



# Patient Information Guide

Pulmonary Fibrosis

FOUNDATION

## Contents

- 2** About This Guide
- 4** About IPF
- 7** Prevalence
- 8** Symptoms
- 9** Diagnosing IPF
- 12** Treatments
- 17** Quality of Life
- 20** About the PFF

“Pulmonary  
fibrosis will  
not define  
who I am.”

**This educational guide is provided by the Pulmonary Fibrosis Foundation (PFF) as a public service to our patient community.**

To offset our publication costs and to ensure that all patients and families continue to receive this guide for free, please consider making a donation to the PFF by visiting [www.pulmonaryfibrosis.org](http://www.pulmonaryfibrosis.org) or mailing your donation to:

**Pulmonary Fibrosis Foundation  
811 West Evergreen Avenue, Suite 204  
Chicago, Illinois 60642**

# About this Guide

**The patients and family members who call** the Pulmonary Fibrosis Foundation with news that they or a family member have just been diagnosed with pulmonary fibrosis (PF), or idiopathic pulmonary fibrosis (IPF), often feel frightened, confused, and concerned. Most patients find themselves frustrated by the lack of available information. Physicians often don't have the necessary time or resources to explain to their patients the details of the disease, or help patients deal with the trauma of being told that they have an illness for which there is no cure.

The Pulmonary Fibrosis Foundation is deeply aware of these concerns and strives to provide patients, family members, and health care providers with the resources necessary to more fully understand PF and IPF, and to provide patients with the tools necessary to live with their disease and improve their quality of life.

It is important to note that there is no consistent standard of care for IPF in the medical community, and disease progression varies greatly in patients — your physician may have discussed this challenge with you. Because of these issues, it is critically important for patients to understand their condition and ask their physicians important questions to ensure they are being treated appropriately based on their individual symptoms. This brochure is intended to help patients achieve this goal.

It is important to understand that the terminology concerning these diseases is often confusing to patients. IPF is a specific disease within a classification of diseases referred to as interstitial lung diseases (ILD). IPF implies that there is no obvious or discernible cause of the PF, thus it is called “idiopathic.” If there is a clear association with another disease such as scleroderma or rheumatoid arthritis, or a side effect resulting from a medication an individual

“Living with PF has been a good thing, one of the best things that has ever happened to me! It has made me live life to the fullest!”

– **LESLIE HOWARD**

NEW ROCHELLE, NEW YORK

may have been prescribed, then the cause of the disease is no longer considered idiopathic.

PF clearly associated with another disease (such as scleroderma, etc.) would be referred to as pulmonary fibrosis secondary to scleroderma, or secondary to rheumatoid arthritis. As we learn more about the genetic and peripheral biomarkers of IPF, new, more precise terminology may develop.

Please remember that this information is a brief overview of IPF and the Pulmonary Fibrosis Foundation, and is for educational purposes only. It is not intended to be a substitute for professional medical advice. Always consult your personal physician or health care provider with any questions you may have regarding your specific medical condition.

Also please know that we are here to help you. You may contact the Pulmonary Fibrosis Foundation with any questions or concerns you have about PF or IPF during the course of your care. Our staff can be reached at 888.733.6741 or by email at [info@pulmonaryfibrosis.org](mailto:info@pulmonaryfibrosis.org).

# About IPF

## **What is Idiopathic Pulmonary Fibrosis, or IPF?**

According to the National Institutes of Health (NIH), PF is a condition in which over a period of time the lung tissue becomes thickened, stiff, and scarred.<sup>1</sup> The development of the scar tissue is called fibrosis. As the lung tissue becomes scarred and thicker, the lungs lose their ability to transfer oxygen into the bloodstream. As a result, the brain and other organs don't get the oxygen they need. In some cases, doctors can determine the cause of the fibrosis, but in most cases, there is not a known cause. When there is no known etiology (cause) for the development of lung fibrosis (and certain radiographic and/or pathologic criteria are met) the disease is called idiopathic pulmonary fibrosis or IPF. IPF affects approximately 200,000 Americans and an estimated 40,000 Americans pass away from IPF each year.<sup>1</sup>

## **How is IPF Related to Interstitial Lung Disease?**

There are more than 200 related diseases of the lung known as interstitial lung diseases (ILD). ILDs can also be referred to as diffuse parenchymal lung diseases (DPLD). These diseases can often have similar characteristics to IPF and most result in lung scarring. Further, IPF belongs to a subgroup of ILDs called idiopathic interstitial pneumonias (IIP). IIP is further broken down into a number of pathological subtypes. The pathological pattern most commonly seen in IPF is referred to as usual interstitial pneumonia (UIP). There are a number of other subtypes of IIP. Two of the more common subtypes are nonspecific interstitial pneumonia (NSIP) and acute interstitial pneumonia (AIP).

Although this “alphabet” soup is confusing and complicated, it is important for doctors to differentiate the cause and pattern of fibrotic disease, since standards of care and prognosis can vary. IPF is the most common of the all ILDs and IIPs. One recent study estimated the prevalence of all interstitial lung diseases in the United States at about 500,000.<sup>9</sup>

## What Causes IPF?

The origin (e.g. epidemiology) and development (e.g. pathogenesis) of IPF is still not completely understood. The current thinking is that there is an abnormal fibrotic and inflammatory response to microscopic injury that ultimately results in scarring of the lung. There are also epidemiological and genetic factors that may contribute to the development of IPF, and as these are more clearly defined, the disease process should be better understood. Ultimately, this will lead to new pathways to treat the disease.

**Epidemiological Factors:** There are certain environmental and occupational exposures that can be prevalent in the medical histories of patients diagnosed with fibrotic lung diseases, and as a result doctors may cite these exposures as contributing factors to a diagnosis of PF. (If there is a clear causal relationship, then the disease would no longer be considered IPF).

These exposures may include the following:

- Cigarette smoking
- Prolonged exposure to occupational or environmental contaminants or dusts (inorganic dusts such as asbestos, silica, beryllium, and hard metal dusts; organic dusts such as bacteria and animal proteins)
- Viral or bacterial lung infections

- Certain medicines such as antibiotics (Nitrofurantoin, Sulfasalazine), antiarrhythmics (Amiodarone, Propranolol), anticonvulsants (Phenytoin), chemotherapeutic agents (Methotrexate, Bleomycin, Oxaliplatin, Erbital), and therapeutic radiation
- Acid reflux disease (GERD)

In addition, PF may be associated with upper respiratory infections such as pneumonia and tuberculosis. The specific connection between PF and these diseases remains largely unknown.

Pulmonary fibrosis (PF) has also been associated with connective tissue diseases including rheumatoid arthritis, scleroderma, lupus, and sarcoidosis.

**Genetics and IPF:** There is a growing body of clinical evidence suggesting that genes or genetic variants may predispose certain patients to developing IPF. Approximately 10–15% of cases are considered to be familial and is highly suggestive of a genetic predisposition. Recent studies have found a mutation in the SP-C protein that exists in families with a history of more than two cases of IPF.<sup>6</sup> Another recent study suggested that the presence of specific genes may predict which IPF patients will have a more severe, rapidly progressing form of the disease.<sup>5</sup> Another recent study showed that shortened telomeres (which protect the fragile ends of chromosomes from deterioration) may be the cause of PF in certain patients as they grow older.<sup>5-5a</sup> Yet another recent study reported that a genetic variant in the MUC5B gene may increase risk of developing PF between 6-22 times depending on family history.<sup>12</sup> There is limited availability of genetic testing to identify genes that may contribute to IPF. It is important for patients to discuss the potential risks and benefits of genetic testing with a qualified genetic counselor and their health care provider.

# Prevalence

## How Many People Have IPF?

There is a lack of newly published data to demonstrate an accurate estimate for the incidence of pulmonary fibrosis in the United States. The most recent estimates indicate that approximately 200,000 Americans have IPF.<sup>7</sup> Varying terminology, lack of standardized diagnostic criteria, and lack of a national surveillance registry have complicated the accrual of accurate data.

More importantly, it is anticipated that the number of individuals diagnosed with IPF will continue to increase. This is thought to be a result of people living longer, and/or an improved clinical understanding of IPF, as an improved understanding of IPF will lead to earlier and more accurate diagnosis.

The lack of clinical understanding of IPF remains a concern in the medical community. Limited awareness of the epidemiology (causes) and pathogenesis (disease progression) has made misdiagnosis of IPF a common problem. In fact, a recent study showed that more than 50% of IPF patients may be initially misdiagnosed.<sup>3</sup> Further complicating the difficulty in diagnosis is the fact that there are more than 200 different types of interstitial lung diseases (ILD), and it has not been until recently that the American Thoracic Society (ATS) recognized IPF by its specific clinical and pathological characteristics.<sup>8</sup> At times, progress was slowed by an incorrect understanding of the pathophysiology, inability to perform adequate clinical trials, and a failure to communicate and collaborate within the research community.<sup>11</sup>

IPF has no strong demographic profile; it is found in equal proportions in urban and rural environments. A history of smoking and certain genetic factors has been associated with an increased risk of IPF, and a variety of published studies show that, on

average, two-thirds of those with IPF have a history of smoking.<sup>3</sup>

IPF affects more men than women and most commonly occurs between the fifth and seventh decades. The median age at time of diagnosis is approximately 63 years old according to a variety of published studies; however, IPF has been diagnosed from early adulthood into the late eighties.

# Symptoms

## What are the Symptoms of IPF?

Symptoms aren't always present when the disease starts and may not be present until the disease has progressed. The most common symptom is shortness of breath, also known as dyspnea. Many patients describe it as a feeling of "breathlessness." Many individuals, especially older patients, often ignore the occasional difficulty with breathing, attributing it to just getting older or being out of shape. As the condition progresses and the damage to the lungs becomes more severe, breathlessness may occur with minor physical activity such as showering and getting dressed. Speaking on the phone and eating may also cause breathlessness with advanced disease.

Other common symptoms include:

- Chronic dry, hacking cough
- Fatigue and weakness
- Discomfort in the chest
- Loss of appetite
- Rapid weight loss

# Diagnosing IPF

## How is IPF Diagnosed?

**History and Physical Exam:** The physician should take a detailed history to learn if there were any environmental, occupational, familial, or other medical conditions that could have contributed or predisposed a person to the disease's development. When listening to the lungs with a stethoscope, the physician may hear "crackles" or Velcro-like sounds. These are "opening" sounds made by the small airways during inspiration. About 50% of patients with IPF may have "clubbing" of the fingertips. Due to a lack of oxygen in the blood, clubbing is a thickening of the flesh under the fingernails, causing the nails to curve downward. Clubbing of the fingertips is not specific to IPF and occurs in other lung disorders, heart and liver disease, and can also be present from birth.

**Chest X-Ray:** A routine chest X-ray may be used as a screening test. However, 5–15% of patients with significant scarring will have a normal chest X-ray and IPF cannot be diagnosed from a chest X-ray alone.

**High Resolution Computerized Tomography (HRCT):** This test provides a detailed image of the lungs to help physicians more clearly identify certain radiographic patterns in the lung tissue that may indicate disease. In IPF a radiologist may identify a "honeycombing" pattern that suggests lung scarring and damage to the air sacs or "ground-glass opacity" which refers to the hazy appearance of lung tissue that is most associated with inflammation.

**Pulmonary Function Tests:** These are breathing tests that measure the total amount of air in the lungs and assess the flow of air in and out of the lungs. Additionally, they can also measure the lungs' ability to exchange oxygen and carbon dioxide properly. These tests are usually done in a hospital or clinical laboratory and consist of breathing into a spirometer; they are sometimes done in a "body box" which resembles a glass telephone booth.

There are two important components to a pulmonary function test: (1) spirometry, which measures inspired and expired lung volumes and the rate at which this occurs, and (2) diffusion capacity, or DLCO, which measures the ability of oxygen to diffuse into the blood stream.

**Pulse Oximeter:** This is a screening test that indicates the amount of oxygen saturation in the blood. An oximeter is placed on the finger or earlobe and transmits light at different wavelengths through small blood vessels. Normal ranges are 95–100% on room air. Pulse oximetry does not measure carbon dioxide levels so a blood gas level measurement may be necessary in some patients.

**Arterial Blood Gas (ABG):** This test is a direct measurement of arterial pH, oxygen, and carbon dioxide through a direct arterial puncture. Arterial blood has recently been oxygenated by the lungs and thus indicates how much oxygen is available to the body. Venous blood has a lower oxygen concentration and indicates how much oxygen has been extracted.

**Bronchoscopy:** This involves an examination of the main airways of the lungs through the use of a small, flexible tube called a bronchoscope. Bronchoscopy helps to evaluate lung problems or blockages and provides a means to sample tissue or fluids. Unfortunately, the lung tissue samples obtained through bronchoscopy are small and are usually inadequate for definitive diagnoses.

**Bronchoalveolar Lavage (BAL):** BAL is done through the bronchoscope and is a way to remove a tiny sampling of cells from the lower respiratory tract. A small amount of saline is injected through the bronchoscope and when withdrawn removes a sample of cells from the respiratory tract. Usually this is not helpful in making the diagnosis of IPF but may be beneficial in other clinical situations.

“Before I take my last breath I am going to make this a household word. I’m going to go out and tell everybody I possibly can talk to and say, ‘are you familiar with pulmonary fibrosis?’”

– BOB O’ROURKE

PASADENA, CALIFORNIA

**Surgical Lung Biopsy:** Surgical lung biopsy is the most revealing diagnostic tool in the evaluation of patients suspected of having idiopathic pulmonary fibrosis and is considered the “gold standard.” Since there are many diseases that mimic IPF, and there can be significant differences in the treatment and prognosis, it is important to get a correct diagnosis. A lung biopsy in conjunction with the HRCT can also help determine how far the disease has progressed. Usually the biopsy can be obtained minimally invasively with video assisted thoracoscopic surgery (VATS). VATS is usually well tolerated, but it may not be recommended for all individuals.

**Exercise Testing:** Exercise testing is used to measure how well the lungs function during exertion. The methods used for exercise testing vary from hospital to hospital, but usually include the use of a stationary bike or treadmill. The most common method of exercise testing is the six-minute walk test, where the distance a patient can walk in six minutes is measured. Blood pressure, electrocardiogram, and oxygen saturation levels (recorded by an electronic device placed on the ear or finger) are monitored during exercise testing.

# Treatments

## How is IPF Treated?

The clinical course of idiopathic pulmonary fibrosis (IPF) is highly variable and may be difficult to predict. As a result, strategies to treat IPF are highly individualized, based upon the specific patient's medical history and other conditions (comorbidities).

While there are currently no effective treatments, or a cure for IPF, there are a variety of therapeutic options to help patients manage their condition and maintain their quality of life and activities of daily living. Typical standards of care may include prescription therapies, supplemental oxygen, pulmonary rehabilitation, lung transplantation, and/or referral for clinical trial participation. Lung transplantation remains the most viable course of treatment to extend the lives of those with IPF; this option should be discussed with your physician as soon as you are diagnosed.

**Therapeutic Options:** For some patients depending on their diagnosis and biopsy, medications may stabilize their disease and there may be a benefit to continuing usage. While there remains no consistent standard of care in the IPF community, the following medications are commonly prescribed in an attempt to treat symptoms:

- **Corticosteroids (prednisone):** Prednisone is used for suppressing the immune system and inflammation. It mimics the action of cortisol which is produced by the adrenal glands. Depending on the dose, prolonged therapy can cause the adrenal glands to stop producing its own cortisol. For this reason when prednisone is discontinued, it may be necessary to gradually lower or taper the dose to allow time for the adrenal glands to recover. Since prednisone suppresses the immune system, it can potentially increase the frequency and severity of infections. Prednisone has

many side effects including sugar intolerance (can worsen or cause diabetes), weight gain, swelling, depression, anxiety, fatigue, and peptic ulcer to name just a few. Individuals receiving prolonged treatment or higher doses need to be carefully monitored.

- Cyclophosphamide (Cytoxan): Cytoxan is an anticancer drug frequently given in conjunction with prednisone or may be given alone. While it is usually taken daily by mouth, in some instances it may also be administered intravenously.
- Azathioprine (Imuran): Imuran is used to suppress the immune system and is commonly used to treat autoimmune diseases such as rheumatoid arthritis. It is also used to help prevent the body from rejecting organs following transplantation. Although there have been some successful reports in a small number of individuals, Imuran's effectiveness to treat IPF has not been confirmed in a randomized clinical trial to date.
- N-acetylcysteine (NAC): NAC is a naturally occurring antioxidant. It can be taken orally and theoretically could prevent some of the oxidative injury that precedes fibroproliferation. A small, non-randomized study demonstrated some improvement in lung function in patients with IPF. There are a number of ongoing studies investigating the efficacy of NAC in combination with other drugs to treat IPF.

**As with any medicine for any condition, patients should discuss specific treatment options directly with their physician to determine the best approach for their care.**

**Supplemental Oxygen Therapy:** All the body's functions depend upon delivery of a steady supply of oxygen. Because PF inhibits an adequate transfer of oxygen into the blood stream, some patients may require supplemental oxygen. This helps to reduce breathlessness, enabling the patient to be more active. Some patients may need oxygen therapy all the time while others may only need it during sleep and exercise. By testing the saturation level of oxygen in a patient's blood, a physician can determine if a patient requires supplemental oxygen.

If your doctor has prescribed supplemental oxygen, it is important to use it as prescribed. Many patients are fearful that they will become "addicted" to supplemental oxygen. Supplemental oxygen is not "addictive," the proper amount of oxygen in the bloodstream is necessary to maintain normal body functions. Low blood oxygen levels can lead to additional health problems.

**Pulmonary Rehabilitation:** Pulmonary rehabilitation has become the standard of care for people with chronic lung disease, and recent studies have demonstrated improvements in both exercise capacity and health-related quality of life in patients with IPF.<sup>10</sup> The goal of pulmonary rehabilitation is to restore the patient's ability to function without extreme breathlessness. These programs offer a variety of services and can be inpatient, outpatient, or home/community based. The programs are "multidisciplinary," meaning that the team includes nurses, respiratory therapists, physical therapists, social workers, dieticians, etc. The range of services includes: exercise training breathing exercises and retraining; anxiety, stress, and depression management; and nutritional counseling to name a few. Another recent study recommended that pulmonary rehabilitation be considered as a standard of care for those with ILDs like IPF because of its potential to improve functional status and dyspnea.<sup>11</sup>

**Lung Transplantation:** IPF is now the leading indication for lung transplantation in most large centers. In 2009, at the Cleveland Clinic, University of Pittsburgh Medical Center, and a number of other large transplant centers, over 50% of the lung transplants performed were for IPF. Transplantation can improve both longevity and the quality of life in properly selected patients who have no other significant health problems. Previously it was uncommon for individuals over the age of 70 to receive transplants. However, as surgical techniques and outcomes have improved, more individuals over 70 are receiving transplants, and many medical centers have updated their age requirements to now include those over the age of 70.

Until recently, because of long pre-transplant wait times, early referrals were essential so that patients could begin accruing time on the transplant waiting list. Fortunately with a new lung allocation system (LAS) used by the United Network for Organ Sharing, or UNOS ([www.unos.org](http://www.unos.org)), candidates are evaluated based on the severity of their disease, and as a result wait times for those with IPF have been dramatically reduced. Transplantation is not without risk, and patients should discuss all the potential risks and benefits of lung transplantation with their physician.

**Clinical Trials:** Today, more than ever before, researchers are aggressively investigating new treatments for idiopathic pulmonary fibrosis (IPF). While the long term goal of IPF research is to prevent and cure the disease, present therapeutic approaches consist of attempts to slow disease progression, and to extend the life expectancy of patients with IPF. There are a variety of therapeutic approaches currently being studied, including:

- Anti-fibrotic therapies that may slow, or inhibit, the body's ability to produce scar tissue, or fibrosis.

- Endothelin receptor antagonists (ERAs) that may help IPF patients manage pulmonary arterial hypertension (PAH), which is a serious condition commonly associated with IPF; also in experimental models, ERAs have been shown to inhibit the formation of collagen and scar tissue.
- Inhibitors of “growth factor” proteins that may, alone, or in combination with other similar proteins, contribute to the formation of scar tissue, or fibrosis.
- Pulmonary vasodilators, such as sildenafil, that may help IPF patients process oxygen more efficiently.
- Some blood pressure lowering medications, such as losartan which is an angiotensin receptor blocker, may function similar to ERAs and allow patients to process oxygen more efficiently.
- Genetic research to identify genes that may be associated with IPF and help identify individuals and families that are prone to IPF. The markers may also predict the rate of disease progression.

While some studies are in advanced stages of development, others are in much earlier stages. There are a variety of clinical trials that are actively seeking the participation of patients.

**Since there are currently no FDA approved therapies to treat IPF, many patients choose to participate in clinical trials after consulting with their physician. New, experimental therapies are tested for their effectiveness through clinical trials. It is very important that patients discuss the possibility of participating in a clinical trial with their physician upon diagnosis. It is through clinical trials that a cure for the disease will be found. Please visit the research section of our website at [www.pulmonaryfibrosis.org/research](http://www.pulmonaryfibrosis.org/research) to learn more about active clinical trials in the United States.**

# Quality of Life

## What Can You Do?

What can patients do to stay healthy? There are a variety of things that patients can do to maintain or improve their quality of life while living with PF or IPF. The National Institutes of Health ([www.nih.gov](http://www.nih.gov)) and the Mayo Clinic ([www.mayoclinic.org](http://www.mayoclinic.org)) offer a variety of recommendations for patients, some of which we have referenced in this section.

**Stay in Shape.** The most damaging consequence of lung disease and its sensation of “breathlessness” is the development of an inactive lifestyle. For many patients, activities of daily living like bathing and dressing can create overwhelming fatigue. Air hunger can create panic attacks, and produce negative psychological effects. People with chronic respiratory problems sometimes limit their physical activities in an attempt to avoid shortness of breath. The lack of exercise works against you. Inactivity weakens your muscles and they become less efficient. Deconditioning can make even the simplest daily activities more difficult. Through regular exercise muscles become stronger and more resistant to fatigue. With practice and training you can learn to perform tasks in a more efficient manner. By being more efficient you need less oxygen for the same amount of work. The result is that you may find that you have more energy to accomplish daily tasks and that you are less short of breath. A formal rehabilitation program (pulmonary rehabilitation) is preferred because it allows for observation during exercise and it can be tailored to your specific needs.

**Eat Well.** A healthy diet includes a variety of fruits, vegetables, and whole grains. It also includes lean meats, poultry, fish, beans, and fat-free or low-fat dairy products. A healthy diet is low in saturated fat, trans fat, cholesterol, sodium (salt), and added sugar. Eating smaller, more frequent meals may relieve stomach fullness, which can make it hard to breathe. If you need help with your diet, ask your doctor to arrange for a dietitian to work with you. A nutritionally rich diet that contains adequate calories is essential. A dietitian can give you further guidelines for healthy eating.

**Get Plenty of Rest.** Getting at least eight hours of quality rest every night can boost your immune system and sense of well-being.

**Stop Tobacco Use.** Avoiding environmental irritants, like cigarette smoke, is a good way to prevent further damage to your lungs. If you are still smoking, the most important thing you can do is to stop. Due to the addictive nature of tobacco, this is can be difficult. Seek the help of your physician to find a smoking cessation class or other beneficial methods to help you. Secondhand smoke can be as harmful to you as if you were smoking yourself. Ask your family and friends to refrain from smoking around you as well.

**Learn and Practice Relaxation Techniques.** When you are physically and emotionally relaxed, you avoid excessive oxygen consumption caused by tension of overworked muscles. Additionally, learning relaxation techniques can help you manage the panic that often accompanies shortness of breath. Joining a support group and/or seeing a counselor can help you cope with your feelings and the anxiety and depression that are common in people with chronic breathing disorders. These feelings may

aggravate the underlining disease. Many fear losing the ability to function and becoming dependent on others. The restriction on activity due to shortness of breath may lead to isolating oneself from family and friends, adding to the depression.

**Join a Support Group.** Just knowing that there is someone “out there” that knows just how you feel is comforting. Share ideas, share fears, and share joys. A detailed listing of face-to-face and online support groups can be found at [www.pulmonaryfibrosis.org](http://www.pulmonaryfibrosis.org).

**Participate in Your Health Care.** Remember you are part of a health care team that includes doctors and nurses. They will be asking you a lot of questions. As a member of that team you have a responsibility to do your part. Be prepared to ask your own questions. Be a participant. Bring someone with you to each appointment and prepare a list of questions to be answered by your physician during your visit.

**Help Others with IPF.** Consider participating in the Pulmonary Fibrosis Foundation’s advocacy program. You may gain strength in knowing that you are helping future patients and researchers by lobbying your members of Congress to do more to help the PF community.

**Keep a Positive Attitude!** Actively participating in all parts of the management of your disease is greatly enhanced by a positive attitude. A positive attitude can help you and your loved ones cope with your disease.

“A strong positive mental attitude will create more miracles than any wonder drug.” – Patricia Neal

# About the PFF

## Our Mission

The mission of the Pulmonary Fibrosis Foundation (PFF) is to help find a cure for idiopathic pulmonary fibrosis (IPF), advocate for the pulmonary fibrosis community both locally and in Washington, D.C., promote disease awareness, and provide a compassionate environment for patients and their families.

Our staff is always available to discuss your individual needs. Please use our contact information freely, and if you know of a patient or family that could use our help, please share our contact information.

## Pulmonary Fibrosis Foundation

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As a 501(c)(3) public benefit organization, the Pulmonary Fibrosis Foundation provides information to the PF community free of charge; we rely on public support to provide this valuable resource to patients and their families throughout the United States and internationally. If you have found our educational tools, website, or staff helpful please consider making a gift to the PFF so that we may continue to be a source of compassionate support for patients and families in need while we help to find a cure for IPF by funding new research.

## Our Objectives

The Pulmonary Fibrosis Foundation has an ambitious, forward looking agenda to achieve our mission:

- Developing caring relationships with patients and their families throughout the course of their disease
- Substantially increasing funding for IPF research and assist in creating partnerships between the academic research community and the biotech industry to drive new treatments
- Hosting a national conference on IPF to improve clinical knowledge of PF in a collaborative environment — *IPF Summit: From Bench to Bedside* — Chicago, Illinois (December 1–3, 2011)
- Implementing new web-based patient education and disease awareness programs including webinars, online support services, and social media platforms
- Establishing nationwide Affiliate Groups to expand our outreach while enabling the PF community to participate in the PFF’s advocacy, awareness, education, and patient support initiatives
- Representing the needs of our constituents in Washington, D.C. through national advocacy
- Aggressively pursuing an increase in public awareness through a series of public service announcements (PSA), social networking, and traditional media exposure

## Our Partners in the Medical Community

The Pulmonary Fibrosis Foundation continues to be committed to creating partnerships between the academic research community and the biotech industry to drive new treatments for IPF, while fostering collaboration in the clinical community to share information and ideas. We are proud to work with the following organizations in funding research programs to find a cure for IPF:

American College of Chest Physicians, Northbrook, Illinois  
American Lung Association, New York, New York  
American Thoracic Society, New York, New York  
Baylor College of Medicine, Houston, Texas  
Cincinnati VA Medical Center, Cincinnati, Ohio  
Coalition for Imaging and Bioengineering Research,  
Washington, District of Columbia  
Brigham Women's Hospital, Boston, Massachusetts  
Duke University, Durham, North Carolina  
Emory University, Atlanta, Georgia  
Instituto Nacional de Enfermedades Respiratorias,  
Mexico City, Mexico  
Inova Fairfax Hospital, Falls Church, Virginia  
Irish Lung Fibrosis Association, Dublin, Ireland  
Mayo Clinic, Rochester, Minnesota  
Medical University of South Carolina, Charleston, South Carolina  
National Heart, Lung and Blood Institute, Bethesda, Maryland  
National Jewish Health, Denver, Colorado  
Northwestern University, Chicago, Illinois  
Office of Rare Diseases, NIH, Bethesda, Maryland  
Pulmonary Fibrosis Research Institute, Chicago, Illinois  
Rare Diseases Clinical Research Network Patient Advocacy  
Consortium, Bethesda, Maryland

Rare Lung Disease Foundation Consortium, Cincinnati, Ohio  
Research! America, Washington, District of Columbia  
Royal Brompton Hospital, London, United Kingdom  
Rush University Medical Center, Rush Generations,  
Chicago, Illinois  
Stanford University, Stanford, California  
The CHEST Foundation, Northbrook, Illinois  
The Centre for Respiratory Research at University College,  
London, England  
Tulane University, New Orleans, Louisiana  
Univ-Klinik fur Innere Medizin IV, Vienna, Austria  
University of Iowa, Iowa City, Iowa  
University of California at Los Angeles, Los Angeles, California  
University of Chicago, Chicago, Illinois  
University of Illinois, Chicago, Illinois  
University of Miami School of Medicine, Miami, Florida  
University of Michigan, Ann Arbor, Michigan  
University of Pittsburgh, Pittsburgh, Pennsylvania  
University of Southern California, Los Angeles, California  
University of Vermont, Burlington, Vermont

**Join the PFF Community – It's Free!**

The Pulmonary Fibrosis Foundation is committed to supporting research to find a cure for idiopathic pulmonary fibrosis (IPF), advocating for the pulmonary fibrosis community both locally and in Washington, D.C., promoting disease awareness, and providing a compassionate environment for patients and their families. We need your help to do it! Joining the Pulmonary Fibrosis Foundation is free of charge, and will help you better connect with the pulmonary fibrosis community as it strives to cure this devastating disease. Benefits include:

- Invitations to PFF-sponsored educational events, including webinars
- Participation in PFF online communities and support groups
- PFF's quarterly *Breathe Bulletin* newsletter
- Emails about news and updates important to the PF community
- Support group announcements
- Fundraising announcements and invitations
- Clinical trial announcements
- Participation in PFF national advocacy efforts

Simply visit our website and click the "JOIN" button!

### **How Can You Invest in Helping to Find a Cure for IPF?**

- Make a gift of cash
- Make a gift of marketable securities
- Purchase a PFF "Breathe" bracelet and related products
- Name the PFF in your family wills and bequests
- Establish a charitable gift annuity for the benefit of the PFF
- Become a volunteer

Regardless of which method you choose, you will be making an important contribution to Pulmonary Fibrosis Foundation's goal of finding a cure for IPF.

**Call 888.733.6741 or visit [www.pulmonaryfibrosis.org](http://www.pulmonaryfibrosis.org) to make a gift or join the PFF community.**

## **Our National Patient Advocacy Program –**

### **We Need Your Help!**

Since 2001, the Pulmonary Fibrosis Foundation has had an increasing presence in Washington, D.C. to effectively represent the needs of our constituents. Now more than ever, we need to expand our efforts to effectively advocate for patients and families, and for researchers who face immense challenges in finding new treatments for this devastating disease. While the Pulmonary Fibrosis Research Enhancement Act (PFREA) is central to our efforts to help our community, the PFF is committed to other advocacy and disease awareness efforts that can have a positive, enduring impact on the lives of the patients we serve, including:

- Building awareness of PF at the Centers for Disease Control and Prevention (CDC) to improve public education and awareness of PF
- Working closely with the Food and Drug Administration (FDA) to improve the design of clinical trials for PF and broker an improved collaboration between the FDA and private industry to foster new investment in the development of PF treatments
- Influence state and federal legislation that has an impact on PF patients, families, and researchers, such as the PFREA, Medicare coverage issues, Social Security benefits, National Institutes of Health (NIH) funding, and other legislation affecting our community
- Working with state and federal agencies to improve awareness of organ donation to increase the availability of donor lungs for PF patients needing transplant
- Be a source of expertise, and advocate around, the broader public health issues surrounding 9/11 first responders and the potential long term impact of their environmental exposures attributed to their work at Ground Zero

“I don’t waste time getting caught up in statistics or life expectancy. If I worried about that I wouldn’t be able to live life the way I want to live it. I am still going 12 years after I first remember symptoms and 9 years after being diagnosed. I don’t think about stats. I expect to live a long life.”

– KERRY GERON  
NEW ALBANY, INDIANA

**To accomplish our goals, the PFF NEEDS YOUR HELP – collectively, we can be effective advocates for PF issues!**

**Our national advocacy program relies on the participation of patients and families that have been impacted by PF and are interested in joining our efforts to advocate for PF issues.**

**Please visit our website at [www.pulmonaryfibrosis.org](http://www.pulmonaryfibrosis.org) or call **888.733.6741** to become a member today!**

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