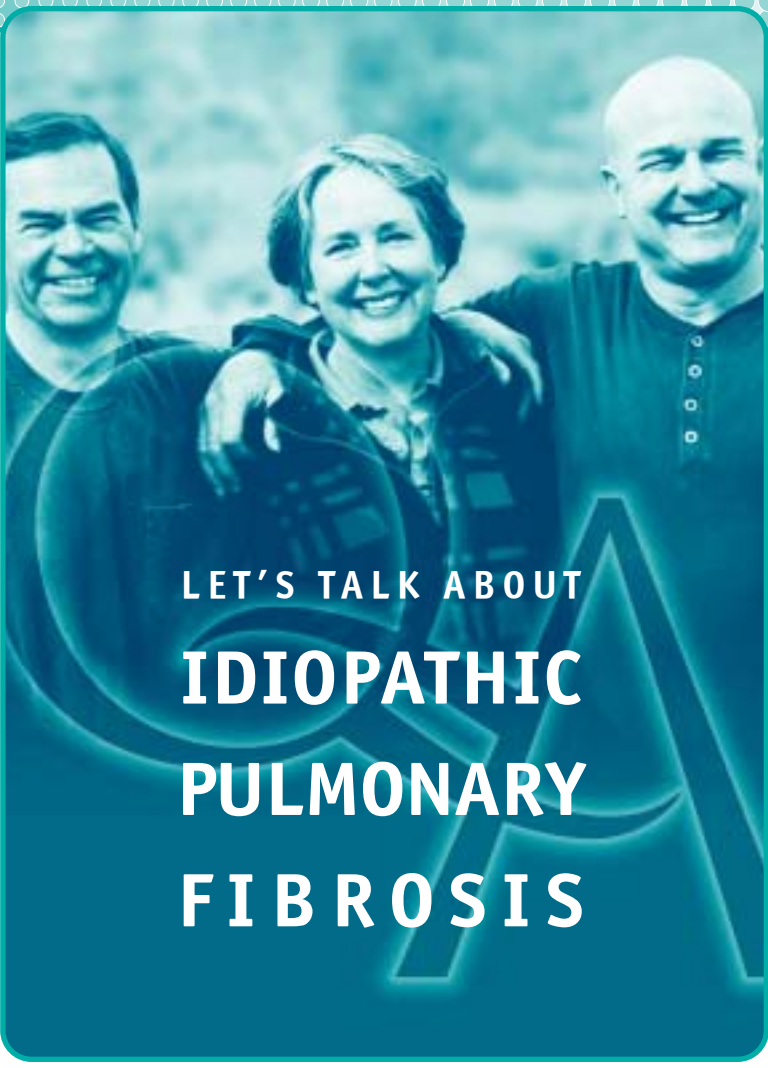

LET'S TALK ABOUT

IDIOPATHIC PULMONARY FIBROSIS

Questions and Answers for Patients and Caregivers

COALITION FOR PULMONARY FIBROSIS

CPF™

Education. Support. Hope.

This brochure is designed to provide useful information to patients newly diagnosed with idiopathic pulmonary fibrosis—with the goal of helping them and their caregivers better understand and manage this potentially life-threatening condition. The information supplied here is presented in the form of answers to commonly asked questions regarding the disease, and its symptoms, diagnosis, and treatment.

WHAT IS IDIOPATHIC PULMONARY FIBROSIS?

Idiopathic pulmonary fibrosis (*IPF*) is a debilitating disease—marked by progressive scarring of the lungs—that gradually interferes with a person's ability to breathe. IPF belongs to a family of approximately 200 related diseases, called *interstitial lung diseases* (*ILDs*), that have similar characteristics and can result in scarring. The lung scarring, a condition typical of these disorders, is referred to as *pulmonary fibrosis* (*PF*).

Sometimes PF can be linked to a particular cause, such as exposure to metal dust, wood dust, gases, or fumes; chemotherapy or radiation therapy; residual infection; or a connective tissue disease, such as systemic lupus erythematosus or rheumatoid arthritis. In the majority of PF cases, however, no known cause can be estab-

lished. When pulmonary fibrosis has no known cause, it is called "idiopathic pulmonary fibrosis" or "IPF." The word "idiopathic" means "of unknown cause."

Your doctor may have referred to your condition by one of a number of names that are sometimes used to describe IPF. This confusion may be due, in part, to similarities between IPF and the other ILDs. Only recently has IPF been recognized as a distinct clinical disorder.

Approximately 83,000 people in the United States have IPF, and prevalence is on the rise with an estimated 15,000 new cases developing each year. The disease tends to affect men more than women and usually strikes people between the ages of 50 and 70.

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

WHAT CAUSES IPF?

While the cause of IPF remains a mystery, what we do know is that IPF involves changes in the lung's normal healing process. IPF's chronic cycle of injury leads to an exaggerated or uncontrolled healing response that, over time, produces fibrous scar tissue. 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 appears to play a major role. Researchers are investigating a number of potential risk factors that may make a person more likely to develop IPF. These risks may include:

- Occupational exposure to metal dust or wood dust
- Cigarette smoking
- Viral infection
- A family history of pulmonary fibrosis

WHAT SYMPTOMS MIGHT YOU EXPERIENCE?

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

- Shortness of breath during or after physical activity
- Spasmodic, dry cough
- Weight loss and fatigue

Patients may also develop a buildup of tissue in their fingertips. This condition is called *clubbing*.

HOW IS IPF DIAGNOSED?

Because IPF has symptoms and an underlying pattern of scarring similar to those of other lung disorders, it can be difficult to diagnose. In fact, in most cases, identifying IPF is usually a matter of ruling out other diseases. Also, until recently, the medical community had no agreed-upon standards for the diagnosis of IPF. Consequently, other related diseases may have been mistakenly classified as IPF. With new diagnostic standards now in place, the recognition and management of IPF should be substantially improved.

To diagnose IPF, 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 Procedure	*Description*	*Purpose*
Chest imaging	Use of radiologic machines to take pictures (x-ray or CT scan) of the lungs	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 test	A measurement of oxygen levels in blood taken from an artery	To determine how well the lungs are performing vital gas exchange
Exercise test	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
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 look for signs of inflammation in the lungs
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 between the ribs (open-lung biopsy)	To obtain a sample of lung tissue for direct examination

WHAT MAY HAPPEN AS IPF PROGRESSES?

IPF 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 IPF makes it difficult for a person to fight infection.
- IPF 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*).
- IPF 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 IPF TREATED?

Current treatments are intended to improve symptoms and slow disease progression. As yet, there is no cure for IPF. A panel of experts sponsored by the American Thoracic Society and the European Respiratory Society has developed guidelines for the treatment of IPF. This panel recommends the following as standard IPF therapy:

- An anti-inflammatory corticosteroid (such as prednisone)

used in combination with:

- A drug to suppress the body's immune response (azathioprine or cyclophosphamide)

This treatment approach is effective in only a small number of cases and tends to work best when started early in the course of the disease. These drugs can also cause side effects—some minor and some more serious. Be sure to contact your doctor or nurse if you have any negative reactions to your medications.

In some cases, doctors may consider lung transplantation. This procedure is most often performed in patients under 60 years of age whose IPF has not responded to other treatments.

New medications to treat IPF are under investigation.

WHAT CAN YOU DO?

If you have been diagnosed with IPF, 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 IPF.

- Take all prescribed medications as instructed by your doctor.
- 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.

- Eat a well-balanced diet. 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.
- Try some moderate exercise, such as walking or riding a stationary bicycle. If you're already exercising, keep up your regular workout. This helps you maintain strength and lung function. Talk to your doctor before starting a new exercise program. Those who need to can generally use oxygen during this kind of activity.
- Consider enrolling in a pulmonary rehabilitation program to help increase your strength, learn breathing techniques, and expand your social support network. Ask your doctor or nurse for more details.
- 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.

- Join a support group in your community—or start one of your own. It's a great way to get the emotional support you need.
- 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.

ARE THERE ANY NEW TREATMENTS ON THE HORIZON?

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

- *Antifibrotic* or *antifibrogenic agents* (such as interferon and certain blood-pressure–lowering medications) to suppress the scarring process
- *Antioxidants* (such as N-acetylcysteine and glutathione) to prevent damage to lung tissue
- *Monoclonal antibodies* to inhibit "bad" *cytokines* (protein growth factors that activate inflammation)

A promising new approach to treatment involves the cytokine *interferon gamma-1b*. An advanced clinical study is currently under way to confirm its effectiveness in slowing or reversing the scarring associated with IPF and in potentially improving lung function and patient survival.

Further research is needed to determine the effectiveness and safety of these new therapies for IPF. If you have any questions about experimental treatments or ongoing clinical trials, ask your doctor, contact the Coalition for Pulmonary Fibrosis, or visit www.coalitionforpf.org/patient/ipfresearch.asp for an up-to-date listing of active investigational research.

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.

PATIENT SUPPORT ORGANIZATIONS

Coalition for Pulmonary Fibrosis (CPF)

The Coalition for Pulmonary Fibrosis (CPF) is a 501 (c) (3) non profit organization, founded in 2001 to further education, patient support and research efforts for pulmonary fibrosis, specifically idiopathic pulmonary fibrosis (IPF). The CPF is governed by the nation's leading pulmonologists, individuals affected by pulmonary fibrosis, medical research professionals and advocacy organizations. The CPF's partners include the Mary D. Harris Foundation, The Pulmonary Paper, the Pulmonary Fibrosis Association, the Caring Voice Coalition, Second Wing Lung Transplant Association, and over 20 leading medical and research centers nationwide.

Coalition for Pulmonary Fibrosis (CPF)
1685 Branham Lane, Suite 227
San Jose, CA 95118
(888) 222-8541 *www.coalitionforpf.org*

Caring Voice Coalition

Caring Voice Coalition is dedicated to building relationships with charitable organizations founded to help individuals and families affected by serious chronic disorders and diseases including IPF.

Caring Voice Coalition
POB 1384
Meridian, ID 83680
(877) 455-3374 *www.caringvoice.org*

Mary D. Harris Memorial Foundation

This nonprofit organization supports efforts to find a cure for pulmonary fibrosis as well as educational initiatives that help to improve the lives of those living with the disease.

Mary D. Harris Memorial Foundation
1500 Ashbury Street
Evanston, IL 60201
(847) 869-5276

Pulmonary Fibrosis Association (2001-2003)

The Pulmonary Fibrosis Association ceased operations in June, 2003. Services previously offered by the PFA are now available through the CPF.

The Pulmonary Paper

This nonprofit organization publishes a newsletter with the latest information on respiratory care and products for people with chronic lung problems. For more information, contact:

The Pulmonary Paper
P.O. Box 877
Ormond Beach, FL 32175
(800) 950-3698 *www.pulmonarypaper.org*

Second Wind Lung Transplant Association

This nonprofit organization publishes a newsletter with the latest information on respiratory care and products for people with chronic lung problems. For more information, contact:

Second Wind Lung Transplant Association
300 South Duncan Avenue, Suite 227
Clearwater, Florida 33755-6457
888-855-9463 *www.2ndwind.org*

The CPF also partners with leading advocacy groups, medical and research centers, and other organizations committed to helping the IPF community. For a complete list of CPF partners, please visit **www.coalitionforpf.org/aboutus/recognition.asp**

RECOMMENDED READING

The following books offer information on lung disorders, and are available through the CPF web page at www.coalitionforpf.org/patient/resources.asp:

The Breathing Disorders Sourcebook
By F.V. Adams, MD

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
J.N. Parker & P. Parker

Share the Care: How to Organize a Group to Care for Someone Who Is Seriously Ill
C. Capossela & S. Warnock

Provided by

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The CPF relies on the contributions of individuals, corporations, and associations who share our commitment to improving IPF awareness and education. To learn more, or to make a contribution, please call (888) 222-8541.