial tree:** The series of airways connecting the trachea (wind-pipe) to the alveoli.

**Bronchus:** One of the airways of the bronchial tree (plural: bronchi)

**Bronchoscope:** A tool usually passed through the nose or mouth used for inspecting the inside of airways (bronchial tubes) of the lungs. Biopsies of the lungs can be performed by bronchoscopy.

**Comorbidity:** A disease or other medical problems that occurs simultaneously with PF. A comorbidity is typically neither a cause nor a consequence of PF.

**Diffuse parenchymal lung disease (DPLD):** Another name for interstitial lung disease

**Diffusion capacity (DLCO):** A measure of the ability of gases to diffuse into the bloodstream

**Dyspnea:** Difficulty breathing, shortness of breath, or breathlessness

**Fibroproliferation:** The growth of fibroblasts, the cells that makes scar tissue

**Fibrosis:** Scar tissue

**Forced expiratory volume (FEV1):** The amount of air you can blow out in one second after filling up your lungs with air as much as possible. Measured by a test called spirometry

**Forced vital capacity (FVC):** the amount of air you can blow out of your lungs after filling up your lungs with air as much as possible. Measured by a test called spirometry

**Gastroesophageal reflux disease (GERD):** A medical condition defined by passage of stomach contents into the esophagus (food pipe) and often into the throat. GERD can cause discomfort (heartburn or acid indigestion) and sometimes injures the lining of the esophagus. Also called acid reflux disease.

**Hospice care:** focuses on caring, not curing with an emphasis on comfort and support for patients, hospice is designated for patients with a life expectancy of six months or less certified by a physician.

**Idiopathic:** of unknown cause

**Idiopathic interstitial pneumonias (IIP):** A family of nine types of ILD of unknown cause.

**Interstitial lung disease (ILD):** A broad category of over 200 lung diseases that affect the lung interstitium. Typically, ILDs cause inflammation, fibrosis (scarring), or an accumulation of cells in the lung not due to infection or cancer.

**Interstitial:** The walls of the air sacs of the lung. Your lung is made of air, interstitium, and blood vessels. The word “interstitial” refers to the interstitium.

**Palliative care:** Non-curative therapy that treats symptoms and focuses on improving quality of life. It can be received at the same time as curative therapy

**Pathologist:** A physician specializing in disease-associated changes in tissues and organs. Pathologists look at lung tissue under a microscope to aid in medical diagnosis.

**Pulmonary:** Relating to the lungs

**Pulmonary hypertension:** Abnormal high blood pressure in the pulmonary arteries, which connect the heart to the lungs.

**Pulmonologist:** A physician specializing in the lungs

**Radiologist:** A physician specializing in using radiology tests (e.g., X-rays) to diagnose illness

**Rheumatologist:** A physician specializing in rheumatic diseases, which may include autoimmune diseases and joint diseases

**Spirometry:** A test that measures the amount of air inhaled and exhaled with each breath

**Trachea:** Windpipe that connects your voice box to your bronchial tree

**Usual interstitial pneumonia (UIP):** A specific abnormal radiologic or pathologic pattern

**Video-assisted thoracoscopy surgery (VATS):** A surgical procedure on the lungs using tools and cameras

# abbreviation list

**AIP:** acute interstitial pneumonia

**CTD-ILD:** connective tissue disease interstitial lung disease

**FPF:** familial pulmonary fibrosis

**FVC:** forced vital capacity

**GERD:** gastroesophageal reflux disease

**HRCT:** high-resolution computed tomography

**ILD:** interstitial lung disease

**IPF:** idiopathic pulmonary fibrosis

**NSIP:** non-specific interstitial pneumonia

**OSA:** obstructive sleep apnea

**PFTs:** pulmonary function tests

**RA-ILD:** rheumatoid arthritis ILD

**UIP:** usual interstitial pneumonia

**VATS:** video-assisted thoracoscopy surgery

The Pulmonary Fibrosis Foundation is thankful to the following for writing and reviewing this guide:

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