

Idiopathic Pulmonary Fibrosis and You

Patient Education Guide



AMERICAN
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FOUNDATION

Idiopathic Pulmonary Fibrosis and You

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The American College of Chest Physicians is the leading resource for the improvement of cardiopulmonary health and critical care worldwide. Its mission is to promote the prevention and treatment of diseases of the chest through leadership, education, research, and communication.

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What Is Idiopathic Pulmonary Fibrosis?

Idiopathic pulmonary fibrosis (IPF) is a buildup of scar tissue in the lungs. This scar tissue damages the lungs and makes it hard for oxygen to get in. Not getting enough oxygen to the body can cause serious health problems and even death.

“Idiopathic” is the term used when no cause for the scarring can be found. In these cases, doctors think the scarring starts by something that injures the lung. Scar tissue builds up as the lungs try to repair the injury. In time, so much scarring forms that patients have problems breathing.

IPF usually worsens over time. However, while some patients get sick quickly, others may not feel sick for years. There is no cure for IPF, but there are treatments that may be able to slow down the lung scarring.

If you have been diagnosed with IPF, this information is for you. Understanding your condition will go a long way to help you cope with the effects it has on your body. Read on to find out more.

Symptoms of IPF

The two major symptoms of IPF are shortness of breath and cough. Other symptoms may include:

- Fatigue and weakness
- Chest pain or tightness in the chest
- Loss of appetite
- Rapid weight loss



What Are the Causes of IPF?

The causes of IPF are unknown. There are other conditions that cause lung scarring. Lung scarring that is the result of other conditions is often called “pulmonary fibrosis” but should be called by the name of the cause. These other causes include the following:

- Diseases, like rheumatoid arthritis and sarcoidosis.
- Medicines, such as those used for certain heart conditions. (For more information on drugs that cause lung diseases, visit www.pneumotox.com.)
- Breathing in mineral dusts, such as asbestos or silica.
- Allergies or overexposure to dusts, animals, or molds. There are many names for this condition, such as “bird breeder’s lung,” “farmer’s lung,” or “humidifier lung.” These conditions are called hypersensitivity pneumonitis.



The job of your lung specialist (pulmonologist) is to determine if you have IPF or one of these other diseases. A more detailed list of the diseases your pulmonologist might test for is available at the American College of Chest Physicians (ACCP) Interstitial and Diffuse Lung Disease NetWork Web page (www.chestnet.org/networks/lung_disease).

Who Gets IPF?

Five million people worldwide have IPF, and it is estimated that up to 200,000 people in the United States have this condition. It usually occurs in adults between 40 and 90 years of age and is seen more often in men than in women. Although rare, IPF can run in families.

How Is IPF Diagnosed?

Patients with any symptoms of IPF should see a pulmonologist to rule out similar conditions. The doctor will use a number of tests, including:

- Breathing tests: to measure how well your lungs are working.
- CT scan: to get a detailed image of your lungs, and to see if scarring has started.
- Blood tests: to see if you have an infection, problems with your immune system, or to see how much oxygen is in your blood.
- Bronchoscopy: to test a small sample of lung tissue. A tube is inserted through the nose or mouth into the lung. A light on the end of the tube lets the doctor see where to go. The doctor then takes a small piece of lung tissue to be tested (this is called a biopsy). You usually do not need to stay overnight in the hospital to have this done.
- Thoracoscopic biopsy: to obtain larger tissue samples. This is a surgical procedure in which small incisions are made in between the ribs. It usually requires a hospital stay and general anesthesia.



Treatment for IPF

Once lung scarring forms, it cannot be removed surgically. Also, at this time, there are no medications that remove lung scarring. However, there are treatments, such as the ones that follow, that may be able to help.

Smoking Cessation

Cigarette smoke not only damages the lining of the lungs, it can also make you more likely to get a lung infection. While some studies suggest that patients with IPF who smoke actually live longer, these studies are not accepted by everyone, and most experts agree that you should stop smoking.



Supplemental Oxygen



As lung scarring gets worse, many patients need extra oxygen to help them go about their daily lives without getting too out of breath. You get this oxygen from a tank that you carry around with you. In later stages of IPF, oxygen may be needed even while sleeping or resting.

Oxygen is not addictive, so you do not have to worry about using it too much. To help maintain your oxygen levels, ask your doctor about a small, easy-to-use device called a pulse oximeter. This device helps you to know just how much oxygen flow you need, especially during activity.

If you use oxygen, you may qualify for a discount on your electric bill. Contact your electric company for more information. Also, to prepare for a power failure, be sure to register as a “priority” with the electric company, so that your power will go back on as soon as possible. In some areas, you are required to register with your emergency medical services in case of an emergency.

Exercise

Regular exercise can help patients with IPF. Staying in shape not only keeps your breathing muscles strong, it also gives you more energy. This is because healthy muscles need less oxygen to perform work. Many communities have exercise centers (usually associated with hospitals) that have programs especially planned for patients with lung conditions.



Nutrition

Many patients with IPF lose weight because of their disease. If you lose too much weight, your breathing muscles can become weak. You also may not be able to fight off infections very well. A well-balanced diet is important to keep up your strength. Be wary of supplements and other nutrition treatments that claim to improve IPF.

Medications

There is a variety of drugs used to treat IPF. They are often given for long periods of time (3 to 6 months or longer), and regular check-ups may be needed to see how well they are working. The following are examples of some of the medications used for IPF:

- **Corticosteroids.** This is a common treatment for many lung conditions. However, corticosteroids have many side effects and do not usually work well for patients with IPF. In fact, they may do more harm than good. Most experts agree that these drugs should only be used for patients who have had improvement while taking them.

- Cytotoxic and immune suppressing drugs. These medicines are sometimes used for cancer and transplantation patients. Bone marrow problems may occur, and white blood cell counts must be watched closely to avoid levels becoming too low. How well these drugs work for IPF has never been well established.
- Other medications. Colchicine (used to treat gout) may help stop scar tissue from forming; however, it has not been shown to be better than the more commonly used drugs. Another drug, called N-acetyl cysteine (an antioxidant), may have some benefit, but experience with the drug is limited, and more studies are needed. Many experts think that acid reflux disease (when stomach acid backs up into the throat) is an important factor in IPF. Patients with this condition should take medicines to help control it.
- Experimental treatments and clinical trials. Many FDA-approved studies, called clinical trials, are being performed to find new medications for IPF. In a clinical trial, a new drug is compared to a “placebo” (an inactive medicine, such as a sugar pill). Participants do not know which treatment they are getting, the experimental medicine or the placebo. Clinical trials are important, because they can speed the discovery of new drugs for IPF. Talk to your doctor about information on new treatments for IPF.

Lung Transplant

For some patients with severe IPF, the only way to improve their quality of life or increase their survival may be a lung transplant. However, the decision to have a lung transplant must be made carefully. On average, only 50% of patients live for 5 years after a lung transplant. Not all patients qualify for lung transplantation, depending on age or other medical problems. Talk to your doctor to find out more.

Specific Issues

Lung Infections

Due to lung damage, lung infections can cause serious problems for patients with IPF.

Call your doctor right away if you notice the following symptoms:

- Worsened cough
- A change in the color or amount of sputum you produce
- Fever
- Chills



Your doctor will prescribe an antibiotic if you have a bacterial lung infection.

To avoid other infections and illnesses, you should also get the pneumococcal pneumonia vaccine and a yearly flu shot. These vaccinations may help keep you from becoming severely ill. Talk to your doctor about how and when to get these shots.

Coping With Cough

A nagging, dry cough is one of the most common complaints of patients with IPF. Although cough is often caused by IPF, there are other things that can make it worse. These include the following:



- Postnasal drip
- Acid reflux disease
- Allergies and pollutants
- Cigarette smoke
- Some medications, such as beta-blockers or angiotensin-converting enzyme (ACE) inhibitors, for blood pressure control

Nasal sprays, cough medicines, and cold remedies may help control cough. Ask your doctor if your cough is treatable.

Leg Swelling

Patients with IPF may develop swelling in their legs. Lung scarring makes it difficult for the heart to pump blood through the lungs. When this happens, fluid can back up, causing the legs to swell. This is called right-sided heart failure and can get worse if blood oxygen levels are too low. If you have swelling in your legs, see your doctor right away.



Sexuality

Because sexual intercourse requires more energy, patients with IPF may have problems with shortness of breath during sex. The following tips may help:

- Choose a time for intercourse when you are relaxed and rested; rushing things uses up more energy.
- Try positions that place less stress on the heart and lungs, such as side by side or on your back.
- Avoid sexual intercourse after a heavy meal.
- Use oxygen if it has been prescribed for you.
- Most importantly, talk to your partner about your concerns. Work together to find the most comfortable way for both of you to enjoy having sexual intercourse.

Travel

A loss of oxygen pressure during air travel often makes flying very difficult for patients with IPF. If you are planning to fly, your doctor may be able to test you to see if you need oxygen during your flight.

The airline rules for oxygen use often change. Contact the airline, and let them know that you will need oxygen during the flight (they may charge you a fee for this). Also, make sure you can get oxygen, should you need it, when you get off the plane, particularly if it is in an area of higher altitude.



Life Support

If your lung disease becomes very severe, you may need life support (ventilation) to keep you alive. Newer forms of mask ventilation, called bilevel pressure ventilation (BiPAP), may help for a short time. However, most patients with IPF, who are placed on a ventilator, cannot be taken off. You may need this type of ventilation for the rest of your life. It is up to you to decide if you want this treatment.

End-of-Life Issues

Talking about end-of-life issues is not easy. However, it is important that you let your family and your doctor know how you feel. There are legal forms that you can fill out that state what treatments you want or do not want, should you become too ill to make your wishes known. You should also choose someone as a power of attorney. This person can make decisions about your treatment should you become so ill that you are unable to speak for yourself.

Information Brings Hope

When patients are diagnosed with IPF, they are understandably worried and upset. Keep in mind that there are treatments available that can help you, and you should not consider your situation hopeless. Also, knowing more about the condition can help you understand and cope with this disease. We hope that the information in this booklet will help. You may want to share it with your loved ones so that they have a better understanding of what you are going through. Having a good support system will help you stay positive and maintain your quality of life. For more information about IPF, see below.

For More Information

American Lung Association

61 Broadway, 6th Floor
New York, NY 10006
(800) LUNGUSA or (212) 315-8700
www.lungusa.org

American Thoracic Society

61 Broadway
New York, NY 10006-2755
(212) 315-8600
www.thoracic.org

Pulmonary Fibrosis Foundation

1440 West Washington Blvd.
Chicago, IL 60607
(312) 377-6895
www.pulmonaryfibrosis.org

Coalition for Pulmonary Fibrosis

1685 Branham Lane, Suite 227
San Jose, CA 95118
(888) 222-8541
www.coalitionforpf.org

National Heart, Lung, and Blood Institute

PO Box 30105
Bethesda, MD 20824-0105
(301) 592-8573
www.nhlbi.nih.gov

National Organization for Rare Disorders

55 Kenosia Avenue, PO Box 1968
Danbury, CT 06813-1968
(800) 999-6673
www.rarediseases.org

International Society for Heart and Lung Transplantation

14673 Midway Road, Suite 200
Addison, TX 75001
(972) 490-9495
www.ishlt.org

Other Useful References

- Idiopathic pulmonary fibrosis: proceedings of the 1st annual Pittsburgh International Lung Conference. October 2002. *American Journal of Respiratory Cell and Molecular Biology* 2003; 29(suppl):S1-S105
- American Thoracic Society. Idiopathic pulmonary fibrosis: diagnosis and treatment. International consensus statement. *American Journal of Respiratory and Critical Care Medicine* 2000; 161:646-664





About the author

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Dedication

Dedicated in memory of Dr. Henry Vyskocil. Dr. Henry Vyskocil, the father of Dr. James J. Vyskocil, died of idiopathic pulmonary fibrosis in 1994, the year his son began his pulmonary specialty practice.

**Other patient education guides available from the ACCP
in print and on the ACCP Web site (www.chestnet.org) are:**

Controlling Your Asthma (English and Spanish versions available)

Flexible Bronchoscopy

Cough: Understanding and Treating a Problem With Many Causes

Mechanical Ventilation: Beyond the ICU

Pulmonary Rehabilitation: A Team Approach To Improving Quality of Life

Living Well With COPD

Additional Resources:

www.thoracic.org

www.yourlunghealth.org

www.goldcopd.com

www.copdresourcenetwork.org

www.nationaljewish.org

www.lungusa.org

www.smokefree.gov/talk.html

www.centerwatch.com

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