



Caregivers Guide

to Living with

Pulmonary Fibrosis



PF Warriors' Caregivers Guide to Living with Pulmonary Fibrosis

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This publication was made possible by generous funding from Three Lakes Foundation. Special thanks to the generous support of the many PF Warriors who gave their time and contributions to this effort.

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This guide is intended for a U.S.-based audience. Please check with your medical team and local support groups to recognize variations in care.

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Welcome Letter

Dear friend,

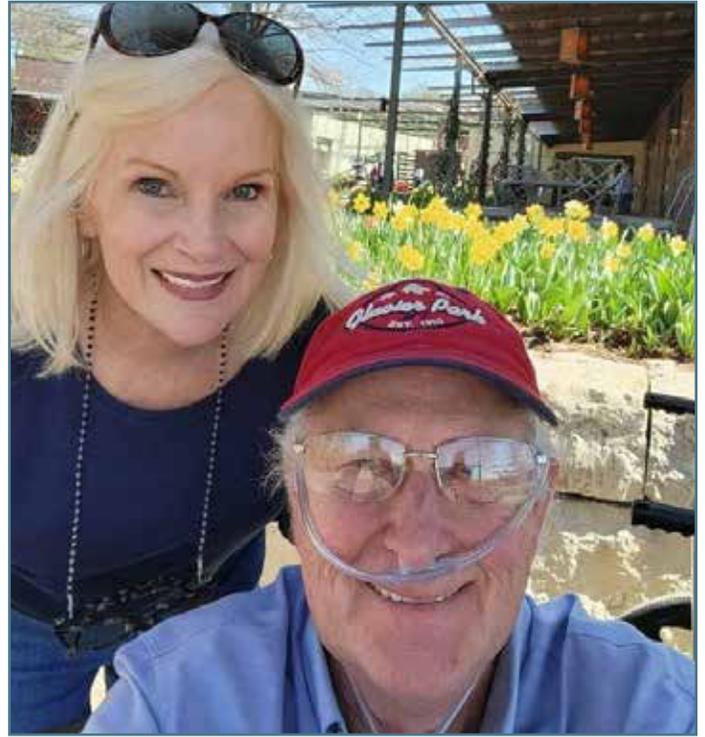
We are sorry that your loved one received a diagnosis of progressive pulmonary fibrosis (progressive fibrosing ILD). At this point, you may have never heard of this condition, and you may be overwhelmed. It is okay to feel this way. It is a major change and a new journey for you, but you have entered a very caring community. Many others are in your situation and are willing to share the journey with you. Having walked in your shoes, we know the challenges that are ahead. We built this guide to help you along the way. In it, we offer you education, inspiration, and support. Please know, you are not alone.

We remember those early days, especially that first appointment to discuss pulmonary fibrosis. It felt like we walked in one door, and everything was fine. When we walked out the other door, our whole lives had changed. One caregiver described her feelings like this, “It was like falling off the edge of a cliff. You didn’t mean to fall, but there you are, wondering to yourself, “What’s going to happen when I hit the bottom? Is there water? Will I survive?”

Some of us wondered, “What can I do to help? What is my role in this? Do I even have a role?” Many of us were in shock. We’d never heard of pulmonary fibrosis. We wanted to know what caused it, and what it meant for them...and for us. What could we do to have an impact on those scary numbers on the Internet?

We also felt some relief. We thought, “Maybe the hypoxia was causing the irritability,” or “Maybe that’s why the medicine didn’t seem to be working (it was treating a disease they didn’t have).” We finally knew what was wrong, after spending too much time wondering and worrying.

The people who contributed to this guide include those who live with patients, as well as those who live nearby or across the country



Shelly Mathis, Caregiver and Executive Director of the PF Warriors, with her husband, Scott Calhoun.

from the patients in their lives. We are spouses, loved ones, or adult children of those who need our support. Our loved ones have a variety of interstitial lung diseases, along with other health concerns. We are also joined by those who have gone through successful lung transplant. We have shared what we learned along the way with the hope that this guide can make your journey a little easier.

As you care for your loved one, remember that we’re here. PF Warriors are “Patients helping Patients” and “Caregivers helping Caregivers.” You are a valued member of the PF Warrior community and play a very important role in our mission. Visit www.PFWarriors.com so you may access additional resources on our website and participate in virtual meetings with peers and medical community experts. Also, please join the ongoing conversation in our very active Facebook group, [PF Warriors](#).

**With best wishes from the
PF Warriors Caregiver Program Team**



What is it?

What You Need to Know About PF and ILD

You came to this point in your life from one of two directions: your loved one has a disease that has progressed, and it is now affecting their lungs. Or the two of you are new to this disease altogether. In either case, you will benefit from understanding what pulmonary fibrosis (PF) is and how it may affect the patient, you, and those around you.

Diseases that cause pulmonary fibrosis belong to the family of interstitial lung diseases (ILD). Not everyone living with an ILD will develop pulmonary fibrosis. And not everyone living with pulmonary fibrosis will have the same experience.

Unlike people living with some other diseases, no two lung disease patients are alike. The medical community calls pulmonary fibrosis that gets more severe over time progressive pulmonary fibrosis or progressive fibrosing ILD.

Before going any further, please note that the scary statistics that you find on the Internet about pulmonary fibrosis are unlikely to apply to you. This is a rapidly evolving field for researchers. Great strides are being made, especially over the last couple of years. **The prognosis statistics online generally represent outdated information. Tell this to your family and friends.** There are many good ways to stay current with the latest news in this field. See the list of recommended sources in Appendix A.

Pulmonary Fibrosis vs. Idiopathic Pulmonary Fibrosis

Pulmonary Fibrosis simply means “scarring in the lungs.” There are many conditions and diseases that cause scarring. For example: Your loved one may have developed pulmonary fibrosis as a result of their advancing case of scleroderma. Scleroderma is the disease, and it is causing scarring in the lungs, or pulmonary fibrosis.

Idiopathic Pulmonary Fibrosis (IPF) is categorized as a disease. When you have IPF, you develop scarring in the lungs, but no one knows why. For the other diseases that may cause scarring, such as autoimmune diseases like rheumatoid arthritis, only some of those patients will develop pulmonary fibrosis.

This is important to know in case you have friends with one of those diseases. Don't assume that they will eventually develop pulmonary fibrosis. Each patient's journey is unique.

Although most of the symptoms of pulmonary fibrosis look similar across the ILD family of diseases, **treatments and outcomes are often different, depending on the cause.** The wrong treatment may be detrimental to your loved one's health. That is one reason why it is so very important to make sure the diagnosis is correct.



What You May be Wondering About

“What does having pulmonary fibrosis feel like?”



Symptoms

People who have pulmonary fibrosis most often report shortness of breath, fatigue, and coughing. But what does it **feel** like? Some patients have said that it's like breathing through a drinking straw, all the time. It isn't too hard to do while sitting down but try climbing the stairs quickly while breathing through a straw!

Patients have said:

“Having PF means being fatigued all the time. It's not a ‘normal tired’ that you can go to sleep and wake up and it will be gone.”

“I feel like I have a young anaconda snake wrapped around my chest. As he grows, I will need lots of help to carry him around. One fear is that every time I breathe out, the anaconda will tighten and further reduce the air coming in.”

“I get panic attacks when I can't catch my breath. It's truly frightening.”

“When I am sitting still, I sometimes forget about pulmonary fibrosis. However, when I stand up, leave the shower, or walk out to get the mail, I feel like I'm underwater trying to breathe.”

“An acute exacerbation could come at any time, so I'm on guard constantly.”

“The reality of what I'm living with angers and frightens me and I ask myself “why?”

“I can leave a room to give respite to the others from my distress. But everywhere I go, there I am.”



“How did this happen?”

Potential Causes

Those people who were already aware of a chronic condition, like sarcoidosis that can lead to lung scarring, may have fewer questions about how they developed this condition. For those who are new to pulmonary fibrosis, they will wonder:

- Was I exposed to damaging substances without knowing it?
- Did I do something wrong that hurt my lungs?
- Are any of my current health issues creating or exacerbating my lung problems?
- Is it genetic?

You two will work with the doctor to learn as much as possible about your loved one’s unique medical and lifestyle history, and what may have contributed to the scarring. Each person’s body and experiences are different, and your loved one may spend a lot of time over the coming months trying to put together his or her own, unique puzzle. Unfortunately, the answer is often not completely clear and you may not determine the exact cause of PF.

Many pulmonologists believe that there are potential lung irritants all around us, and some people may not be able to fend off, or recover from, the small injuries they cause. Environmental exposures are the subject of a lot of ongoing research. This is an area that you can stay on top of to make sure your home and lifestyle aren’t part of the problem.

Common Comorbidities

Pulmonary fibrosis may not be the only condition you two are living with. Here is a partial list of common comorbidities, or diseases and conditions that co-exist in people living with PF. You may already know that your loved one has one or more of these, and new treatments will have to be coordinated with the ones already in use. Some of these items are potential causes, and some are effects, of pulmonary fibrosis. All should be prevented or addressed.

- GERD (gastroesophageal reflux disease)
- Inflammation
- Infection
- Congestive heart failure
- Sleep apnea
- Hypothyroidism
- Anemia
- Cancer
- Anxiety or depression
- Blood clotting risk
- Emphysema
- Obesity
- Pulmonary arterial hypertension
- Venous thromboembolism

“What else might be going on?”



Staging the Disease (Not)

You may have already heard the term “progressive, fibrosing disease,” or “progressive pulmonary fibrosis.” People with an ILD who develop scarring in their lungs may progress (develop more scars over time) quickly or slowly. Talk with the medical team about what they anticipate for your loved one’s unique situation. Be prepared for ongoing medical testing to track changes.

Pulmonologists don’t “stage” most interstitial lung diseases. There are two main reasons it is very difficult to predict your future: **too many variables** and the unpredictability of **acute exacerbations**.



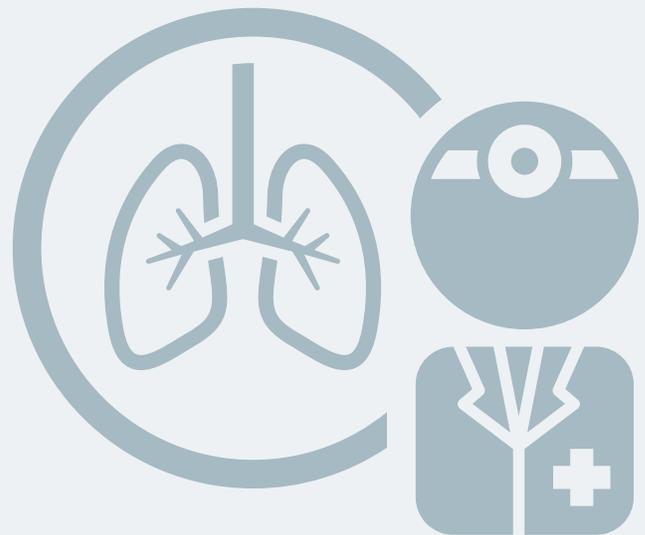
“Why won’t anyone tell me my spouse’s prognosis?”

Too Many Variables

To be able to predict outcomes, doctors need to follow a lot of people with the same condition over time. There are not enough patients in any one category with this rare condition to make that effective.

For example, age, gender, percentage of lung fibrosis, degree of issues with GERD and sleep apnea, medical history and additional underlying conditions, genetics, smoking history, and of course, cause of fibrosis all play into outcomes. Some patients take medicine to address one issue, and that same medicine has a negative effect on one of their other issues.

In addition, having pulmonary fibrosis puts patients at greater risk for developing other diseases, such as lung cancer, which comes with its own prognosis. Then there are factors that are ongoing, such as body weight and nutrition, sleep patterns, stress management, exposure to environmental triggers or infections, compliance and response to medications, exercise, and the development of new diseases. **The takeaway message: your loved one’s situation is unique.**



Acute Exacerbations

Another very important reason it isn't practical to stage this disease is because, unlike many other chronic diseases, a patient may develop an acute exacerbation at any time. An acute exacerbation is a sudden increase in respiratory symptoms (more shortness of breath, more cough, more fatigue).

In some cases, it may result in permanently reduced lung capacity and can even be fatal. Many exacerbations are set off, or triggered, by infection or exposure to something that harms the lungs. The medical team needs to act quickly to identify and address causes and symptoms.

Sometimes, however, no obvious cause is found, but physicians will often still prescribe medications like steroids to try to calm the inflammation.

Warning signs for exacerbations include more pronounced PF symptoms (shortness of breath, coughing, fatigue, weakness), and can include flu-like symptoms, including fever or chills. The symptoms may follow a negative trend over several weeks' time. Caregivers or support partners often see patterns even before their loved ones notice a change. It is very important to speak to the doctor early on about what to be looking for and how you should respond. In general, patients with PF **should not** "wait until Monday to call the clinic" when feeling ill. When in doubt, call the doctor.

Not all exacerbations can be prevented. However, taking preventative steps may not only help the patient stay healthy, but may also help you feel less anxious. See Appendices E and F for suggestions.



Talk with each other about setting expectations with your family and friends.

Although many patients can go through an exacerbation and carry on, there are others who will become critically ill. Some of those people come home from the hospital, but they will live a "new normal." It will be valuable to educate your close friends and family members about exacerbations **now** so that you don't need to have that conversation when in the throes of managing one. Plan ahead about who you will contact if you need to go to the emergency room, and what you might want them to do to be helpful.



What Can be Done About It?

Modern health care is more than just medicine. The pulmonary fibrosis community is a leader in recognizing the importance of caring for the whole person: mind, body, and spirit. That includes you, too. You will participate in many interventions, several of which will also benefit you.

The goal of the items listed below is to protect your loved one's lung capacity. That means controlling disease progression, preventing exacerbations, and managing symptoms. All may help improve outcomes.

Managing Pulmonary Fibrosis

Medicine

There are about 200 diseases in the ILD family, so details about appropriate medical treatments can't be covered here.

“What do I need to know about my spouse's medicine?”



- Two antifibrotic medicines are available for PF and some other progressive ILDs: OFEV™ (nintedanib) and Esbriet® (pirfenidone). The purpose of the antifibrotics is to slow progression. Work with the doctor to determine the best time to start the medications.
- Medicine that is helpful for one disease may be harmful for another. For example, prednisone and other steroids **may** work **against** the goals of other treatments. It is crucial to get an accurate diagnosis, so your loved one is taking the right medicine. Be sure to discuss comorbidities and treatments with all of the specialists.

“We want access to the very latest research and thinking about PF therapies. Where can we find it?”

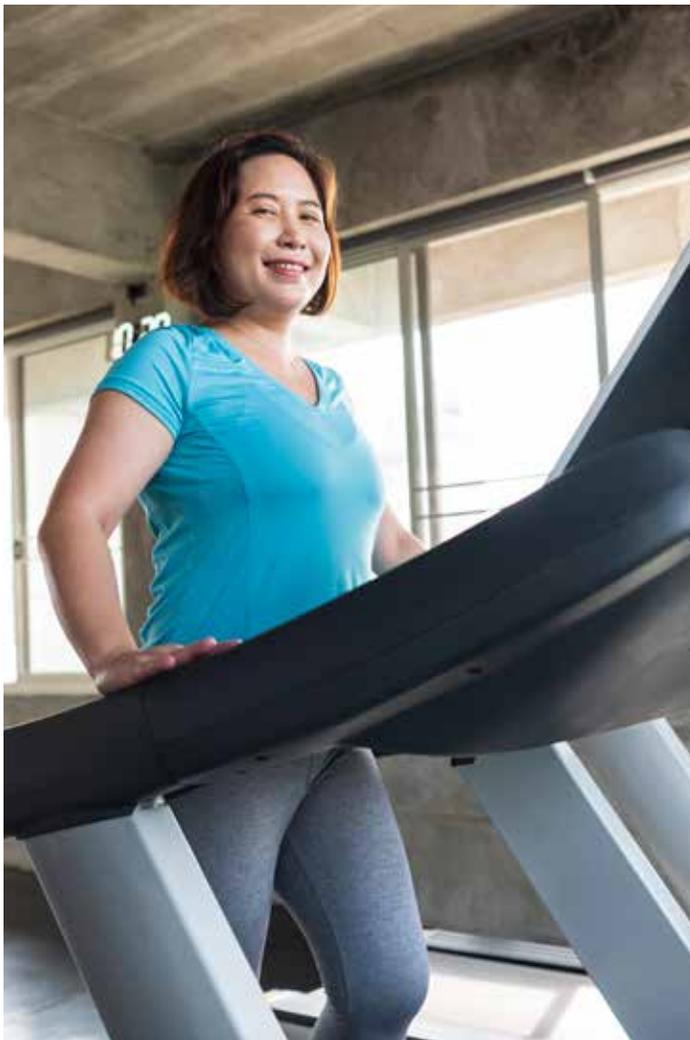


- Stay current by following doctors, researchers, and other key opinion leaders on Twitter, reading vetted or critically examined sources of PF information, and participating in PF meetings and conferences. See Appendix A for a list of vetted resources.
- Go to www.ClinicalTrials.gov to learn about trials that are available. Speak with the medical team about participating in a trial.



Stop Smoking and Vaping

Not only should a person living with PF stop smoking, but everyone in the household should quit. Second-hand smoke is dangerous, too. People with a PF diagnosis are at increased risk for other lung diseases, including lung cancer. Also, if your loved one needs to use supplemental oxygen, smoking around that equipment is hazardous.



Supplemental Oxygen Therapy

Many people living with serious, chronic lung diseases need supplemental oxygen therapy. Patients with **progressive** disease will need increasing levels of oxygen.

A lot has been written about oxygen; it can be confusing and overwhelming. People who are ahead of you on this journey recommend learning all that you can about oxygen therapy **before** your loved one needs it. Take your time to understand it **now** before it arrives on your doorstep. See the list of vetted oxygen resources in Appendix B.

Exercise

Doing the right type of exercise is one of the best ways to manage the symptoms of pulmonary fibrosis. Ask the doctor to prescribe a pulmonary rehabilitation program to introduce your loved one to specific moves and activities that may preserve or enhance lung health. In addition, the two of you should get more active, even when one of you is wearing oxygen. See Appendix A for a list of vetted resources.

Good Eating Habits

Good nutrition affects our overall health, and this is very important with chronic disease. Now is the time to follow general nutritional guidelines for a balanced diet. This includes eating plenty of fresh fruits and vegetables, lean proteins, and avoiding sugar and processed foods.

It is very important for PF patients to manage their weight: not too heavy, but also not too thin. Staying a **little** heavy can mean staying on the safe side of living with PF, as a decline can be hard to address and may get out of control. Those who are considering transplant will need to pay particular attention to weight management, as weight-related comorbidities may affect the ability to get “listed.”

It is not only what they eat and when, but also how they eat and what they do after they eat that becomes important for people living with pulmonary fibrosis. This is partly because so many PF patients also experience GERD. However, all of us should know our food sensitivities and how to manage our microbiome (the bacteria, viruses, fungi, etc. that live in our bodies).

See Appendix I for recommended GERD eating guidelines. See Appendix A for vetted resources on nutrition and pulmonary fibrosis.



Clean Living

- **Clean up the home and work environment.** Most people are surprised to learn that there are things all around us that can potentially harm our lungs. Also, your loved one may need to curtail certain activities while away from home, such as attending bonfires and big barbecue events. See Appendix E for suggested ways to improve living and working conditions.
- **Make the home more hospitable.** Bending, lifting, and climbing are difficult for someone with chronic lung disease. Also, people using supplemental oxygen equipment need to avoid tripping hazards and other challenges with tubing. See the suggestions for improvement in Appendix G.
- **Minimize the risk of infection, which is a common cause of exacerbations.** Develop new, stricter cleanliness habits in the home, car, and workplace. Also, talk with the doctor about both of you getting vaccinated against pneumonia and influenza. See Appendix F for recommendations on ways to clean up your environment.

Support for Mental Health

Stress, anxiety, and depression can go hand-in-hand with any chronic, debilitating medical condition for patients, as well as those around them.

For all of us, the levels of mood-stabilizing hormones in our brains naturally decrease as we age. That means that it is not unusual for older people to experience some depression. Add the stress and anxiety of managing the symptoms of pulmonary fibrosis and it is not hard to see that good mental health is at risk for both of you. This can have a real impact on your overall well-being.

Speak to the medical team about getting a referral to a counselor who can help. Join a support group so you can spend time with people who are familiar with your challenges. Finally, learn about additional ways to manage mental health, such as meditation, spiritual fulfillment, and good nutrition, as well as exercise, including qi gong and yoga.

Transplant

A lung transplant is a wonderful gift of life, but one that requires careful consideration by the two of you, your family, and the medical team. In 2021, only about 3,100 transplants were done in the United States. Knowing that there are more than 50,000 new ILD patients diagnosed every year in this country, and cystic fibrosis, COVID-19, and COPD patients may also qualify for lung transplants, the small number of transplants should set everyone's expectations.

If you are interested in pursuing a transplant, know these basics:

- Getting a lung transplant is expensive financially, physically, and emotionally. Some people consider a lung transplant as trading one type of chronic issue for another. Keep your family involved in the decision-making process.
- It is better to be evaluated for a transplant sooner rather than later. That will give you time to improve on any items that may be in question. Possibilities include getting to the desired weight, addressing osteoporosis and other comorbid conditions, giving up bad habits, and/or moving into a manageable living situation where you have a strong support system.
- Not everyone will qualify for a lung transplant. Age, comorbidities, and level of available support are but a few of the variables that the transplant team will take into consideration.
- Your loved one will need to be seen as “compliant”: one who carefully follows the doctors’ instructions. That is something that will be tracked over time.
- Lung transplant centers can have differing guidelines for approval. If you get turned down from one center, you may investigate the possibilities at other centers.
- Many transplant patients move (temporarily) so they may live within proximity to the hospital for several months following surgery.

PF Warrior Caregivers' Top 10 Tips

**1**

Don't believe everything you read on the Internet or watch on YouTube. There is a lot of outdated, and even wrong, information out there. In general, be wary of anything that was written or posted more than three years ago.

2

A Center of Excellence is a medical center that has been recognized by the Pulmonary Fibrosis Foundation (PFF) as a leader in the care of patients living with pulmonary fibrosis. The PFF refers to them as their “**Care Center Network.**” Usually located in or near a large city, the centers are where you will find experts who specialize in your loved one's condition. If at all possible, patients should go to one of the centers to get an accurate diagnosis.

3

Shortness of breath does NOT equal hypoxia. Depend on a pulse oximeter to help you understand oxygen saturation. A pulse oximeter is an easy to use, affordable device you can use at home that clips on a finger to measure the level of oxygen in the blood. You will read that your loved one should stay above 88 percent. However, not all oximeters are calibrated accurately. By staying above 90 percent, your loved one will benefit from a 2 percent safety net.

4

Most PF patients struggle with gastroesophageal reflux disease, or GERD, and it needs to be addressed. Lack of heartburn does not mean that they don't have GERD.



5

If your loved one does not want CPR performed in the event of an emergency, he or she may need a legal document valid in your jurisdiction. Several U.S. states have two very different Do Not Resuscitate (DNR) forms: one for in-hospital care, and one for out-of-hospital (paramedics).

It may be valuable to check the rules in the states where you travel often, as well as the local rules. In addition, look into Medical Orders of Life-sustaining treatment (MOLST) and Portable Medical Orders (POLST). Those forms address more options than a DNR form.

6

Palliative care is care that your loved one can and should receive all along the journey. It is not “hospice-lite.” However, it is important to understand what your Medicare plan will cover.

Knowing this may help you navigate the payment system. Also, some medical systems require the patient to switch medical teams when receiving palliative care, which may not be desirable. Families living with PF have been known to pay palliative care nurses directly (no insurance reimbursement) because they find them to be so valuable.



7 When your loved one starts oxygen, and is on Medicare, you will be required to sign a five-year contract with the supplier. “Local” durable medical equipment suppliers (DMEs) may promise good customer service. But they are very unlikely to help you obtain oxygen and equipment in other states when you travel. The “national” companies (e.g., Apria, Lincare) are unlikely to service you when you travel if you are **not** in their system, **even** if you offer to pay out-of-pocket.

8 Medicare will provide a home concentrator and either a portable oxygen concentrator (POC) OR canisters for mobility. Some durable medical equipment suppliers have a “try before you buy” option, which is worthwhile.

9 Portable oxygen concentrators typically cannot provide more than 3 liters per minute of continuous flow oxygen. Don’t be fooled by misleading advertising.

10 **Warning:** The numbers on the portable oxygen concentrator dial are NOT liters per minute (LPM). They are simply levels set by the manufacturer and they only indicate “more” or “less.” Use the pulse oximeter for guidance regarding turning oxygen up or down, and always follow your medical team’s direction.

How Can You Live with It?

The Good News

Your Loved One is Not This Disease

Let's not forget that, before your loved one became a patient with PF, they were someone who was leading a fulfilling life. They continue to be that person. Do not let this disease redefine them... or you. Of course, during the early days after diagnosis and during exacerbations and transitions, your days and your thoughts will be focused on pulmonary fibrosis. But the many days in between can and should be enjoyed.

There is Hope

While several sites on the Internet list scary statistics about pulmonary fibrosis, that does not need to be your story. Every PF patient's health situation is unique, so care and outcomes are also unique. Here are some positive comments from patients.



"I was diagnosed in early 2014. I'm still going strong (2021) without oxygen. Fight for every step and keep moving!"

"There is always hope! I'm 10 years in and I'm planning trips around the U.S. now! I'm not on oxygen yet. I'm a walker and have always walked 2 or 3 miles every day. I'm still doing it. I think that has really helped me, but I also must have a slow progressing type. I know I'm fortunate if that's possible with IPF".



"I try to walk a line between knowing the truth and realities of our disease and maintaining enough hope and positivity. PF isn't pretty. It's hard and it's debilitating but it gives us a unique opportunity to truly value things so many take as a given".

"After the initial blow, I started reading peoples' posts on here (PF Warriors Facebook group feed) and saw that with a positive attitude, exercise, good diet and other things you can go on for many years. Don't let this disease consume you."



Travel

There are many ways to continue this wonderful hobby, even with portable oxygen. Airlines, cruise ships, and some train travel will require additional planning. Note that airlines and cruise ships will not allow high-flow oxygen equipment, so make distant travel your priority while it's still a possibility. Be sure to review the travel resources listed in Appendix J.



We have made multiple trips across the states to visit family and friends. From Texas to Florida, South Carolina, Tennessee, Virginia, New York, Connecticut, California, Las Vegas, Washington state and even Alaska. We've been to the Grand Canyon, Mount Rushmore, Yellowstone, and the Balloon Festival in Albuquerque. Now, we avoid higher altitudes. This past summer we went to California and played golf at Pebble Beach. It was awesome!

— Victoria R.



The Tough Stuff

Dealing With the Diagnosis

Patients and caregivers have shared that they went through Dr. Elizabeth Kübler-Ross' Five Stages of Grief™ (denial, anger, bargaining, depression, acceptance) soon after diagnosis. Not everyone goes through all stages, and not always in the same sequence. Importantly, many people find themselves revisiting stages too. Being able to recognize that those feelings are common may help you feel better.

The challenges seem to come when you and your loved one are at **different** stages of grief. Be patient with your loved one. Try to be aware of when this happens and consider adjusting your actions and reactions, as possible, to match the needs of your loved one.

Many patients and caregivers report that it took them about six to eight months to get through the trauma of the news, during which time there may be moments of loneliness. However, many people feel that they continue to have some difficult days, even years into the journey. It is likely that your emotions and those of your loved one will become more closely aligned as you adjust, and you will get back to functioning as a team.

Some patients go through the stages of grief all over again when they start using supplemental oxygen. The lessons learned after diagnosis will be helpful at that milestone.

Maintaining a Good Quality of Life

Talk honestly with each other about quality of life. Some people want to do everything they can, including pursuing all clinical trials or transplant, to fight the disease. Others want to make some modifications to daily living and focus on staying on track toward meeting certain personal goals. Some value their privacy and don't want to open up to their inner circle. Others enlist friends and family into a team approach. What do you want? Do your values align? If not, what will you try to do to keep both of you happy? Start this conversation, and revisit it often. Thoughts on this subject will probably change over time for both of you. Remember to share key points with your inner circle.

Some topics you may consider:

- Privacy about disease and progression
- Level of attention to diet and exercise
- Prioritizing travel
- Quitting smoking and other habits
- Sanitizing the home: how much is too much? (E.g., give away the family pet, remove the carpeting, etc.)
- Questioning the medical team's direction and advice: second opinions? Which doctor leads the care?
- Managing the appointments and medications, advocating for the patient: whose job is this?
- Interest in pursuing transplant if that's an option



My loved one had visited 49 of the U.S. state capitols; only Alaska was missing from her list. After her diagnosis with interstitial lung disease, we wondered how we could check Alaska off her bucket list.

We tentatively selected a cruise liner and read the fine print on their policies for medical support, accessibility, and use of supplemental oxygen. Most cruise ships allow travelers to bring along portable oxygen concentrators.

Choosing port excursions turned out to be fairly easy. Our cruise line provided detailed descriptions of each excursion, noting whether it was wheelchair accessible or not and providing a general idea of the level of activity to expect (easy, moderate, strenuous). We chose a bear-watching adventure that was wheelchair accessible because we could see from photos that the path was a boardwalk without rough terrain or elevation. We also checked the excursion length to be sure that the oxygen concentrator batteries would last the entire trip.

We only made one mistake in our planning. The walk to the state capitol building in Juneau was uphill from the boat dock. In hindsight, we should have taken a cab from the dock. But with the frequent stops to rest, we made it, took a photo on the steps of the capitol, and checked Alaska off the list of all 50 states.

— Kathy P.

Managing Perceptions

Telling The Story: Your Family and Close Friends

Who needs to know what? How and when do you want to tell them? Who will do the telling? The two of you might consider making a list so you don't accidentally leave someone out. Keep the list handy to make sharing news easier. You may want to create a text group chat to be used for that purpose.

Here are some things you may want to prepare an answer to:

- Do the doctors know what caused it?
- Is it contagious?
- Can they get treatment? Is there a cure?
- What about lung transplant?
- Did they get a second opinion? See a specialist? Are they getting the best care?
- What exactly does that disease do to their lungs? What's happening inside?



Elevator Speech: Everybody Else

Neighbors, work colleagues, acquaintances.

When others hear that this is a serious condition, they will tend to ask the care partner about it in the hope of avoiding sending unintended, negative messages to the patient. Do yourself a favor and develop a way to explain the disease quickly and simply, what is often called an “elevator speech.” You can work on this together.

What do you both want others to know? Here are some examples:

“She has a rare lung disease called pulmonary fibrosis. It causes scarring in her lungs, which makes it hard for her lungs to work properly. So, she has difficulty getting enough oxygen. She gets short of breath easily and has a chronic cough. No, it’s not contagious.”

“He has IPF, or Idiopathic Pulmonary Fibrosis. It is a serious, rare disease that will affect him for the rest of his life. Basically, he has scars in his lungs that prevent him from getting enough oxygen. No one knows why people get IPF. We are so lucky because he sees some of the very best doctors in the country. You will see us out often because exercise is really good for him... and me.”



Paramedics, Emergency Room Personnel

Develop a different, short speech that you can use in case of emergency. Pulmonary fibrosis is rare; assume that the people in the ambulance and the ER do not know what it is or what to do about it. You may want to show a draft to the doctor to make sure it includes the most important information.

Also, consider having it available in written form so you can ask an ER nurse to attach it to the chart. One example:

“My husband has Idiopathic Pulmonary Fibrosis, or IPF. It is a rare, restrictive lung disease. He needs supplemental oxygen to keep his O₂ sats up above 90%. He’s been using 6 liters per minute continuous flow, but that doesn’t seem to me to be enough. We are here because he has been struggling to catch his breath for nearly 15 minutes and needs help right away. Just now, his pulse ox said only 74%. He has GERD and Type 2 diabetes, both very well managed. He doesn’t have any pulmonary hypertension. I already called his pulmonologist, Dr. Smith. Here is Dr. Smith’s phone number and a list of my husband’s medications.”

What Did You Just Say?

You may have noticed that your loved one has become increasingly irritable over the past several months. Some patients follow a conversation clearly on one day, but not the next. Others have displayed a dangerous change in focus and reaction time when driving. You may be surprised and relieved to know that some changes in your loved one's personality and abilities may be temporary due to low oxygenation. Hypoxia can alter a person's personality or even their ability to think clearly. If that is the case with your loved one, supplemental oxygen may result in improvement.

Of course, getting this diagnosis can be life altering, and everyone will have a different reaction to the news. There may be more bad days than good days as you both adjust to living with PF. Because of that, you may not want to jump to conclusions about oxygen saturation. Also, some patients experience emotional side effects when taking steroids. Although you may not be able to change that, knowing that it may be the medicine talking can help you cope.

You are Not the Boss

One of the hardest parts of living with someone who has a serious, chronic disease is resisting the temptation to "remind" them about something they should or should not be doing. You want to save them; you want the best for them. Your urge to nag or push is coming from the best intentions. Of course, there will be times when you may need to emphasize what is best for their health. Pick your battles.

It is not unusual for the caregiver to know more about managing the disease than the patient. Try to avoid slipping into a parent-child dynamic. A patient may claim to be very private about their condition. Are you sure this isn't their way of saying they want to handle it on their own? Your loved one is an adult who is going to live with this the best they can. One patient said, "I don't want you to fix me. I want you to support me."



Here are some recent comments from caregivers:

"You can't change your husband. You can only change how you respond to him! You can obtain the information and provide it to him. Ultimately, that's ALL you can do. Then step back and let him make the decision whether to use it or not. Take care of YOU."

"I have to remember that it is HER life after all. All I can do is be there for her, love her, spend as much time as I can with her and make her feel good."

"Remember that you can't make her take better care of herself. You can't make people do what they don't want to do. If things look grim, make an emergency plan so you can begin to let go."

"I know at some point with my dad there will be a time when it's just going to be what he decides to do, and I'll just have to accept it. As long as he is okay with his decision, then I will have to be okay with it, too."



PF ON THE MOVE

I took a warrior challenge and made a commitment to myself to make it to the rim of Crater Lake National Park in Oregon, approximately 8,000-foot elevation. The last part of our journey up was a 0.8-mile long hike up about 150 feet from the trail head.

It took my Pulse POC working on overdrive; every pulmonary breathing exercise I have learned over the past three years; slowing down my exertion pace to that of a sloth, while constantly monitoring my O2 levels... but it was well worth it. The view was awesome and magical.

Living my bucket list.

— Keith B.



Evolving Roles Around the House

As the disease progresses, it will be necessary to redistribute the household chores. Every caregiver and patient have their own way of approaching this subject. Just because someone **can** do something doesn't mean that they **should**. That advice goes for both of you. Here are some tips to consider:

- Support their need for maintaining status quo and independence as best you can.
- Some patients do not want to be reminded that they are vulnerable. Giving up responsibilities may be perceived to be a sign of weakness.
- If your loved one has tasks taken away, how will that affect their self-esteem?
- Use “I” phrasing when offering to do more around the house. For example, “Here is what I am seeing. When you mow the lawn, I see you get out of breath, and I worry about you. It looks taxing. It’s hard seeing you use your energy on something I can do.”
- Is it necessary to have a conversation about the chores, or will your relationship fare better if you slowly take on more responsibility over time?

Keeping Your Morale Up When the Chips Are Down

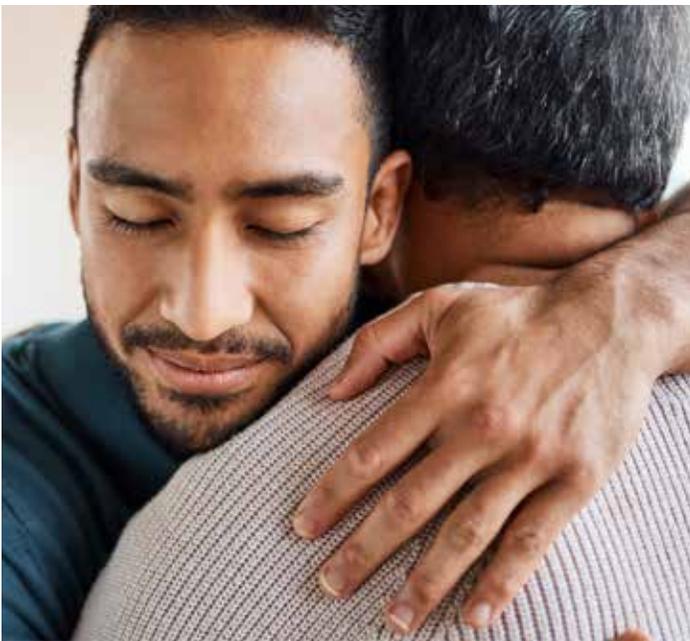
Living with a serious illness can be devastating to the person with the diagnosis, for obvious reasons. It can be very hard on you, too.

When looking for ways to keep your spirits up, recognize that, in general, there are two main things that contribute to your morale*:

1. Feeling valued, important, and appreciated
2. Feeling in control of your near-term future

Feeling Valued

You participate in the management of this disease, in addition to whatever you were already doing prior to diagnosis. Needing to ask for words of appreciation from your loved one can lead to feelings of resentment. This strategy is rarely successful over the long term. Instead, seek alternate sources of appreciation, such as volunteering, participating in a support group, or helping with your grandchildren. It does not need to be a huge time commitment; even small acts of kindness can generate great appreciation.



*Original concept developed by Suzy Norman. Reprinted in this guide with permission.

Feeling in Control

Attempting to correct (control) your loved one's actions is not fun for either of you. When they "cheat" and have a sugary breakfast, for example, they know what they are doing. They may be cheating on their diet, "just this once," to regain their own sense of control. It is not your job to police their compliance with the doctors' orders. As one care partner said, "I am not the breakfast sheriff." Over the long run, your efforts to control them may lead to a parent-child relationship that may come with a side dish of passive-aggressive behavior. This pattern is common, especially with people who need assistance in daily living.

Instead of trying to control your loved one (or the disease), improve your morale by controlling your own activities. What can you change about your own situation to make yourself feel better? Look for small windows of time to do something just for you. This can be as simple as going for a drive, walking around the block, playing cards with friends, doing yoga, reading a book, or talking on the phone. You don't have to go away physically to get away mentally. The goal is not to escape. Rather, it is to improve the sense of control you feel in your life. When looking for ideas, remember: the more flexible in terms of scheduling, the better. Only you will know when you need that morale booster.

On a broader scale, another way to help yourself feel more in control is to educate yourself about the disease, treatments, options, and what may lie ahead. Care partners advise staying at least one step ahead to manage your stress levels. Consider sharing what you've learned with your family and close friends so they may also feel more in control.

Deciding When to Turn to Hospice

There is not a right or wrong time to turn to hospice services. Every patient, family situation, and hospice provider has different requirements. This is partially true because each patient will define quality of life differently.

Officially, hospice care is for people who are expected to live less than six months. “Going on hospice” means that a cure is no longer being sought. Some caregivers have said that they knew it was time to consider hospice when they observed their loved one losing motivation to keep going.

There may come a time when the challenges of managing high-flow oxygen and/or getting **enough** oxygen, issues with comorbidities, paying close attention to eating habits, and dealing with medical side effects, for example, become overwhelming and the desire to “keep up the good fight” diminishes. You will see that the patient’s quality of life has diminished. By doing some homework about hospice ahead of time, you will be better prepared to recognize when it is time to have the conversation.

Consider looking into the various hospice options in your area long before you might need them to avoid the need to make hasty decisions when under stress. Options may include in-home care weekly or daily, or inpatient care in a hospice facility that may have been converted from a former large home, for example. Some hospice programs state that people can “go into” hospice, then decide it’s not for them and “step back out” of hospice.

So, because it is not an exact science, explore options early on. Be sure to get a clear understanding of what is included and what is not, as well as the patient’s portion of the costs involved. Read online reviews of the various local programs. And don’t forget to reach out to your support group peers to learn from their experiences.

See Appendix L to see a comparison of the different types of medical care that are available to support you and your loved one.



How You Can Be Helpful

Sharing is Caring

Go With Your Loved One to a Center of Excellence

See Appendix A for a link to the Centers of Excellence.



Together, Engage With a Support Group

PF Warriors is the largest international PF support group. Go online and join the PF Warriors Facebook group to give and receive information and support.

Share in a Healthier Lifestyle

Look for ways to incorporate healthy choices every day. Some examples include cooking healthy meals, taking walks or exercising together, and building good sleep habits.

Encourage Use of the Pulse Oximeter

Buy extra pulse oximeters (bedroom, living room, work, car, etc.). Allow the pulse ox to help manage your anxiety and the patient's anxiety. Shortness of breath does not necessarily equal hypoxia!

Respect Their Choices and Decisions

Demonstrate respect for your loved one's method of managing their illness:

- Support and encourage the management of oxygen saturation.
- Look for ways to be helpful, not pushy or judgmental.
- Avoid discussing the disease every day. You are not this disease!

Learn About Potential Side Effects of All Their Medicines

Be aware of the possibility of rare side effects and be prepared to talk to your loved one if you spot difficult-to-manage changes in personality, eating and/or bathroom habits. The medical team needs to know and will help you manage them.

Manage Foundation Grants

Antifibrotic medicines are expensive. But, there are foundations that can help shrink your portion of the cost. The medical team should be able to provide guidance on how to access this money. Learn the nuances of each grant so you can anticipate lapses in coverage.

Complete Advanced Planning Documents

See Appendix K for a list of important legal paperwork. Consider completing the documents for yourself at the same time.

Prepare a “Go Bag”

Bring all medications, the medical history paperwork, and advanced planning documents with you if your loved one goes to the hospital. It may be difficult for the hospital pharmacy to obtain the necessary medicines in a timely manner. Note that the antifibrotics are only available from specialty pharmacies.

Go to Doctor Appointments Together

Prepare questions ahead of time, take notes, and advocate for your loved one. It is helpful to take your notebook to all appointments. Ask the doctor if you may record the conversation on your phone to review after you get home. It can be difficult to remember everything that is said during the appointment. Demonstrating a team approach to care may help bolster a transplant evaluation in the future too.

Help With Symptom Management

Check the list of suggestions for managing cough at the back of this guide: Appendix H.

Take the Lead in Planning a Getaway

Plan travel in line with disease management requirements. See Appendix J for additional information about traveling with pulmonary fibrosis.

Redistribute Responsibilities

These household chores can be difficult for someone living with PF. Do them yourself or ask someone to help around the house.

- Emptying the clothes dryer
- Emptying the dishwasher
- Taking the trash out
- Changing bed sheets
- Vacuuming
- Unloading groceries
- Mopping
- Carrying heavy items

A word of caution: not all patients need assistance, especially early in this disease progression. Taking on too much of the patient’s responsibilities can make them feel useless, handicapped, or disabled. It can also affect their sense of control, which affects morale.

“



Hubby has RA-ILD and has been using supplemental oxygen for several years. We went to Italy for a month with a POC, and the following year, we went on a river cruise in France. Then, we went on a coach tour of the entire Hudson River Valley. We even went from the southwestern United States to an island off the coast of Massachusetts for a week in the summer heat and humidity. The planning for those trips seemed endless, but after the first trip to Europe, we had a little less anxiety. We are so happy we made the effort!

— Shelly M.





Become an Expert in Oxygen

Spend the time **now** to understand the details related to supplemental oxygen therapy. You will be better prepared when it is time for your loved one to require more oxygen. If you are lucky enough to have someone show you how to set up and use the oxygen equipment, ask them if you may record their explanation so you can refer to it later, if needed. All of this work will help you with troubleshooting, and it may reduce your stress in case of emergency. See Appendix B for vetted resources.

Prepare for Emergencies When Using Oxygen

Watch the PF Warriors meeting video on Disaster Preparedness. Be sure you have extra cannulas, tubing, batteries, and sufficient oxygen to withstand a power outage, shipping delays, or natural disaster.

Revisit the Discussion About Quality of Life Often

How involved do they want you to be in their care? What do they want your inner circle to know? What has changed? What needs to change to better reflect your needs and their wishes?

Communicate With Friends and Family

Recognize when you pass milestones in the disease journey. Let your inner circle know about the changes and how they can be helpful.

Give Back

Get involved in awareness campaigns, fundraisers, legislative activities. Promote organ donation.

How You Can Help Yourself

Help Yourself First so You Can be Helpful to Others

Information is Power

Participate in virtual meetings about pulmonary fibrosis. PF Warriors hosts experts every month who teach us about various aspects of living with this disease. The meeting topics vary from news about new treatments to nutrition, functional medicine, and emerging research. Videos of past meetings may be found on the PF Warriors website. Also, see Appendix A for a list of additional resources.

Stay One Step Ahead

Caregivers recommend that you become an expert in the next phase of the disease. For progressive disease, it is important to be “over-prepared” because it is often difficult to anticipate changes. That goes for your inner circle too: prepare them for the next milestone as well.

Get Organized

You have accumulated a wealth of information about this disease, insurance, medical bills, and lists of recommended doctors. Because the odds of exacerbations increase as the disease progresses, getting control of your files, books, websites, and other sources of information will help you in a pinch.

Add Family Members to the HIPAA Forms so They Have Access to the Doctors

This may help you delegate future tasks. Consider adding your adult children and/or sisters- and brothers-in-law. This should be a joint decision because it is the patient’s privacy that is potentially being shared.



I am on 3 lpm O₂ with exertion with a wheeled POC. It’s a heavy unit at 22 lbs. Mid-2020, we did a 24,000-mile road trip in a small two-door car across 27 states that took six months. We had the opportunity to witness the beauty of 24 national parks.

Last fall we flew (with the heavy POC) to Boston, to do another road trip around New England to see the beautiful fall foliage across seven states over three weeks.

We have plans for another road trip up the west coast to Seattle from San Diego and back down slightly inland to pick up Walla Walla, Wash., and then down through Oregon to our last stop in Death Valley National Park. We have to manage our routes on these trips and plan for full-time O₂ when traveling at higher elevation, but we never let our disease stop us from traveling.

Make the memories while you can. See your loved ones and hug them tightly.

Peace, love, and friendship!

— Keith B.

Convert Your Exercise Routine to be More Flexible

Instead of setting yourself up for frustration because you missed your yoga class, join a gym, buy an exercise bike or treadmill, or start power walking when you find free time. Try to squeeze in the same amount of exercise but let go of when and where you do it.

Pay Attention to Your Mental Health

Living with someone who has a serious, chronic disease is not easy. Your medical team can recommend a therapist who can help you process the sense of loss you may be facing, along with the myriad of other issues you face at this point in your life. Participate in a support group, where you will meet others who are walking the same path.



Be Good to Yourself: Manage Your Morale

As the disease progresses, your loved one will become more dependent on you, possibly leaving less time to focus on your own well-being. Find ways to get out and do something for yourself, alone or with friends, where you can let go for a bit. Also, remember the two basic contributors to morale: feeling valued and feeling in control. When you are down, doing something to improve one or both contributors will do wonders for your outlook.

Ask for Help

Friends and neighbors will want to help. Let them! Many families living with PF ask for help with chores that aren't easy to do anymore, such as mowing the lawn, clipping the hedges, changing light bulbs in the ceiling, changing furnace filters, or fence repair. You might ask someone to bring your morning paper up to the front porch when they are out walking their dog. Asking for help is hard, especially at first. Try weaving just one or two people onto your team so you have less anxiety about the process. Over time, either you, or they, can recruit additional help if needed.



Appendices

A: Vetted Resources for Living With PF

These are materials that a team of PF Warrior caregivers found especially informative. Also, visit [PFWarriors.com](https://www.pfwarriors.com) to access links to a variety of additional resources for friends and family on the members-only part of the website.

Easy-to-read book about how our lungs work, major lung diseases, and risks our lungs face every day	<i>Breathing Lessons: A Doctor's Guide to Lung Health</i> , MeiLan K. Han, MD, November 2021, W. W. Norton & Company
Detailed information about interstitial lung disease	Johns Hopkins https://www.hopkinsmedicine.org/health/conditions-and-diseases/interstitial-lung-disease-pulmonary-fibrosis?
Detailed information about idiopathic pulmonary fibrosis	Pulmonary Fibrosis Foundation https://www.pulmonaryfibrosis.org/understanding-pff/types-of-pulmonary-fibrosis/idiopathic-pulmonary-fibrosis
Slide show about IPF acute exacerbations	Insights in IPF https://www.insightsinipf.com/about-ipf/acute-exacerbations/
Pulmonary Fibrosis Foundation Care Center Network medical centers (Centers of Excellence)	Pulmonary Fibrosis Foundation https://www.pulmonaryfibrosis.org/docs/default-source/medical/care-center-network/full-list-of-ccn-sites.-.pdf?Status=Master&sfvrsn=b09662df_6
Excellent nutrition and exercise information for those living with pulmonary fibrosis	<i>Guide to Pulmonary Fibrosis & Interstitial Lung Diseases: FOR Patients, Caregivers & Clinicians</i> BY Patients, Caregivers, & Clinicians by Noah Greenspan PT DPT CCS EMT-B and Robert Kaner MD Mar 14, 2022, BookBaby; 1st edition (January 17, 2022)

<p>Online pulmonary rehabilitation bootcamp</p>	<p>Dr. Noah Greenspan’s Ultimate Pulmonary Wellness https://pulmonarywellness.org/bootcamp/</p>
<p>Intimacy and chronic lung disease</p>	<p>National Jewish Health https://www.nationaljewish.org/NJH/media/pdf/pdf-Understanding-BeingClose.pdf</p>
<p>Getting paid for caregiving</p>	<p>American Association of Retired Persons (AARP) https://www.aarp.org/caregiving/financial-legal/info-2017/you-can-get-paid-as-a-family-caregiver.html?intcmp=AE-CAR-CRC-LL</p>
<p>Family Medical Leave Act and paid family leave</p>	<p>U.S. Department of Labor https://www.dol.gov/agencies/whd/fmla AARP https://www.aarp.org/caregiving/financial-legal/info-2019/paid-family-leave-laws.html?intcmp=AE-CAR-CRC-LL</p>
<p>Legal documents for advanced planning (e.g., living wills)</p>	<p>AARP: scroll down to the Caregiver Resource Center menu https://www.aarp.org/caregiving/financial-legal/?intcmp=AE-CAR-SUBNAV-LEG</p>
<p>Clinical trials</p>	<p>National Institutes of Health https://clinicaltrials.gov</p>



To follow the latest medical and research news, join Twitter and start following community key opinion leaders.

Look for Twitter recommendations about additional people to follow, particularly health care professionals in this field.

- **@ActionPFCharity** – UK patient organization committed to serving people living with PF
- **@BrwdBreaths** – PF and organ donation advocate
- **@DollyPFstrategy** – Dolly Kervitsky, PF Strategies
- **@drdayjee** – Scientific Director for the ILD at the University of Chicago
- **@EU_IPFF** – The EU-IPFF is a European federation of national PF patient organizations
- **@ild_inn ILD_IN** – a group of ILD health care professionals aiming to support interdisciplinary teams and those in their care
- **@ILFA_Ireland** – Irish Lung Fibrosis Association
- **@LungFoundation** – Lung Foundation of Australia
- **@LungPT** – Dr. Noah Greenspan, Pulmonary Wellness Foundation
- **@lungtaskforce** – Taskforce for Lung Health in England
- **@meilan_han** – Chief of Pulmonary and Critical Care at the University of Michigan
- **@NIH** – NHLBI National Institutes of Health institute on heart, lung, blood and sleeping disorders research
- **@PatientMPower** – Works with PF nurses and pulmonologists, enabling them to monitor PF patients’ spirometry, oxygen and quality of life at home
- **@PFFORG** – Pulmonary Fibrosis Foundation
- **@PFTpeter** – Peter Bryce, Chairman of the Pulmonary Fibrosis Trust
- **@PulmWellness** – Pulmonary Wellness Foundation
- **@resmedjournal** – Journal devoted to rapid publication of respiratory research
- **@ResponsumforPF** – Free patient platform designed to help patients take control of their health
- **@SclerodermaCO** – National Scleroderma Foundation
- **@SjogrensOrg** – Sjogren’s Foundation
- **@TeresaLungLover** – Teresa Barnes, respiratory research advocate
- **@thetelomereproj** – The Telomere Project
- **@ThreeLakes_TLF** – Three Lakes Foundation: Serves as a catalyst for uniting research, industries, and philanthropy in PF to improve time to diagnosis and accelerate new therapies
- **@Wescoeforpf** – Wescoe Foundation for Pulmonary Fibrosis

B: Vetted Oxygen Resources

<p>Oxygen safety</p>	<p>Oxygen should never be used near an open flame or anything that can produce intense heat, flames or sparks, such as a burning cigarette, a lighted match, heaters, heating pads, hair dryers, a stove or a pilot light. Anything that can produce hot flames or sparks during operation should be kept at least 5 feet away from your oxygen equipment. (credit to UCSF)</p> <p>From the World Health Organization’s oxygen safety site:</p> <p>https://cdn.who.int/media/docs/default-source/oxygen-scale-up-initiative/cylinder_safety_print.pdf?sfvrsn=aa1595fa_5&download=true</p>
<p>Living and traveling with supplemental oxygen</p>	<p>American Lung Association</p> <p>https://www.lung.org/lung-health-diseases/lung-procedures-and-tests/oxygen-therapy/traveling-with-oxygen</p>
<p>Supplemental oxygen quick start guide</p>	<p>Pulmonary Fibrosis Foundation</p> <p>https://www.pulmonaryfibrosis.org/docs/default-source/programs/educational-materials/quick-start-guide-for-supplemental-oxygen/quick-start-guide-for-supplemental-oxygen-english.pdf?sfvrsn=b9694a62_13</p>
<p>The science behind maintaining good oxygen saturation</p>	<p><i>Guide to Pulmonary Fibrosis & Interstitial Lung Diseases: FOR Patients, Caregivers & Clinicians BY Patients, Caregivers, & Clinicians</i> by Noah Greenspan PT DPT CCS EMT-B and Robert Kaner MD Mar 14, 2022, BookBaby; 1st edition (January 17, 2022)</p>
<p>Detailed descriptions of features and functions of all portable oxygen equipment</p>	<p>The Pulmonary Paper</p> <p>https://www.pulmonarypaper.org/portable-oxygen-concentrators-comparison-chart-2021/</p>

C: Checklist of Things to Do Soon After a Diagnosis of Pulmonary Fibrosis

- Create a health history document and make copies. Upload information into an automated tool.
- Make a list of medications, dosages, instructions and prescribing doctor, including over the counter and herbal medicines.
- Get a second opinion from a Center of Excellence.
- Clean the house and office space to be more lung friendly.
- Organize medical insurance paperwork: Keep plan coverage summary, deductibles, out-of-pocket maximums, deductible rules, contact info, Health Savings Account (HSA) information.
- Plan to share the news with your family and close friends.
- Create a system to manage contact info, medical bills, health insurance claims, list of medical expenses, receipts, explanation of benefits statements, and correspondence about denials of coverage and appeals, Family Medical Leave Act (FMLA), social security, notes from doctor appointments.
- Learn about the disease.
- Share a healthy lifestyle including good nutrition, adequate sleep, and reasonable exercise.
- Ask about potential treatment options, such as pulmonary rehabilitation.
- Inform any other healthcare providers of the diagnosis, such as your primary physician, allergist, and other specialists.
- Join a support group.

Items to Consider for Purchase

Some people like to have extras on hand, especially when the patient goes to work or visits someone often:

- Pulse oximeter
- Oxygen flow meter
- Thermometer, plus a different type to use as backup in case you question the results of the first one
- Spirometer
- Humidifier
- Nebulizer, tubing, and mask if the doctor has prescribed the medication
- Air purifiers
- N95 or KN95 masks, disposable vinyl gloves
- Hand sanitizer and disinfecting wipes
- Ayr[®] gel and saline nose spray

D: Recommendations for Managing Health Information

Health history may include:

- Name, age, marital status
- Contacts in case of emergency
- Significant diagnoses, dates
- Surgeries and hospitalizations: reasons, dates
- List of hobbies, description of past and present occupations, past and present hometowns, and anything else that may provide a clue regarding lung health
- Immunizations (helpful for international travel)
- Most recent lab tests and results
- All specialists' names and contact information
 - Pulmonologist, ILD center staff, pulmonary rehab, rheumatologist, sleep specialist, cardiologist, gastroenterologist, endocrinologist, psychiatrist, osteoporosis specialist, allergist, etc.
- Pharmacy name, phone, fax
- List of current medications, dosing, and purpose
- List of current supplements, dosing
- Allergies: drug, environmental, and food
- Significant family health history and how related, cause and age of death
- Behavioral health status: smoking, drinking, recreational drugs
- Women's health history
- Long-term planning: is there a Living Will? DNR?



Consider putting together a three-ring binder where you can organize the following documents:

- Multiple copies of the health history, including current medications and supplements, to hand to medical staff.
- Ongoing list of questions for the doctors and important notes from appointments
- Health insurance information
- Patient portals login data
- History of laboratory and imaging tests, dates, locations
- Business cards from all doctors and pharmacies: buy a clear plastic baseball card organizer page to store the cards in the binder.
- Legal paperwork: Living Will, Advanced Directives, Do Not Resuscitate Order (DNR), Health Care Power of Attorney, MOLST, POLST.

E: Suggestions for Managing Lung Irritants in Your Environment

The goal of each of these suggestions is to minimize fumes or airborne particles that may be problematic, such as smoke, animal dander, or sawdust.

- If you or your loved one smokes, you should stop, and you should both avoid second-hand smoke.
- Get your home's air quality tested by a Home Environmental Testing professional.
- Open the windows when practical, as long as the outdoor air quality is good. You can check this at any time in your area at www.airnow.gov. Let the fresh air in and the stale air out.
- Repair or remove water leaks, standing water, and damp areas, all of which can develop mold.
- Install a whole-house dehumidifier
- Get air ducts cleaned.
- Employ air purifiers. Change filters on schedule.
- Use distilled water in the portable humidifier.
- Exchange carpet and rugs for hard surface flooring or deep clean carpets regularly.
- Replace or remove down-filled pillows, comforters.
- Evaluate your window treatments for the best way to manage dust.
- Keep the blades of your ceiling fan free from dust.
- Make sure all cleaning products used in your home meet the new EPA Safer Choice standards. Buy natural cleansers to protect from caustic fumes. Suggestions may be found at the Environmental Protection Agency site: Safer Choice | US EPA – <https://www.epa.gov/saferchoice>.
- Keep the oven clean. Cooking fumes can be irritating. Consider running the stove exhaust while cooking with either the stove or the oven.
- Consider upgrading your kitchen ventilation system and keep it clean. Also, open the windows when cooking if possible.
- Consider using baking soda and vinegar to clean the oven rather than using the self-cleaning option. Self-cleaning ovens often emit harsh fumes for many hours.
- If you want baked goods, consider buying them instead of making them. The dust from the flour may be troubling to your loved one. Or, bake when he or she is not in the kitchen with you.
- Learn to use an air fryer to prepare foods. Generally, they give off fewer fumes than conventional frying.
- Stop using heavy fragrances, including scented candles, potpourri, and air fresheners. Also, petroleum-based candle wax can release chemicals into the air. Use soy-based products instead.



- Avoid using your wood fireplace. If you use a wood-burning stove to heat your home, make sure it meets the most recent EPA standards to keep emissions low.
- Suggest taking a bath instead of sitting in the hot tub. Bacteria that can develop in the hot tub may affect the lungs when breathing in the vapor.
- Rehome your pet bird. Bird droppings and feathers are serious lung irritants for some people.
- Avoid dust and fine particles that result from wood working and other construction work, especially countertops (silica); including asbestos, plastics, resin, wall and model paints, roofing materials, vinyl tile work.
- Remove mold and mildew. Stay on top of cleaning the shower stall with gentle, natural cleansers. The hot water can “kick up” irritants in the stall.
- Find someone, other than your loved one, to rake the leaves.
- Find someone, other than your loved one, to clean the kitty litter.
- Follow weather predictions about air quality, humidity, flu risk, and allergy risk for your area. See www.airnow.gov.
- Protect against the dust that blows around after street sand and salt dries on the pavement during the winter months.
- Change air filters in cars regularly.
- Set up an area outside of your bedroom for your dogs to sleep at night. Dog fur, dander, and bacteria brought indoors can be lung irritants.

Avoid the Following Additional Irritants

Household

- Radon (have your home checked if this has never been done)
- Gas appliances (run the exhaust hood when using)
- Hair spray
- Diesel gas fumes
- Strong cologne and perfume
- Coffee bean dust from grinding
- BBQ grilling smoke
- Wood-burning fires
- Bug spray and pesticides used with gardening
- Fireworks

Occupations and Hobbies

- Farming
 - Tobacco plants
 - Moldy or green hay, or silage
 - Grain
 - Barley
- Sugar-cane refining
- Wine processing or vineyards
- Cheese-making
- Grinding granite kitchen countertops
- Carpentry and woodworking

An extensive list of irritants that may contribute to hypersensitivity pneumonitis may be found at www.hplung.com.



F: Minimizing Infections

- Whenever your loved one may be exposed to germs, viruses, or bacteria, they should wear a mask. Buy several packages of the most protective mask that they'll wear regularly, fits well, and is comfortable. You should also wear a well-fitting mask when you are out and about; you don't want to bring diseases home to share. Finally, ask visitors to wear masks to protect both of you.
- When wearing a mask, don't touch the outside and then touch your face, etc. Store masks properly between uses. The CDC recommends storing masks in a clean paper bag between uses. Replace masks when they become soiled or wet.
- Ask the doctor about pneumonia, influenza, COVID-19, Tdap, and shingles vaccines.
- Wash hands several times each day for at least 20 seconds.
- Use hand sanitizer.
- Protect hands from minor cuts or cracks due to dry skin.
- Minimize shaking hands and hugging.
- Reinforce a good household rule: throw away tissue as soon as it is used.
- Routinely clean surfaces in home that are touched often: doorknobs, countertops, knobs, and handles.
- Avoid large crowds, especially during flu season or when viruses are circulating.
- Leave your shoes near the entryway to avoid bringing dust in from outdoors.
- Suggest that someone else do the gardening or offer a protective mask and gloves.
- Suggest that someone else clean the cat litter, walk the dog (poop scoop).
- Use distilled water for medical and household devices such as a neti pot, humidifier, CPAP, and other similar equipment.
- Practice good dental hygiene.
- Minimize exposure to particles and odors associated with manicure and pedicure, which can be irritants.
- Maintain foot care to avoid blisters and cuts.
- Clean and replace the oxygen cannula regularly to avoid germs and irritation.
- Wash fresh fruits and vegetables well to avoid contaminants. Be on the lookout for recalls due to listeria or salmonella and other food-borne diseases.

G: Suggestions for Making the Home More Hospitable

- Gas stoves are a hazard for oxygen users. Consider swapping yours out for an electric stove or keeping oxygen canisters at a safe distance. See the oxygen safety tips in Appendix B for more information.
- Buy laundry baskets on wheels.
- Replace clothes that require raising arms overhead when dressing and undressing.
- Switch dresser drawers so your loved one doesn't have to bend to reach lower drawers
- Replace lace-up shoes with loafers, mules, clogs, or replace laces with elastic lace closures.
- Buy a long shoehorn to help with getting into shoes.
- Get a raised or high-seat toilet to minimize the exertion needed when sitting and standing.
- Buy a cotton terry cloth robe that may be used instead of a towel when exiting the shower.
- Open a window in the bathroom or install an exhaust fan above the shower to remove humidity when showering.
- Install a grab bar in the shower or near the bathtub.
- Get a shower chair (or transfer bench) with a back on it.
- Buy a long-handled scrub brush for the shower.
- Set a chair or walker with a seat outside the shower stall to sit on after showering.
- Install a hand-held shower wand.
- Install a bidet or bidet system in your toilet.



H: Managing Cough

Consider Potential Causes

- Check oxygenation level
- Post-nasal drip
- Air too dry/low humidity
- GERD
- Bronchitis
- Allergies
- Side effects of medications
- Heart disease
- Eating too much or too fast

Try Home Remedies

- Reduce acidic foods, fried foods.
- Improve eating habits: volume, quality, speed, frequency.
- Avoid very cold foods, like ice cream.
- Drink pineapple juice. (The bromelain in pineapple can be helpful and may reduce inflammation. But be careful if your loved one has GERD due to the acidic nature of pineapple.)
- Drink fluids often to keep the throat moist.
- Set up a fan to blow at your loved one's face. The cool breeze can really help.
- Drink hot tea with honey.
- Suck on peppermint candy.
- Add ginger to foods.
- Suck on Fisherman's Friend cough drops.
- Sip apple juice after eating.



Ask the Doctor About...

- Tools to help with productive cough, such as the Aerobika®
- Huff coughing
- Mucinex®
- Delsym®
- Saline nebulizer to loosen up phlegm
- Sinus wash of sodium chloride with Mupirocin (ointment melts in the liquid).
- Nebulizer with budesonide and Performist®
- Tessalon Perles
- Phenergan with codeine
- Gabapentin
- Thalidomide
- Opioids
- Steroids

I: Managing GERD

This is a list of general guidelines for anyone who is affected by gastroesophageal reflux disease (GERD).

Good Eating Habits

- Eat several small meals rather than three big meals. Smaller meals will also help them avoid getting too full, which can make it harder to breathe.
- Eat slowly to minimize swallowing extra air that may add pressure in the stomach and abdomen.
- Sit up with good posture to help the food go down.
- Minimize carbonated drinks because the resulting gas can push up on the diaphragm and put pressure on the lungs.

Good Sleeping Habits

- Keep the head of the bed elevated. Prop up the head of the bed or use a wedge pillow to elevate the head and upper airway while sleeping.
- Eat the last meal 3 hours before bedtime.

Helpful Additions to the Diet

- Apple cider vinegar
- Ginger

GERD-friendly Foods

Eat high-fiber foods such as whole grains, root vegetables, and green vegetables. Also:

Alkaline Foods

- Bananas
- Melons
- Cauliflower
- Fennel
- Nuts

Watery Foods

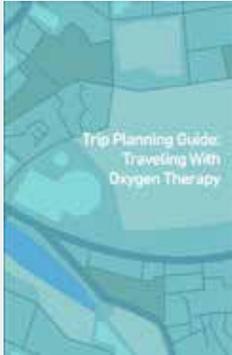
- Celery
- Cucumber
- Lettuce
- Watermelon
- Broth-based soup
- Herbal tea

Heartburn Triggers

- Alcohol, particularly red wine
- Black pepper, garlic, raw onions, and other spicy foods
- Chocolate
- Citrus fruits
- Coffee and caffeinated drinks
- Peppermint
- Tomatoes



J: Traveling with Pulmonary Fibrosis



Recommended Guide
CHEST Trip Planning Guide:
https://foundation.chestnet.org/wp-content/uploads/2021/10/Travel_Guide_single.pdf



Additional General Tips

- For families living with progressive disease, plan distant travel first. Travel options become limited when your loved one needs high-flow oxygen.
- Higher altitudes can affect shortness of breath and hypoxemia. Speak with the doctor about your travel plans to ensure your loved one will be able to enjoy the location. Ask the doctor about the High-Altitude Simulation Test (HAST).
- It is never too soon to begin planning your trip. This is especially important if an updated oxygen prescription is needed and/or a HAST test needs to be scheduled.
- When arranging for tanks at your destination, be sure to ask for all supplies needed, e.g., O₂ regulator, extra washers, cannulas, etc.
- Consider options for obtaining or transporting a floor concentrator (stationary oxygen concentrator) for your trip.
- See Appendix E to remind yourselves about things to look for regarding lung triggers when planning your trip.
- Bring the following with you in your carry-on when you travel: medicines and supplements, health history binder (see Appendix D), and contact information for the local oxygen supplier and emergency contacts.
- Find the Center of Excellence that is closest to the place you will be visiting, in case of emergency. See Appendix A for a link to the list of centers.
- Carry the after hours' phone number for the oxygen supplier with you at all times.
- 14 U.S. states require annual or semi-annual renewal of the oxygen prescription. Plan ahead so the necessary paperwork is up to date when you travel.
- Practice changing POC batteries prior to travel. If you are borrowing or renting a POC from a supplier, ask your loved one to get used to it before leaving for the trip. This will help to make sure you both know how it operates and that the batteries are good.
- Consider buying a flow meter to help your loved one alter their oxygen levels more easily throughout the trip.



Tips for Traveling by Plane with Oxygen

- It may take several weeks to secure oxygen supplies on the other end. Consider calling the supplier a second (third?) time a week or so prior to departure to ensure your requests haven't been lost or misunderstood.
- Some airlines require that you complete a special form in order to travel with oxygen equipment. See your carrier's website for more information.
- Bring these items with you on board when you travel by plane: oxygen prescription, letter from doctor indicating your loved one is fit for travel, medicines and supplements, medical history binder (see Appendix D), and key contacts at your destination. Also, bring the oxygen equipment and batteries that will support the POC for at least 1.5 times the length of the flight. Finally, don't forget the travel wipes.
- Ask the porter at the airport to push your loved one in a wheelchair once inside the airport. Request a wheelchair to be waiting for you at the gate at your destination. Conserve their energy and POC battery.
- Check in at the ticket counter, then again at the gate.
- Medical equipment doesn't count as your carry on.
- When boarding: give crew paperwork and request/use early boarding.
- Airplanes are pressurized to 8,000 feet above sea level. Some people breathe faster while on board. It may be necessary to bring along more than the required number of batteries, to allow for contingencies.
- The air on commercial jets gets recirculated from the area under the window and near the floor. To breathe fresher air, your loved one should try to place the POC away from that location.
- In an emergency, when the plane's oxygen masks drop from above, your loved one should keep the POC cannula in place, and place the airplane oxygen mask over it.

Lodging Considerations

- See the tips in Appendices E, F, and G when researching options.
- Request a hotel room lower than the 7th floor because that is the highest that the rescue team can reach quickly in case of emergency.
- Request a hotel room close to the elevator so your loved one doesn't need to expend energy or oxygen tank capacity and battery life needlessly.
- Ask the hotel to "de-feather" the hotel room. This is common practice, and you shouldn't be concerned about this special request.

Traveling by Cruise Line

Sea Puffer Cruises are designed to support those traveling with supplemental oxygen. Respiratory therapists are on all ships.

Call: (866) 673-3019, or

Visit: <https://www.getupandgo2.com/pages/cruise-with-oxygen-29629>

For all cruises: contact the cruise line to learn about rules regarding using oxygen on board.



K: Important Documents Checklist

- HIPAA forms
- Forms for paramedics (some states require out-of-hospital DNR forms if your loved one chooses to decline CPR)
- Living Will or Advanced Directive
- Revocable Living Trust
- Durable Power of Attorney
- Ethical Will

L: Support for Living With Pulmonary Fibrosis

This is not a precise description of types of medical practices. Also, the adjoining chart is meant to serve as a guide for seeking additional services. Check with the professionals in your region for specific offerings.

Traditional care includes primary and specialty care professionals such as a primary care physician (PCP), gastroenterologist, pulmonologist, psychiatrist, psychologist, ear/nose/throat doctor, etc.

Precision medicine doctors evaluate a person's genetic makeup and make recommendations that are unique to each patient's very specific needs based on predictive response or risk of disease. Therapies include addressing nutritional or enzyme deficiencies, recognizing differences in ability to respond to pain relievers, and other targets.

Alternative medicine includes internists, doctors of osteopathy (DO), chiropractors, and nurse practitioners and their teams who perform acupuncture, meditation, therapeutic massage, and other forms of stress management with the intent to improve response to injury or disease.

Pulmonary rehabilitation is available by prescription from the medical team. It is also available online through various programs. Led by a respiratory therapist specifically trained in lung diseases, it is much more than just targeted exercises. It includes disease education and support for managing symptoms. Some programs are offered in coordination with support groups.

Palliative care is a multidisciplinary approach designed to minimize suffering and improve quality of life for patients with serious illnesses. Palliative care is not "hospice-lite," although some medical professionals continue to confuse the two. Palliative care can begin at diagnosis. Learn about how Medicare covers palliative care and hospice care prior to researching providers.

Support groups are organized groups of people sharing something in common where all can benefit from peer-to-peer support. Most support groups are online, but some smaller groups hold face-to-face meetings. PF Warriors is the largest international PF support group and benefits from the daily, vibrant interaction of people from around the world.

Hospice is formally defined as care for people who are recognized as having less than six months to live. It is symptom management, including psychological support, that is offered once a cure is no longer being pursued.

	Traditional Care	Precision Medicine	Alternative Medicine	Pulmonary Rehab	Palliative Care	Support Groups	Hospice
Diagnose and treat pulmonary fibrosis and comorbidities medically	✓						
Identify and recommend treatments for nutritional deficiencies and genetic tendencies		✓					
Provide disease education about pulmonary fibrosis, general lung health, including general suggestions about dietary health	✓			✓		✓	
Teach and practice beneficial exercises and breathing techniques to promote lung health				✓			
Provide suggestions for ways to manage symptoms	✓	✓	✓	✓	✓	✓	✓
Provide medicine to manage symptoms	✓				✓		✓
Provide massage, acupuncture, and other therapies meant to reduce stress and improve immune response		✓	✓		✓		
Prescribe anti-anxiety, depression and other mental health medications	✓				✓		✓
Diagnose and treat sleep disorders and GERD	✓						
Provide psychological counseling and support in an individualized and private manner	✓				✓		✓
Provide psychosocial support, mostly in a public forum				✓		✓	
Provide help in navigating the medical system						✓	



Please consider a donation to the PF Warriors which will help ensure patients 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.

The PF Warriors Fund is a component fund of The Dallas Foundation under a fiscal sponsorship agreement. You can donate either online or via mail:

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

Mail to: The Dallas Foundation
PF Warriors Account
Dept# 42321
PO Box 650823
Dallas, TX 75265-0823

To donate stock, other assets, or from your estate plan, contact the Dallas Foundation directly at 214-741-9898 and ask them for guidance for making a directed contribution to the PF Warriors.

Your donation demonstrates vision, commitment and caring.