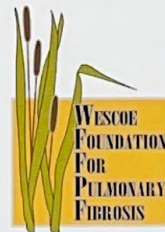


IDIOPATHIC  
PULMONARY  
FIBROSIS

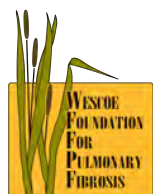


# IPF Carebook

Lung Center  
 **TEMPLE HEALTH**



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- *Care Team*
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- *What Do I Need to Know About Traveling with Supplemental Oxygen?*
- *Why Did My Provider Prescribe Supplemental Oxygen?*
- *Oxygen Basics*



# Questions for your doctor

The Pulmonary Fibrosis Foundation (PFF) has developed questions that may be helpful to you in managing your pulmonary fibrosis (PF). It is important to note that some of the questions may not be applicable depending on your situation. We encourage you to address relevant questions as you develop a relationship with your physician over multiple visits.



## DIAGNOSIS AND PROGNOSIS

- ☐ What is my specific diagnosis and what does it mean? Is a lung biopsy necessary to confirm my diagnosis?
- ☐ What other options are there for confirming the diagnosis?
- ☐ What could have caused my pulmonary fibrosis?
- ☐ How severe is my condition and what is my prognosis?
- ☐ How will pulmonary fibrosis impact what I can and cannot do?
- ☐ Will my disease progress and what are the signs that I would see to know? If it does progress what things would I notice and will I need oxygen? What kind of tests will I need and how frequently to monitor for progression?



## TREATMENT OPTIONS

- ☐ What treatments are available for my condition? How will these treatments help me?
- ☐ What are the side effects?
- ☐ Is pulmonary rehabilitation an option for me?
- ☐ Is palliative care appropriate at this time or in the future?
- ☐ Is lung transplantation an option for me? If so, when should I be referred or what is keeping me from being referred?
- ☐ Is a clinical trial of experimental therapy right for me? Where do I get information?
- ☐ What can I do about the cough?
- ☐ Do you have a support group or know of one?



## SUPPLEMENTAL OXYGEN (IF PRESCRIBED)

- ☐ Should I use oxygen all the time? Or just some of the time? What oxygen settings should I use?
- ☐ Am I able to travel with my current supplemental oxygen prescription?
- ☐ Should I be monitoring my oxygen saturation levels at home?



## FOLLOW-UP

- ☐ What should I do and whom should I contact if I have any problems with my PF?
- ☐ What should I do if I get a cold, flu, pneumonia?
- ☐ How often should I schedule follow-up appointments to monitor my condition?
- ☐ What tests or treatments can I expect during follow-up appointments? What do these tests show you and why are they important?
- ☐ What complications might I experience?
- ☐ What should I do if my symptoms suddenly worsen? Who should I call?



PA IPF  
SUPPORT  
NETWORK

# Idiopathic Pulmonary Fibrosis (IPF) Patient Checklist

A new diagnosis can be an overwhelming and scary time. You may be asking yourself, “what next?” The following IPF Patient Checklist serves as a helpful roadmap for you and your care partner to navigate your diagnosis, treatment options, and care.

## Support

☐

### Join a support group.

You're not alone! Support groups can offer additional care and connection with those who share your journey and who can relate to what you're going through.

## Medical Care and Treatment

☐

### Schedule an appointment with a pulmonologist who has experience caring for patients with IPF.

#### PFF CARE CENTER NETWORK

- **Temple Health Lung Center:** <https://www.templehealth.org/services/lung>
- **University of Pennsylvania Pulmonary, Allergy, and Critical Care Division:** <https://www.pennmedicine.org/>
- **Jane and Leonard Korman Respiratory Center at Thomas Jefferson University:** <https://hospitals.jefferson.edu/departments-and-services/jane-and-leonard-korman-respiratory-institute/>
- **Penn State Health Milton S. Hershey Medical Center, Division of Pulmonary, Allergy and Critical Care Medicine:** <https://hmc.pennstatehealth.org/pulmonary>
- **The Dorothy P. and Richard P. Simmons Center for Interstitial Lung Disease at UPMC:** <https://www.upmc.com/>

☐

### Research drug therapies approved to treat IPF.

- Pirfenidone (Esbriet®, Pirfenex®, Pirespa®)
- Nintedanib (OFEV®)

## Pulmonary Rehabilitation & Oxygen Management

☐

### Explore pulmonary rehabilitation education programs.

☐

### When oxygen levels go below 88%, talk to your doctor about a prescription for oxygen.

## Lifestyle Changes

☐

### If you are overweight, adjust your diet to have a well-balanced diet and limit your caloric intake as being overweight can contribute to shortness of breath.

☐

### If you are a smoker, tobacco cessation is imperative.

## Lung Transplantation

☐

### Schedule a lung transplant evaluation.

#### PENNSYLVANIA LUNG TRANSPLANT CENTERS:

- **Temple Health Lung Transplant Program:** [www.templehealth.org](http://www.templehealth.org)
- **Penn Lung Transplant Center:** [www.pennmedicine.org](http://www.pennmedicine.org)
- **UPMC Lung Transplant Program:** [www.upmc.com](http://www.upmc.com)

## Palliative Care

☐

### Explore palliative care options to address symptoms and advanced care planning.



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<https://paipfsupportnetwork.org>





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## Idiopathic Pulmonary Fibrosis (IPF) Patient Resources

The following are resources to supplement the checklist above. A wealth of information and support exists for IPF patients and their care partners. These resources, combined with your medical team, will help you to live your best life while managing this disease.

### Resources, Support Groups, and Online Communities:

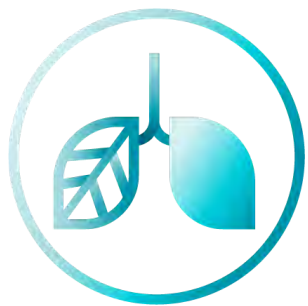
- **PA IPF Support Network:**  
[paipfsupportnetwork.org](https://paipfsupportnetwork.org)
- **Wescoe Foundation for Pulmonary Fibrosis:**  
[wescoefoundationforpulmonaryfibrosis.org/](https://wescoefoundationforpulmonaryfibrosis.org/)
- **Pulmonary Fibrosis Foundation:**  
<https://www.pulmonaryfibrosis.org/>
- **PF Warriors:**  
<https://pfwarriors.com/>
- **American Lung Association:**  
<https://www.lung.org/>
- **American Thoracic Society:**  
<https://www.thoracic.org/>
- **CHEST Foundation:**  
<https://www.thoracic.org/>
- **Responsum for PF:**  
<https://responsumhealth.com/pulmonary-fibrosis/>
- **Pulmonary Wellness Foundation:**  
<https://pulmonarywellness.org/>



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# Idiopathic Pulmonary Fibrosis (IPF) Provider Checklist

A helpful roadmap to help your patient and their care partner navigate their IPF diagnosis.

## Medical Care and Treatment

- ☐ Obtain previous CT scans or biopsy slides
- ☐ Encourage them to maintain a healthy lifestyle that includes smoking cessation, achieving a healthy weight, monitoring oxygen levels, and maintaining oxygen levels over 90% 24 hours a day.
- ☐ Discuss approved treatment options:
  - ☐ Pirfenidone (Esbriet® , Pirfenex® , Pirespa® )
  - ☐ Nintedanib (OFEV® )

## Pulmonary Rehabilitation & Oxygen Management

- ☐ Arrange pulmonary rehabilitation
- ☐ Prescribe oxygen when levels go below 88%

## Lung Transplantation

- ☐ Schedule a lung transplant evaluation

### PENNSYLVANIA LUNG TRANSPLANT CENTERS

- Temple Health Lung Transplant Program: [www.templehealth.org](http://www.templehealth.org)
- Penn Lung Transplant Center: [www.pennmedicine.org](http://www.pennmedicine.org)
- UPMC Lung Transplant Program: [www.upmc.com](http://www.upmc.com)

## Palliative Care

- ☐ Educate them on palliative care -- the benefits, including symptom management, and the importance of advanced care planning.
- ☐ Indicate a Palliative Care program within your medical center.

## Support

- ☐ Suggest support groups and resources
  - ☐ See back of document for the full list of Resources, Support Groups, and Online Communities.

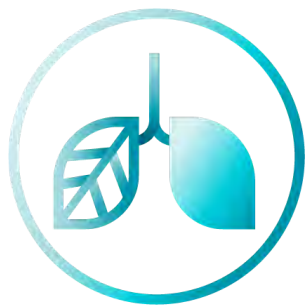


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## Idiopathic Pulmonary Fibrosis (IPF) Provider Resources

A helpful roadmap to help your patient and their care partner navigate their IPF diagnosis.

### Resources, Support Groups, and Online Communities:

- **PA IPF Support Network:**

[paipfsupportnetwork.org](https://paipfsupportnetwork.org)

- **Wescoe Foundation for Pulmonary Fibrosis:**

[wescoefoundationforpulmonaryfibrosis.org/](https://wescoefoundationforpulmonaryfibrosis.org/)

- **Pulmonary Fibrosis Foundation:**

<https://www.pulmonaryfibrosis.org/>

- **PF Warriors:**

<https://pfwarriors.com/>

- **American Lung Association:**

<https://www.lung.org/>

- **American Thoracic Society:**

<https://www.thoracic.org/>

- **CHEST Foundation:**

<https://www.thoracic.org/>

- **Responsum for PF:**

<https://responsumhealth.com/pulmonary-fibrosis/>

- **Pulmonary Wellness Foundation:**

<https://pulmonarywellness.org/>



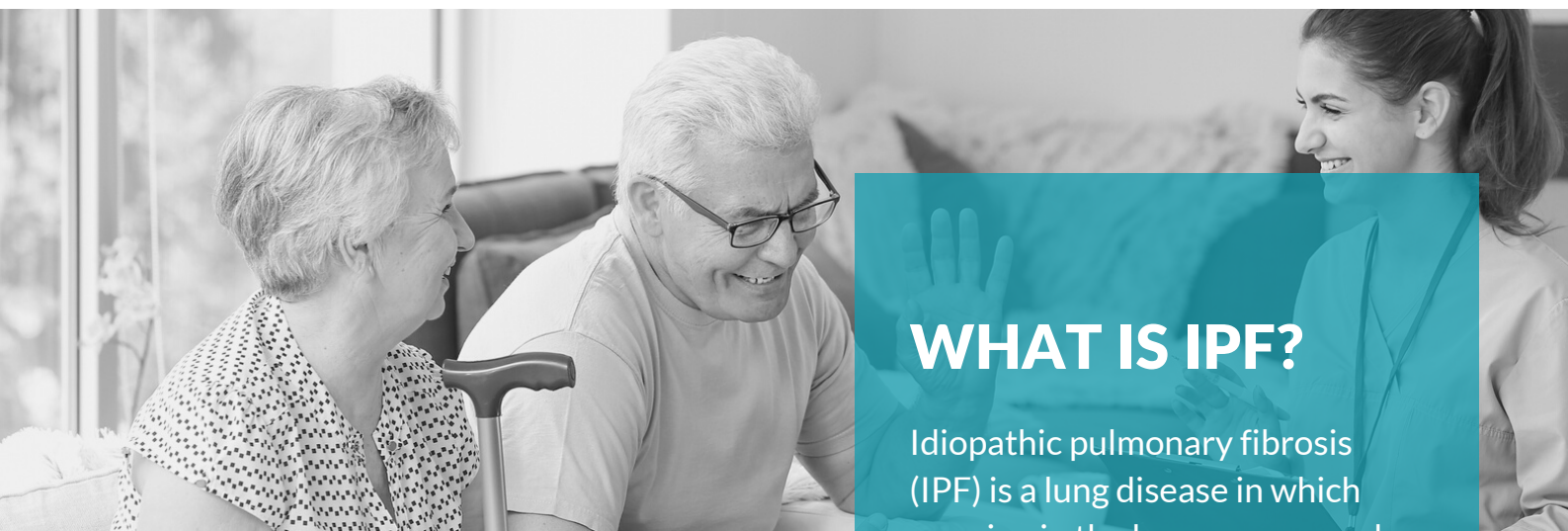
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A guide to help you with your new diagnosis



## WHAT IS IPF?

Idiopathic pulmonary fibrosis (IPF) is a lung disease in which scarring in the lungs occurs and the cause is unknown. It is part of a larger group of lung diseases characterized by thickening of the walls of the air sacs known as alveoli. As the disease progresses, the air sacs gradually become replaced by fibrotic tissue or scar tissue. As the scar tissue becomes thicker, it leads to stiffness in the lungs, making it difficult to breathe.

## QUICK FACTS:

- IPF impacts 5,000,000 patients globally each year
- More than 132,000 US citizens have IPF
- More than 50,000 new cases are diagnosed each year

## The Beginning of Your IPF Journey

A new diagnosis can be overwhelming and frightening. You may be looking for information and answers. You may be looking to get a jump start on treatment options. You may also be looking for others who know what you're going through.

## What's Next?

You have options. Knowing what comes next can help you and your health care provider determine the next best steps in your course of treatment. The following are the options you should begin to consider as you move forward on your treatment path:

- Medical Treatment
- Pulmonary Rehabilitation/Oxygen Management
- Lung Transplantation
- Palliative Care
- Support for Patients and Care Partners



## Medical Treatment

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

Several organizations have more information on Pirfenidone and Nintedanib as well as other medications that help to treat symptoms of IPF including the Pulmonary Fibrosis Foundation and the American Lung Association.

## Pulmonary Rehabilitation/Oxygen Management

Pulmonary rehabilitation is a program of education and exercise that helps you manage your breathing problem, increase your stamina, and decrease your breathlessness. You will be educated on taking charge of your breathing--learning how to pace your breathing with your activities, how to take your medicines, and even how to talk to your health care provider.

learning how to pace your breathing with your activities, how to take your medicines, and even how to talk to your health care provider.

Oxygen is prescribed when oxygen levels go below about 88%. Your doctor may choose to measure your oxygen level in different circumstances: while sitting, while walking in the hallway or on a treadmill, and during sleep. If your oxygen level drops below about 88%, your doctor may prescribe supplemental oxygen to prevent your oxygen level from dropping further.

## Lung Transplantation

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.

More than 15% of all lung transplants in the United States performed in Pennsylvania hospitals.

## Palliative Care

Palliative care—also referred to as supportive care—is a 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.



**Transplant Centers within the state include:**

- Penn Lung Transplant Center: [www.pennmedicine.org](http://www.pennmedicine.org)
- UPMC Lung Transplant Program: [www.upmc.com](http://www.upmc.com)
- Temple Health Lung Transplant Program: [www.templehealth.org](http://www.templehealth.org)

## Support for Patients and Care Partners

Support groups can offer additional care and connection with those who share your journey and who can relate to what you're going through. In addition to helping put you at ease, talking with others who are in the same situation can help you to learn new information or ask questions you hadn't thought about. In addition to receiving additional information about IPF, studies have shown that participation in support groups helps patients and care partners feel less isolated and improves the outlook on their condition.

In addition to in-person support groups, many online communities exist where patients can connect virtually.

*In affiliation with the Wescoe Foundation for Pulmonary Fibrosis.*

## Resources:

- Wescoe Foundation for Pulmonary Fibrosis: [wescoefoundationforpulmonaryfibrosis.org/PA](http://wescoefoundationforpulmonaryfibrosis.org/PA)
- IPF Patient and Caregiver Support Network: [paipfsupportnetwork.org](http://paipfsupportnetwork.org)
- Pulmonary Fibrosis Foundation: [pulmonaryfibrosis.org](http://pulmonaryfibrosis.org)
- American Lung Association: [lung.org](http://lung.org)
- American Thoracic Society: [thoracic.org](http://thoracic.org)
- CHEST Foundation: [thoracic.org](http://thoracic.org)

## PFF Care Center Network:

- Temple Health Lung Center: <https://www.templehealth.org/services/lung>
- University of Pennsylvania Pulmonary, Allergy, and Critical Care Division: <https://www.pennmedicine.org/>
- Jane and Leonard Korman Respiratory Institute at Thomas Jefferson University: <https://hospitals.jefferson.edu/departments-and-services/jane-and-leonard-korman-respiratory-institute/>
- Penn State Health Milton S. Hershey Medical Center, Division of Pulmonary, Allergy and Critical Care Medicine: <https://hmc.pennstatehealth.org/pulmonary>
- The Dorothy P. and Richard P. Simmons Center for Interstitial Lung Disease at UPMC: <https://www.upmc.com/>

## Support Groups and Online Communities:

- PF Warriors: <https://pfwarriors.com/>
- Responsum for PF: <https://responsumhealth.com/pulmonary-fibrosis/>



## PENNSYLVANIA IPF Support Network

### What is the Pennsylvania Idiopathic Pulmonary Fibrosis (IPF) Support Network?

The PA IPF Patient and Caregiver Support Network is dedicated to raising awareness of IPF and providing support and education to patients, families, and healthcare providers throughout the Commonwealth.

### Supporting Patients, Families, and Providers

We know that being diagnosed with a disease can be confusing and overwhelming. Our support network was created to educate patients and their care partners about IPF and interstitial lung diseases (ILD), while providing support, resources, and connecting them to the treatment and care they need.

### Who we are

We are a support network dedicated to connecting IPF patients, their care partners, and healthcare providers to each other as well as important resources and information.

### Who we support

We focus on providing information and resources to patients, care partners, and healthcare providers.

### Connect With Us

Find us on Facebook, Twitter, and Instagram!



@paipfsupportnetwork



@paipfnetwork



@paipfsupportnetwork

Subscribe to our newsletter:  
Visit [paipfsupportnetwork.org/](https://paipfsupportnetwork.org/)



### Resources We Provide

We offer patients, families, and providers one place to find more information about:

- IPF and interstitial lung disease (ILD)
- Providers including the Pennsylvania Centers of Excellence
- Treatment options including oxygen management, transplant, and clinical trials
- Pennsylvania IPF support groups
- Awareness events and organizations that support IPF patients and families

### Downloadable Patient and Provider Resources

Also available on our website are helpful resources that patients, providers, and care partners may access online or download and print.

- Patient Guide: a helpful one-sheet for newly diagnosed patients that provides important information on managing their disease including medical treatment, pulmonary rehabilitation and oxygen management, lung transplant, palliative care, and finding support.
- Patient Checklist: a helpful checklist for patients and care partners to navigate and manage IPF. This includes where to find specialized treatment through the PA Centers of Excellence, when to start oxygen, recommended lifestyle changes, lung transplant centers, and more.
- Provider Checklist: a checklist to help providers navigate the treatment and management of IPF for their patients.

### Listen in!



The PA IPF Support Network is proud to present the Pulmonary Fibrosis Podcast, a first-of-its-kind! Tune in on Apple Podcasts, Google, Spotify, Amazon Music, Deezer, Stitcher, and Tunein.



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<https://paipfsupportnetwork.org>



# pulmonary fibrosis information guide



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

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10	diagnosis & treatment options
25	clinical trials
27	maintaining your health
31	for the caregiver
35	about the foundation
38	glossary
41	abbreviation list



# 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.

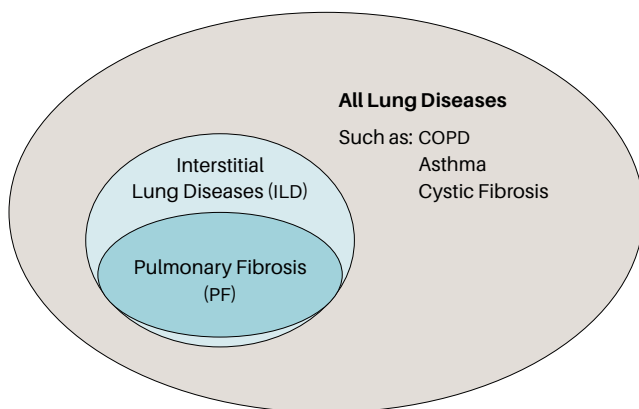
We are here to help. Contact the **PFF Help Center** with any questions or concerns you have about PF and available resources during the course of your care. Our staff can be reached at **844.TalkPFF** (844.825.5733) or [help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org).

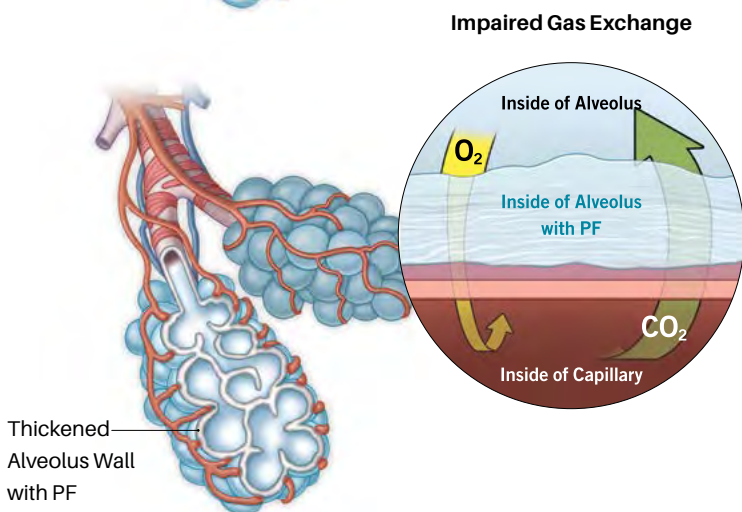
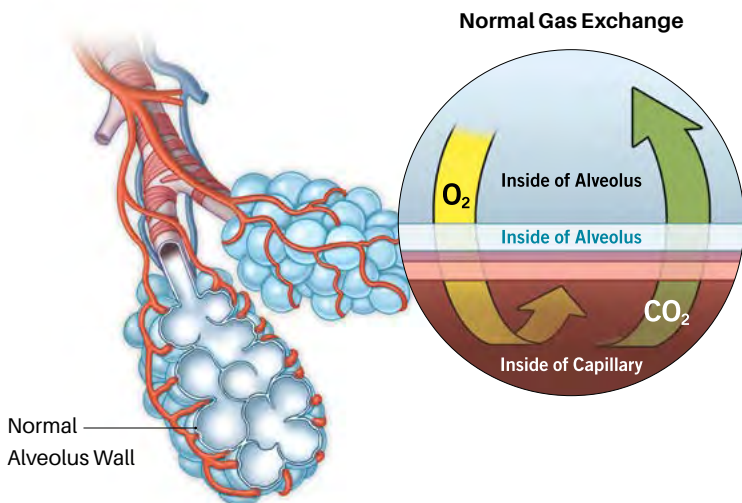
# about pulmonary fibrosis

## 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.

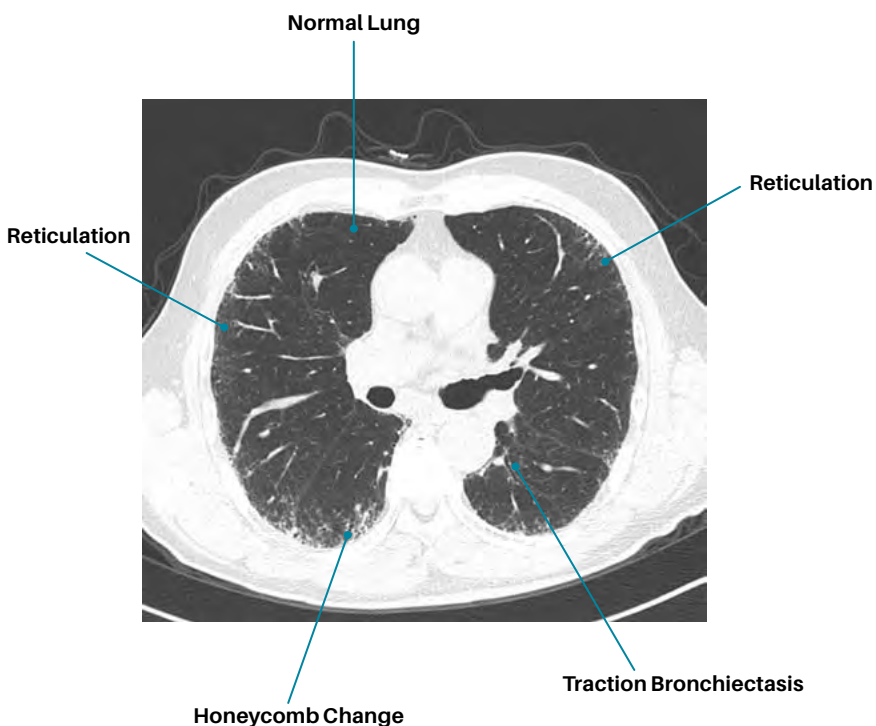
- **“CRACKLES”** – Your doctor may have told you that **“crackles” were heard** in your lungs. Crackles (also called “rales”) sound like Velcro being pulled apart. They are heard in many lung diseases because any type of problem affecting the air sacs (such as PF, pneumonia, or a buildup of fluid in 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		
PIF	L/sec	4.17	2.46	59
FIF50%	L/sec	3.73	2.23	60
FET100%	Sec		7.21	
Lung Volumes				
VC	Liters	3.45	1.64	48
TLC	Liters	5.37	2.55	47
RV	Liters	2.01	0.91	45
RVTLC	%	38	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	mLmHg/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	<p>• <b>"Ref" stands for "Reference.</b> Sometimes this column is also labeled "Predicted." These are the test results we expect if your lungs are healthy.</p>
2.74	1.40	51	
79	86		<p>• <b>This is the actual result of each test.</b> 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.</p>
2.69	2.23	83	
5.64	4.08	72	<p>• <b>This column is often called "Percent Predicted."</b> 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%.</p>
3.61	5.25	145	
1.32	0.64	48	<p>• <b>In this case, the FVC is 48% of the predicted value.</b> 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.</p>
6.09	5.25	86	
95			<p>• <b>In this case the TLC is 47% of the normal value.</b> Most people with PF have a low TLC.</p>
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		

### 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 (heart-burn 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:

**DAVID J. LEDERER, MD, MS**

FORMER PFF SENIOR MEDICAL ADVISOR, EDUCATION AND AWARENESS

**THE PFF MEDICAL TEAM**

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University of Michigan Health System

Ann Arbor, MI

STEERING COMMITTEE CHAIR, PFF CARE CENTER NETWORK

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San Francisco, California

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Special thanks to **Myrna Taylor** for writing the "For the Caregiver" section.





**PULMONARY FIBROSIS FOUNDATION**

230 EAST OHIO STREET, SUITE 500

CHICAGO, ILLINOIS 60611

**phone** PFF Help Center

844.TalkPFF (844.825.5733)

**email** [help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org)

[pulmonaryfibrosis.org](http://pulmonaryfibrosis.org)

## Order FREE educational resources

The PFF Help Center, a dedicated call center, offers patients, caregivers, and healthcare providers the most up-to-date medical information, communicates the availability of support services, and information about other essential resources. Call or email us today to order FREE educational resources!



844.TalkPFF



Oxygen Information Line  
(844.825.5733)



help@pulmonaryfibrosis.org

**FOLLOW US!** Keep up with the latest news from the PFF by following us at [@pfforg](#) on social media!



230 East Ohio Street, Suite 500  
Chicago, Illinois 60611  
**844.TalkPFF** (844.825.5733)  
help@pulmonaryfibrosis.org  
[pulmonaryfibrosis.org](#)

## You can make a difference

You have the power to make an impact and create hope for PF patients everywhere. Start by making a donation to support the PFF in its mission to help find a cure.

**PFF Government Affairs:** Advocate and engage with lawmakers regarding the needs of the PF community.

**PFF Walk:** Build a team, raise funds, and lead the way toward a world without PF by joining one of our national walks. You can participate in-person or virtually.

**Fundraise for Team PFF:** Create and host an event that is personally meaningful to you. Organize an online fundraiser, hold a bake sale, or host a community walk.

**PF Awareness Month:** Throughout September, get involved by participating in disease education days, support group events, fundraisers, or with #BlueUp4PF.

**Shop PFF:** Support the cause when you wear wristbands, t-shirts, caps, and more. Visit [Shop-PFF.com](#).

### Make a Gift

You can support the PFF's important programs by making a donation today. Visit [pulmonaryfibrosis.org](#) to learn about the many ways you can give.

## Our research

The Foundation places enormous importance on creating an environment that will assist in the development of effective treatments for pulmonary fibrosis. The PFF is committed to funding research to identify new therapies and one day, a cure.

**PFF Patient Registry:** A resource of data gathered from more than 2,000 patient volunteers across the country to help researchers further understand the disease and develop treatments. The data collected can be used to better diagnose, treat, and potentially cure pulmonary fibrosis.

**PFF Scholars:** A new grant program to support and provide career development opportunities for early stage investigators.

**PFF Clinical Trial Finder:** A tool to help patients navigate feasible and relevant clinical trials. This searchable platform filters the type of trial, patient characteristics, and proximity to one's home thereby accelerating the development of new treatment options for patients. Visit [trials.pulmonaryfibrosis.org](#) to get started.

**PF Drug Development Pipeline:** A tool that allows viewers to filter interventions, such as novel drugs or devices that are in development or have been approved for the U.S. market, by relevant areas of interstitial lung disease. Visit [pulmonaryfibrosis.org/clinicaltrials](#) for more information.

We imagine a world without pulmonary fibrosis



Pulmonary  
Fibrosis  
Foundation



trusted resource



research



education



advocacy



collaboration



fundraising

[pulmonaryfibrosis.org](#)



# About the Pulmonary Fibrosis Foundation

The **Pulmonary Fibrosis Foundation (PFF)** mobilizes people and resources to provide access to high quality care and leads research for a cure so people with pulmonary fibrosis (PF) will live longer, healthier lives.

## The Trusted Resource

The PFF is the nation's leading pulmonary fibrosis patient education and advocacy organization. From providing disease education materials to creating the largest patient registry focused on all-cause PF and establishing a nationwide care center network, the PFF is dedicated to improving all aspects of a patient's quality of life.

We work with leading experts in the field of pulmonary fibrosis. The physicians on our world-class medical team, investigators at each Care Center Network site, and members of the Medical and Scientific Advisory Committee, Research Review Committee, and Medical Advisory Board champion collaborative efforts in the care of patients and research.

The PFF is at the forefront of research for different lung diseases that make up the family of pulmonary fibrosis diseases, including idiopathic pulmonary fibrosis (IPF).

**As the leading advocate for the PF community, we are building better tools and resources for diagnosing and treating people living with PF. We invite you to join us in the drive toward a cure.**

William T. Schmidt  
PFF President and Chief Executive Officer

# About pulmonary fibrosis

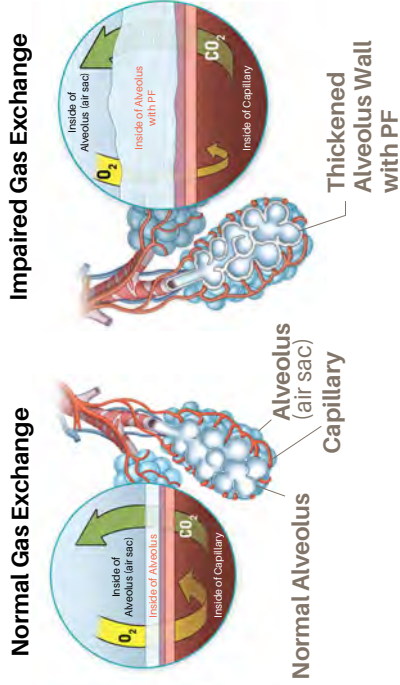
**Pulmonary fibrosis** is a family of more than 200 different lung diseases that share common symptoms of cough and/or breathlessness. Idiopathic pulmonary fibrosis is one form of PF. The PF family of lung diseases falls into an even larger group of diseases called "interstitial lung diseases (ILD)."

The word "pulmonary" means "lung" and "fibrosis" means scar tissue - so in its simplest sense, pulmonary fibrosis means scarring of the lungs.

Pulmonary fibrosis may cause progressive scarring in the lungs, limiting a person's oxygen intake. However, every individual diagnosed with pulmonary fibrosis has a unique experience with the disease and there is no "standard" or expected clinical course.

**With no known cure**, certain forms of PF may take the lives of patients within three to five years from diagnosis.

The good news for people living with PF is that there are treatments designed to specifically manage the symptoms of the disease and researchers are studying new ways to halt its progression.



# Community programs

**PFF Care Center Network:** 68 medical centers nationwide offering comprehensive and individualized care to those living with PF.

**PFF Summit:** Our biennial healthcare conference had over 950 attendees in 2019, including patients, caregivers, physicians, nurses, and researchers, with 10 countries and 44 states represented.

**PFF Help Center** The hub for information and resources for patients, families, and healthcare professionals. The PFF Help Center gets inquiries from nearly every U.S. state and 40 countries.

**PFF Support Group Network:** Over 150 volunteer groups around the country providing a safe environment for those affected by PF to share their experiences.

**Virtual Support Groups:** Four unique telephone-based support groups for those who are unable to attend in-person meetings including: PFF Voices for everyone in the PF community; PFF Caring Conversations for caregivers; PFF Coloquio for Spanish speakers; and the Lung Transplant Community Support Group for lung recipients or those awaiting transplant.

**PFF Ambassadors:** Volunteers impacted by PF who offer hope and inspiration to the community and promote disease awareness.

**PFF Educational Videos:** Watch videos from our Life With Pulmonary Fibrosis Video Series, Disease Education Webinar Series, and Advocacy in Action Webinar Series to learn more about a variety of topics.

**PFF Oxygen Information Line:** Provides information and resources to individuals using supplemental oxygen and to caregivers.

# Quick facts



## DIAGNOSIS

**One in 200** over the age of 70 are living with IPF.



## CASES

**50,000** new cases are diagnosed annually.



## PREVALENCE

**200,000+** people are living with IPF in the U.S.



## SYMPTOMS

- Shortness of breath
- Dry, hacking cough
- Fatigue and weakness
- Discomfort in chest



## POSSIBLE CAUSES

- Environmental
- Autoimmune diseases
- Drug-induced
- Radiation-induced
- Occupational

Often a cause cannot be identified. These diseases are called idiopathic interstitial pneumonias. One type is idiopathic pulmonary fibrosis.



## TREATMENTS

- Supplemental oxygen
- Prescription medication
- Pulmonary rehabilitation
- Lung transplantation
- Medications

To all of our patients and families living with Idiopathic Pulmonary Fibrosis (IPF),

When you are diagnosed with IPF, there are a lot of questions about the disease and your care. This IPF Carebook serves as a personal resource to help you and your carepartner(s) communicate with your healthcare professional team and to maintain important information for your IPF care needs.

The IPF Carebook is designed to keep you organized, empowered and supported through your journey. Bring your Carebook to each of your clinic and healthcare appointments and share this book to help coordinate your care.

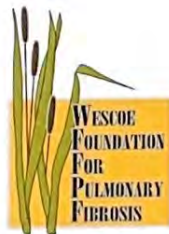
We hope you will find this resource helpful as we work together as partners in your care. Please feel free to contact us at 484-553-6340 or [jennifer@wescoefoundationforpulmonaryfibrosis.org](mailto:jennifer@wescoefoundationforpulmonaryfibrosis.org), if you have any questions.

Most sincerely,



Jennifer H. Wescoe, M.Ed., NCC

*Executive Director  
Wescoe Foundation for Pulmonary Fibrosis*



#### ABOUT THE WESCOE FOUNDATION:

The Wescoe Foundation for Pulmonary Fibrosis is a 501(c) 3 nonprofit organization that provides support, education, resources and advocacy for patients and their carepartners living with Idiopathic Pulmonary Fibrosis (IPF) in order to sustain the highest possible quality of life.



#### ABOUT THE TEMPLE LUNG CENTER:

The Temple Lung Center helps people with serious lung disease breathe easier and live more active lives. The Center is a national leader in diagnosing and treating both common and complex lung problems.







# My Care Planning

## About Me

NAME \_\_\_\_\_

DIAGNOSIS \_\_\_\_\_



# Medications

MY PHARMACY \_\_\_\_\_

ADDRESS \_\_\_\_\_

PHARMACY PHONE NUMBER \_\_\_\_\_

MEDICATION NAME \_\_\_\_\_

TIME OF DAY \_\_\_\_\_ DOSAGE \_\_\_\_\_

MEDICATION NAME \_\_\_\_\_

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## Medications

### Notes on medications



## Care Team

**PRIMARY CARE PHYSICIAN** \_\_\_\_\_

CONTACT INFO \_\_\_\_\_

**PULMONOLOGIST** \_\_\_\_\_

CONTACT INFO \_\_\_\_\_

**NURSE PRACTITIONER** \_\_\_\_\_

CONTACT INFO \_\_\_\_\_

**RESPIRATORY THERAPIST** \_\_\_\_\_

CONTACT INFO \_\_\_\_\_

**NUTRITIONIST** \_\_\_\_\_

CONTACT INFO \_\_\_\_\_





## Care Team

**OTHER** \_\_\_\_\_

CONTACT INFO \_\_\_\_\_

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## Appointment Notes

DATE \_\_\_\_\_

APPOINTMENT DESCRIPTION \_\_\_\_\_

NOTES \_\_\_\_\_

\_\_\_\_\_

\_\_\_\_\_

FOLLOW-UP FOR NEXT APPOINTMENT \_\_\_\_\_

\_\_\_\_\_

QUESTIONS FOR MY PHYSICIAN \_\_\_\_\_

\_\_\_\_\_

DATE \_\_\_\_\_

APPOINTMENT DESCRIPTION \_\_\_\_\_

NOTES \_\_\_\_\_

\_\_\_\_\_

\_\_\_\_\_

FOLLOW-UP FOR NEXT APPOINTMENT \_\_\_\_\_

\_\_\_\_\_

QUESTIONS FOR MY PHYSICIAN \_\_\_\_\_

\_\_\_\_\_







My Insurances







My Contacts



My PFT's  
(Pulmonary Function Tests)



*One step at a time*



FILL YOUR PERSONAL PAGE WITH THE LOVE AND  
INSPIRATION YOU RECEIVE ALONG YOUR SUPPORTED  
JOURNEY!

WHEN YOU FEEL  
*loved and supported*  
YOU CAN DO  
*anything*





# Resources & Information

RESOURCES &  
INFORMATION





## **PULMONARY FIBROSIS SUPPORT GROUPS 2023**

*A place and time where Patients and Care Partners can connect, talk and support each other while caring for their loved one living with pulmonary fibrosis.*

### **Meetings Held:**

Robert Wood Johnson Rutgers- 1st Tuesday of the month  
Wellspan Health York Hospital- 1st Wednesday of the month  
Geisinger Med Center Danville- 1st Thursday of the month  
Coffee with Care Partners- 1st Friday of the month  
Robert Wood Johnson Rahway- 1st Friday of the month  
SLHN Anderson Campus- 2nd Wednesday of the month  
Veteran's ILD Support Group- 2nd Thursday of the month  
Project Positivity- 2nd Friday of the month  
LVHN Cedar Crest- 3rd Tuesday of the month  
Christiana Med Center-ILD Support- 3rd Weds. of the month  
Monmouth PF Support Group- 3rd Thursday of the month  
Robert Wood Johnson Trinitas- 3rd Friday of the month  
SLUHN Lung Transplant Mentorship- 4th Tues. of the month  
Geisinger Med Center Wyoming Valley- 4th Weds. of the month  
Encompass Toms River- 4th Thursday of the month  
Grief Support Group- 4th Friday of the month

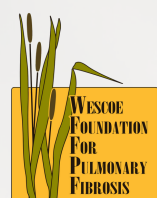
# **RSVP**

Jennifer H. Singley, M.Ed., NCC, Executive Director,  
Wescoe Foundation for Pulmonary Fibrosis

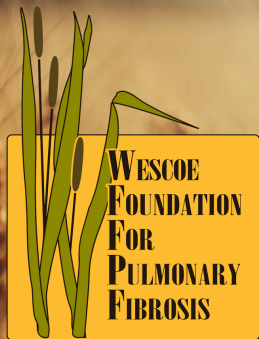
**[jennifer@wescoe.org](mailto:jennifer@wescoe.org)**

**484.553.6340**

*The Pulmonary Fibrosis Support Group will continue to meet virtually until further notice.*







# Quarterly Seminar 2023

## Understanding IPF and PF-ILD?

**Presented By:**

**Dr. Ross Summer**

**Jefferson Health**

Section Chief of Pulmonology, TJUH, Thomas Jefferson University Health Hospital  
Professor of Medicine, Division of Pulmonary, Allergy & Critical Care Medicine

**DATE:** Tuesday, March 21, 2023

**TIME:** 12:00 - 1:30 PM

**LOCATION:** Online

Register online at the link below or scan the qr code with your smartphone.



<https://wescoe.wufoo.com/forms/m1xoq1jd1vdqq4p/>

**Questions?** Please contact Jennifer Wescoe at **484-553-6340** or by email at [jennifer@wescoe.org](mailto:jennifer@wescoe.org).

**Wescoe Foundation for Pulmonary Fibrosis**

484-553-6340  
[www.wescoe.org](http://www.wescoe.org)



WescoeFoundationForPulmonaryFibrosis



@wescoefoundationforpf



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## "Improving Access to Care through Technology"



Thursday, May 4, 2023

12:00 - 1:30 PM

VIRTUAL EVENT



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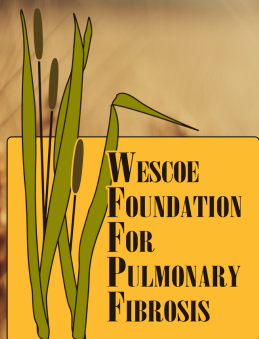
**Dr. Ryan Boente**

Director of Interstitial Lung  
Disease (ILD) Program  
Indiana University Health



**Dr. Patricia Fogelman, DNP**

Director of Palliative Medicine  
Mt. Nittany Medical Center



# Quarterly Seminar 2023

What is Palliative Care and How Can it be Helpful to You

**Presented By:**

**Dr. Patricia Fogelman, DNP**

Director of Palliative Medicine  
Mount Nittany Medical Center



**DATE:** Tuesday, September 12, 2023

**TIME:** 12:00 - 1:30 PM

**LOCATION:** Online

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at [jennifer@wescoe.org](mailto:jennifer@wescoe.org).

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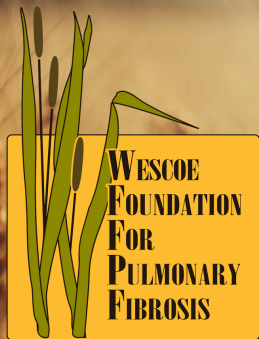
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# Quarterly Seminar 2023

Clinical Trials and Current Research

## Presented By: Aamir Ajmeri, MD

Temple Lung Center  
Pulmonology, Critical Care Medicine  
Assistant Professor, Clinical Thoracic  
Medicine and Surgery



**DATE:** Tuesday, November 7, 2023

**TIME:** 1:00 – 2:30 PM

**LOCATION:** Online

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at [jennifer@wescoe.org](mailto:jennifer@wescoe.org).

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Kevin Carney, MSN, CRNP, CCTC  
Temple Lung Center  
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# PATIENT-ENGAGED RESEARCH AND PULMONARY FIBROSIS



## What is pulmonary fibrosis (PF)?

PF is a lung disease where tissue becomes thick and scarred, making it more difficult to breathe.

## What is patient-engaged research?

Patient-engaged research is an approach to research that includes patients and care partners as **members of the research team**. They have a seat at the table with researchers and **share their expertise** about living with PF to help design and conduct a research study.

## What are the requirements to be part of patient-engaged research with PF?

The only requirement is to be a PF patient or care partner who is **willing to share** what it's like to live with PF.

## Why is patient-engaged research valuable?

Patient-engaged research creates studies that are more **meaningful** to patients and care partners and may **improve quality of life** for future generations of patients.

## How is this different from other types of research?

In most other types of research patients and care partners contribute only by having information collected from them. In patient-engaged research, **patients and care partners help direct the research**.

### PATIENT-ENGAGED RESEARCH IS:

- A collaborative approach to research with patients as team members.
- Conducted with input from patients and care partners.
- Key to creating more meaningful research.
- Currently uncommon in the field of PF.
- Created when patients and care partners share their ideas for research studies.

### TO LEARN MORE, INCLUDING HOW YOU CAN HELP, VISIT:

[www.pfpatientengagement.org](http://www.pfpatientengagement.org)



# What is Pulmonary Rehabilitation?

Pulmonary Rehabilitation (PR) is a multidisciplinary program consisting of education and exercise for individuals who have chronic lung disease, and/or have difficulty breathing while performing normal everyday activities. These programs are designed to improve lung function, reduce symptom severity and improve quality of life.

The PR team may include pulmonary doctors, nurses and specialists, such as a:

- Respiratory Therapist
- Exercise Physiologist
- Dietician or Nutritionist
- Psychologist or Social Worker

## What to Expect:

When you begin pulmonary rehabilitation, your rehab team will create a plan specific to your particular needs and abilities. Features of PR include:

### Exercise

**Cardio-Pulmonary/Endurance Training**—You will use equipment such as treadmills, stationary bicycles, arm ergometers (cycles) and recumbent elliptical machines.

**Strength Training**—Build strength using resistance bands and free weights.

### Education

Our rehab program involves education about your disease and how to manage it. Part of this includes instruction on how to take medications, or may include instructions for how to use oxygen therapy. The education component of your rehab may also include advice on how to quit smoking or ways to avoid substances that irritate the lungs. You will also be taught strategies to help you relieve symptoms, and also when to call your doctor or get emergency medical care.

### Breathing strategies

Our rehab team will teach you strategies that can improve your breathing, such as pursed lip and diaphragmatic breathing.

### Energy conservation techniques

You will learn tips on how you can conserve energy in your day-to-day life so that you may do more activities for longer periods of time without getting as tired as you do now.



# Understanding Oxygen Therapy

Your doctor can determine if you need therapy by measuring your blood oxygen levels (oxygen saturation) with either a blood sample or a small, electronic monitoring device called a pulse oximeter.

- A blood sample is usually drawn from an artery in your wrist for an arterial blood gas (ABG) measurement.
  - The ABG measurement is the most accurate because it measures your oxygen level directly from your blood.
- A pulse oximeter estimates your oxygen level by measuring your oxygen saturation externally.
  - The pulse oximeter is usually clipped on a finger, and is noninvasive—no needles are used.

Always use your oxygen as prescribed. Too little oxygen can strain your heart and brain, causing heart failure, fatigue, or memory loss, and too much can slow your breathing to dangerously low levels. Never change your oxygen prescription without consulting your doctor.

There are two types of oxygen supply systems primarily used today:

- The **compressed gas** is widely used either in stationary or portable units.
- **Oxygen concentrators** come in stationary units for in-home use and portable ambulatory units.
- A **home concentrator** provides a continuous flow of oxygen and pulls oxygen from room air and filters out nitrogen.
- **Portable concentrators** weigh from 3 to 20 pounds, and come in varied styles and sizes ranging from small units that can be worn with a strap like a handbag, to larger units on wheeled carts.
- Portable concentrators may deliver a lower percentage of pure oxygen than larger, home concentrators.
- Some portable concentrators deliver a pulse of oxygen and not a continuous flow. Consult with your doctor before purchasing or renting a portable oxygen concentrator to make sure the system meets your needs.





Oxygen is compressed under high pressure and stored in either large or small steel or aluminum cylinders (tanks).

- **Large cylinders** are not portable and primarily used in the home.
- **Smaller cylinders** are used as portable oxygen outside the home and can serve as backup during a power outage or equipment problem.
- Oxygen cylinders come in three sizes—M, D, and E tanks.
  - Weight, size, and liter flow will determine your oxygen needs
  - E tanks are large and heavy, but can be wheeled on a cart
  - D tanks are a more manageable size for higher liter flow
  - M tanks are the lightest and easiest to transport

The most common methods of delivering oxygen are:

- Nasal cannula – a two-pronged tube that is placed in your nose while the other end of the tube is attached to your oxygen system.
- Nasal pendant – which provides effective oxygen delivery to patients at supply flows substantially less than the standard nasal cannula.
- Simple mask – a clear plastic, disposable facemask used by patients who can breathe on their own but need a higher oxygen flow than a nasal cannula mask.
- Non-rebreather – a facemask which allows for the delivery of higher concentrations of oxygen; usually required for critically ill patients.

Oxygen delivery safety tips:

- No smoking while on oxygen therapy – ever!
- Keep oxygen at least 10 feet away from any sources of ignition.
- Always store oxygen containers upright in an open and well ventilated area.
- Be alert to avoid kinks in your tubing or disconnections from your oxygen source.
- Secure oxygen tanks upright in the car and NOT in the trunk.

Traveling with oxygen:

- You can absolutely travel by car, train, cruise ship, and airplane with oxygen.
- Be sure to discuss travel plans well in advance with your doctors, oxygen company, and airline to allow for appropriate travel documentation.
- Check with your insurance carrier as well as your durable medical equipment (DME) provider for out-of-town coverage.





# Understanding Clinical Trials and Research

## There are two types of clinical research—clinical trials and observational studies.

- A clinical trial is a research study that tests a new medical treatment (procedure or drug) or a new way to use an existing treatment.
- In an observational study, researchers assess health outcomes in participants in a normal setting to gather more information and better understand a disease.

All clinical trials are conducted according to a protocol—an established action plan—that ensures the safety of trial participants and the accuracy and trustworthiness of the information collected.

## Clinical trials are conducted in four phases.

- **Phase I:** Includes 20–80 healthy participants to test an experimental drug or treatment for the first time to evaluate its safety, determine appropriate dosage range, and identify possible side effects.
- **Phase II:** Includes 100–300 participants with a particular disease or condition to further evaluate safety and effectiveness.
- **Phase III:** Includes 1,000–3,000 participants with a particular disease or condition to confirm safety and effectiveness, monitor side effects, and compare to current standard treatments.
- **Phase IV:** Includes thousands of participants with a particular disease or condition to provide additional information, including risks, benefits, and optimal use for a new drug or treatment.

## The benefits of being in a clinical trial include:

- Access to new drugs and other treatments that aren't yet available
- Close and consistent attention and monitoring from doctors who are renowned experts at a leading academic (teaching) medical institution
- Contributing to research that may help others with your disease and save lives in the future
- Feeling empowered by taking an active role in your health care



### **Before participating in a clinical trial you should consider:**

- The purpose of the trial
- The phase of the trial
- Where you will have to go for the study visits
- Who will be in charge of your care
- How long the trial will last
- What kinds of tests and treatments are involved
- How your safety will be protected and your risks minimized
- If you will be compensated (paid) for your time and travel to study visits
- If you will get the trial results
- The possible risks and side effects of the study, including:
  - the treatment or drug being tested may not improve your condition
  - the study protocol may be challenging and take more time, effort, travel, and hospital stays than your current treatment
  - you may experience negative and unpleasant side effects despite constant and careful medical attention

### **For more information about clinical trials, visit the following sites:**

<http://www.clinicaltrials.gov>

<http://www.fda.gov>

<http://www.patientinform.com>

<http://www.centerwatch.com/patient/trials.html>

<http://www.clinicalstudyresults.org>



# Lung Transplant

## What Is Lung Transplantation?

Lung transplantation involves surgically replacing one or two diseased lungs with healthy lungs, usually from a deceased donor. In some cases, a heart transplant is performed at the same time. New lungs help the body get the oxygen it needs to survive. Following surgery, most patients must take powerful immunosuppressant drugs for the rest of their lives to reduce the risk of organ rejection.

## Who Should Have Lung Transplantation?

Lung transplants are a last-resort treatment for irreversible lung failure brought on by disease. Lung failure occurs when the lungs are damaged and unable to transfer oxygen and carbon dioxide to and away from cells. Only patients who have severely damaged lungs that cannot be helped by other therapies are eligible for transplantation. It is always better to try all the available treatments with your own lungs before considering a transplant.

If no other treatments are possible, and if the patient does not have certain risk factors (e.g., body weight too high or low; serious heart, liver, or kidney disease; current smoker; inability to follow a long-term health plan), then they may be a candidate for lung transplantation. Testing as described in the previous section is still required.

The conditions that most commonly lead to end-stage lung disease requiring transplantation include:

- Chronic obstructive pulmonary disease (COPD), including emphysema
- Pulmonary fibrosis (including IPF)
- Pulmonary hypertension
- Cystic fibrosis
- Sarcoidosis

## Steps to Lung Transplantation

**EVALUATION:** The first step to transplant is finding out if you are sick enough to need a new lung yet healthy enough to receive a new lung. This involves a careful evaluation by specially trained Temple staff members who will consider your physical and psychological health and suitability for major surgery. This team will also determine whether another treatment other than transplantation is more appropriate for you. In some cases, adjusting medications or a different procedure can improve symptoms and avoid the need for transplant.





**PREPARATION:** Once you have been cleared for transplant, you will prepare for the operation and wait for a donor match. During this period, you may need to reduce certain risks or undergo further tests or procedures. You will also be added to the national waiting list for a donor lung. The list is administered by the United Network for Organ Sharing (UNOS). As organs become available, UNOS makes the most appropriate match based on factors such as blood type, organ size, distance between donor organ and patient, and the patient's overall health and likelihood of success.

**TRANSPLANTATION:** When a lung becomes available, you are contacted and must report to the hospital immediately so that the transplant team can perform a final evaluation to make sure you are still in good condition for transplantation. Once approved, you will be prepared for surgery. Prior to surgery, the donated organ must be examined carefully by the retrieving surgeon to determine whether it is still functioning properly. This last-minute check is important and improves the likelihood of a successful transplant. In some cases, an organ will fail inspection and the transplant will be cancelled. Should the organ pass inspection, the transplant team will begin surgery. Depending upon the complexity of the case, surgery can last from 4 to 12 hours. The operation usually involves a full sternotomy (splitting of the breastbone), although Temple surgeons have developed a new minimally invasive technique that is appropriate for some patients. After surgery, you will stay in the hospital (first the ICU and then a special pulmonary care unit) for about 2 to 3 weeks. Some stays extend for even longer.

**RECOVERY:** Soon after surgery, you will begin education and physical rehabilitation. This continues after you leave the hospital. Transplant patients require approximately 6 to 8 weeks of recovery time. Recovery and rehabilitation includes: physical activity, medications (e.g., antirejection drugs, antibiotics as needed, and drugs for blood pressure, diabetes, and cholesterol), home testing/monitoring, and follow-up appointments for chest X-rays and screening bronchoscopies at 1, 3, 6, and 12 months after transplantation. (Bronchoscopy with biopsy is an exam and sampling of the lung through a small scope inserted through the nose to test for possible signs of rejection or infection.) The Temple transplant team also helps you start and stick to a long-term plan of exercise, diet, and mental health to ensure the best possible outcome.



# Care for the Caregiver

## Understanding the Importance of Self-Care

When it comes to the health of a loved one, the caregiver's role is critical and often complex—from buying groceries and cleaning the house to booking appointments and administering medication.

At the Temple Lung Center, we often see caregivers struggle to prioritize taking care of themselves when they're focusing all their time, energy and emotion on someone else's well-being. However, self-care for a caregiver is important because you may not be able to function, and then you may not be able to provide the care that is needed for the patient. Here are some tips to incorporate self-care into your day-to-day life.



### 1 Be Aware of Burnout

Sometimes you may not realize the toll your responsibilities are taking on your mind and body. Uncertainty, change, money, legal matters and emotions are all stressors that can unexpectedly creep up. This is why it's important to watch out for the warning signs of caregiver burnout, such as:

- Sleep deprivation
- Poor eating habits
- Little to no exercising
- Putting off your own medical appointments

### 2 Gather Information

#### Ask questions.

For many caregivers, the initial stages of a loved one's disease may be the most demanding because information and expectations are still evolving during the diagnosis and development of a treatment plan. This may also make it hard to concentrate during appointments, leaving you feeling uncertain and insecure. Don't be afraid to ask questions of your medical team. Plus, if you bring a notebook with you during appointments, you can document all the details and then revisit the information anytime.

#### Research resources.

If you're searching for tips on caregiving, remember to utilize reliable resources. URLs endings in *.gov*, *.org* and *.edu* are typically good indicators that a website is secure. When reading an article, pay attention to dates and authors; ask yourself: is this article current? Is this author a trusted professional? Again, always circle back to your care team if you have any questions or concerns.





### **3** Take Time for Yourself

#### **Try something new.**

Breaks from caregiving are essential, which is why we encourage you to maintain a life outside of caregiving. Join a class to teach yourself a new skill. Book a monthly massage appointment. Treat yourself to a quiet cup of tea at a nearby cafe. When you take time to rest and recuperate, you're also giving your loved one an opportunity to restore as well.

#### **Change your thinking.**

Sometimes, negative thoughts can be a roadblock to positive behavior. The next time you find yourself saying "There's no way I have the energy to exercise" or "I will never find time to read that new book" try to reverse them into positive statements: "I can muster the strength to exercise for 15 minutes while my loved one takes a nap" or "I'm going to read one chapter before bed each night."

#### **Ask for help from friends and family.**

Delegate simple tasks like laundry or meal preparation and you'll find the few regained minutes add up to a larger chunk of time.

### **4** Seek Out Support

Caring for someone with a chronic lung disease like Chronic Obstructive Pulmonary Disease (COPD) or Pulmonary Fibrosis is a challenge, but the right support can help improve quality of life for both you and your loved one.

#### **Consider connecting with a local support group or joining an awareness event.**

Here you can meet people who understand what you're facing because they may be facing similar situations. Specialists in the Temple Lung Center host several free monthly support groups, including one for COPD patients, friends and family. If you would like more information on Temple's COPD support group, please contact Susan Estrella-Eades, CRNP, FNP-BC at **215-707-5864** or [Susan.Estrella@tuhs.temple.edu](mailto:Susan.Estrella@tuhs.temple.edu). More information and a full listing of support groups available for pulmonary patients can be found at [lung.templehealth.org](http://lung.templehealth.org).



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# What is a clinical trial?

**Clinical trials are research studies designed to test whether new treatments are safe and effective to treat a disease.** Clinical trials follow strict scientific standards to evaluate safety and effectiveness, protecting volunteers and ensuring reliable results. Each potential participant should make an informed and voluntary decision regarding participation in clinical trials.

There are typically three stages or phases of clinical trials that must be performed before a drug or treatment may be submitted to regulatory agencies, such as the U.S. Food and Drug Administration (FDA), for approval.

- 1 Phase I:** Researchers test a new drug or treatment in a small group of people (usually 20-100 volunteers) for the first time to evaluate its safety, determine a safe dosage range, and identify side effects. Doses start small and increase in different patient groups until the desired effect of the treatment is observed or side effects of the treatment become too severe.
- 2 Phase II:** The drug or treatment is given to a larger group of people (up to several hundred volunteers) to see if it is effective and to further evaluate its safety. Most Phase II studies have randomized control (placebo) groups and treatment groups. Most of these trials are “double-blind” which means that neither the patients nor researchers know whether the patient is receiving the treatment or placebo.
- 3 Phase III:** The drug or treatment is given to large groups of people (several hundred to thousands of volunteers) to confirm its effectiveness, monitor side effects, compare it to commonly used treatments or placebo, and collect information that will allow the drug or treatment to be used safely. Phase III trials are also randomized and most are double-blind. Positive and significant results in this phase often lead to FDA approval.



# Interested in finding a clinical trial?

**The PFF Clinical Trial Finder** is a search tool on the PFF website that allows users to browse US-based studies that are recruiting patients with pulmonary fibrosis. Visitors may also fill out a short questionnaire and receive a cultivated list of clinical research opportunities closest to them.

**The PF Drug Development Pipeline** is a tool that allows users to filter drugs in development for, or already approved for the treatment of, pulmonary fibrosis and related conditions in the United States. Trials can be filtered by study phase, intervention type, and more.

Locate a trial at

[trials.pulmonaryfibrosis.org](https://trials.pulmonaryfibrosis.org)

Find more resources at

[pulmonaryfibrosis.org/  
clinicaltrials](https://pulmonaryfibrosis.org/clinicaltrials)

For more information, visit [pulmonaryfibrosis.org](https://pulmonaryfibrosis.org) or contact the PFF Help Center at **844.TalkPFF** (844.825.5733) or [help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org).



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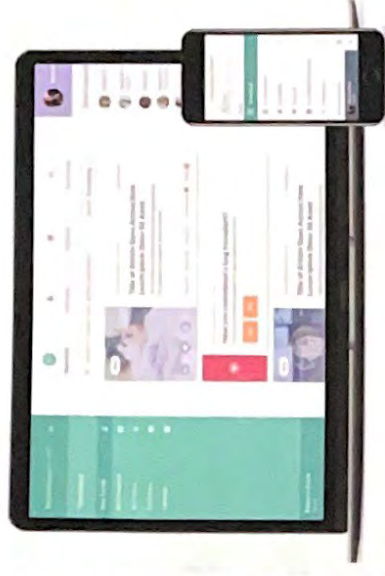


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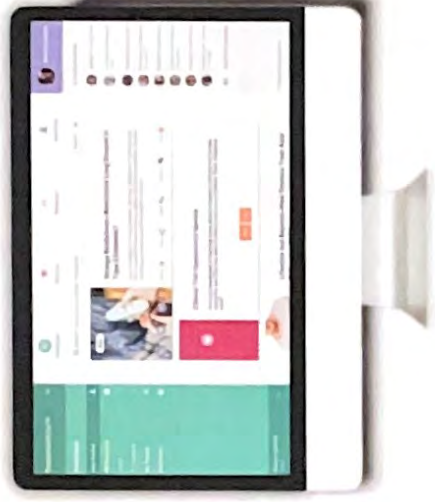
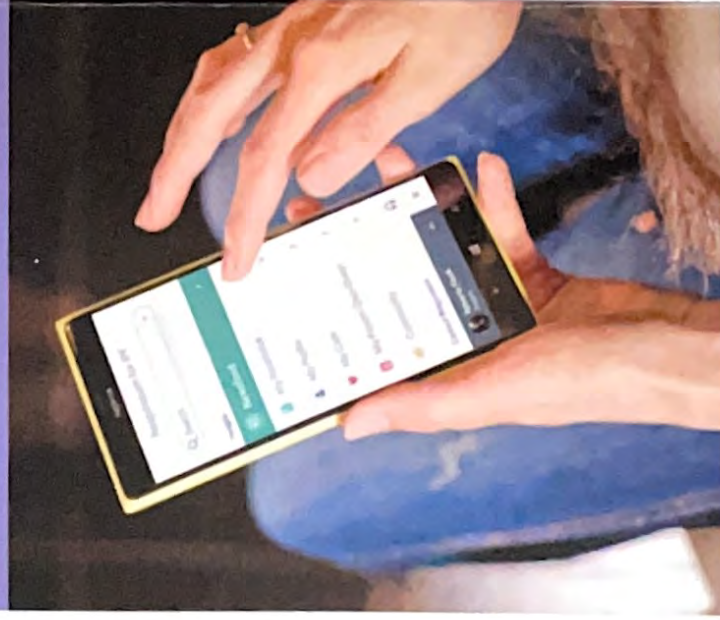
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## WHAT IS RESPONSUM FOR PF?

Responsum is a FREE online platform designed by patients that provides a daily stream of easy-to-understand articles summaries and resources—all you need to know about discoveries, treatments, and healthy living tips.

Our mission is to provide PF patients with all the tools, knowledge, and support necessary to better tackle their own health and lifestyle challenges.



## PFF VETTED CONTENT

Responsum searches and summarizes the most relevant information from the world's leading medical journals, newswires, patient blogs, and trusted sources of health news.

Our partnership with the Pulmonary Fibrosis Foundation provides Responsum users with the confidence to know that the content they are reading has been selected and vetted by some of the leading medical minds in the field.

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- Find medical, emotional, and financial support
- Communicate directly with your care team

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## PATIENT EDUCATION | INFORMATION SERIES

# Pulmonary Function Tests

Pulmonary function tests (PFT's) are breathing tests to find out how well you move air in and out of your lungs and how well oxygen enters your blood stream. The most common PFT's are *spirometry* (spy-RAH-me-tree), *diffusion studies*, and *body plethysmography* (ple-thiz-MA-gra-fee). Sometimes only one test is done, other times all tests will be scheduled on the same day.



## Lung function tests can be used to:

- Compare your lung function with known standards that show how well your lungs should be working.
- Measure the effect of chronic diseases like asthma, chronic obstructive lung disease (COPD), or cystic fibrosis on lung function.
- Identify early changes in lung function that might show a need for a change in treatment.
- Detect narrowing in the airways.
- Decide if a medicine (such as a bronchodilator) could be helpful to use.
- Show whether exposure to substances in your home or workplace may have harmed your lungs.
- Determine your ability to tolerate surgery and medical procedures.

## To get the most accurate results from your breathing tests:

- Do not smoke for at least 1 hour before the test.
- Do not drink alcohol for at least 4 hours before the test.
- Do not exercise heavily for at least 30 minutes before the test.
- Do not wear tight clothing that makes it difficult for you to take a deep breath.
- Do not eat a large meal within 2 hours before the test.
- Ask your health care provider if there are any medicines that you should **not take** on the day of your test.

## What is spirometry?

Spirometry is one of the most commonly ordered tests

of your lung function. The spirometer measures how much air you can breathe into your lungs and how much air you can quickly blow out of your lungs. This test is done by having you take in a deep breath and then, as fast as you can, blow out all of the air. You will be blowing into a tube connected to a machine (spirometer). To get the "best" test result, the test is repeated three times. You will be given a rest between tests.

The test is often repeated after giving you a breathing medicine (bronchodilator) to find out how much better you might breathe with this type of medicine.

It can take practice to be able to do a spirometry test well. The staff person will work with you to learn how to do the test correctly.

It usually takes 30 minutes to complete this test.

## What should I know before doing a spirometry test?

- You may be asked not to take your breathing medicines before this test.
- Instructions will be given on how to do this test. If you do not understand the instructions, ask the staff to repeat them.
- It takes effort to do this test and you may become tired. This is expected.
- If you become light-headed or dizzy during this test, immediately stop blowing and let the staff know.

## What are diffusion studies?

Diffusion tests find out how well the oxygen in the air you breathe in moves from your lungs into your blood.



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Like spirometry, this test is done by having you breathe into a mouthpiece connected to a machine. You will be asked to empty your lungs by gently breathing out as much air as you can. Then you will breathe in a quick (but deep breath), hold your breath for 10 seconds, and then breathe out as instructed.

You will do the test several times. It usually takes about 30 minutes to complete this test.

### What should I know before doing a diffusion test?

- Do not smoke and stay away from others who are smoking on the day of the test.
- If you are on oxygen, you will usually be asked to be off oxygen for a few minutes before taking this test.

### What is body plethysmography?

Body plethysmography is a test to find out how much air is in your lungs after you take in a deep breath, and how much air is left in your lungs after breathing out as much as you can. No matter how hard you try, you can never get all of the air out of your lungs. Measuring the total amount of air your lungs can hold and the amount of air left in your lungs after you breathe out gives your healthcare provider information about how well your lungs are working and helps guide your treatment. This test requires that you sit in box with large windows (like a telephone booth) that you can see through. You will be asked to wear a nose clip and you will be given instructions on how to breathe through the mouthpiece. You will be asked to take short, shallow breaths through the mouthpiece when it is blocked for a few seconds, which may be uncomfortable. If you have difficulty with being in closed spaces (claustrophobia), mention this to your provider ordering the test. This will avoid any misunderstanding and discomfort to you. It usually takes about 15 minutes to complete. Some PFT labs will use other tests instead of plethysmography to measure the total volume of air in your lungs.

### What should I know before doing a plethysmography test?

- If you are on oxygen, you will usually be asked to be off oxygen during this test.
- Let the staff know if you have difficulty in closed spaces.

### What are normal results for lung function tests?

Because everyone's body and lungs are different sizes, normal results differ from person to person.

For instance, taller people and males tend to have larger lungs whereas shorter people and females have smaller lungs. It is normal for your lung function to fall slightly as you age.

These standards that your healthcare provider uses, are based on your height, age, and sex at birth. These numbers are called the "predicted values". Your measured values will be compared to these predicted values.

**Authors:** Bonnie Fahy, RN, MN; Marianna Sockrider, MD, DrPH; Suzanne Lareau, RN, MS

**Updated:** Marianna Sockrider MD, DrPH

**Reviewers:** Brian Graham PhD, David Kaminsky MD

## Rx Action Steps

- ✓ Ask questions if you do not understand the instructions for the lung function test.
- ✓ If you have a cold or flu, let the test center know because you may need to reschedule your test.
- ✓ If you have difficulty with closed spaces (claustrophobia), let the test center know in case one of the tests involves being enclosed.
- ✓ Ask if there are any medicines you should stop taking before being tested and for how long you should stop it.
- ✓ After your pulmonary function testing is over, you can return to your normal activities.

### Healthcare Provider's Contact Number:

## Other Resources

### American Thoracic Society

- <http://www.thoracic.org/patients/>

### Canadian Lung Association

- <https://www.lung.ca/lung-health/lung-disease/spirometry>

### National Lung Health Education Program

- <http://www.nlhep.org/Pages/Spirometry.aspx>

### WebMD.com

- <https://www.webmd.com/lung/copd/living-with-copd-17/video-spirometry-test>

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PATIENT EDUCATION | **INFORMATION SERIES**

# Pulmonary Rehabilitation

If you have shortness of breath because of lung problems, you may have asked yourself:

- Can I exercise?
- How can I get in better shape?
- What medications should I be taking?
- Can I do anything to improve my overall well-being?



Pulmonary rehabilitation (PR) can help answer these and other questions. Participating in a PR program will help decrease your shortness of breath and increase your ability to exercise. You may have heard that pulmonary rehabilitation is only for people with COPD (chronic obstructive pulmonary disease). We now know that people with other lung conditions such as pulmonary hypertension, interstitial lung disease, pre/post transplant, and cystic fibrosis can benefit as well.

## What is Pulmonary Rehabilitation?

Pulmonary rehabilitation is a program of education and exercise that helps you manage your breathing problem, increase your stamina (energy) and decrease your breathlessness. The education part of the program teaches you to be “in charge” of your breathing instead of your breathing being in charge of you. You will learn how to pace your breathing with your activities, how to take your medicines and how to talk with your healthcare provider.

The exercise sessions are supervised by a pulmonary rehabilitation staff that prepares an exercise program just for you. The exercises start at a level that you can handle (some people start exercising while sitting and others on a treadmill). The amount of time you exercise will be increased in time and the level of difficulty will change based on your ability. As your muscles get stronger, you will exercise longer with less breathlessness and be less tired.

## How much time does a Pulmonary Rehabilitation Program take?

Most programs meet two to three times a week and programs can last 4 to 12 weeks or more. Because the program staff are constantly monitoring your progress and increasing your exercises as you are able, attending every session is important.

## How will I know if Pulmonary Rehabilitation is right for me?

Your healthcare provider will determine if you qualify for pulmonary rehabilitation by:

- Evaluating your current state of health and lung function test results
- Discussing your current activity level and your ability to do the things you want to do
- Determining your willingness and ability to attend.

Pulmonary rehabilitation programs are limited in the number of people who can attend so that you get close supervision. You will be evaluated before you begin the program to make sure you do not have health issues that would limit your ability to join. This evaluation may take place at the rehabilitation site or in a clinic by a physician, advanced practice nurse, or physician assistant.

Once the program begins, a team of healthcare professionals (nurses, respiratory therapists, physical therapists, occupational therapists, psychologists, dietitians, social workers, spiritual advisors such as a chaplain and others) will work with you to put you in charge of your breathing.

## What will I learn in Pulmonary Rehabilitation?

The education part of the program happens both in a classroom, one-on-one with the professional staff, and during each exercise session. During group meetings, you will learn new ways to breathe during stressful times and while being active. You will practice these new breathing techniques during your exercise sessions. You will learn about your medications; what the medications do and how to use your inhalers to get the most benefit from them. During the program, you may be given an Action

Plan that outlines what you should do when you are having a lung flare-up (exacerbation).

Some people with breathing problems need to use oxygen. During pulmonary rehabilitation you will be tested at rest and with exercise to see if oxygen may help you. You will learn the reasons why some people with shortness of breath use oxygen and others do not need it.

If you smoke, the program will provide support for you to stop or guide you to a program that can help you to stop smoking. You will also learn how and when to call your healthcare provider, including what key points to share, and what questions to ask. Also during the program, you can expect to meet others that also have breathing problems. You will have the time to share concerns and learn from others living with lung disease.

### What will I do in the exercise sessions?

You may not think that you can exercise when just walking across the room makes you breathless. There are however, standard exercises that have been found to work well for people with breathing problems. The type and amount of exercise you will do will depend on what you can do now and as you get stronger, your exercises will increase. Exercise sessions begin with stretching exercises or warm ups, followed by exercises for your arms and legs. Usually you will do both exercises to build your strength and exercises to build your endurance (stamina). To build your strength, generally weights and lifting devices are used. For endurance, activities might include walking on a treadmill or in a corridor and/or using a stationary cycle. The amount of time you exercise depends on what you can handle. After attending pulmonary rehabilitation, people are frequently amazed at how much they can exercise and how much less short of breath they experience.

### How can I find a Pulmonary Rehabilitation Program and what will it cost?

Ask your healthcare provider for a referral to a qualified program. Programs are often offered in an outpatient department of a hospital, including Veterans Administration hospitals. Some programs are certified by the American Association of Cardiovascular and Pulmonary Rehabilitation (AACVPR). The ATS maintains a directory of programs at <https://www.livebetter.org>. Other national and local directories are available through AACVPR, COPDF, and PERF. The American Lung Association (ALA) can also help you find Better Breathers and PR programs in your area.

The cost to you and insurance coverage of pulmonary rehabilitation can vary greatly depending on where you live and what program you choose. Medicare covers pulmonary rehabilitation for COPD, providing you meet certain requirements. Medicare may also cover rehabilitation for other lung conditions, but this varies with different regions of the country. The pulmonary rehabilitation program coordinator can tell you if you qualify and what the cost to you will be.

### What happens after I finish a Pulmonary Rehabilitation program?

It is so very important that you continue to exercise after finishing your rehabilitation program or you will lose all of the benefits you have gained. Before you “graduate”, the pulmonary rehabilitation staff will design for you a long-term plan of exercise for you. Many programs offer a “maintenance” plan so that you can continue to exercise with others with breathing problems.

**Authors:** Suzanne C. Lareau RN, MS, Bonnie Fahy RN, MN

**Reviewers:** Richard ZuWallack MD, Linda Nici MD, Marianna Sockrider MD, DrPH, Mary Gawlicki

#### Resources:

##### American Thoracic Society

- [www.thoracic.org/patients](http://www.thoracic.org/patients)
- [www.livebetter.org](http://www.livebetter.org)

##### American Lung Association

- <http://www.lung.org/lung-disease/copd/living-with-copd/pulmonary-rehabilitation.html>  
or call Help Line at 1-800-586-4872.

##### American Association of Cardiovascular and Pulmonary Rehabilitation (AACVPR)

- <http://www.aacvpr.org/Resources/SearchableProgramDirectory/tabid/113/Default.aspx>
- <https://www.aacvpr.org/Resources/Resources-for-Patients/Pulmonary-Rehab-Patient-Resources>

##### COPD Foundation (COPDF)

- <https://www.copdfoundation.org/>

##### Pulmonary Education & Research Foundation (PERF)

- <https://perf2ndwind.org>

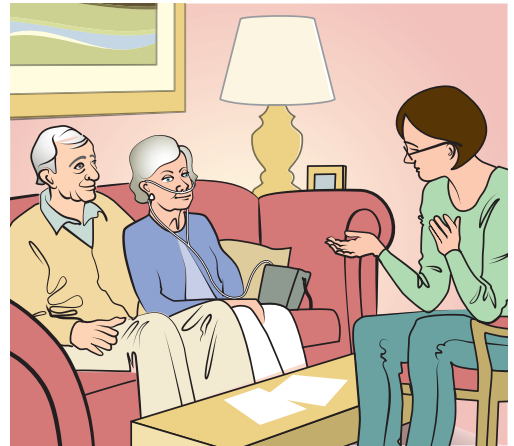
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PATIENT EDUCATION | **INFORMATION SERIES**

# Palliative Care for People with Respiratory Disease or Critical Illness

When you have a serious illness, you may suffer from physical discomfort, such as pain, difficulty breathing, nausea (stomach upset) or fatigue (tiredness). You may also have feelings of psychological distress, such as anxiety or depression, or feelings of spiritual distress. Your healthcare providers can offer medical therapies to manage your illness, for example, antibiotics to treat pneumonia. Your healthcare providers can also offer care that is focused on improving your quality of life and helping you feel more comfortable. This type of care is called palliative (PAL-lee-uh-tiv) care.



## What is palliative care?

The World Health Organization (WHO) defines palliative care as an approach that improves quality of life for patients and families facing life-threatening illness. People with serious lung problems or who are critically ill may suffer from symptoms of pain, breathlessness, or cough. Palliative care works to improve quality of life by preventing or relieving these symptoms and can also help with a person's psychological, spiritual, or emotional needs.

## When should I receive palliative care?

You can receive palliative care at any time during the course of your illness, from the time you are diagnosed throughout the treatment of your disease. Palliative care can be provided at the same time that you are receiving medical treatments such as chemotherapy, radiation therapy, bronchodilators (inhalers), oxygen therapy, or intensive care. You do not need to be in a hospital to receive palliative care. This care can be provided in many different settings, such as a clinic, nursing home, hospital, or at your home.

## Could I benefit from palliative care?

Persons of any age, from children to the elderly, may benefit from palliative care. If a serious illness

negatively affects you and your quality of life, you may receive palliative care. You may already be receiving palliative treatments. Some examples are:

Condition	Medical Treatment	Palliative Care
Lung cancer	Chemotherapy or radiation therapy	Medication to treat side effects of your medical treatments. Side effects managed by palliative care might include fatigue, nausea, poor appetite, pain, or difficulty breathing.
Chronic lung disease, such as Chronic Obstructive Pulmonary Disease (COPD) or Idiopathic Pulmonary Fibrosis (IPF)	Oxygen and bronchodilators (inhalers)	Medicines and counseling to help with anxiety and difficulty breathing. A fan blowing cool air to reduce shortness of breath.
Severe pneumonia	Antibiotics and care in an intensive care unit (ICU)	Medications to reduce pain, help with sleep, or reduce feelings of anxiety.



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Palliative care may help if you are feeling sad or anxious about your disease, the side effects of your medical treatment, or the effects your illness is having on your family. Palliative care can also help address your spiritual needs.

### Who provides palliative care?

Your healthcare provider may be able to provide palliative care or may refer you to a team of palliative care specialists. Your healthcare provider and your palliative care specialists make up your palliative care team. Palliative care specialists may be physicians, nurse practitioners, nurses, social workers, chaplains, or counselors. For children, a child life specialist can also be an important member of the team. If you are receiving care at home, providers may be able to make home visits and help coordinate care with other members of your team.

### What kind of care is right for me?

In addition to helping you address the symptoms and feelings of stress related to your illness, palliative care providers can help you explore your values, goals, and preferences for medical care. This process is called advance care planning. The palliative care team can work with you to address important questions, such as: "What are my hopes for the future?" or "What do I value most in my life?" Answering these questions can help you with decision-making throughout the course of your illness. The palliative care team can also help you document your preferences for care in the form of advance directives, such as a durable power of attorney for healthcare or a living will.

### How can palliative care help if I am at the end of my life?

Palliative care also includes care at the end of life. Similar to palliative care that can be provided throughout the course of a serious illness, palliative care provided at the end of life focuses on improving your quality of life by managing feelings of physical, psychological, and spiritual distress. This type of care is also called end-of-life care. People who are critically ill may receive end-of-life care in an intensive care unit (ICU). Some people receive end-of-life care from hospice providers. Hospice care focuses on comfort

and relief from suffering, when medical treatments used to prolong life are no longer helpful. Hospice care is generally for people who have months to live and can be provided in a home, nursing home, or hospital. After a person has died, palliative care providers can also provide bereavement care to support the family during the grieving process.

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\*Authors of original 2013 version.

## Rx What to Do

- ✓ If you have a serious illness that affects your quality of life, you should ask your healthcare provider about the role of palliative care in your treatment plan.
- ✓ Ask your healthcare provider how palliative care could help improve your quality of life.
- ✓ Discuss palliative care with your family members, who may also benefit from the services provided by a palliative care team.

**Healthcare Provider's Contact Number:**

\_\_\_\_\_

### Additional Resources:

#### Get Palliative Care

- <https://getpalliativecare.org/>

#### Palliative Doctors

- <http://palliativedoctors.org/>

#### Making Your Wishes Known

- <https://www.makingyourwishesknown.com>

#### American Thoracic Society

- [www.thoracic.org/patients/](http://www.thoracic.org/patients/)

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PATIENT EDUCATION | **INFORMATION SERIES**

# What is Lung Transplantation?

Lung transplants are performed as a life saving treatment for people with end-stage lung disease. A lung transplant is a surgery to remove diseased lungs and replace them with lungs from a deceased donor with healthy lungs. There are only a few specialized centers around the world where lung transplants are performed. You will need to be referred to one of these centers to be considered for the surgery.



## Do I need a lung transplant?

Lung transplantation is an option for people with 'end stage' lung diseases such as chronic obstructive pulmonary disease (COPD), pulmonary fibrosis, cystic fibrosis (CF), pulmonary arterial hypertension (PAH), sarcoidosis, and other more rare lung diseases (See "Recipient Selection for Lung Transplantation"). Lung transplantation should only be considered when you and your physician have exhausted all other forms of available treatment. However, lung transplant should be performed before you become too sick to tolerate the surgery. Hence, the timing of evaluation and the surgery itself is very important.

In order to qualify for surgery, you need to be evaluated at a lung transplant center. During your evaluation, you will meet with a transplant surgeon, transplant pulmonologist (lung doctor) and other staff. They will inquire about other medical problems that you may have such as heart or kidney disease. They will make sure that you have been screened for cancers that are relevant for your age, such as colon, prostate (in men), breast and cervical cancer (in women). You will have a thorough evaluation that may include a CT scan of your chest, pulmonary (lung) function tests, an echocardiogram (ultrasound of your heart) and possibly a heart catheterization. You may have testing to see if you have gastroesophageal (acid) reflux. Finally, they will also make sure that you have good social support from those who will take care of you after your transplant. At the end of the evaluation, the transplant team will determine if surgery is the right option for you and whether to put your name on the lung transplant waiting list.

## What is the "list"?

The waiting list includes the names of all people who have been accepted by a transplant program and are currently waiting for a transplant. United Network for Organ Sharing (UNOS) maintains a list of accepted candidates from all centers across the United States. If your transplant team decides that you are a candidate for lung transplantation, you will be assigned a score called the Lung Allocation Score (LAS). The LAS is used to help direct donated organs to people who would most benefit from a transplant. Some (but not all) of the factors that are used in calculating the score are your age,

oxygen requirement, pulmonary function test results, distance walked in 6 minutes and the type of lung disease you have. Your transplant team may update your score over time if some of these factors change.

Other factors used when matching donor lungs to a transplant recipient include:

- where the donor is,
- donor and recipient blood types,
- antibodies you may have against certain donors' blood and tissues,
- donor lung size and your chest size (often related to your height).

The time spent on the waiting list by an adult is no longer a factor in deciding who receives the transplant.

## What do I do while I am waiting?

After your name is added to the lung transplant waiting list, the waiting time varies depending on the availability of suitable donor organs and your position on the list determined by your LAS. The typical waiting time can be a matter of weeks to several months. However, during this time you should not be idle. You should use this time to exercise as tolerated to get your body in the best shape possible for the surgery. Your transplant team may recommend you work with a pulmonary rehabilitation program. In general, the better shape you are in before the surgery, the easier and quicker your recovery will be. Your team may also ask you to move closer to the lung transplant center to follow you closely as your disease progresses, and to shorten the travel time to the center when organs becomes available. It is very important during this period to let your team know when your health changes.

When a suitable organ becomes available, you will receive a phone call telling you to come into the hospital. Because this call can come at any time, you should have a bag packed and be ready to get to the hospital immediately. Sometimes the call may result in a "dry run", in which the donor organ is found to not be suitable for transplantation after further evaluation.

**What is the surgery like?**

The operation can vary between each person and center. Your team will decide if either a single lung or a double lung transplant is better for you and explain the surgery to you. A single lung transplant may be better tolerated in sicker and older people. The operation itself usually lasts about six to eight hours but people are typically in the operating room for much longer. Some people may temporarily require heart-lung bypass support and other invasive measures to get through the operation. You will be asleep for the entire surgery, with medicines given by an anesthesiologist. The incision is generally located below your breast. When you wake up, you will have chest tubes in your sides and a breathing tube in your mouth. You will be given medicine to control the discomfort from the surgery until you fully recover.

**How long is recovery?**

After the surgery, you may be in the ICU for 3-5 days if no complications occur during the surgery. In that case, you should be discharged from the hospital after about 2-3 weeks. However, the duration of the hospital stay can extend to several months if problems occur during the recovery. Much of your hospital stay will be focused on getting the right doses of immune suppressing medicines into your body to prevent your body from rejecting the lung transplant. The most common medications your doctors will prescribe to suppress your immune system are tacrolimus (or cyclosporine), mycophenolate mofetil (or azathioprine), and prednisone. The other major part of your hospital stay is rehabilitation. A physical therapist will start working with you to get you out of bed and walking as soon as is medically possible. This pulmonary rehabilitation may be continued after you are discharged from the hospital in an outpatient monitored setting.

You will have regular blood tests, chest x-rays and spirometry after your transplant. You may also need to have a bronchoscopy done from time to time (see "Flexible Bronchoscopy"). In general, your initial pulmonary function testing will continue to improve during the first year after transplant as long as your new lungs stay healthy. You will also be asked to monitor your pulmonary function on a regular basis with a portable micro-spirometer. This may help you detect problems early, even before you have symptoms. You will have blood tests to be sure your immune system is adequately suppressed and that other organs (such as your kidneys and liver) are not affected by your new medications.

**What different problems might I encounter after a lung transplant?**

While a lung transplant is often a life saving treatment, there are certain problems that recipients may face. The two major problems are infections and rejection of the transplanted lung (see "Rejection after Lung Transplantation"). Because you are taking medications to suppress your immune system, you are more prone to infections. Your team will prescribe preventative antibiotics for some common infections. Unfortunately, not all infections can be completely prevented. You should do what you can to avoid contact with ill people and wash your hands well and often. Your team will tell you what vaccines to take and how else to avoid infection.

In addition to infection, acute rejection can occur early on after lung transplantation. Acute rejection occurs when your immune system recognizes your new lungs as foreign and starts to attack them. Acute rejection requires prompt attention and may need adjustment of your immune suppressing medicines.

The main limiting factor in the long-term survival for patients who have had a lung transplant is chronic lung allograft dysfunction (CLAD). Bronchiolitis obliterans syndrome (BOS) and restrictive allograft syndrome (RAS) are forms of CLAD (see ATS Patient Information Series piece "Bronchiolitis Obliterans Syndrome" at [www.thoracic.org/patients](http://www.thoracic.org/patients)). It also can occur many years after your transplant and is usually identified by specific changes in your pulmonary function tests. If these changes occur, your transplant team may perform a bronchoscopy with biopsies of your transplanted lungs before considering specific treatments for Chronic Lung Allograft Dysfunction (CLAD).

Other problems can also occur in recipients, including scarring or stenosis in the windpipe at the attachment site of the transplanted lung or in other areas of your airways. You will also be more likely to develop skin cancer and other types of cancers because of the immune suppressing medicines you will be taking. However, with regular visits with your transplant team, you will be able to watch out for and manage these complications if they arise.

Considering lung transplantation for your lung disease can be a scary process. However, a lung transplant can offer people with end stage lung disease a longer life expectancy and improved daily quality of life.

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**Reviewers:** Hrishikesh Kulkarni MD, Marianna Sockrider MD, DrPH, Jeffrey Goldstein

**Rx Action Steps**

- ✓ Talk with your lung specialist and primary healthcare provider about whether you should consider a lung transplant.
- ✓ If you have end-stage lung disease and want to consider a transplant, arrange to talk with or visit a lung transplant center.
- ✓ Take part in pulmonary rehabilitation to help stay as well as possible before and after transplant.
- ✓ Work with your transplant team to get necessary tests before and after transplant.
- ✓ Enlist friends and family who can help support you along the way.

**Healthcare Provider's Contact Number:**

**For Additional Information:**

**Organ Procurement and Transplantation Network (OPTN)**

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

**United Network for Organ Sharing (UNOS)**

<http://www.unos.org>

**Lung Transplant Foundation**

<http://www.lungtransplantfoundation.org>

**American Thoracic Society Patient Education Documents**

<http://www.thoracic.org/patients>

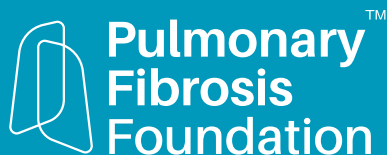
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# what do I need to know about traveling with supplemental



## TRAVELING WITH SUPPLEMENTAL OXYGEN

Many people with pulmonary fibrosis (PF) can safely travel by air, but for some, air travel can be dangerous. The atmosphere is made of 20% oxygen and 80% nitrogen whether you are at sea level or living high up in the mountains. At higher elevations, there is simply less air (the atmospheric pressure is lower), so there is less oxygen for you to breathe.

Since commercial airplane cabins are usually pressurized to an elevation equivalent to about 1,500-2,500 meters (about 5,000-8,000 feet), you will be breathing less oxygen while in-flight.

If you have PF, it is important that you discuss your travel plans with your health care provider weeks or months before planned travel. After completing any required medical tests, your healthcare provider will determine whether you need oxygen while in-flight. You will then need enough time to notify the airline, have your doctor fill out paperwork for the airline, and coordinate with an oxygen supplier.

## Do I need to bring oxygen on the plane?

You might need to use oxygen during your flight, even if you do not use oxygen at home. Your healthcare provider can help determine whether you need oxygen on the plane. Some tests that your doctor might order include:

1. Pulse oximetry to check your oxygen level
2. Six-minute walk testing
3. Pulmonary function testing
4. Arterial blood gas measurement to check your oxygen and carbon dioxide levels
5. Echocardiography
6. "Hypoxia altitude simulation test" (HAST) – a test where your doctor measures your oxygen level while you breathe air with a reduced oxygen level (15% oxygen instead of 20%).

Based on these test results, your doctor might prescribe oxygen for you to use on the plane.

If your oxygen requirements are too high or if you have other medical conditions, your healthcare provider may instead advise you to not travel by air, since your oxygen levels may drop dangerously low in-flight.

If you do need oxygen during your flight, your doctor will tell you what oxygen flow setting you should use.

## Will the airplane supply oxygen if my doctor prescribes it?

In the United States, airlines are required to allow passengers to use battery-powered portable oxygen concentrators that have been approved by the Federal Aviation Administration (FAA).

Most airlines require you to bring your own portable oxygen concentrator, but not all concentrators are allowed by all airlines. Each airline maintains a list of which portable oxygen concentrators they will allow on board. A small number of airlines will provide you with oxygen on board. There is usually a charge to use an airline's oxygen. Airlines will not allow you to bring filled oxygen tanks (green cylinders) or liquid oxygen onto the plane.

Your healthcare provider must complete paperwork ahead of time that instructs the airline about how and when you should use oxygen. Your healthcare provider can also help to arrange for a short-term oxygen concentrator rental from an oxygen supply company.

## Looking for more information?

The PFF Patient Communication Center, a dedicated call center, provides patients, caregivers, and healthcare providers with the most up-to-date medical information, communicates the availability of support services, and provides information about other essential resources. Call or email today at **844.TalkPFF (844.825.5733)** or **[help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org)**.



## Checklist for traveling with oxygen

- ☐ Prepare weeks or months ahead of time. See your doctor and notify the airline as early as possible. While some airlines may only need 48 hours of advance notice, it is advisable to prepare much earlier.
- ☐ Be aware of the airline's specific requirements regarding which oxygen concentrators are allowed and what paperwork is required. Contact your airline for further information.
- ☐ Be sure to bring enough batteries – and make sure they are fully charged! The FAA requires you to have battery life equal to 150% of your expected travel time. You should also factor in time needed to travel to the airport, waiting to board, layovers, and traveling from the airport to your destination after arrival. The airplane may or may not have an electrical outlet available if your batteries run out.
- ☐ Ask your doctor if you should monitor your oxygen level in flight with a portable pulse oximeter.
- ☐ If you are traveling outside the United States, different regulations may apply. Contact your airline for guidance. And remember to bring the correct electrical plug adapter for the country you are visiting.

## What else should I know when I travel by air?

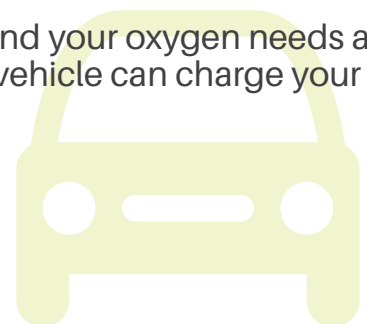
If your condition has worsened or you feel ill on the day of travel, you should talk to your doctor about your travel plans. Talk to your doctor to see if you should bring extra medication on your trip. Many people are more active while traveling. You may find that you are unable to participate in some activities that require a good deal of effort. Consider this possibility when planning your trip.

## Can I travel to high elevations?

It is important that you discuss your travel plans with your healthcare provider. If your destination is at an elevation above sea level, you may suffer from breathlessness with small degrees of exertion or even while resting. Your healthcare provider may advise you to not travel to destinations at high elevation.

## What about traveling by car?

Contact your oxygen supplier to tell them about your travel plans and your oxygen needs at your destination. The 12V DC outlet power source charger in your vehicle can charge your portable oxygen concentrator.



## My doctor said I need oxygen at my destination. How can I obtain oxygen while away from home?

Your doctor may determine that your portable oxygen concentrator is sufficient for your travel needs while away from home. If you need an additional oxygen delivery device while away from home, your oxygen supplier may be able to coordinate with an oxygen supplier at your destination to provide the device(s) you need. Be sure to plan ahead of time.

### Resources for traveling with supplemental oxygen:

American Airlines  
1.800.433.7300

Delta Airlines  
1.800.221.1212

Southwest  
1.800.I.FLY.SWA (1.800.435.9792)

United Airlines  
1.800.UNITED.1 (1.800.864.8331)



Boehringer  
Ingelheim

**Genentech**  
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**Pulmonary  
Fibrosis  
Foundation**

230 East Ohio Street, Suite 500  
Chicago, Illinois 60611  
**844.TalkPFF** (844.825.5733)  
[help@pulmonaryfibrosis.org](mailto:help@pulmonaryfibrosis.org)

The mission of the Pulmonary Fibrosis Foundation is to accelerate the development of new treatments and ultimately a cure for pulmonary fibrosis. Until this goal is achieved, the PFF is committed to advancing improved care of patients with PF and providing unequalled support and education resources for patients, caregivers, family members, and health care providers.

# why did my provider prescribe supplemental oxygen?





## why did my provider prescribe supplemental oxygen?

People living with lung disease often have lower than normal levels of oxygen in their blood. When levels drop to 88% or lower, a healthcare provider may order supplemental oxygen. The benefits may include:

- Reducing breathlessness
- Increasing your ability to maintain an active lifestyle
- Helping to reduce stress on other organs

Oxygen is not addictive. Use it as prescribed by your healthcare provider.

## what does my oxygen prescription include?

1. When to use your oxygen (during sleep, rest, activity, or at altitude)
2. How much oxygen you need for each activity (number setting or liter flow)
3. What type of oxygen equipment fits your lifestyle and oxygen requirements
4. A “Certificate of Medical Necessity” (required by Medicare and many other insurances)

There are many oxygen delivery systems available. What you receive is determined by your lifestyle, the level of oxygen your provider orders, and what’s available through the oxygen company. Before you enter into a Medicare contract with an oxygen company, ask your healthcare provider to make sure the equipment offered by the supplier is correct for you.



## why is oxygen needed?

Every cell in your body needs oxygen to work. Oxygen moves from your lungs into your blood, which carries it to the rest of your body.

Oxygen levels are measured using a fingertip device called a pulse oximeter, or by taking a blood sample.

## how do I know I am getting enough oxygen?

Your healthcare provider may recommend a pulse oximeter to monitor your oxygen levels at home. Ask what your target oxygen level should be. If directed by your healthcare provider, you can adjust the flow of oxygen depending on what you’re doing to keep your level in the recommended range.

- Some people need oxygen 24 hours a day. Others may not need oxygen while sitting, but require higher flows with activity.
- Some people need oxygen while sleeping.
- When traveling in an airplane or to a higher elevation, ask whether you will need oxygen or different settings.
- Airlines only allow oxygen with specific devices and require a prescription.
- Plan ahead to avoid running out of oxygen or battery life.

## what is the difference between continuous flow and pulse or demand flow?

**Continuous flow** means that oxygen flows continuously into your nose through the nasal cannula. With **pulse or demand flow**, a special regulator senses when you inhale and ONLY delivers a pulse of oxygen when you breathe in, allowing tanks and batteries to last longer.

Continuous flow and pulse settings are both prescribed as a number (1, 2, 3, etc.). A continuous flow setting of 2, 3, or 4 liters per minute is not the same as a pulse or demand flow setting of 2, 3, or 4.

## how do I use oxygen safely?

- Avoid flames, sparks, cigarettes, matches, lighters, gas stoves, pilot lights, hair dryers (use the cool setting only), and frayed electric cords.
- Remember that oxygen is not flammable and does not explode on its own, but it will make a fire burn faster.
- Place a sign on your front door stating that oxygen is in use.
- Store tanks in an upright position. Do NOT use broken or cracked tanks. Do NOT store tanks in a closet, in direct sunlight, or next to heat sources.
- Do NOT use oily (petroleum-based) substances on your lips, nose, or face, including ChapStick, Vaseline, or Blistex. Check online for nasal lubricants designed for oxygen users.



## I just received my oxygen equipment. What do I do now?

Make sure you received all the equipment your provider prescribed, which may include the following items.

### OXYGEN CHECKLIST

- ☐ **Stationary Oxygen Concentrators**  
Stationary concentrators concentrate oxygen from the air to a more pure form of oxygen. They are powered by electricity and typically not portable. Units are designed primarily for home or work.
- ☐ **Portable Oxygen Concentrators (POCs)** POCs are smaller concentrators designed to be used for walking and activities outside the home. Units range from 3 lb. (worn over the shoulder) to 25 lb. (pulled on wheels). Generally, the smaller the unit, the lower the oxygen output (liter flow or pulse settings) and the shorter the battery life. POCs can be recharged via wall plug or car battery.
- ☐ **Oxygen Tanks** These are green and silver (aluminum) tanks filled with oxygen. They can be very large or small enough to fit in a backpack. Some compressed tanks can be filled at home with a concentrator filling system, often referred to as a “Trans-Fill” unit. As with POCs, the smaller the tank, the more limited the amount and duration of oxygen.
- ☐ **Liquid Oxygen** This is oxygen that has been supercooled, and turned to a liquid form. Liquid oxygen is stored in a canister that resembles a large thermos. As the oxygen warms, it turns back to gas that is available for use. A stationary liquid oxygen reservoir tank is used at home and can fill a smaller, portable tank for use outside. Weighing 3–11 lb., these tanks provide higher continuous flows than POCs. Not every oxygen company offers liquid oxygen.
- ☐ **Cannula and Tubing** All stationary systems connect to tubing (25 or 50 ft) for use. Oxygen set above 6 liters per minute uses special “high-flow” tubing. The part that goes in your nose is called a cannula. Some who need high-flow rates use a small mask placed over the nose and mouth instead of a cannula.
- ☐ **Oxygen Conserving Device (OCD)** This is a device that delivers oxygen by pulse or demand flow. It conserves oxygen by only delivering oxygen during a breath, and turning off until the next breath.
- ☐ **Regulator and Flow Meters**  
Traditional oxygen regulators and flow meters deliver continuous-flow oxygen.

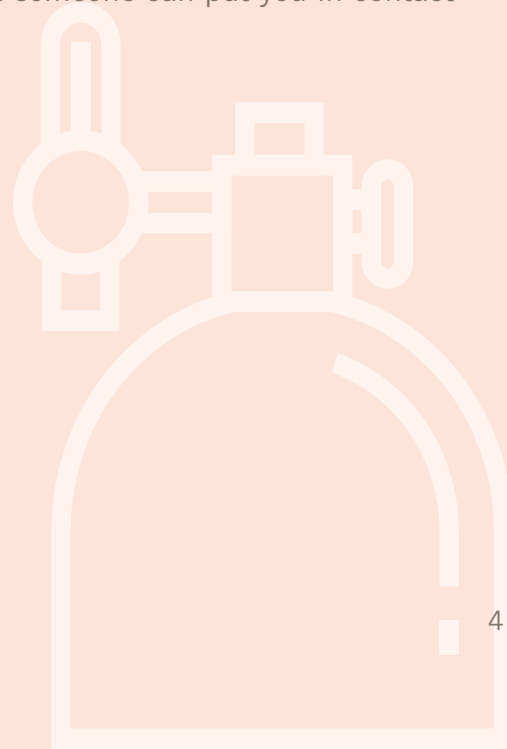
## Make sure you get instructions for operating the equipment, including how to:

- ☐ Turn it on and off
- ☐ Make sure oxygen is flowing
- ☐ Attach flow meter and/or OCD and adjust flow rates
- ☐ Check whether oxygen is in the tank
- ☐ Know when the tank is empty
- ☐ Attach a humidifier (if needed)
- ☐ Understand your prescription, including how much oxygen (liters per minute or pulse setting) is ordered and what the settings are for rest, exertion, and sleep
- ☐ Properly wear a cannula
- ☐ Know the battery life of your POC and how to prolong it
- ☐ Maintain and care for your equipment
- ☐ Take safety precautions

## Make sure you know whom to call for problems with equipment or deliveries.



Know your rights. If you have continuing problems with your equipment, service, or reimbursement, call **1-800-MEDICARE (633-4227)** and someone can put you in contact with a Competitive Acquisition Ombudsman (CAO).



For more information, contact:



[pulmonaryfibrosis.org](http://pulmonaryfibrosis.org)  
[pcc@pulmonaryfibrosis.org](mailto:pcc@pulmonaryfibrosis.org)  
844.TALKPFF (844.825.5733)



**American Association  
for Respiratory Care**

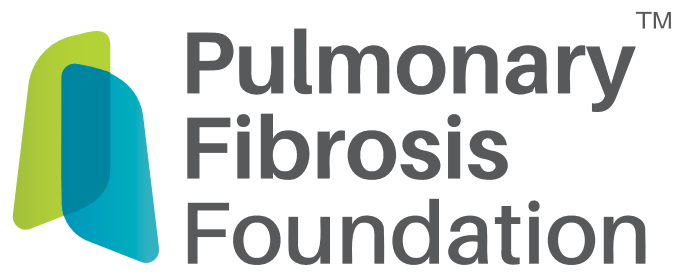
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## Oxygen Basics

A step-by-step guide to using supplemental oxygen





## FIND IT FAST

Oxygen fundamentals	1
Oxygen equipment	2
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Medicare and supplemental oxygen	5
Traveling with oxygen	6

## Have questions about oxygen that aren't answered in this guide?



Call the PFF Oxygen Information Line Monday to Friday between 9:00 a.m. and 5:00 p.m. CT at 844.825.5733. The PFF Oxygen Information Line is staffed by trained professionals who can provide further information about topics such as oxygen safety and equipment.



## INTRODUCTION



Every cell in your body needs oxygen to function. When you take a breath, oxygen moves down your airways into the small air sacs and then into your bloodstream. The area where the oxygen crosses into the blood is called the interstitium. Interstitial lung disease (ILD) or pulmonary fibrosis (PF) cause this area to be swollen or scarred, which can decrease your oxygen levels, especially when you are active. Oxygen levels can be measured by taking a blood sample or, more often, by placing a device called a pulse oximeter on your finger or forehead.

Use of supplemental oxygen may prevent breathlessness, increase your ability to maintain an active lifestyle, and help decrease stress on organs. Although no studies have shown that supplemental oxygen prolongs life in pulmonary fibrosis patients, some studies demonstrate that supplemental oxygen extends life in patients living with chronic obstructive pulmonary disease (COPD). For some PF patients with prolonged low oxygen levels (or who need oxygen 24 hours a day), use of oxygen can lower the stress on the heart and blood vessels in the lung.

The Pulmonary Fibrosis Foundation (PFF) is pleased to provide these step-by-step tips for starting and safely using oxygen at home and when traveling, as well as detailed information on Medicare coverage of supplemental oxygen.

## OXYGEN FUNDAMENTALS

### Possible signs and symptoms

Many people who need supplemental oxygen experience shortness of breath or fatigue. Other possible signs that you're not getting enough oxygen include irritability, blue fingertips or lips, rapid heart rate with activity, and ankle swelling. However, some people may not experience symptoms even when they're not getting enough oxygen into their bloodstream. Some patients with PF can experience breathlessness during exertion even when their oxygen levels are normal. This is due to scarring, which makes the lungs stiffer, requiring more "work" to breathe.



### BOOST YOUR OXYGEN IQ

New users sometimes ask if supplemental oxygen is addictive. The answer is no—use oxygen as prescribed by your healthcare provider.

### Determining your need for supplemental oxygen

Your healthcare provider can test your need for supplemental oxygen by measuring your oxygen levels while you are at rest, during walking or exercising, or during sleep. The six-minute walk test is commonly used to determine whether you need supplemental oxygen while walking. Your healthcare provider will decide which tests are right for you. You'll need a face-to-face visit with your healthcare provider and testing for insurance purposes.

If your healthcare provider finds your oxygen levels are low during testing, he or she will determine whether you need supplemental oxygen, when you need it (at rest, during activity, and/or during sleep), and how much you need in each setting.

### Your healthcare provider's role

In addition to identifying your supplemental oxygen needs, ordering appropriate testing, and providing a prescription and supporting documentation, your healthcare provider will work with the company supplying your supplemental oxygen and assist you in determining your oxygen equipment needs for use at home, with activities outside the home, and for travel.



### BOOST YOUR OXYGEN IQ

A recent study found that patients who received education from healthcare providers about supplemental oxygen were less likely to have problems with their oxygen than those who didn't receive education or were only educated by the person delivering their oxygen.

### Components of an oxygen prescription

Only your healthcare provider can change your oxygen prescription, which will include:

- When to use your oxygen (during sleep, rest, or activity, or at altitude)
- How much oxygen you need for each activity (the number setting if you use pulse flow, or liters per minute if you use continuous flow)
- What type of oxygen equipment fits your lifestyle and oxygen requirements
- A Certificate of Medical Necessity (required by Medicare and many other insurers)

### Your supplemental oxygen company's role

The company supplying your supplemental oxygen will provide a basic oxygen system, including equipment and disposable supplies like nasal cannulas or humidifiers. The company will also share information on using your equipment safely and properly, provide 24-hour emergency service for equipment malfunctions or power outages, verify insurance coverage, and bill your insurer.

# OXYGEN EQUIPMENT

## Types of oxygen systems

Work with your healthcare provider and supplemental oxygen supplier to identify the oxygen system that best fits your lifestyle and oxygen requirements. Some things to consider: how active you are, how many steps or stairs are in your home, how often you leave home and how long you're gone for, your physical strength, and your personal preferences. The types of systems available vary between companies.

Most patients will receive an in-home, stationary unit as well as a portable system that allows them to leave home.

### Types of stationary systems

#### Stationary concentrators



- A stationary concentrator pulls oxygen from the air in the room through special filters in the machine to produce a stream of concentrated oxygen through the tubing.
- The machine usually stays in the room where it's placed. You can use different lengths of oxygen tubing to move around your home or workplace.
- The cost of electricity for running the concentrator is not covered by insurance.
- There are two types of oxygen concentrators. Normal-flow devices go up to 5 liters per minute and high-flow devices can go up to 10 liters per minute.

#### Stationary liquid systems



- When oxygen is cooled to a very low temperature, it becomes liquid. The liquid oxygen is stored in a container. As it leaves the container, it warms up to room temperature and becomes a gas, which then flows into tubing.
- Stationary liquid systems are quiet, don't have major moving parts, and don't require a power source to operate.
- Not every supplier offers liquid oxygen, so it's important to check first before you start a contract with that company.
- The large liquid oxygen reservoir usually needs weekly refilling from your oxygen company.

### Types of portable systems

#### Compressed gas tanks



- These green and silver aluminum tanks are filled with compressed oxygen gas and come in a variety of sizes. During use, smaller tanks run out of oxygen more quickly than larger tanks.
- Some compressed gas tanks can be filled at home with a concentrator filling system; these are often referred to as transfill or home fill units and are placed on top of your stationary concentrator.
- You may choose to use a backpack or rolling option for carrying smaller compressed gas tanks. Work with your supplier and healthcare provider to find a good option for your needs.

#### Portable oxygen concentrators



- A portable oxygen concentrator (POC) is a smaller version of a stationary concentrator.
- POCs run on batteries, so you may need to carry extra batteries to keep the unit running. Battery life depends on factors such as the amount of oxygen used and POC size. POCs can be recharged via wall plug or car battery.
- Generally, the smaller the unit, the lower the oxygen output and maximum pulse or continuous flow setting, and the shorter the battery life.
- The larger POCs usually have a maximum setting of 3 on continuous flow and

6-9 on a pulse setting.

- POCs vary in weight from 3-22 lbs; the larger ones require a stroller unit on wheels.
- This type of oxygen system can be used in flight, although you must contact your airline ahead of time to ensure you can bring your POC on the plane.
- Many people requiring high-flow oxygen need more oxygen flow than a POC can provide. If you're considering a POC, please talk with your healthcare provider first to make sure this type of device will meet your needs.



\*

### Liquid portable tanks

- Liquid portable tanks are filled from a stationary liquid system. To avoid the risk of cold burns, training is required before filling a liquid portable tank from the stationary system. Work with your supplier to make sure you understand the system.
- Weighing 3-11 lbs, these units provide a larger reservoir capacity and higher continuous flows than portable oxygen concentrators.
- Keep liquid oxygen units upright to prevent leaks.
- Not every supplier offers liquid oxygen.

### Other equipment

#### Nasal cannulas and tubing

- Stationary oxygen systems connect to 25-foot or 50-foot tubing. The part that goes into your nose is called a cannula or nasal cannula.
- Oxygen set above 6 liters per minute uses special high-flow tubing and may require a face mask instead of a cannula.
- If you use high-flow oxygen, your healthcare provider may recommend a reservoir nasal cannula. These cannulas may come in a pendant or a mustache style. Also known as an Oxymizer®, this type of cannula helps maximize delivery of oxygen. Reservoir nasal cannulas cannot get wet and shouldn't be worn in the shower.



#### Humidifiers

If you use high-flow oxygen, ask your healthcare provider about whether you need a humidifier, which can help prevent nasal membranes from drying out. Flows of 4 liters per minute and above require humidification. Humidifiers cannot be used with reservoir nasal cannulas.



### Types of oxygen flow

There are some differences between pulse or demand flow and continuous flow:

- With pulse flow (also known as demand flow), your portable oxygen system senses when you inhale and only delivers a pulse of oxygen when you breathe in, allowing tanks and batteries to last longer.
- Continuous flow provides a continuous flow of oxygen into your nose, even when you are not breathing in.
- Pulse flow and continuous flow are both prescribed as numbers, but the numbers do not mean the same thing. A continuous flow setting of 2, 3, or 4 liters per minute is different from—and usually more than—a pulse flow setting of 2, 3, or 4.
- Pulse flow isn't appropriate for all patients. Talk with your healthcare provider about the right options for you. You should be tested on a pulse flow device before using it.

\* Photo courtesy of CAIRE Inc.

## GETTING STARTED

### Safety ABCs

#### Follow these tips to stay safe around oxygen:

- Avoid flames, sparks, cigarettes, matches, lighters, gas stoves, pilot lights, and frayed electric cords.
- Keep compressed tanks 8-10 feet from open flames or sparks.
- Avoid petroleum-based products to moisturize your face or nose. Ask your oxygen company or your healthcare provider for names of nasal lubricants designed for oxygen users.
- Don't store tanks in closets, in direct sunlight, or next to heat sources.
- Ensure that stationary concentrators are well ventilated with no items stored on top.
- Contact your electrical company and fire department to let them know you use supplemental oxygen. It's important they're aware of your need for oxygen in case of a power outage.
- Keep smoke detectors and fire extinguishers in your home.
- Don't use cracked or broken tanks.
- Secure oxygen tanks to prevent them from falling and becoming projectiles.

### Ensuring the right amount of oxygen

Your healthcare provider may ask you to periodically monitor your oxygen levels using a pulse oximeter. Ask him or her what your target oxygen level should be. If your healthcare provider asks you to, you can adjust the flow of oxygen depending on what you're doing to keep your oxygen level in a recommended range. It's very important to alert your healthcare provider of changes in your oxygen saturations or an increase in oxygen needs.



### Maintaining oxygen equipment

Ask your supplemental oxygen supplier for instructions for cleaning and maintaining your specific equipment. Some basics you may need to consider:

- Change the nasal cannula and long tubing for the stationary system regularly according to guidelines from your oxygen company.
- Regularly clean the filter if your oxygen concentrator requires one.
- If you're using a humidifier, wash it regularly. Fill it only with distilled water, not tap water.
- Clean face masks regularly.
- Ask your supplemental oxygen supplier to schedule a yearly servicing for your concentrator.



#### BOOST YOUR OXYGEN IQ

Oxygen isn't flammable and doesn't explode on its own—but it will make a fire burn faster. Always keep oxygen away from sparks, sources of flames, and flammable items.



### More PFF oxygen resources

The PFF offers additional resources for learning about and using supplemental oxygen:

- Check out oxygen-related webinars (visit [pulmonaryfibrosis.org/webinars](https://pulmonaryfibrosis.org/webinars)).
- Several patient and caregiver sessions at PFF Summits cover supplemental oxygen issues. Find slides from these presentations at [pulmonaryfibrosis.org/summitarchives](https://pulmonaryfibrosis.org/summitarchives).

# MEDICARE AND SUPPLEMENTAL OXYGEN

Here are answers to frequently asked questions about Medicare and supplemental oxygen.

## How is supplemental oxygen covered by Medicare?

Oxygen equipment and accessories are covered through Medicare Part B on a five-year cycle. If you have original Medicare Part B and a medical need for oxygen, you'll rent your equipment from a supplier for 36 months. The supplier must then continue to provide the equipment, oxygen tank refills, and accessories like tubing and filters for an additional 24 months after the end of the initial 36-month rental period. Your supplier also must service and repair the equipment throughout the whole five-year cycle.

## What if I have a Medicare Advantage Plan?

Medicare Advantage Plans must cover supplemental oxygen. If you have one of these plans, review its benefits to understand how supplemental oxygen coverage works.

## Can my supplier decide to stop providing equipment and services in the middle of the five-year cycle?

Under Medicare guidelines, your supplier is obligated to continue providing your equipment and services for the length of the five-year cycle, except under a few circumstances (for example, in emergency situations).

If your supplier refuses to provide equipment or services, get their intentions in writing, then file a complaint at 1-800-MEDICARE (1-800-633-4227).

## Can my supplier change the type of oxygen system I'm receiving during the five-year Medicare contract?

Your oxygen supplier cannot change the type of oxygen system you receive—or the number of tanks you receive—unless your healthcare provider orders the change.

## What happens if my oxygen needs change during the five-year Medicare contract?

Keep up with recommended medical appointments and be sure to talk with your healthcare provider if you notice a change in your oxygen needs. If your needs change, you should be retested and your healthcare provider will change the Certificate of Medical Necessity as needed. The supplier is required to provide the equipment and service that meet your current medical needs.

## What happens at the end of a five-year cycle?

At the end of the five-year cycle, your supplier is no longer required to provide your oxygen equipment and service. You may choose to remain with the same supplier or switch to a different one. Your healthcare provider will need to complete a new Certificate of Medical Necessity.

Remember: before you enter into a Medicare contract with an oxygen supplier, ask your healthcare provider to make sure the equipment offered by the supplier is correct for you.

## What happens if I move in the middle of the five-year cycle?

If you move during the initial 36-month rental period, you can ask your current supplier to help you find a supplier in your new area. You can also use Medicare's supplier directory, found at [medicare.gov/supplier](https://www.medicare.gov/supplier), to identify a new supplier.

If you move after the initial 36-month rental period, your current supplier generally has a responsibility to make sure you have a supplier in the new area.

## Who owns the equipment when the five-year cycle ends?

The oxygen supplier owns the equipment both during and at the end of the five-year cycle.

## Can I decide which brand of oxygen equipment I receive?

You can ask for a specific brand, but a supplier is only required to provide you with the type of system prescribed by your healthcare professional, not a specific brand. Before you enter into a Medicare contract with an oxygen company, ask your healthcare provider to make sure the equipment offered by the oxygen supplier is correct for you, as different companies may offer different choices.

## If I have original Medicare, what will my costs be?

You're responsible for 20% of the Medicare-approved amount. A Medicare supplemental insurance plan (also known as a Medigap plan) may be able to help with the 20% coinsurance.



## TRAVELING WITH OXYGEN

### Air travel and PF

Whether you're at sea level or high in the mountains, the atmosphere is made of 20% oxygen and 80% nitrogen. But at higher elevations, there is simply less air—the atmospheric pressure is lower—so there is less oxygen for you to breathe.

Since commercial airplane cabins are usually pressurized to an elevation equivalent to about 1,500-2,500 meters (about 5,000-8,000 feet), passengers are breathing less oxygen while in flight. Many people with PF can safely travel by air, but for some, air travel can be dangerous. If you have PF, it's important that you discuss your travel plans with your healthcare provider weeks or months before planned travel, even if you don't normally use oxygen at home.

### Determining your ability to travel by air

To determine whether you should travel by air, your healthcare provider may order tests including:

- Pulse oximeter to check your oxygen level
- Six-minute walk testing
- Pulmonary function testing
- Arterial blood gas measurement to check your oxygen and carbon dioxide levels
- Echocardiography
- High-altitude simulation test (HAST) measuring your oxygen level while you breathe air with a reduced oxygen level (15% oxygen instead of 20%)

Based on these test results, your healthcare provider will determine whether you need oxygen while in flight. You'll then need enough time to notify the airline, have your doctor fill out paperwork for the airline, and coordinate with an oxygen supplier. If your oxygen requirements are too high or you have other medical conditions, your healthcare provider may advise you to not travel by air, since your oxygen levels may drop dangerously low in flight.

### Airline requirements for oxygen

In the United States, airlines are required to allow battery-powered portable oxygen concentrators (POCs) that have been approved by the Federal Aviation Administration (FAA). Most airlines require you to bring your own POC, but not all POCs are allowed by all airlines. Each airline maintains a list of which POCs they will allow on board. Airlines won't allow you to bring filled oxygen tanks (green cylinders) or liquid oxygen onto the plane.

Your healthcare provider must complete paperwork ahead of time that instructs the airline on how and when you should use oxygen. Your healthcare provider can also help arrange for a short-term POC rental from an oxygen supply company.



#### BOOST YOUR OXYGEN IQ

A small number of airlines will provide you with oxygen on board rather than requiring you to bring your own. There's usually a charge to use an airline's oxygen. Check with your airline for details.

## Checklist for air travel with oxygen

- ✓ Prepare weeks or months ahead of time. See your healthcare provider and notify the airline as early as possible. While some airlines only need 48 hours advance notice, it's advisable to prepare at least a month in advance.
- ✓ Be aware of the airline's specific policies for which oxygen concentrators are allowed, what paperwork is required before you travel, and what documentation you need to have with you when you travel. Contact your airline for further information.
- ✓ Be sure to bring enough batteries—and make sure they're fully charged. The FAA requires you to have battery life equal to 150% of your expected travel time. Remember to factor in time for traveling to the airport, waiting to board, layovers, deplaning and picking up baggage, and traveling from the airport to your destination after arrival. The plane may or may not have an electrical outlet available if your batteries run out. Always "plug in" while waiting for your flight.
- ✓ Ask your healthcare provider if you should monitor your oxygen level in flight with a portable pulse oximeter.
- ✓ If you're traveling outside the United States, different regulations may apply. Contact your airline for guidance, and remember to bring the correct electrical plug adapter for the country you're visiting.



## Planning for battery time

**When planning battery life for a flight, be sure to include your total travel time. Here's an example:**

Travel time to airport.....	1 hour
Checking in, going through security, waiting to board .....	2 hours
Flight.....	2 hours+ 1 hour*
Deplaning, picking up baggage .....	30 minutes
Travel time after arrival to your destination.....	1 hour

**Minimum battery time  
needed for this trip: ..... 7.5 hours**

\*The FAA requires battery life equal to 150% of your expected flight time. For this example, a two-hour flight, the FAA would require battery life of three hours for the flight itself.

## Traveling to high elevations

If your destination is at an elevation above sea level, you may suffer from breathlessness with small degrees of exertion or even while resting. Your healthcare provider may advise you to not travel to destinations at high elevation.

## Traveling by car

Inform your oxygen supplier of your travel plans and your oxygen needs at your destination. The 12V DC outlet power source charger in your vehicle can charge your portable oxygen concentrator.

## Oxygen at your destination

Your healthcare provider may determine that your portable oxygen concentrator is sufficient for your travel needs. If you need an additional oxygen delivery device while away from home, your oxygen supplier may be able to coordinate with a supplier at your destination to provide the equipment you need. Be sure to plan ahead of time.

## HAVE QUESTIONS ABOUT SUPPLEMENTAL OXYGEN THAT AREN'T ANSWERED IN THIS GUIDE?

The PFF Oxygen Information Line provides information and resources to individuals using supplemental oxygen and to their caregivers. The Oxygen Information Line is staffed by trained representatives who can help you understand how to access oxygen, how to use it safely, and what to do if you have a problem.



Call the PFF Oxygen Information Line Monday to Friday between  
9:00 a.m. and 5:00 p.m. CT at 844.825.5733.

## ACKNOWLEDGMENTS

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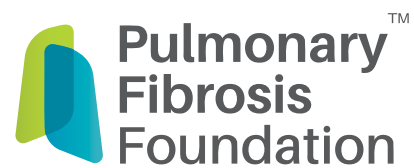
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Please note that any information contained in this guide is for informational and/or educational purposes only. It is not intended to be a substitute for professional medical advice. Always consult your personal physician or healthcare provider with any questions you may have regarding your specific medical condition.

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