



LET'S TALK ABOUT PULMONARY FIBROSIS (PF):
Questions and Answers for Patients and Caregivers

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WHAT IS PULMONARY FIBROSIS?

Pulmonary Fibrosis (PF) is a debilitating disease—marked by progressive scarring of the lungs—that gradually interferes with a person’s ability to breathe. PF belongs to a family of approximately 100 related diseases, called *interstitial lung diseases*, which have similar characteristics and can result in lung scarring. This scarring is most often referred to as pulmonary fibrosis.

Your doctor may have referred to your condition by one of a number of names that are sometimes used to describe PF. This confusion may be due, in part, to similarities between PF and the other forms of interstitial lung disease. Only since 2001 has PF been recognized as a distinct clinical disorder, meaning that specific clinical criteria were developed to determine a diagnosis. Your physician combines clinical information derived from a medical evaluation and certain diagnostic tests to diagnose PF according to these criteria.

Sometimes pulmonary fibrosis can be linked to a particular cause, such as certain environmental exposures, chemotherapy or radiation therapy, residual infection, or autoimmune diseases such as scleroderma or rheumatoid arthritis. However, in many instances, no known cause can be established. When this is the case, it is called idiopathic pulmonary fibrosis or IPF.

This educational tool was designed to provide useful information to patients newly diagnosed with pulmonary fibrosis—with the goal of helping them and their caregivers better understand and manage their condition. The information supplied here is a guide to answer many of the most commonly asked questions regarding the disease, and its symptoms, diagnosis, and treatment.

The information is not intended to replace the advice or services of trained health professionals. You should seek the advice of your physician or healthcare professional in all matters relating to your health, particularly with respect to the diagnosis and treatment of any medical condition.



Education. Support. Hope.

Whether you are an PF patient, a family member of a patient, or are close to someone with PF, the CPF is always just a phone call away. Our staff have counseled thousands of patients, and we are always here to provide you with the resources and support you need. Please call (888) 222-8541 to learn more.

PULMONARY REHABILITATION AND OXYGEN MANAGEMENT FOR THE PF PATIENT:
Helping You Make the Most of Every Day

LUNG TRANSPLANTATION:
What Every Patient with Pulmonary Fibrosis (PF) Should Know

PF hinders a person's ability to take in oxygen. It causes shortness of breath and is usually associated with a persistent dry cough. The disease progresses over time, leading to an increase in lung scarring and a worsening of symptoms. Unfortunately, PF is ultimately disabling and fatal.

WHAT CAUSES PF?

While the cause of PF remains a mystery, what researchers suspect is that PF involves changes in the lung's normal healing process. PF patients may have an exaggerated or uncontrolled healing response that, over time, produces excessive fibrous scar tissue – or fibrosis – in the lungs. This scarring, in turn, causes the lung's tiny air sacs, called alveoli, to thicken and stiffen—rendering them less able to function and provide the body with the oxygen it needs.

Exactly what sets this abnormal tissue-repair process in motion is unclear. The body's own immune response may play a major role. Researchers are investigating a number of potential risk factors that may make a person more likely to develop PF. These risks may include:

- Cigarette smoking
- Occupational exposure to dusty environments (e.g. wood or metal dust)
- Genetic predisposition (10-15 percent of cases)
- Viral or bacterial lung infections
- Acid reflux disease

WHAT SYMPTOMS MIGHT YOU EXPERIENCE?

Symptoms of PF usually have a gradual onset and may include:

- Shortness of breath, particularly during or after physical activity
- Spasmodic, dry cough
- Gradual, unintended weight loss or weight gain
- Fatigue and weakness
- Chest discomfort
- Clubbing, or enlargement of the ends of the fingers (or sometimes the toes) due to a buildup of tissue

As you can imagine, PF symptoms can greatly impact quality of life. Fortunately, there are things you and your doctor can do to help minimize lifestyle-altering affects of the disease

DID YOU KNOW?

Approximately 128,000 people in the United States have PF, and prevalence has risen dramatically since 2000. An estimated 48,000 new cases are diagnosed each year. The disease tends to affect men more than women and usually strikes people between the ages of 50 and 70.



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PF can be difficult to diagnose. An estimated 50% of cases are initially misdiagnosed as another form of respiratory disease.

HOW IS PF DIAGNOSED?

Because patients with PF experience symptoms and also have an underlying pattern of scarring similar to those of other lung disorders, PF can be difficult to diagnose. In fact, it was not until 2000 that PF was classified as a distinct clinical disorder by a panel of experts sponsored by the American Thoracic Society (ATS) and European Respiratory Society (ERS). Until recently, the medical community has had no agreed-upon standards for the diagnosis of PF. Consequently, other related lung diseases were often incorrectly classified as PF. With new diagnostic standards now in place, the recognition and management of PF should be substantially improved.

To diagnose PF, your doctor will take a complete medical history and perform a thorough physical examination. During this exam, the doctor will use a stethoscope to listen to your chest to try to determine if your lungs produce any abnormal sounds when you breathe. He or she may then order one or more of the following diagnostic tests or procedures:

DIAGNOSTIC	DESCRIPTION	PURPOSE
Chest imaging	Use of radiologic machines to take pictures of your lungs, such as x-ray or High Resolution Computer Tomography (HRCT)	To view lung structures look for scar tissue and assess patterns of scarring
Pulmonary function test	A test using a device with a mouthpiece to measure a patient's breathing capacity	To measure the degree of impairment in lung function
Arterial blood gas test	A measurement of oxygen and carbon dioxide levels in blood taken from an artery in the wrist	To determine how well the lungs are performing vital gas exchange
Exercise Test (or desaturation study)	A test in which the patient is monitored while using a treadmill or stationary bicycle	To measure how well the lungs and heart respond to physical activity and evaluate oxygen levels with exertion
Six Minute Walk Test (SMWT)	A test where a patient walks on a flat surface as far as possible in six minutes	To measure the distance you are able to walk as well as lung function during the walk.
Bronchoalveolar lavage (BAL)	A "lung-washing" procedure conducted through a flexible tube (bronchoscope) inserted into the airways through the nose or mouth; fluid (salt water) is injected into the lungs and then removed for inspection	To examine cells and fluid to look for signs of inflammation in the lungs, or markers of disease activity
Lung biopsy	A procedure in which a tissue sample is obtained through a bronchoscope (see BAL, above) or by means of a small surgical incision (VATS- video-assisted thoracic surgery) between the ribs (open-lung biopsy)	To obtain a sample of lung tissue for direct examination



WHAT MAY HAPPEN AS PF PROGRESSES?

PF affects each person differently and progresses at varying rates. Generally, the patient's respiratory symptoms become worse over time. Activities (such as walking or climbing stairs) become more difficult.

In addition:

- The patient may require supplemental oxygen
- Advanced PF makes it difficult for a person to fight infection.
- PF causes a lack of oxygen in the blood. This condition (called hypoxemia) puts a strain on the heart and on the blood vessels in the lungs, and may lead to high blood pressure in the lungs (pulmonary hypertension).
- PF has also been associated with these potentially life-threatening conditions: heart attack, respiratory failure, stroke, blood clot in the lungs (pulmonary embolism), lung infection, and lung cancer.

HOW IS PF TREATED?

Physicians generally follow standards of care intended to improve symptoms and hopefully slow the progression of the disease. In 2001, a panel of experts sponsored by the American Thoracic Society and the European Respiratory Society recommended that if therapy was given, it should consist of a trial of corticosteroids and an immunosuppressive agent (e.g. azathioprine or cyclophosphamide). Recent evidence has suggested that adding a third therapy called N-acetylcysteine (NAC) to this regimen may be beneficial. *It is important to recognize, however, that there are no definitive studies showing that this treatment approach is effective, and there is no consensus regarding the use of this approach in the pulmonary community. For a complete listing of active clinical trials for PF, please visit www.coalitionforpf.org.*

Importantly, these therapies can cause side effects—some minor and some more serious. The potential risks and benefits of therapy should be discussed with your physician in detail. Be sure to contact your doctor or nurse if you have any negative reactions to any medications you have been prescribed.

In all cases, doctors should consider referring their patients to a clinical trial to gain access to experimental treatments and also refer patients for lung transplantation evaluation.

There remains no FDA-approved treatment for PF, and as of yet, there is no cure.

Please see the Lung Transplantation section of this educational tool for more information, and consult your physician to determine if lung transplantation is an option for you.

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Funding for new research into PF remains unacceptably low. While PF is five times more common than Cystic Fibrosis (CF), for comparison purposes, CF receives an estimated \$85 Million in NIH funding each year while PF receives an estimated \$18 Million. You can help solve this challenge. Please contact the CPF to learn how you can help increase funding for PF research through a gift, or by participating in our national patient advocacy campaign.

ARE THERE ANY NEW TREATMENTS ON THE HORIZON?

Researchers are developing and testing a variety of new ways to treat PF. These approaches target the various steps in the disease process. Therapies under investigation include:

- Antifibrotic or anti-fibrogenic agents to suppress the scarring process
- Antioxidants (such as N-acetylcysteine-NAC) to prevent damage to lung tissue
- Endothelin antagonists (such as certain medications for pulmonary arterial hypertension (PAH))
- Monoclonal antibodies to inhibit “bad” cytokines (protein growth factors, such as TGF-beta, TNF-Alpha, or CTGF) that activate inflammation.

There are several emerging therapies currently in clinical trials for PF patients. Contact the CPF or visit www.coalitionforpf.org for a complete up-to-date listing of active investigational research. The National Institutes of Health (NIH) also offers information at www.clinicaltrials.gov for those interested in identifying PF-related research at medical centers across the United States. It represents the most up-to-date research information for patients including clinical trial sites, patient inclusion/exclusion criteria, and contact information for each trial.

As with any course of care, please consult your physician to determine what current treatment options, including participation in clinical trials, may be appropriate for you.

A WORD TO CAREGIVERS

Caring for someone who has a debilitating illness can be both physically and emotionally demanding. You are doing your best to ensure that your loved one gets the best possible care—staying informed about potential treatment options and doing all you can at home to make sure his or her day-to-day needs are met. In the process, you need to remember to take care of yourself as well.

You and your loved one may be coping with a great deal of stress and anxiety as a result of your current situation. If you ignore these feelings, they can have a negative impact on your health and well-being. To help reduce the stress in your life, try to get regular exercise and learn some relaxation techniques. You may also want to join a support group or to see a counselor to help you cope with your feelings. The important thing is for both of you to ask for and get the help that you need. There are support groups specifically designed for caregivers-only.

PROVIDING SUPPORT TO PATIENTS AND THEIR FAMILIES

Joining a local support group can help patient and their families better cope with their disease, while sharing information and support in an open, caring environment. The CPF has helped to create more than 50 PF support groups across the country. Visit the CPF web site for a listing of support groups at www.coalitionforpf.org, or call (888) 222-8541 to learn more.

Dozens of patients have helped create PF support groups in their communities. It's a great way to get the emotional support you need. If there is not yet an PF support group in your area, contact the CPF for your free copy of the "PF Support Group Coordinator's Kit" and we'll guide you through the process of starting an effective and lasting group.

WHAT CAN YOU DO?

If you have been diagnosed with PF, there are a number of things you can do to take part in your own treatment and help yourself stay healthy. Caregivers may also be interested in the following information, to assist a family member diagnosed with PF:

- Call your doctor or nurse with any questions about your condition or its treatment. If you notice anything unusual about how you are feeling or how your medicines are working, call your doctor right away.
- Take all prescribed medications as instructed by your doctor.
- Get your influenza (flu) vaccine every year, and also make sure that your pneumococcal vaccine (Pneumovax) is up to date.
- If you smoke, it is very important that you stop as soon as possible. Ask your doctor or nurse about smoking cessation programs and products that can help.
- Your doctor may have prescribed supplemental oxygen, which can provide your body with the oxygen it needs but your lungs can no longer supply. Some patients fear that they will become addicted to oxygen, but this is not true. Supplemental oxygen can help you feel less breathless and more energetic, while protecting your heart and other vital organs.

- Consult your physician about enrolling in a pulmonary rehabilitation or respiratory therapy program to help increase your strength, learn breathing techniques, and expand your social support network. Ask your doctor or nurse for more details. Many patients report improved breathing and quality of life after adding education and exercise to their treatment. Please note you can participate in a respiratory therapy program even if you are currently taking supplemental oxygen.
- Eat a well-balanced diet to maintain in ideal body weight. This helps support your body and keeps up your strength. Discuss any special nutritional concerns with your doctor or nurse.
- Consider eating smaller, more frequent meals during the course of your day. Many patients find it easier to breathe when their stomach isn't completely full.
- Talk about PF: Get the word out to raise awareness about the disease. Tell your friends, colleagues and local newspapers about your experience – your story can make a difference. Speak and write to members of Congress about PF and join the CPF in its work to increase Congressional attention to the disease.



PULMONARY REHABILITATION AND OXYGEN MANAGEMENT FOR THE PF PATIENT:

Helping You Make the Most of Every Day

LUNG TRANSPLANTATION:

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“I am full of energy for this cause. I feel as if I have just planted several small, young trees on Capitol Hill, and over the next many years they will grow into a nice big canopy of protection, comfort and love for the generations of PF patients and their families to come.”

— Bill Rhodes, CPF member and delegate to Washington D.C.

CAMPAIGN ACT – THE CPF AND ITS MEMBERS SERVE AS A NATIONAL VOICE FOR THE PF COMMUNITY

The CPF and its volunteers tirelessly voice the needs of patients, their families, and all those who are affected by PF, through the CPF’s campaign ACT national advocacy program. The CPF regularly advocates for increased federal funding for pulmonary fibrosis and for legislation in Congress that is important to the community we serve.

Since 2002, the CPF has been leading a national advocacy effort to increase research funding for PF and accelerate efforts to find a cure for this devastating lung disorder. This has included a close collaboration with the late Congressman Charlie Norwood - who lost his battle with PF in 2007 - to secure passage of H.R. 182 in 2007, which was the first-ever Congressional recognition of the need for increased research funding and improved public awareness of PF in the United States. This Resolution laid the groundwork for the CPF’s collaboration with Congressmen Brian Baird (D-WA) and Mike Castle (R-DE) to introduce the historic Pulmonary Fibrosis Research Enhancement Act (H.R. 6567) in June, 2008. This landmark Bill has further established the CPF as a national voice for the PF community, and we remain committed to advocating for several legislative efforts in Washington that would impact our patients and researchers.

Campaign ACT - Ask. Challenge. Take Action!

By participating in the CPF’s campaign ACT national advocacy program, you will be part of an army of volunteers who will contact Congress, national health policymakers and the media to educate them about PF, and communicate to them the desperate needs of our patients and researchers around the country. The CPF regularly advocates for increased federal funding for pulmonary fibrosis and for legislation in Congress that is important to the pulmonary fibrosis community.

Continue your fight against PF, and help others who are suffering, by joining the CPF’s campaign ACT program today! To learn more or participate, please call us at (888) 222-8541 or visit our Web page at www.coalitionforpf.org/cpf_advocacy.

ACT
Ask.
Challenge.
Take Action.

RECOMMENDED READING

The following books offer information on lung disorders, and are available through most major booksellers online:

- The Breathing Disorders Sourcebook, *by F.V. Adams, M.D.*
- Shortness of Breath: A Guide to Better Living and Breathing, *by A.L. Ries, et al*
- The Lung Transplantation Handbook, *by K.A. Coulture*
- Coping with Prednisone, *by E. Zukerman and J.R. Ingelfinger, M.D.*
- The Official Patient’s SourceBook on Idiopathic Pulmonary Fibrosis, *by J.N. Parker & P. Parker*
- Share the Care: How to Organize a Group to Care for Someone Who Is Seriously Ill, *by C. Capossela & S. Warnock*
- Taking Flight - Inspirational Stories of Lung Transplantation, *Compiled by Joanne Schum, authored by lung recipients around the world*
- Adventures of an Oxy-phile, *by Thomas L. Petty, M.D.*

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PARTNERING WITH WORLD-CLASS ORGANIZATIONS WHO SHARE OUR COMMITMENT TO THE PF COMMUNITY

The CPF's nonprofit partners include many of the most respected medical centers and healthcare organizations in the U.S., including the American Thoracic Society, the National Organization for Rare Disorders (NORD), The Pulmonary Paper, Mary D. Harris Memorial Foundation, National Coalition of Autoimmune Patient Groups, The Genetic Alliance, The Anne Harroun Landgraf Foundation, the Caring Voice Coalition, and more than 40 medical centers specializing in the treatment and study of PF.

For a complete listing of our partners, please visit www.coalitionforpf.org.

You can have a profound impact on the future of pulmonary fibrosis patients – and lead the charge to cure PF – by contributing to the CPF.

To learn more, call us at (888) 222-8541 or visit www.coalitionforpf.org

ARE YOU A PATIENT WITH A FAMILIAL FORM OF PULMONARY FIBROSIS?

The Coalition for Pulmonary Fibrosis (CPF) and National Jewish Medical and Research Center partnered to launch the first genetic counseling program for patients and families affected by FPF. The telephonic counseling program is operated by National Jewish, and funded by CPF. An estimated 10-15 percent of PF patients have a form of the disease that runs in families, known as familial pulmonary fibrosis (FPF).

The genetic counseling program provides a qualified genetic counselor, who has expertise in FPF, to discuss by phone various issues surrounding FPF. These can include preparation for and interpretation of genetic tests, and various life decisions, such as having children and planning for the future. Experts recommend talking to a counselor prior to having any genetic tests, so that people are prepared to learn the results.

For further information on the FPF Genetic Counseling Program or to speak with a genetic counselor, call (800) 423-8891, ext. 1097.



PULMONARY REHABILITATION AND OXYGEN MANAGEMENT FOR THE PF PATIENT:

Helping You Make the Most of Every Day

LUNG TRANSPLANTATION:

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“When a treatment or a cure comes, the CPF will be involved. I feel strongly that the CPF is the most likely vehicle that will lead to hope for PF patients and researchers in the United States.”

— Dave Steffy, CPF major contributor

BECOME A CPF MEMBER AND CONNECT TO THE PF COMMUNITY!

When you register with the CPF, free of charge, you'll join a national community of patients, families, and researchers committed to curing PF. You'll begin receiving a complimentary subscription of the CPF's quarterly *Action Alert* newsletter, the definitive source of education, information and resources for PF patients and their families in the U.S. Just send us your name, address, phone number and email address to become a member! You can also call, email, or send your contact information to us:

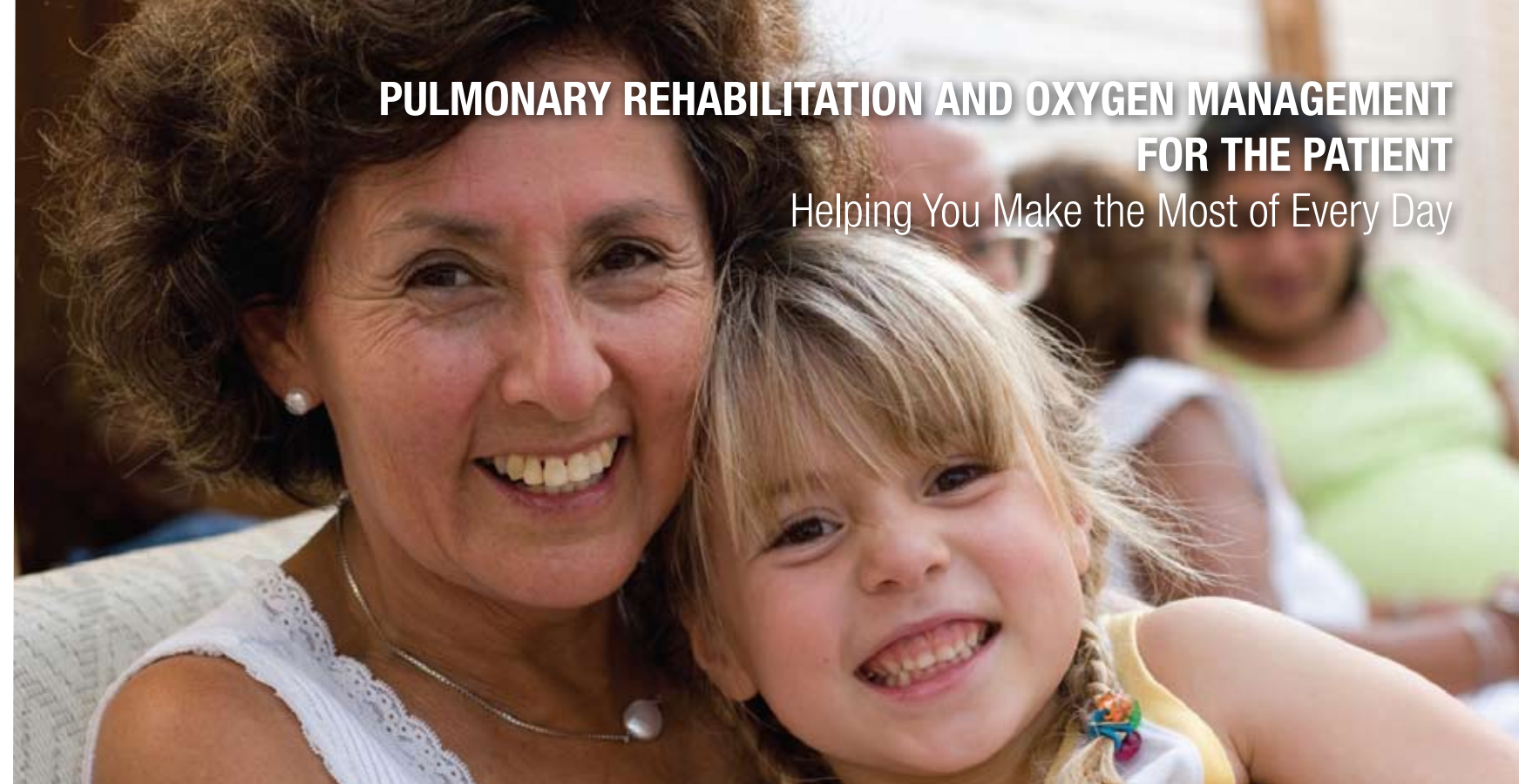
Coalition for Pulmonary Fibrosis

Suite F, #227
1659 Branham Lane
San Jose, CA 95118-5226

info@coalitionforpf.org
(888) 222-8541

The CPF touches the lives of thousands of patients and families each year. If you need help, contact the CPF and we will work with you to assess your needs. If necessary, we'll refer you to one or more of our partner organizations for special assistance.

Contact us at (888) 222-8541, or by email at info@coalitionforpf.org



PULMONARY REHABILITATION AND OXYGEN MANAGEMENT FOR THE PATIENT

Helping You Make the Most of Every Day

PULMONARY REHABILITATION AND OXYGEN MANAGEMENT FOR THE PF PATIENT:

Helping You Make the Most of Every Day

LIVING LIFE TO ITS FULLEST

For the pulmonary fibrosis (PF) patient, managing symptoms of respiratory distress can make even the simplest activities of daily living a challenge. Not only does PF gradually rob you of your ability to breathe properly, but low oxygen levels in your bloodstream can leave you feeling weak, tired and uncomfortable, and compromise your quality of life. You may find yourself curtailing activities that previously brought you pleasure, from walking around the block or visiting the supermarket to attending family outings or enjoying concerts, plays and movies.

Yet if you are a PF patient, there is no need to put your life on hold. Pulmonary rehabilitation and oxygen therapy are two treatments designed to help people with chronic lung disease become more active and vibrant while better understanding how your body uses oxygen.

Through pulmonary rehabilitation, you can build your strength and endurance – resulting in a richer, more enjoyable life experience. If your oxygen levels are low, supplemental oxygen therapy can supply your body with the oxygen it needs and possibly avoid the risk of future cardiac (heart) stress.

Why observe life from the sidelines when you can again
be an active participant?



Pulmonary rehabilitation and oxygen therapy can help you live life to its fullest. Make it a point to discuss these options with your physician.

WHAT'S GOING ON IN YOUR LUNGS?

PF is a debilitating condition that involves scarring of the lungs. The lungs' air sacs develop scar – or fibrotic – tissue, gradually interfering with the body's ability to transfer the oxygen you breathe into the bloodstream, and preventing your vital organs and tissue from obtaining enough oxygen to function normally.

What happens when your lungs lose the ability to transfer oxygen into the bloodstream?

The following symptoms can result:

- Shortness of breath, particularly during or after physical activity
- Spasmodic, dry cough
- Gradual, unintended weight loss or weight gain
- Fatigue and weakness
- Chest discomfort
- Clubbing, or enlargement of the ends of the fingers (or sometimes the toes) due to a buildup of tissue

As you can imagine, these symptoms can greatly impact the quality of life. Fortunately, both pulmonary rehabilitation and oxygen therapy can help minimize lifestyle-altering effects of PF.



Education. Support. Hope.

PULMONARY REHABILITATION: BRINGING BACK LIFE'S PLEASURES

Pulmonary rehabilitation programs are designed to help you be as active and functional in your daily life as possible while living with PF. Most programs include medical management, exercise training to improve endurance and strength, breathing retraining, education, emotional support and nutritional counseling. Physicians, physical therapists, exercise physiologists, nurses, respiratory therapists, psychosocial professionals and nutritionists work together to help you reach your functional goals.

An important component of pulmonary rehabilitation is exercise, which helps reduce the effects of inactivity, increases your exercise capacity, and allows you to maintain activities of daily living. Emotions associated with PF – including depression and anxiety – may interfere with your daily activities. Psychosocial counseling and emotional support are also key components of pulmonary rehabilitation and can help alleviate these feelings.

Pulmonary rehabilitation also addresses nutritional issues, as severe lung disease may cause either weight loss or weight gain. Nutritionists can teach you how to eat properly to maximize your health. Finally, education will help empower you – teaching you how to get the most out of life while living with PF.



Pulmonary rehabilitation programs are designed to help prolong life, reduce disability and raise your level of physical and social functioning to the highest possible level. Since each individual with PF presents unique challenges, pulmonary rehabilitation programs are individually tailored to your needs. You will receive individual assessment and instruction, as well as an opportunity to take part in group activities.

ARE YOU A CANDIDATE FOR PULMONARY REHABILITATION?

If you answer yes to any of the following questions, you may be a candidate for pulmonary rehabilitation:

- Is your shortness of breath compromising your lifestyle?
- Do you want to learn more about your disease and improve your self-care?
- Do you want to discover ways to improve your breathing?
- Do you want to increase your exercise endurance?
- Do you want to maintain your activities of daily living?

Through breathing retraining, regular exercise, proper nutrition, psychosocial support and ventilatory therapy, you can dramatically improve your breathing ability – and your ability to make the most of every day.



Supplemental oxygen can:

- Decrease your shortness of breath – especially with exercise
- Improve your ability to perform activities of daily living (ADL's)
- Improve your overall level of fitness
- Improve your quality of life
- Increase life span by decreasing the extra work your heart is doing because of low oxygen saturation levels

SUPPLEMENTAL OXYGEN THERAPY: ANOTHER VITAL TOOL

When PF progresses to a point where your blood-oxygen levels are low, another vital tool that can help people with PF lead fuller, more active lives is supplemental oxygen therapy. Oxygen is prescribed by your physician, and can be provided by a local medical equipment company for home use, allowing you to be more active in your life.

Oxygen is a gas that you can't see, taste or smell, yet it is in the air all around us and is necessary in order to survive. Why is oxygen so important? Oxygen is vital in the production of chemical energy in the cells of your body that all organs and tissues need to function optimally.

Reduced levels of oxygen in your body can injure cells and may cause cell death. Indeed, this lack of oxygenation has been shown to decrease life span by making your heart work too hard.

The amount of oxygen in the air is always the same – 21%. This is enough for people with normally functioning heart and lungs to thrive. However, a person with PF – whose disease prevents an adequate transfer of oxygen into the bloodstream – will benefit from breathing air with a higher concentration of oxygen because the scarring that occurs in PF patients can slow the movement of oxygen into the bloodstream, requiring the lungs to work harder. Supplemental oxygen helps increase the amount of oxygen that is available to be transferred from your lungs into the bloodstream, thereby producing more energy to be used by the cells of your body. This is why you and your doctor should discuss supplemental oxygen as part of your overall treatment plan.

WHO NEEDS SUPPLEMENTAL OXYGEN?

If you are unable to maintain the necessary amount of oxygen by just breathing room air, you may require supplemental oxygen. To determine if you need oxygen, your doctor will order the following tests:

Arterial Blood Gas

Blood is drawn from an artery in your wrist to measure the amount of oxygen in your blood.

Pulse Oximetry

This is a test performed by placing a probe attached to your fingertip, ear lobe or forehead that is attached to an oximeter to indirectly measure the amount of oxygen in your bloodstream – referred to as 'oxygen saturation' – during rest, with exertion, or even during sleep.

You will require supplemental oxygen therapy if your results breathing room air oxygen at rest, with exertion or with sleep are:

- PaO₂ (a measure of oxygen pressure in the blood): < 55 mm HG measured by arterial blood gas
- SaO₂ (a measure of oxygen saturation in the blood): < 88% on room air measured by pulse oximetry

Once your doctor has your test results, a prescription for supplemental oxygen will be written with different recommended amounts while you are resting, exercising, and sleeping, as each level will differ. Oxygen is extremely important to your quality of life, and should be thought of as one of your medicines.

Supplemental oxygen helps increase the amount of oxygen that is available to be transferred from your lungs into the bloodstream, thereby producing more energy to be used by the cells of your body.

PORTABLE AND CONVENIENT

There are many types of oxygen supply systems, and several of the newer types are smaller, lighter, and more efficient – allowing you to benefit from oxygen therapy both at home and while traveling. PF patients may only need supplemental oxygen support during activity or sleep, but patients most often benefit from a continuous oxygen delivery system that provides consistent oxygen saturation levels when in use.

Your pulmonary doctor will help you determine the appropriate amount of supplemental oxygen necessary to support your needs.

Of course, oxygen use also requires heightened awareness of safety issues. For example, oxygen itself is not flammable, but it will support fire. That means that if anything nearby the oxygen source should ignite, it will flame quickly. Consequently, no one should smoke near you while you are on oxygen therapy. In addition, you should keep oxygen 10 feet away from sources of ignition such as gas stoves, candles, fireplaces or space heaters.

Traveling by air with oxygen presents unique challenges for pulmonary fibrosis patients. Please contact your airline prior to making travel reservations to ensure that your oxygen needs can be coordinated. Individual airlines may have different rules and regulations regarding traveling with portable oxygen. Breathin' Easy (www.breathineasy.com, 925-891-5017) also provides guides, referrals, tips and advice on traveling with oxygen.



TALK TO YOUR DOCTOR

Both pulmonary rehabilitation and oxygen therapy require a prescription from your doctor. If you are an individual with PF, make it a point to discuss these options with your physician upon your next office visit. In most instances, both “prescriptions” are covered by insurance, and both can help enhance the quality of your life.

There is absolutely no need to suffer with shortness of breath or dramatically decreased activity when such valuable and effective therapeutic options are available. Be proactive and find out more from your physician. And make the commitment to enjoy every single precious day.



Education. Support. Hope.

Contributing to the CPF allows us to provide educational materials and resources free of charge to those we help. If you have found our materials helpful in your fight with PF, consider donating to the CPF so that we can continue helping thousands of newly diagnosed patients and their families each year.

ADDITIONAL RESOURCES FOR PULMONARY REHABILITATION AND OXYGEN MANAGEMENT

American Association of Cardiovascular and Pulmonary Rehabilitation (AACVPR)

www.aacvpr.org
401 North Michigan Avenue, Suite 2200
Chicago, IL 60611
(312) 321-5146
aacvpr@sba.com

American Association of Respiratory Care (AARC)

www.aarc.org
9425 N. MacArthur Blvd. Suite 100
Irving, TX 75063-4706
(972) 243-2272
info@aarc.org

Breathin' Easy

www.breathineasy.com
4848 Massive Peak Way
Antioch, CA 94531-8310
(925) 891-5017
info@breathineasy.com

National Home Oxygen Patients Association

www.Homeoxygen.org
5454 Wisconsin Avenue, Suite 1270
Chevy Chase, MD 20815-6920



LUNG TRANSPLANTATION:

What Every Patient with Pulmonary Fibrosis (PF) Should Know



WHEN YOUR LUNGS FAIL

For certain individuals (usually 65 years old or younger, although program criteria may differ between transplant centers), lung transplantation may also be an appropriate therapy. Lung transplantation can both extend life and enhance the quality of life.

PF patients should discuss the possibility of lung transplantation as a treatment option with your physician as soon as possible during the course of their care.

The need for transplant evaluation early in an PF patient's disease course stems from the potentially long waiting time on the transplant list combined with the progressive nature of PF. According to United Network for Organ Sharing (UNOS), approximately 30% of PF patients currently listed for transplantation will succumb to their disease prior to transplantation.

"The course of PF is so unpredictable that you never know when you're going to take a turn and need a transplant."

– Steven Nathan, MD, Inova Fairfax Hospital

WHAT IS A LUNG TRANSPLANT?

Lung transplantation is the placement of one (unilateral) or both lungs (bilateral) from a deceased donor into a recipient with end-stage lung disease. The donor lung must be matched for blood type and size to that of the recipient. The decision to transplant one or both lungs is dependent upon the extent of disease and the results of pre-transplant testing.

In a single lung transplant, an incision is made on the side of the chest. The patient's old lung is removed and the new lung is then sewn in, connecting the blood vessels to the lung and from the lung to the heart and the main airway. At the end of the operation, the ribs are brought back together and the incision is closed with layers of stitches. Chest tubes are inserted to drain air, fluid and blood out of the chest for several days to allow the lungs to remain fully re-expanded.

In a double lung transplant, an incision is usually made side-to-side across the middle of the chest just under the nipple line and both chest cavities are entered between the ribs. The lung on one side is removed and the new lung sewn in place. The opposite lung is then removed and the second new lung sewn in place. At the end of the operation, the ribs are brought back together and the incision is closed in layers with stitches.

Controversy exists regarding single versus bilateral lung transplantation for certain lung diseases. Patient outcomes for single or bilateral transplantation should be discussed on an individual basis with the transplant center, in the context of their specific experience performing the procedure.



Quality of life can be moderately to substantially enhanced by lung transplantation, and life may be extended beyond one's life expectancy prior to transplantation.



BENEFITS AND RISKS OF LUNG TRANSPLANTATION

It is impossible to predict how long a patient may survive after transplantation. Current survival rates are as high as 80% at one year following transplantation and 60% at four years. Further, survival statistics may vary between individual transplant centers. Individual institution's survival statistics are available for review at UNOS's web page (www.unos.org). The most critical period for survival of both the patient and the donor lung(s) is the first year after transplantation – when surgical complications, acute rejection (when a patient's immune system recognizes the lung as 'foreign' and attacks it) and infection are the greatest threats to survival. Rejection and infection are the two potential major complications of lung transplantation. Adherence to treatment plan and maintaining a close alliance with the transplant team is essential to minimize these potential complications.

INFECTION

Because you will be taking immunosuppressive medications, your immune system will be less able to fight off invading bacteria, fungi and viruses. You will be much more susceptible to infection, and infections are more likely to become severe.

You have an important role in the prevention of infection by following instructions to avoid exposure and immediately reporting any symptoms of infection to your physician.

REJECTION

Because your transplanted lung(s) is 'foreign' to your body, your body's immune system will try to destroy it, just as it tries to destroy 'foreign' bacteria and viruses when they invade. This process of foreign-tissue rejection helps protect you from illness, but the process has to be "turned off" to keep your transplanted lung(s) from being destroyed.

Immunosuppressive (anti-rejection) medications prescribed by your doctors will help keep the rejection process "turned off." Other medications or an increase in the dosage of your current medications may be necessary to control and treat rejection if your immune system breaks through the immunosuppressive blockade. Following your doctors' orders and taking all medications as prescribed help to prevent or control rejection. However, sometimes rejection can begin despite your adherence to your medications.

The most likely time for rejection to begin is during the first three months after transplantation surgery. Symptoms of rejection are non-specific and might include shortness of breath, changes on your breathing tests or chest X-ray, or reduced oxygen saturation levels, or in rare cases fever, chills and flu-like aches.

Rejection can also occur without any apparent symptoms. That is one of the reasons why regular check-ups are necessary and why some programs may elect to perform "surveillance" fiberoptic bronchoscopy with biopsies of your lung. Your transplant team will instruct you regarding whom to call to immediately report any symptoms.



Did You Know?... that a \$100 gift allows the CPF to provide a print version of our PF Resource Kit, free of charge, to 15 families in the United States who cannot access the Internet? Every gift Counts! Please consider the CPF to be a part of your annual charitable giving plans.



While criteria differs among medical centers, most single lung transplant candidates are 65 years or younger, and double lung transplant candidates are 60 years or younger. They also have an acceptable nutritional status, and a satisfactory psychological profile and support system.

ARE YOU A CANDIDATE?

According to the American College of Chest Physicians, in order to be considered for lung transplantation, you must:

- Have a condition for which transplantation is considered an effective treatment.
- Have severe and progressive lung disease that no longer responds to medical treatment and may be fatal within two years.
- Be willing to accept the risks of surgery and subsequent medical treatment.
- Be physically capable of undergoing surgery and subsequent medical treatment including pulmonary rehabilitation.
- Have an ideal body weight.

There are a number of potential reasons that may preclude patients as appropriate transplant candidates. For example, severe, coexisting medical conditions may be worsened by the surgical procedure or by the powerful immunosuppressive drugs that are necessary after the transplant. In addition, patients who are acutely ill, or who have an unstable clinical status, those with an uncontrolled infection, those who continue to smoke cigarettes, and those with a drug or alcohol dependency may not be good candidates. The specific criteria (e.g. age limitations) and contraindications (e.g. prior cancer) to lung transplantation may vary between different centers. Therefore, motivated patients should consider obtaining a “second opinion consultation” if they are not deemed acceptable at a single center.

HOW THE TRANSPLANT SYSTEM WORKS – AN OVERVIEW

Under contract with a branch of the U.S. Department of Health & Human Services, Health Services & Resources Administration (HRSA), the United Network for Organ Sharing (UNOS) maintains a centralized computer network linking all organ procurement organizations and transplant centers.

In the spring of 2005, UNOS adopted a new policy for the distribution of donor lungs incorporating a Lung Allocation Score (LAS) for transplant candidates. The LAS is a numerical scale, ranging from 0 (less ill) to 100 (gravely ill), that is used for lung candidates age 12 and over. It gives each individual a “score” (number) based on how urgently he or she needs a transplant and the chance of success after a transplant. The number is estimated using lab values, test results and disease diagnosis. If a transplant center determines that a patient is a candidate for a lung transplant, it will add the patient’s medical profile to the national patient waiting list for organ transplant. To view the LAS system and calculator, please visit the UNOS Web site at www.unos.org/resources/frm_LAS_Calculator.asp.

Once a patient is listed, UNOS utilizes a sophisticated database and allocation system to match transplant recipients with available organs. The names and medical profiles of newly-accepted transplant patients are added to the UNOS database and the waiting list automatically updates. When an organ donor becomes available, a transplant coordinator from an organ procurement organization accesses the UNOS computer. Each patient in the “pool” is then matched by computer for the donor characteristics and ranked based on the LAS. The computer then generates a ranked list of patients for each specific organ that can be procured from the donor in accordance with organ allocation policies.

Once a potential organ recipient is selected and contacted, he or she is instructed to quickly and safely make their way to the hospital. After routine pre-operative testing is performed, the transplant then proceeds. This entails a highly coordinated team effort with the timing of events at the recipient hospital dependent on events at the donor hospital.

PATIENT EVALUATION

Before you can qualify for a lung transplant, you will receive an evaluation from a lung transplant team, which typically consists of transplant pulmonologists, transplant surgeons, transplant coordinators, a social worker and a pulmonary rehabilitation specialist.

During the evaluation, you will undergo a number of testing procedures that are used to assess your physical and medical condition. These tests may include:

- Ventilation-perfusion lung scan – This test is designed to determine blood and air supply to the lungs (and requires injection of “tracer” into a vein)
- Pulmonary function tests – These are designed to measure lung capacity. The patient is required to breathe in and out of a tube connected to a measuring device that records various volumes of air within the lung, air flow and the ability of the lungs to take up oxygen
- High Resolution Computer Tomography (HRCT) Scan – This imaging test provides detailed images of the lungs
- Echocardiography – A test designed to evaluate the function of the heart and to estimate the pressure of the pulmonary artery
- Cardiac catheterization – A study of the vessels and measurements of pressures of the heart may be required in some candidates to exclude the presence of coronary artery disease.

All information from tests, interviews and your medical history will be considered in determining whether you are a candidate for lung transplantation. Different transplant centers may have additional criteria for transplantation evaluation.



THE WAITING LIST

Waiting times for organ transplants vary from region to region, and may vary depending on the institution that you choose to perform the procedure. Transplant centers also may differ in their “acceptable criteria” in choosing suitable donors for lung transplantation. Overall, the median wait times for PF patients on the lung transplant list have been reduced substantially since the adoption of the LAS system in May 2005, dropping from an estimated 275 days (for all lung transplant candidates) in 2002 to a median of 69 days (for PF patients) in 2007 (*Source: UNOS*). Due to the severity of their disease, patients with PF generally benefit from the new Lung Allocation System because they achieve a higher disease severity score, potentially reducing their wait time.

As of March, 2008, there were over 2,100 people waiting for a lung transplant, including approximately 390 PF patients. Unfortunately, though, the Organ Procurement and Transplantation Network, estimates there are only enough donor organs to provide about 1,000 lung transplants a year.

More than 80% of adult patients who receive a lung transplant experience no limitations with regard to their activities one to five years post-transplant.

THE TRANSPLANT SURGERY

Once a donor lung becomes available, time is critical. Ideally the lung should be transplanted into the patient receiving the organ within four to six hours. A team of surgeons and anesthesiologists performs an operation to remove the lung from the donor. Additional surgical teams may be present to remove other organs.

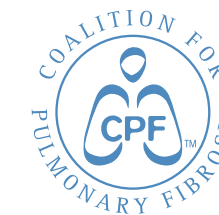
After the lung is removed from the donor, it is preserved and packed for transport. Although the donor is deceased, this procedure is treated like any other operation using standard surgical practices and sterile techniques. Once the operation is complete and the incisions are closed, the donor's body is prepared for burial. Organ donation is considered an incredible gift of life, and the transplant team always treats the donor body with the respect it richly deserves.

Typically, both lungs are removed from the donor together. If the recipient is in need of a double lung transplant, both lungs will be transplanted. Otherwise, the lungs are usually separated after they are removed from the donor and used for two single lung transplant recipients.



In the meantime, the recipient is contacted and then prepared for surgery. Preparation involves administration of general anesthesia, and placement on an artificial breathing machine (e.g. ventilator). Lung transplant surgery may require the use of a "heart-lung machine" to oxygenate blood and maintain adequate blood flow to the recipient's vital organs. The transplant of the lung begins with removal of the diseased lung and the blood vessel attachments. When the lung is placed within the recipient, the blood vessels and airways from the donor lung are connected to the recipient's corresponding blood vessels and airway. Next, the blood flow and airflow are restored. After the transplant is complete, the incision is closed. The patient then recovers in the hospital's intensive care unit.

When a double lung transplant is performed, it is much like two single lung transplants. The lung that is more diseased is transplanted first and then the less diseased lung is transplanted.



Education. Support. Hope.



AFTER TRANSPLANT SURGERY

Following lung transplant surgery, the patient may remain on an artificial breathing machine for the first 12 hours of recovery. However, if the donor lung is functioning properly, the ventilator may be removed at the end of surgery. Depending on their progress, some patients are moved out of the ICU within a few days. Generally, they will also begin eating within the week following surgery. The total hospital stay is usually 10 to 14 days.

Because the recipient's immune system will identify the new organ as foreign, rejection of the transplanted lung is always a possibility. Powerful drugs called immunosuppressants are given starting at the time of lung transplant surgery to try to prevent rejection. Blood tests are done on an ongoing basis post-transplant to confirm that the patient is receiving the correct dosage of medication.

Prior to discharge, the transplant team reviews information with the patient, gives instructions for follow-up care and medications, informs patients of potential side effects and interactions of medications, and answers the patient's questions. A prescribed rehabilitation program will continue at home including physical activity, breathing exercises, nutrition, and the continuation of immunosuppressants and other medications. Signs and symptoms of rejection and infection are also discussed with the patient and family. Frequently, the patients are instructed to buy or are given small breathing machines called microspirometers, to monitor their own breathing capacity at home.



RETURNING TO THE HOME ENVIRONMENT

At-home rehabilitation for lung transplantation is a gradual process, and depends on the individual. The transplant team will give specific instructions. In general, walking is recommended to restore strength and prevent lung complications, but heavy lifting must be avoided for four to six weeks following transplant surgery. Other activities, such as driving, may usually begin when the incisions have healed. Sexual activity can resume when one is comfortable.

Follow-up visits are required for check-ups and begin soon after returning home. Initially, outpatient visits may occur weekly or even more often, and as time progresses the frequency of follow-up visits usually decreases.

As mentioned earlier, current survival rates are an estimated 80% at one year following transplantation and 60% at four years. The most critical period for survival is the first year after transplantation, when patients are at highest risk of surgical complications, rejection and infection.

The functional status of the majority of post-transplant patients is generally improved. More than 80% of adult patients who receive a lung transplant experience no limitations with regard to their activities one to five years post-transplant, while an additional 10-15% require only some assistance with their daily activities. Only a very small proportion of patients (less than 5%) require complete assistance. Patients may return to full or partial employment, and resume participation in sporting activities.

Lung Transplantation is the only treatment option shown to extend the lives of PF patients. Be proactive in discussing this option with your physician – today.



TALK TO YOUR DOCTOR ABOUT LUNG TRANSPLANTATION – TODAY!

It is important to understand that pulmonary fibrosis is a progressive disease for which there is no cure, and that lung transplantation is the only treatment option shown to extend survival for patients with PF. The American College of Chest Physicians (ACCP) provides the following initial questions that patients should ask their physician and/or transplant center:

- Is transplantation my best option for treatment of my condition?
- What are organ and patient survival rates at this institution for the type of transplant I will need?
- How many transplants of this type are done every year at this institution? How many by my physicians and surgeons? How long have they been doing this type of transplant surgery?
- What costs of transplantation and rehabilitation are covered by my insurance? What out-of-pocket costs will I have to pay?
- Am I likely to get a donor lung more quickly if I get wait-listed at more than one medical center, in different parts of the country?

Lung transplantation may be a viable option for patients with PF, but don't wait too long to become educated about the details of this procedure. Become proactive and find out more from your physician. This is a commitment you need to make to yourself – today.

Because of the progressive nature of PF, it is critical for you to discuss lung transplant with your doctor as early as possible to determine if you are a candidate.

RESOURCES FOR TRANSPLANT CANDIDATES AND RECIPIENTS

- Coalition for Pulmonary Fibrosis – www.coalitionforpf.org
- United Network for Organ Sharing (UNOS) – www.unos.org
- U.S. Department of Health and Human Services Organ Donation – www.organdonor.gov
- Second Wind Lung Transplant Association, Inc. – www.2ndwind.org
- American Lung Association – www2.lungusa.org
- Transplant Speakers International, Inc. – www.transplant-speakers.org
- Children's Organ Transplant Association, Inc. – www.cota.org
- National Foundation for Transplants – www.transplants.org
- National Transplant Assistance Fund – www.transplantfund.org
- Medicare Rights Center – www.medicarerights.org

ACKNOWLEDGEMENTS

The CPF is grateful to the following medical professionals for their efforts in peer-reviewing the CPF's educational literature:

- Harold R. Collard, M.D. – University of California, San Francisco (San Francisco, CA)
- Marilyn Glassberg, M.D. – University of Miami/Jackson Memorial Medical Center (Miami, FL)
- Jeffrey Golden, M.D. – University of California, San Francisco (San Francisco, CA)
- Susan Spencer Jacobs, RN, MS – Stanford University Medical Center (Palo Alto, CA)
- Dolly Kervitsky, RCP, CCRC – National Jewish Medical and Research Center (Denver, CO)
- Kathleen Lindell, Ph.D., RN – Dorothy P. and Richard P. Simmons Center for Interstitial Lung Disease at University of Pittsburgh (Pittsburgh, PA)
- Marvin I. Schwarz, M.D. – University of Colorado Health Sciences Center (Denver, CO)
- Timothy Whelan, M.D. – University of Minnesota Lung Center (Minneapolis, MN)

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Information and data from the following organizations and medical publications were referenced by the authors in developing the educational materials herein:

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LUNG TRANSPLANT

Web pages:

- United Network for Organ Sharing
- The American College of Chest Physicians
- The American Society of Transplantation
- Duke University Medical Center & Duke Transplant Center
- Inova Fairfax Lung Transplant Program
- University of Pennsylvania Medical Center
- Tampa General Hospital Transplant Center
- The Mayo Clinic
- Second Wind Lung Transplant Association

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The Coalition for Pulmonary Fibrosis (CPF) accelerates research efforts leading to a cure for pulmonary fibrosis (PF), while educating, supporting, and advocating for the community of patients, families, and medical professionals fighting this disease. Since its inception in 2001, the CPF has worked with thousands of patients and families fighting PF, built a national network dedicated to assisting patients and physicians, served as a national voice for all PF patients and their needs, and funded promising research to better treat - and ultimately cure - the disease.

Generous public support allows the CPF to connect with patients and families – daily – who need information, referrals, and support; to form critical partnerships with PF research centers to identify and fund new discoveries about PF, improve public and physician awareness of the disease, advocate on behalf of the PF community in Washington D.C. Perhaps most important, your generosity allows us to directly fund research, giving hope to thousands of patients that progress is being made. For more information please visit www.coalitionforpf.org or call (888) 222-8541.

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- Raise awareness in your community about PF (ask us how)
- Advocate on behalf of thousands of patients who also suffer from PF
- Access our website at www.coalitionforpf.org
 - ...for patient education and updated information on research efforts
 - ...to learn about how you can be an advocate for all PF patients, and
 - ...to contribute to our cause and help advance the critical mission of the CPF.

CPF membership is free! As a member, you will be updated on latest research, patient experiences, advocacy opportunities, clinical trial and emerging research information, and new educational materials. You'll also receive our Action Alert quarterly newsletter. And please ask your friends, caregivers and doctors to join as well – the more members we have, the more informed and empowered our community will be! Just fill out the attached postcard or call (888) 222-8541 or email us at info@coalitionforpf.org. We look forward to hearing from you!



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