



# Patients Guide

To Living with

# Pulmonary Fibrosis



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## FOREWORD

“Write your obituary.” “Make peace with your family.” “Say your goodbyes.” “Make sure your affairs are in order.” “Take a vacation — you have less than two years to live.” Too many times patients have walked into my office bewildered and confused, told only that they have Pulmonary Fibrosis and two years to live. Newly diagnosed with pulmonary fibrosis, they seek answers, explanations, and support.

The questions running in an endless loop through their heads are released rapid-fire: Why me? Why now? How long do I have to live? Is there a cure? Will my children get this disease? Is there anything I can do differently — eat, exercise, travel? If they are fortunate enough to have a practitioner that is both knowledgeable and compassionate, they may get some answers. Those who are not so fortunate immediately turn to family and the Internet for the answers they so desperately seek.

Providers are not perfect. All too often providers fail to provide answers out of a lack of knowledge and time, not compassion. Sometimes, they get a diagnosis wrong. During this difficult time, patients who have access to others with experience on this journey with pulmonary fibrosis experience a sense of relief in this shared understanding.



I have had the privilege of caring for patients with pulmonary fibrosis (PF) or another interstitial lung disease (ILD) for two decades. While we know significantly more than we did when I began my work, there is still much to be done. We recognize more and more that one critical piece of the puzzle is patient involvement in developing a care plan. Providing patients with the power to control their lives by giving them information about the disease allows them to remain hopeful, controlling the life they have rather than allowing the disease to control them.

PF Warriors — started by Bill Vick — has a mission to see the lives of patients with PF-ILD improve. Led by Bill, assisted by Shelly Mathis and Richard Morgan, PF Warriors has compiled this booklet about topics that most commonly affect patients with PF-ILD, selected based on input from hundreds of patients with PF-ILD.

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There are few people with the tenacity and determination to do everything in their power to improve the lives of PF-ILD patients, to advocate for research, to raise their voices for betterment of patients than these individuals. They are constantly looking for ways to educate, inform and move the search for a cure forward. It was only logical that this booklet with easily understood chapters, covering topics that are rarely addressed in physicians' offices with a simple, in-depth approach answering patients' most pressing questions be published.

I am personally impressed and comforted by this booklet that is designed to be understood by patients living with PF-ILD and their caregivers alike. The many patients who contributed to this project experienced the lack of information available, and by offering up their experiences, these patients advocated for themselves and for patients like you.

As a physician, I cannot understate the incredible value of a source with verifiable and valuable information for patients with PF-ILD. While I cannot hope to truly understand what patients and caregivers go through, I have been blessed and privileged to serve you, and hope that this readable, straightforward, and jargon-free book will support you.

For the established or newly-diagnosed patient, I hope this is a useful segue to help you not only adjust to this new area of your life, but thrive, living the best life you can live.

For the practitioner, this is something you can put in your patient's hands with their new diagnosis to empower them and work with them in a team approach to managing their disease. And as our information changes, and this organization grows and learns and evolves, this guide will evolve too.

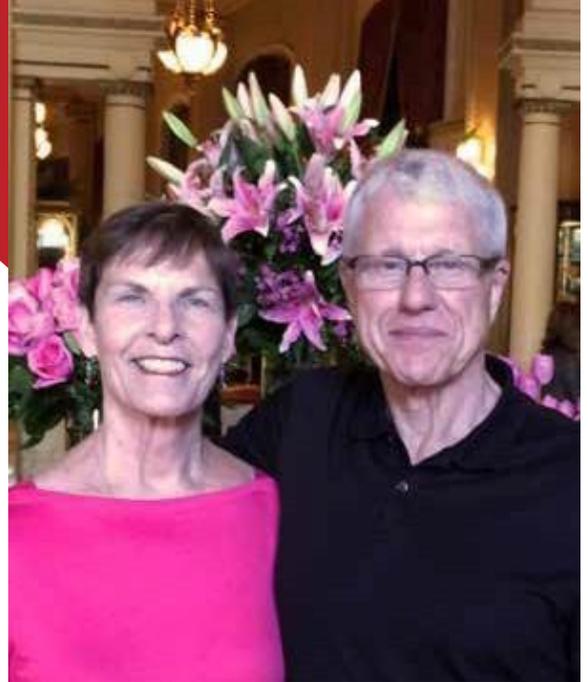
In the end, no one asks to have PF-ILD. But as I have often said to many of my patients, the goal is to learn to live with the disease, not for the disease. And the only way to begin living with PF-ILD and not for it, is to learn about it. I hope that by answering your most important questions, supplying accurate information, and providing you with the culmination of hundreds of patient experiences, this booklet will help you in your journey along this path.

Sincerely,

Yolanda Mageto MD, MPH, ATSF  
Director of Interstitial  
Lung Disease Program  
Baylor University Medical Center  
Dallas, Texas

# WELCOME

Welcome to the PF Warriors, an international group of pulmonary fibrosis/interstitial lung disease patients and caregivers providing meaningful help and support to one another. Our motto is 'Patients Helping Patients' and our mission is to provide the tools and resources for PF patients to live their best life possible despite the diagnosis of PF. We look forward to getting to know you.



My journey with PF took the typical path common to most of us. Although I had always been very athletic, I progressively became shorter of breath for several years before receiving the correct diagnosis of idiopathic pulmonary fibrosis in 2011. At that time, there were no effective medications or treatments, and I was given NO hope for my future life, in fact, I was told to get my affairs in order.

My wife, Patti, and I could not accept that there was nothing we could do to improve this outlook. We were further convinced that no other PF patient should have to experience this bleak diagnosis without hope for their future. We realized patients need so much more than the medical community was offering at that time, such as significantly more information, ideas from other patients living successfully with PF, oxygen resources, group discussions, dietary ideas, involvement of any kind from other patients, caregivers, as well as the medical teams treating PF. So, Patti and I started the PF Warriors support group in 2014 to fill these needs. We have now grown from 3 members in North Texas to over 2,000 in every state in the US and 14 foreign countries.

We believe in the tremendous power of peer-to-peer support. We know what it's like when you wake up every day suffering from lung disease and struggling to live with your "new normal" life. We are here to help you by answering questions and sharing any late-breaking disease management news.

Perhaps most importantly, we support each other through the ups and downs of living with PF. In other words, we will be your "ear to listen and the shoulder to lean on."

The pages in this guidebook are an accumulation of thoughts, wisdom, and comments from our members to help you on your journey.

Bill Vick,  
Founder.

# DIAGNOSIS AND YOUR DOCTORS

You have been diagnosed with a form of interstitial lung disease. You need more information; your doctor may have explained your condition to you, but it is sometimes hard to remember all the details. There are additional actions you can take that will enhance the efforts of your pulmonary specialist on your behalf.

## REMAIN CALM

You have had some tests, and your pulmonologist has made the diagnosis. If you are like many others, you had never heard of pulmonary fibrosis (PF) or interstitial lung disease (ILD) before. Interstitial lung diseases are relatively rare compared to chronic obstructive pulmonary disease (COPD), a much more common breathing-related ailment.

The diagnosis may have shocked you and generated a lot of anxiety and concern to learn that currently there is no cure available for PF or ILD. That's true, but medications are available that have been shown to significantly slow the progression of lung fibrosis.

Medical research and promising clinical trials are underway that may offer even more effective new treatments soon. Many PF Warriors have found that lifestyle changes as well as new drug therapies are helping PF patients not only extend their lives longer than previously expected, but also improve the quality of their lives.

## GET A CORRECT DIAGNOSIS

Pulmonary fibrosis is a relatively rare lung disease that is often misdiagnosed. Many lung-related diseases exhibit similar symptoms. It may have a known or unknown cause (like idiopathic pulmonary fibrosis or IPF). Some patients report being mistakenly treated for bronchitis, allergies, asthma, or COPD (a different type of lung disease).

Pulmonary specialists often must employ multiple tests and a process of elimination to develop a correct diagnosis. It is important to work with your medical professionals and continue to take positive actions that are still under your control.

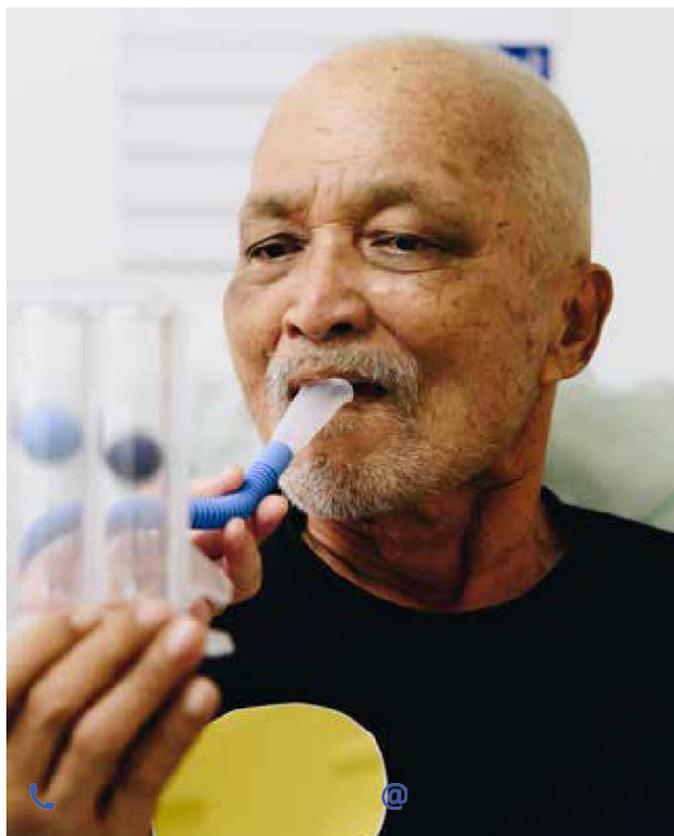
Although it's understandable to feel anxious, it may be wise to get a second opinion from another pulmonary doctor who specializes in PF-ILD lung diseases. The sooner you get an accurate diagnosis, the quicker proper care can be provided.

## TESTS

Respiratory Therapists (RT) perform pulmonary function tests (PFTs) during your doctor visits. One test is the "six-minute walk." A RT uses a pulse oximeter and timer to monitor your oxygen level and pulse rate as you walk at your normal speed for exactly six minutes. The six-minute walk test measures your oxygen saturation during exertion. If your oxygen saturation drops below 88% during the test, it indicates to the doctor that you need supplemental oxygen. It is a necessary test needed to justify insurance coverage to pay for your oxygen service.

Spirometry is another common test conducted by the doctor.

Measurements of lung strength as you breathe normally are taken as you inhale and exhale as hard and as long as you can. Spirometry measures and charts your lung capacity as it changes over time, often stated as forced expiratory capacity (FEV1) and forced vital capacity (FVC). Frequently, your doctor visits will also include blood tests to monitor blood oxygen level and your overall physical health.



## EXACERBATIONS

An exacerbation may have led to your seeking help. An exacerbation is a sudden negative change in your lung function. An exacerbation permanently reduces your previous lung capacity and can shorten your life or even be fatal if not taken seriously and promptly treated.

## VACCINATIONS

Get your vaccinations and stay current. Colds, viruses, and flu are hard on lung patients. Review your medical records and make sure you and your family members are up to date on shots. In addition to the annual flu vaccination in October, ask your pulmonologist about pneumonia shots, shingles shots, and even a tetanus shot. Vaccinations are free or inexpensive, and you just might avoid a hospital stay. Be aware that some vaccinations require booster shots. Your doctor can help you determine if you need a booster shot by testing.



## DOCTOR VISITS

Visits to your doctor and your pulmonary specialist are precious minutes. You will want to glean as much knowledge as possible from each conversation with your pulmonary specialist.

For the appointment, arrive prepared with a written list of questions you want to be answered and concerns you have. Jot down questions as they occur to avoid the inevitable memory lapse during the next doctor visit.

Be sure to take a pen and paper to take notes about the answers you receive and any other recommendations the doctor makes. Having your caregiver with you at your appointments is valuable for insights they can provide to the doctor, and so you can discuss important matters with them later. Some patients and caregivers decide who will take notes and who will listen prior to the doctor's appointment.

## PFF CARE CENTER NETWORK SITE (CCN)

The Pulmonary Fibrosis Foundation (PFF) has established criteria for medical centers to gain PFF certification and listing as a member of their Care Center Network.

These centers are dedicated to improving the lives of those living with pulmonary fibrosis. They must have expertise in treating adult patients with fibrotic diseases and utilize a multidisciplinary approach to deliver comprehensive patient care along with other specific criteria.

For a list of approved sites, visit the Pulmonary Fibrosis Foundation website: <https://www.pulmonaryfibrosis.org/researchers-healthcare-providers/community-engagement/care-center-network>.



### DAILY JOURNAL

Many patients find that a daily journal helps in setting and keeping goals and charting their progress.



# MEDICATIONS AND SUPPLEMENTS

## PRESCRIPTION MEDICATION

Take your medications as prescribed. Develop and maintain a regular schedule for taking any medicines. Also, it may be wise to ask your doctor for emergency prescriptions for cough medicine and an antibiotic in case you get sick during a holiday or a weekend. Some patients find that the many medications they take can leave their skin dry and delicate so moisturizers may be necessary. Medications for PF can cause sensitivity to sunlight. Wear sunscreen when you go out.

FDA-approved PF medications have been available for several years. Drugs by two manufacturers have been shown to retard the progress of lung fibrosis.

**OFEV** and **Esbriet** may slow the progression of lung fibrosis. Other promising drugs are now in clinical trials. Talk to your lung specialist to see if one of these medications is right for you. Both drugs work in a similar manner and have similar side effects. Follow these manufacturer's links for more information:

- Boehringer Ingelheim (nintedanib) <https://ofev.com>
- Genentech (pirfenidone) <https://esbriet.com>

Note: OFEV and Esbriet are registered trademarks of their respective companies.

These approved medications are **very expensive**, with copay amounts that could be unaffordable for many. There are foundations that provide substantial financial aid to cover some or all the copay burden if you are approved.

One such resource is the **Healthwell Foundation** at <https://healthwellfoundation.org>. Another patient advocate firm also may be able to help you apply for aid at <https://copay.org>. In some cases, the drug manufacturer can provide the drug at no cost if insurance and financial conditions are met.

As an example, **Open Doors**®, a complimentary patient support program for OFEV provides copay support. Copay support can make the drugs very affordable, sometimes at no cost.

## OVER-THE-COUNTER SUPPLEMENTS

Over-the-counter (OTC) supplements are available from many sources.

Be aware that the Federal Drug Administration (FDA) does not test or approve the health claims made for supplements and vitamins.

OTC supplements may alter the effectiveness of prescribed drugs, so it would be wise to consult your doctor before adding any OTC supplements. In the absence of federal testing, a reliable testing authority recommended by some doctors is ConsumerLab.com at <https://consumerlab.com>.

**USP** and **NSF** are other marks of certification that supplements have been third party tested for safety and purity. Some of the information on this site is free, however, to benefit from full reports and recommendations, a subscription is required.



### REQUEST A COMPREHENSIVE VITAMIN PANEL

A blood test with a vitamin panel will determine any vitamin or mineral deficiencies, especially your vitamin D level. Discuss proper supplementation for your specific situation with your healthcare professional.





## RESOURCES

### CREATE A TEAM FOR YOUR FUTURE

Your team should include your spouse or another person who will act as your number one supporter and caregiver.

Be sure that you include your primary care physician, your pulmonologist, and other specialists you see regularly, like a cardiologist, gastroenterologist, rheumatologist, or other healthcare professionals. Make sure that your healthcare team shares information with each other. Good communication among your team helps avoid prescription issues or duplication of tests and optimizes the care you receive.

### EDUCATE YOURSELF ABOUT YOUR LUNG DISEASE

There is more information on the Internet than facts. The Internet is filled with opinions, but how reliable and current are they? Recognize that some information on the Internet may be recent, but also you will find outdated data and misleading opinions. Your specialist should be able to provide current medical knowledge



#### **A WORD OF CAUTION...**

Ten-year old studies on the Internet about life expectancy can cause you unnecessary concern.

Such data, based on "averages," may be obsolete. Medications to significantly slow the progress of lung fibrosis have been available only since 2014. Focus on your own situation and avoid over-concern about "averages."

## CONNECT WITH RELIABLE SOURCES OF CURRENT KNOWLEDGE

Specialized foundations act as reliable sources of knowledge about lung diseases. National organizations, like the American Lung Association at <https://lung.org> and the American Thoracic Society at <https://thoracic.org>, cover a wide range of lung diseases and offer helpful information about various disease types. They also provide links to other resources and support groups.

The Pulmonary Fibrosis Foundation: <https://pulmonaryfibrosis.org> focuses on the PF-ILD lung disease types and offers reliable information along with a way to search for support groups, like PF Warriors, by geographic location. Other good sources of information are the pharmaceutical firms who offer FDA approved medications. You and your pulmonologist will decide if a medication should be included in your plan.

The Pulmonary Wellness Foundation: <https://pulmonarywellness.org> is a resource where people with respiratory diseases can find support and information about their disease including education, research, and latest news about respiratory diseases and even pulmonary rehabilitation instruction online that you can benefit from when you cannot attend in-person sessions.





## JOIN A LOCAL SUPPORT GROUP

You will get positive inspiration from the courage and optimism of other patients as well as learn other ways to create your new normal. Large population centers will generally have more than one support group of patients with the same or very similar diagnoses. Support groups provide hope, “an ear to listen to you and, at times, a shoulder to lean upon,” where doctors and other healthcare experts have limited time to spend with individual patients.

Support groups often address your issues, big and small. Support groups put you in touch with other patients and provide information and opinions that come from actual experiences.

Join a local PF support group if one exists in your area. Check out our PF Warriors support group at <https://pwwarriors.com>.



For groups in your local area, links to help you find other support groups are: <https://pulmonaryfibrosis.org/life-with-pf/support-groups> and <https://lung.org/support-community/better-breathers-club>.

## YOU ARE NOT ALONE

Stay engaged with other people and patients. When you are part of an active local support group you can expect to become better informed. Support groups learn about the latest research and the clinical trials being conducted globally.

PF Warriors invites pulmonary specialists and other healthcare experts to make presentations at virtual Internet-based meetings where patients and professionals exchange viewpoints and tackle common issues.

The PF Warriors virtual meetings mean you can tune into sessions no matter where you may be located. You gain a greater sense of personal empowerment which, in turn, reduces stress and anxiety. It really helps to be able to talk with other patients and learn how they have adjusted to cope with their new reality.

To attend virtual meetings, first register at [www.PFWarriors.com](http://www.PFWarriors.com). Once registered, you will automatically be invited to meetings. Membership is free, and as a member you will have the opportunity to meet other patients, caregivers, and families, join meetings and be able to volunteer to help others. We are a volunteer group and there is no charge or obligation of any kind by registering.

If you are a member, you can access high quality video recordings of speaker's presentations at [www.pfwarriors.com](http://www.pfwarriors.com).

If there is no support group in your local area, PF Warriors can help establish one with your local cooperation. One of our goals is to use what we have learned and developed to become the support group for support groups in other geographic areas.

If you need help in establishing or enhancing a support group in your geographic area, please contact us. We will be happy to share our programs, materials, and organizational processes with you. Email us at [support@pfwarriors.com](mailto:support@pfwarriors.com).



## SUPPORT FROM FAMILY AND FRIENDS

Close friends and family can offer more support and encouragement. It's best to let your friends give you emotional and moral support rather than medical or legal advice.

You may find it necessary to modify or curtail certain activities. Don't be shy about asking for a little help from family and friends. Those close to you will usually respond positively when you request help with something they can handle for you. Attempting to do too much in a day can cause stress, frustration and exhaust you.

Review your recurring obligations, club meetings, commuting, etc. and prioritize your various activities. Allow more time for rest, relaxation, doctor visits, or simply doing what you most enjoy. Being pulled in several directions at once can cause depression or mental and physical stress.



**Be mindful that you are still in control. Alter your lifestyle as necessary to best suit your new challenges. Retain important and enjoyable activities and eliminate or delegate other obligations.**



## QUIT SMOKING

You already know about dangers of smoking. Lung disease patients simply must stop all forms of smoking, “vaping” and nicotine...period!

Some quit cold turkey. Others quit over a period of weeks, with medications, patches, nicotine substitutes, group therapy, even acupuncture treatments. There is really no alternative. Get help.

## AVOIDING IRRITANTS AND INFECTIONS

Protect yourself from infections. Drugs that suppress your immune system can make a simple infection grow into a life-threatening event. Your doctors will discuss with you the need to avoid situations where you can get a virus, flu or an infection of your airways from a bad cold. Be doubly careful around crowds or individuals who may be contagious, particularly young children and babies. Be extremely careful in situations where touching is common such as funerals where people often cry and hug.

Some PF patients have allergies, runny nose, excessive phlegm production that easily lead to upper respiratory infections. Make sure you stay in touch with your pulmonologist who can prescribe an antibiotic and other measures to control infection.

If you do get a cold or if you experience flu-like symptoms, don't delay. Contact your pulmonary specialist immediately. If your pulmonologist is not available, ask for a nurse or other professional who can give you advice and help with medications or treatments immediately. Going to the hospital or emergency room are options if you feel like your life is in danger. But you might also expose yourself to other infections from patients with other illnesses. Use your best judgment.

## REMOVE COMMON IRRITANTS

Eliminate feather-filled bedding and any contact with birds, including as pets. Feathers are a common source of airway and lung irritation to those with lung disease. It is best to eliminate feather pillows or natural down filled comforters and bedding, opt for allergy-free substitutes.

Other common irritants to avoid are dust and fine particles that are airborne by construction activities, particularly silica in countertops.

Other irritants include mold, metal dust from brass, lead, steel, pine wood dust, livestock, vegetable or animal dust, hair dressing, asbestos, plastics, resin, paints, metal working fluids, roofing materials, vinyl tile work, working with imported cement pipe and sheeting, moldy or green hay, grain, barley, silage, sugar cane refining, tobacco plants, mushrooms, potatoes, paprika, wine processing/ vineyards, cheese making, coffee bean dust, mildew, BBQ grilling smoke, outdoor fireplaces, secondhand tobacco smoke, and kitchen fumes from frying, toasting, or roasting.



## USE A CLINICAL-STYLE MASK WHEN NECESSARY

Don't reuse a previously worn lightweight disposable mask. If you touch the outside of your mask, then touch your face, you may be defeating the purpose of wearing the mask. It is best to dispose of a used mask then wash your hands. Keep them handy. Carry several with you and change them often. Some masks are washable and reusable but always read the manufacturer's instructions about your particular mask.

A lightweight mask over your nose and mouth really can help you avoid breathing problems associated with all the bad stuff floating in the air. Leaves change in the Fall and provide a colorful show, but you need to avoid raking leaves. Leaves carry microscopic spores, dust, and mold that play havoc with your breathing and cause respiratory difficulties.



## BE AWARE OF YOUR SURROUNDINGS

Large gatherings, crowded shopping malls, young children, adults with colds or flu, or travel by air, rail, ship, or bus also expose you to viruses and bacteria that can cause severe infection of your airways and lungs. Sure, there are times when you cannot avoid exposure to irritants and other people's sicknesses. In such cases, the health benefits of wearing a mask should outweigh any minor concern you might feel. In most cases, others will understand your need to avoid both dangerous indoor and outdoor breathing issues.

## HAND HYGIENE

Wash your hands frequently and don't touch your face. Many bacteria are found on hands. Hand washing with soap and warm water is the best protection from bacteria, germs, and other microbes that cause sickness. Also, try to change your habit of unconsciously touching your face and eyes. Germs are easily transferred from hands to your eyes, nose, and mouth.

Use a hospital grade hand sanitizer. Germs are frequently transmitted by shaking hands or touching public surfaces like faucets, phones, all handheld devices, countertops, keyboards, etc. It only takes a few seconds and a few drops of sanitizer to reduce germs that can otherwise make a pulmonary patient very sick.

Keep hand sanitizer dispensers in the kitchen, your car, purse and in bathrooms to make it convenient to use often.

Avoid shaking hands. Instead, exchange "fist bumps" or simply greet someone by name and give a nod. Acknowledgment without a handshake helps reduce a common source of viral and bacterial illnesses. Elbow bumps work well also. If appropriate, tell people why you can't shake hands. Consider wearing protective medical or vinyl gloves and discard them when you go home. Another suggestion would be to tell your friends "I wish I could give you a big hug right now, but I can't. Thanks for understanding."

## SANITIZE YOUR HOME

Wipe down common use surfaces frequently. You may have observed that hospitals have hand sanitizer dispensers near every door. Healthcare professionals have a habit of cleaning their hands before and after entering each patient's room to help avoid the spread of germs.

At home, your doorknobs, countertops, phones, all hand-held devices, appliances, exercise equipment, chair headrests and arm rests, and TV remote controls are among items to wipe clean with disinfectant or sanitary wipes. Regularly disinfect your cell phone as well, a common source of germs.

Change home air filters often. Most people, especially lung patients, spend a lot of time indoors, so the quality of your indoor air is important year-round. Airborne pollutants, microbes, mold, dust mites, and plain old dust are inevitable, so install high quality pleated air filters on your heat and air conditioner systems to remove significant amounts of irritants from the air you breathe.

Establish a schedule for changing out filters more frequently than a manufacturer may recommend helping ensure clean home air quality. For example, change one-inch thick filters every six weeks. Change four-inch-thick pleated filters every three months even though they are advertised to last six months to a year. There are reputable companies that can clean and sanitize your home duct systems too.

Don't forget the cabin air filters in your automobiles. They filter out much of the fumes and highway pollution. Dirty air filters reduce the effectiveness of your heating and cooling systems and allow more dust and other irritating stuff to circulate around you.

Park shoes in your entry hall or mudroom. You can't know where shoes have been and what they bring into your house. Many cultures have left their shoes at the door for thousands of years. This habit keeps common sources of dirt and many nasty bacteria such as *E. coli* (that can be found on the bottom of shoes) out of your home.

Now, you may not be able to insist that everyone who visits you remove their shoes, but you can reduce shoe-borne grime by asking family members to remove footwear at the door and enjoy the freedom of stockinged feet. Keep in mind, however, that socks can slip on polished floors. Be careful.

# MANAGING SYMPTOMS

## HELP FOR THAT COUGH

This topic is a persistent subject for much discussion among PF patients. Some patient favorites are over the counter cough remedies including Fisherman's Friend and Ricola cough lozenges. Your pulmonologist can give you recommendations too.

Some patients report fresh or unsweetened pineapple juice is a tasty cough suppressant. The juice contains bromelain, an active enzyme that combats inflammation and soothes a cough.

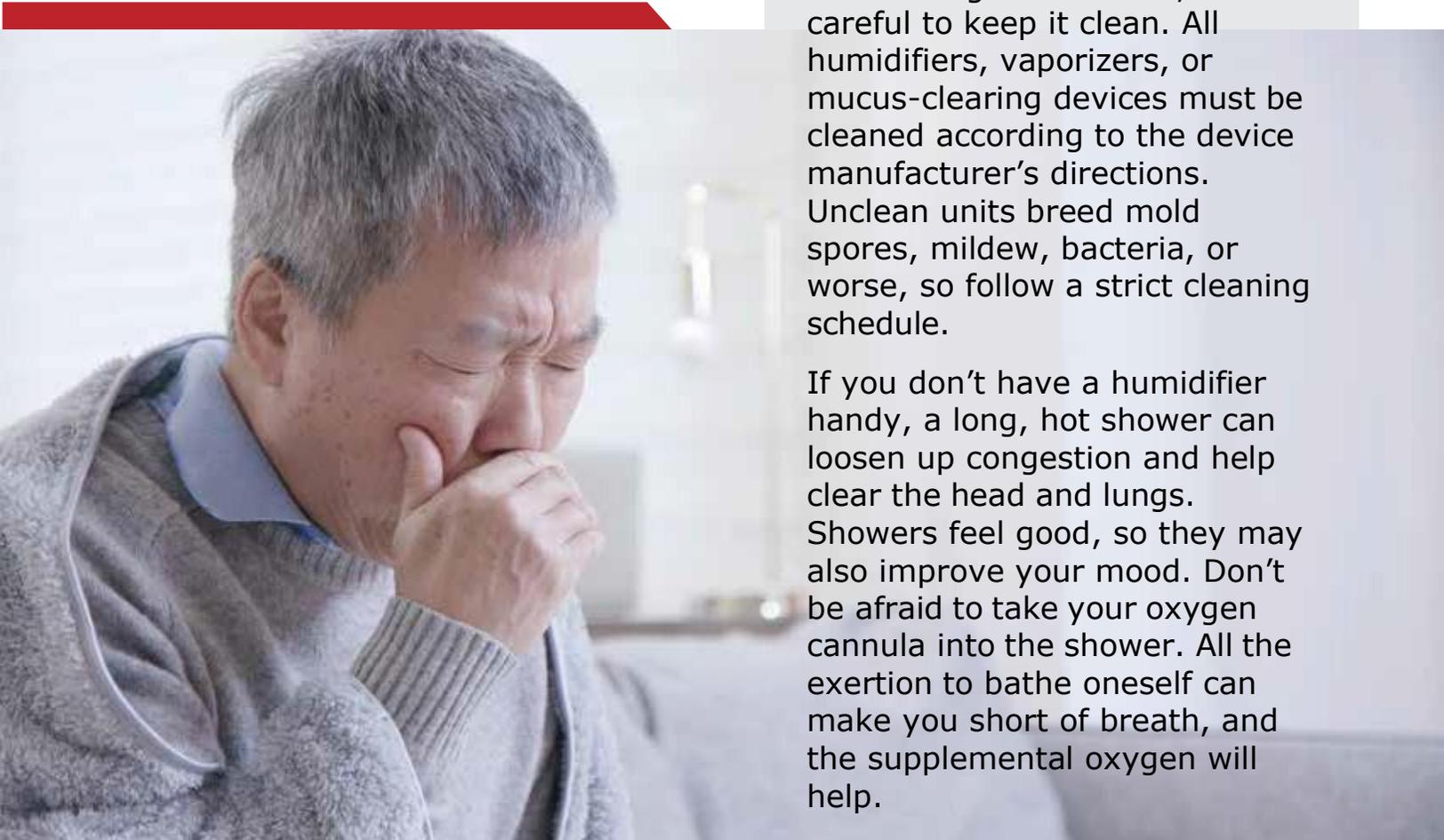
Some patients like a mixture of two parts cider vinegar to three parts honey, while others prefer a cup of hot water with the juice of one lemon and some honey to help with coughs and mucus.



### IMPORTANT

When using a humidifier, be careful to keep it clean. All humidifiers, vaporizers, or mucus-clearing devices must be cleaned according to the device manufacturer's directions. Unclean units breed mold spores, mildew, bacteria, or worse, so follow a strict cleaning schedule.

If you don't have a humidifier handy, a long, hot shower can loosen up congestion and help clear the head and lungs. Showers feel good, so they may also improve your mood. Don't be afraid to take your oxygen cannula into the shower. All the exertion to bathe oneself can make you short of breath, and the supplemental oxygen will help.



## STUFFY NOSE

Many patients wake up with a serious stuffy nose that makes normal breathing difficult. An effective treatment is a sinus rinse. One that is recommended by doctors is **NeilMed**. The rinse is made from a provided additive and distilled water, and the applicator can be a squeeze bottle or a neti pot. This method puts the solution through your nostrils, and the rinse helps remove irritants in your sinus passages. Your doctor also can provide a prescription for sprays that help.

## CONGESTION

Check with your doctor regarding vaporizers, traditional humidifiers, and ultrasonic humidifiers as an aid to reduce congestion.

Vaporizers use steam and a fan to emit a humid mist and reduce congestion at night. Traditional humidifiers use tap water and a device with a fan to emit a cool mist into the room. Modern ultrasonic humidifiers use high speed vibrations and a fan to release a fine vapor that increases room humidity and helps reduce congestion.

Your doctor or respiratory therapist can advise you on the use of mucus clearing devices such as an acapella valve or others. These devices can assist in loosening mucus and its expulsion from your system.



## DIARRHEA

Avoid serious dehydration problems by taking steps to mitigate a significant case of diarrhea. This is a malady that depletes fluid, sodium and electrolytes that your body needs to stay healthy. OFEV or Esbriet and certain supplements may cause diarrhea.

Your doctor or your pharmacist can advise you on managing side effects for these drugs, which may include diet adjustments such as temporarily going on clear liquids for a while if the symptoms are severe and the use of medications like Imodium. Imodium (Loperamide HCl) tablets are a short-term measure to control serious symptoms. They act to quickly halt diarrhea and the associated cramping. They give your body a chance to re-balance using liquids and helpful foods. Imodium or the generic version is available over the counter. Follow the instructions for use and your doctor's recommendations.

Constipation can be uncomfortable and painful. If an over-the-counter laxative, such as Miralax or Colace/docusate (avoid stimulant laxatives like bisacodyl) doesn't work, discuss the problem with your doctor because he or she may have other recommendations for you.



## HEARTBURN

Heartburn (GERD or acid reflux disease) is a common affliction that creates a "burning sensation" in the chest and radiates upward. It is caused when the muscle at the top of the stomach relaxes too much, allowing food to go back up the esophagus and even into the throat. This acidic mess, in turn, irritates the esophagus. At times, gases or even juices can spill over into the lungs. That is bad news for pulmonary fibrosis patients and strictly should be avoided. Your doctor can prescribe an appropriate medication for your situation and can suggest dietary changes you can make. Ask for a list of GERD-friendly foods or try an anti-inflammatory diet. In addition, they often recommend that you do not eat before bed and try to sleep with a raised head or sleep on your left side.

# PSYCHOLOGICAL SUPPORT

## MAINTAIN AND IMPROVE YOUR MENTAL/EMOTIONAL HEALTH

You are now facing a life-altering situation. For most lung disease patients, it is a shock to receive the doctor's diagnosis. You have a rare lung disease that is affecting your ability to breathe normally. You have probably gone through several stages including questioning, denial, grief, and fear similar to the Kübler-Ross stages of grief. Later, you will begin to accept your chronic lung condition, and you will find ways to manage and adapt to your new reality.



### IMPORTANT

Many other patients have used their new situation to rebuild relationships with their loved ones.

Now is not the time to avoid expressing your feelings or shutting others out. That only makes you feel more alone. Take extra time to talk with your spouse, family, and friends and ask for their support.

## STRESS IS YOUR ENEMY

Schedule time to relax and visualize events or experiences, or anything that gives you positive energy, serenity, or comfort. Many patients find that taking time to meditate for only fifteen to twenty minutes each day is just the refreshment they need. Tai Chi, yoga or qi gong have become popular due to the stress relief they provide those who practice the movements each day. As far as dietary supplements for stress relief, before purchasing and taking them, ask your doctor first, as they may interact with your prescribed medications.

## ANXIETY AND DEPRESSION

Anxiety and depression are relatively common among lung patients. There are effective treatments such as counseling and medications that can help. If you are fearful or stressed, talk to your doctor, and describe the things that are troubling you. Trouble breathing at night due to a stuffy nose can create anxiety or a panic attack. A weight gain, or loss, low energy, or irritability may be signs of mild depression. Your doctor may even notice the signs and ask you about your moods. You can safely confide your anxious feelings to your doctor, or another patient who understands. Some recommend meditation or practicing mindfulness.

# NUTRITION AND EXERCISE

## EXAMINE YOUR EATING HABITS

Diet and weight control are things that you can control. Being over or underweight could be detrimental to your overall health and vitality. Crash diets and fads can create health issues. Seek advice from your doctor or a dietary specialist. You cannot manage your genetic makeup, but you can begin developing a sensible diet that you can live with in the future. A proper diet tends to lead to more energy, and it will help you develop a healthy overall body weight and body mass.

## STAY HYDRATED

Choose water as your main drink throughout the day unless you have a medical reason not to do so. Dehydration can be dangerous, cause constipation, heartburn, and other issues. Water will help keep you hydrated and keep your body functioning properly. Living with a serious lung disease becomes easier when you are properly hydrated.



## ASK FOR A PRESCRIPTION FOR PULMONARY REHABILITATION

Your pulmonary specialist can prescribe pulmonary rehabilitation at a local rehabilitation facility. You will gain a better understanding of your disease, how to breathe better, and how best to cope with your symptoms. You will attend monitored sessions for six to twelve weeks. Rehabilitation specialists will measure your physical progress and provide you with practical advice for both physical and mental improvement. Many patients consider this the best thing they did to feel better as the education provided in all aspects of their wellbeing helps to provide confidence that restores a feeling of control.

## MAINTAIN A HEALTHY EXERCISE PROGRAM

All agree that physical exercise helps you maintain a healthy body and helps you extend your ability to handle many daily functions. Physically active people also tend to enjoy better mental health to help cope with breathing issues. Talk to your pulmonologist or pulmonary rehabilitation specialist before starting an exercise program. We can't emphasize enough about the importance of exercise to improve your blood/oxygen exchange and overall health. You will need an exercise plan that fits your capabilities and a plan that you can enjoy performing.

# SUPPLEMENTAL OXYGEN

Oxygen Therapy also known as supplemental oxygen often is prescribed. Pulmonary fibrosis inhibits your lungs' ability to exchange carbon dioxide and oxygen. Oxygen is important for proper functioning of all your organs and wellbeing. Some patients may suffer from low blood oxygen levels only while asleep. Other patients may require extra oxygen almost all the time. Oxygen (O<sub>2</sub>) is considered a medication and requires a prescription. Your lung specialist will be able to determine if you require oxygen supplementation. There are many forms of supplemental oxygen delivery systems including compressed oxygen tanks, home concentrators, portable oxygen concentrators (POC), and liquid oxygen.

## COMPRESSED OXYGEN TANKS

This delivery system is easily recognized in the U.S. by the color of the tank which is primarily silver with a green top. Tank color varies in other countries. It is portable in that it can be mounted on a cart and pulled or pushed around by the user. Depending upon the type of control regulator used it can be low flow (.5 to 5 LPM), high flow (.5 to 15 LPM) and can be used with a "conserving device" which is a term used to describe a special control regulator that delivers a pulse bolus of oxygen triggered by the breathing of the user. The tanks come in a variety of sizes with the most common called an E tank which is about 30" tall depending upon the type of valve configuration it has. The durable medical equipment provider (DME) can provide filled tanks and replace used tanks, but the trend now is to provide the user a "home fill unit" where tanks can be filled by the user.



## OXYGEN CONCENTRATORS

Oxygen concentrators concentrate the oxygen in the ambient air by removing nitrogen and thus deliver almost pure oxygen to your nostrils, typically via plastic tubing and a cannula to supplement the oxygen that you are breathing.

Home oxygen concentrator units can operate 24 hours per day from a normal house electric outlet. They weigh about 40 pounds and have rollers for moving the unit. They typically come in 5 LPM units that deliver what is known as “low flow” and in larger units, the most common is a 10 LPM unit that is considered “high flow.”

## PORTABLE OXYGEN CONCENTRATORS (POC)

POCs are battery powered and weigh about 8 to 20 pounds. The smaller portable units deliver only about 1 LPM in a pulse flow, and if you can get by on that level, it allows you to get out and be more active. The user must keep in mind that the settings (usually 1–5 or 6) on a POC do not correspond to LPM, and as stated above only deliver about 1 LPM total regardless of the settings. Don't be misled.

Larger portable units are available that are heavier (around 18 to 20 lbs.) and these typically can deliver 3 LPM constant flow and have conserving settings from 1–5 or 6. They are usually on a cart though.

## LIQUID OXYGEN

For some patients who require a higher flow, liquid oxygen is a solution that can deliver a higher flow and a longer lasting supply. Liquid oxygen is becoming difficult to obtain due to changes in regulations regarding insurance reimbursement to durable medical equipment distributors (DMEs). DMEs can provide liquid oxygen but many choose not to do so because of the difficulty in getting reimbursed. In some cases, the oxygen delivery solutions described above cannot deliver enough oxygen to some patients due to inadequate capacity as with the small POCs, and even the E tanks at a high flow may not last more than a half hour. Liquid oxygen could provide a longer timeout at a higher flow, however, as above stated it is difficult to obtain.



## IMPORTANT

With POCs, battery life is always an issue and it can limit the user's time away from home. Fast charge technology in newer units that allow one to recharge in an electrical outlet, or in most cars helps. Users should take extra batteries, a car charger, and a power supply (like the one for portable computers) with them when going out or traveling.



**Oxygen therapy is considered a drug and requires a prescription.** Oxygen therapy and concentrators are quite common, but they require a doctor's prescription after noninvasive tests determine your need. Oxygen equipment is often rented from a local compressed gas/durable medical equipment (DME) distributor. Equipment fees are frequently covered by Medicare or other supplemental insurance if the patient is qualified. Your pulmonologist will probably choose a supplier for you or recommend reliable distributors in your local area. Rental agreements have "strings" attached, so be sure to read an agreement before signing. Your insurance company or Medicare may require yearly oxygen testing, usually as a six-minute walk test, to ensure you continue to qualify for supplemental oxygen and may require documentation from your pulmonary provider consistent with their requirements.

**Traveling with oxygen** is certainly possible, but it takes a bit of pre-planning to be sure you are doing so safely. For air travel, remember airlines pressurize cabins to around 8,000 feet elevation. That is marginal for those who need O<sub>2</sub> constant flow above 3 LPM, due to the limitations of portable oxygen equipment. The total output of lightweight shoulder-carried portable oxygen concentrators is only about 1 LPM. Some large POCs can provide 3 LPM constant flow, however, the issue of battery life for a long flight is important to consider. Most airlines require one to carry batteries to provide at least 1½ times the length of the flight including layovers.

If you can stay saturated with your POC at rest, using your POC in an airplane should work. Keep in mind that your POC must be FAA approved and most are. Airlines have various regulations, so it is vital to contact your airline prior to a trip to learn what they provide or approve for use. Also, prior to all air travel, you must notify the carrier that you will carry your portable concentrator on-board and answer any questions they have about the device.

Travel by car is generally not a problem unless your trip takes you to high altitudes such as the Rocky Mountains. High altitude air contains less oxygen, and you will feel the effects which can become dangerous. You may need to take along your home oxygen concentrator when you travel if you need to sleep using constant flow O2. POCs are usually only pulse, and pulse is not recommended for sleep.

Some find that carrying a lightweight home concentrator such as the **Inogen At Home** which weighs only 18 lbs. is a good solution for using at your destination. It doesn't have lithium batteries and can be packed in your baggage and checked when traveling by air, too.

Keep in mind that when traveling, surfaces are full of germs. It is wise to practice increased sanitation. Wash your hands frequently, wipe down surfaces with sanitary wipes, and wear a mask in crowds. It pays to be careful out there.



## PULSE OXIMETERS

Use a pulse oximeter to measure your blood oxygen level. You can usually pick up an inexpensive pulse oximeter at pharmacies for twenty to fifty dollars. They are accurate for home and gym use. Get to know your O2 level and the effects of your daily activities.

The device is noninvasive and indirectly measures the level of oxygen in your blood. The oximeter comfortably fits over one of your fingertips and provides readings of your pulse rate plus your blood oxygen level.

The healthy range for an O2 reading is 90 to 100. Readings below 90 indicate that too little oxygen is available for your body to function normally.

Physical exercise beyond your capabilities will temporarily deplete your oxygen level. Most patients also experience a reduction in blood oxygen during sleep. Your doctor may prescribe O2 in some form for you.



## SLEEP APNEA

Seniors usually need seven to eight hours of restful sleep each night. Unfortunately, lung disease is often accompanied by sleep apnea (interrupted sleep patterns). You may feel more fatigued than usual. It could be a side effect of one or more medications, but if you have sleep apnea, your pulmonologist may recommend that you have an overnight test at a sleep lab to help determine the root cause of your frequent fatigue. Patients who receive treatment for sleep apnea report a significant improvement in energy level.

Continuous Positive Airway Pressure (CPAP) machine users report that they now wake up refreshed. Talk to your pulmonologist if you feel fatigued all day, sleep apnea can be treated in several ways, and you can expect to regain your vitality.

**Insurance coverage for oxygen therapy options varies**, so review your existing health insurance policy, including Medicare and any supplemental coverage to determine if oxygen therapy in various forms is included and if there is a deductible amount you will have to pay for concentrator(s), a CPAP machine, auxiliary items, or bottled oxygen. Avoid nasty surprises by carefully checking your health insurance coverage in advance of possible needs. Your doctor's office staff may be able to help you determine options and to handle the initial steps to acquire the oxygen you need.



## TRAVEL TIPS

**Consult with your physician about your trip.** Start with a phone call to confirm you are well enough to travel. This will provide confidence to you and your travel companion(s). Frequently check the weather and be sure to bring the right clothes to make sure you are always comfortable. Airlines will need to know in advance about any portable oxygen concentrator you will need during the flight. There is no need to add stress to your trip. If you are traveling by air with oxygen, paperwork may need to be filled out by your doctor in advance.

Be sure to include helpful information when packing such as a list of your current medications, list of emergency contacts, your insurance information, doctors' contact information including fax numbers. Make 3 copies: one for you, one for your travel companion, and one to leave at home.

Bring enough of your medications to last the whole trip, plus some time after you return. If possible, bring along an emergency prescription for an antibiotic in case you need it. Oxygen users should pack extra supplies.

Here are a few other suggestions:

- Make your itinerary "doable."
- Consider purchasing travel insurance.
- Do your research and understand where the local hospitals are.

# PERSONAL AFFAIRS

## UPDATE LEGAL DOCUMENTS

Everyone needs to have a valid will that deals with the details related to how you want your estate distributed.

In addition, doctors strongly advise that lung patients prepare and sign a **Directive to Physicians (Living Will)** to provide medical staff and your family or surrogates with your wishes at a future time when you might be suffering in a terminal condition from which you are expected to die within months (even with available life-sustaining treatment) or if in the judgment of your doctors you are suffering with an irreversible condition.

**The Healthcare Power of Attorney** allows you to designate someone to make your medical decisions if you are not able to do so.

**The Statutory Durable Power of Attorney** appoints an individual you choose as your agent (attorney-in-fact) to act for you in any lawful way with respect to all the listed powers you stipulate. The document also provides for additional individual(s), who are successors to that agent. A successor can act for you only if your first appointed agent dies, becomes legally disabled, resigns, or refuses to act on your behalf. This agent will be able to make financial decisions for you so think carefully about who you will appoint. You can change or cancel these documents at any time if you are physically and mentally able.



### IMPORTANT

Legal requirements will vary depending on the laws of the state where you reside. You should consult a licensed attorney to advise you as you prepare your will and the other appropriate documents well in advance of actual need.

**A HIPAA Waiver** (or authorization to release medical information) allows you to appoint certain persons to receive information from your healthcare providers.

**A Declaration of Guardian** allows you to designate the person(s) you want to serve as your guardian if one is ever needed.

It is probably a good time to check beneficiary designations on your insurance policies, bank accounts and retirement accounts. Legal estate documents and directives today will help to resolve unwelcome burdens on your heir(s) and other family members in the future. If your estate is sizable or complex, an estate attorney also may provide advice to minimize estate taxes, changes in asset ownership, setup of trusts, and potential ways to streamline probate.



## TRANSPLANT

Lung transplantation is possible for many on a qualified list, but life after a lung transplant demands more stringent monitoring, life changes, and multiple medications. Lung transplant surgery has become more common as the only cure for pulmonary fibrosis, but the number of actual transplants is still quite small compared with the number of lung disease patients. Each hospital that conducts lung transplants has its own guidelines and methods of qualifying patients.

To qualify for a lung transplant, the patient is evaluated by a pre-lung transplant team that conducts a large number of tests that determine the patient's overall physical condition and determines any comorbidities that may need resolving, and any that could disqualify the candidate from being considered.

The team examination weighs the benefits to the patient against many factors such as age, comorbidities, patient compliance to doctors' instructions as to lifestyle and medication, if you have a caregiver and an adequate support team, and overall health survivability of that patient.

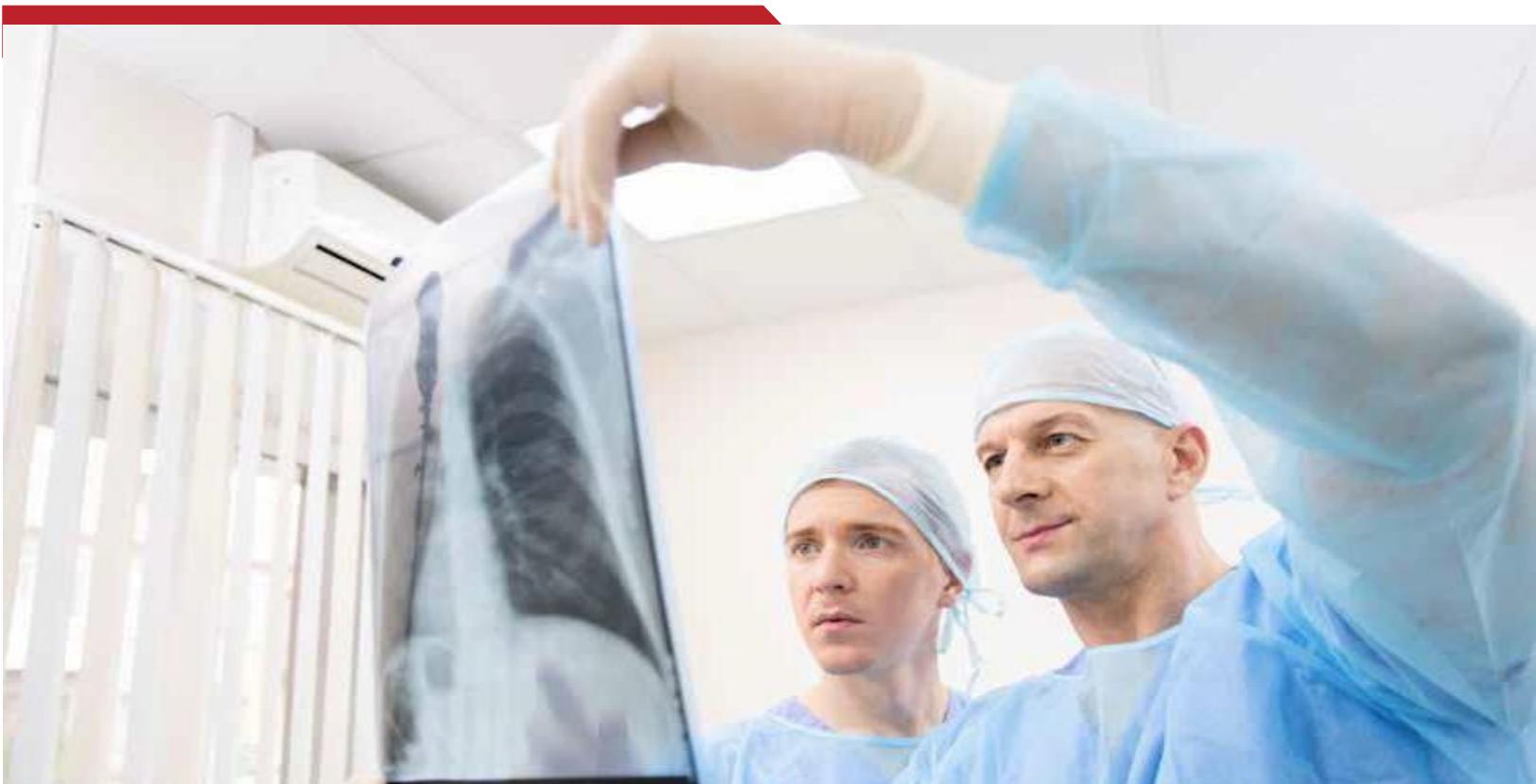
Some who are evaluated for lung transplantation can be disqualified due to serious unresolvable comorbidities or preexisting medical reasons. In some cases, the candidate can be turned down because he/she is "too well" or not sick enough to be listed. In these cases, reevaluation can take place if the patient's condition deteriorates, which is fairly common, as PF is a progressive disease that cannot be cured.

There is a great demand for lungs. Currently, logistics and transportation are serious issues unrelated to the lungs themselves. According to transplant surgeons, the supply of lungs is generally adequate but sometimes geographically distant. First, donor lungs must be a “match” medically. Then, lungs must be procured, and the transplant begun within a period of about six hours. For example, when donor lungs are in Florida and the transplant patient is in Los Angeles, air transportation and a streamlined delivery become vital to a successful transplant.

Currently, there are waiting lists of patients who have undergone careful examination and are rated as qualified. Those on the list must then await a donor with a close match.

Getting on a qualified list does not mean a new lung match will be found at the right time and place.

To consider a lung transplant, a first step is to discuss the possibility with your lung specialist. Your doctor can provide specific information to you about the qualification processes, facilities that perform transplants, the risks involved in the actual procedure, life after transplant, and the likelihood of your qualifying for a single lung or double lung transplant.





## CLOSING THOUGHTS

You are not alone. Pulmonary Fibrosis is not a well-known disease, but 150,000 people in the U.S. are presently living with PF. We encourage you to include your family in your health care decisions and develop a medical care team including a cardiologist, gastroenterologist, rheumatologist, pulmonologist, psychiatrist/ psychologist, and primary care physician.

Reach out to us; PF Warriors is all about Patients Helping Patients. Our goal is to provide education, inspiration, and support to members, and that's the reason we created this guide.

We bring patients and families together to better understand Pulmonary Fibrosis and devise ways to enjoy life as they cope with lifestyle changes. We have a global reach, with members in all states and some 15 countries. We continue to develop alliances with other organizations that help raise public and medical PF awareness. We encourage development of new products and services for the PF community.



Patients Helping Patients



**This guide is not a substitute for the advice of licensed healthcare professionals. Always seek the specific advice of doctors and their staff of registered nurses and other specialists.**



## PATIENTS GUIDE TO LIVING WITH PF

This patient guide was created by PF Warriors members and families.

It was an effort led by and inspired by PF Warrior Dick Morgan. He is the driving force behind this patient guide and, as a pulmonary patient, well understands the importance of patients helping patients living with any interstitial lung disease such as pulmonary fibrosis.

His tireless efforts have made this book possible.

Please consider a donation to PF Warriors which will help ensure that patients will receive the most relevant education, inspiration and support about living as successfully as possible following a diagnosis of pulmonary fibrosis. Compassionate, well-informed health care professionals, researchers, patients and caregivers actively participate within our international group to give meaningful support to patients.

You can donate either online or via mail:

Online at: <https://pfwarriors.com>

Mail to: The Dallas Foundation  
PF Warriors Account  
3963 Maple Avenue, Suite 390  
Dallas, TX 75219

Your donation to PF Warriors demonstrates vision, commitment and caring.

Thank you for your contribution!

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## CONTACT US

 [WWW.PFWARRIORS.COM](http://WWW.PFWARRIORS.COM)

 [SUPPORT@PFWARRIORS.COM](mailto:SUPPORT@PFWARRIORS.COM)

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The PF Warriors Fund is a component fund of The Dallas Foundation, a 501(c)3, publicly supported charity, Federal Tax ID 75-2890371.