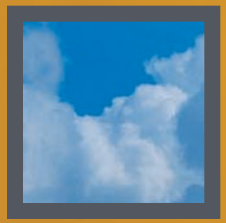

PILOT™

Pulmonary Fibrosis Identification:
Lessons for Optimizing Treatment

# IDIOPATHIC PULMONARY FIBROSIS: PATIENT INFORMATION BROCHURE

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# PATIENT INFORMATION

## CONTENTS

## UNDERSTANDING IPF

### What is Pulmonary Fibrosis?

Pulmonary fibrosis is the progressive scarring of the lungs that occurs when air sacs gradually become replaced by fibrotic (scar) tissue. As the scar tissue becomes thicker, it leads to "stiffness" in the lungs, making it difficult to breathe. Pulmonary fibrosis is found in over 200 lung disorders, so it is important for your health care provider to identify the cause of the fibrosis because different types of fibrosis respond to different treatments.

### What is Idiopathic Pulmonary Fibrosis?

Idiopathic pulmonary fibrosis (IPF), a specific type of pulmonary fibrosis, is a progressive and debilitating lung disease. In some patients, the development of scarring occurs quickly, while in others it happens over a longer period of time. Though IPF usually progresses slowly, episodes of rapid deterioration called acute exacerbations can occur. There are currently about 80,000 people in the United States with IPF and approximately 30,000 new cases will be diagnosed this year. IPF typically occurs between the ages of 40 and 70 years, and affects men more often than women. Patients with IPF typically survive only 3 to 4 years after diagnosis.

### What causes IPF?

Idiopathic means "of unknown cause." The current understanding of IPF involves a change in the lung's normal healing process. It is believed that recurrent injuries to the lungs of patients with IPF result in an exaggerated healing cycle in which extra scar tissue is produced.

While no causative factor has been established for IPF, some of the risk factors associated with the disease include:

- Genetics—10% to 15% of the cases occur in patients who have family members with IPF

## UNDERSTANDING IPF

### The Main Parts of Your Lungs

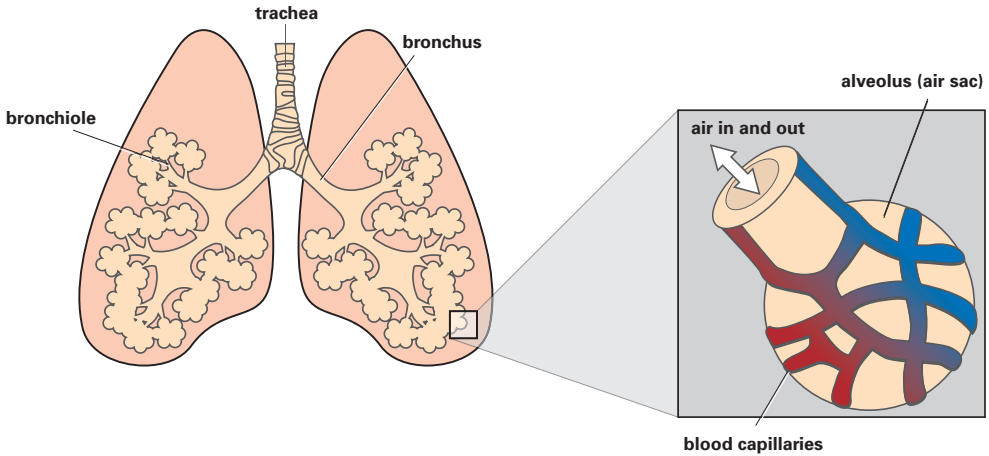



***Figure 1.*** **With pulmonary fibrosis, the air sacs become thicker and stiffer, which in turn makes it difficult to breathe**

## UNDERSTANDING IPF

- Cigarette smoking
- Work-related exposure to dusty environments, especially wood or metal dust
- Medications—certain medications may cause pulmonary fibrosis, you should speak to your doctor for additional information
- Viral infections
- Acid reflux disease or gastroesophageal reflux disease (GERD)

### What are the symptoms of IPF?

The early symptoms of IPF typically include:

- Cough: usually dry and not producing phlegm or mucus
- Shortness of breath with physical exertion
- Sometimes weight loss and fatigue

The late symptoms of IPF may include:

- Shortness of breath while at rest, which may begin to affect eating or talking on the phone
- Clubbing: about half of patients will also develop clubbing, an enlargement of the fingertips

It is important to remember that IPF progresses differently in each patient, and although some patients advance quickly, others remain stable for longer periods of time.

### How is IPF diagnosed?

A number of different tests can be performed by your physician to help diagnose IPF:

- **Blood tests** will assist in ruling out other diseases
- **Pulmonary function tests** measure your breathing capacity, determine the "stiffness" of your lungs, and calculate the amount of oxygen that travels from your lungs to your bloodstream

## UNDERSTANDING IPF

- **Chest X-ray and CT scan** help to identify the pattern of scarring in the lungs
- **Lung biopsy** for diagnosis, in which small pieces of lung tissue are removed by a chest surgeon while you are under anesthesia and analyzed under a microscope

An accurate diagnosis of IPF is achieved on the basis of clinical and radiologic criteria, and by ruling out other diseases. In some cases when the diagnosis is not clear, microscopic examination of small samples from your lung (biopsy) may be necessary.

### Why is IPF difficult to diagnose?

An accurate diagnosis of IPF is achieved on the basis of clinical and radiologic criteria, and by ruling out other diseases. Since the cause of IPF is unknown, a valuable clue is missing and the diagnosis depends on tests and elimination of other causes. If you have "typical IPF", your doctor may be confident of the diagnosis based on your clinical results and radiology. However, the disease is heterogeneous, and doesn't always have the same features.

Diagnosis may also be difficult because the symptoms of IPF, such as cough and shortness of breath, are common symptoms of other diseases including asthma, COPD, and congestive heart failure. In addition, these symptoms can be mild and progress slowly.

### Why is correct diagnosis important?

Your doctor will explain that there is no proven therapy for IPF. However, accurate diagnosis is important for several reasons. Other conditions with similar symptoms can be effectively managed, so it is critical to identify

which condition you are suffering. Also, clinical trials for IPF are available for participation, but these depend on accurate diagnosis.

### How will I be monitored?

Most pulmonologists like to evaluate their patients with IPF every 3 to 6 months, with breathing and physical exertion tests. These may include spirometry (measurement of breath), gas diffusion (efficiency of absorption from air), and a 6-minute walk test. Your health care provider is likely to assess your quality of life with a questionnaire and determine your oxygen requirement. Also, other conditions such as heart disease, psychological well-being, and sleep disturbance may be evaluated.

### How is IPF managed?

There is **no FDA-approved medication for the treatment of IPF**, but several strategies can be helpful for improving your quality of life.

If you smoke, it is important to stop. **Smoking** increases the risk of respiratory infection, heart disease, and lung cancer. Quitting is not easy, but several strategies can be used. Counseling helps manage your lifestyle patterns and gives you tools for coping with the difficult process. Some patients find nicotine replacement, in the form of gum, inhaler, nasal spray, patch, tablet, or lozenge to be helpful. Others use pharmacologic agents such as bupropion, nortriptyline, or varenicline. Your health care provider can help find the best process for you.

**Sleep disturbances and GERD** are common in patients with IPF. A family member or caregiver can observe your sleep, noting frequent awakenings or breathing irregularities. Your doctor may want to do a more formal

sleep study. GERD is common in aging people, and is due to acid from the stomach inappropriately coming up the throat. If this acid is aspirated into the lung it can contribute to tissue damage. Speak with your doctor if you wake in the night with a sour taste in your mouth.

A treatment with wide support is **pulmonary rehabilitation** (PR). It is a personalized exercise program that improves your energy, strength, and endurance. Many patients report that a PR program helps them feel better and have a better outlook. The program is structured and monitored by health care professionals who provide nutritional advice, breathing techniques, information about your disease, and emotional support.

There are also treatments for your IPF symptoms. **Home oxygen therapy** may be used for patients with difficulty breathing. If oxygen is prescribed, it is important that you use it as directed by your doctor. Cough medicine may help reduce the severity of your cough, but it will not cure it.

Some patients believe that when a disease such as IPF is diagnosed that normal life comes to a halt. This is not necessarily so. **Recreational activities** bring pleasure and give rhythm to life. Socializing, cultural activities, family events, sexual activity, and especially exercise should be continued to the extent you can manage them. Enjoy life!

You may be eligible to participate in **clinical trials** for emerging therapies for IPF. Participants typically receive a high standard of care and some will receive medications that may help the disease. Depending on

your location, a research center may be conveniently located. Ask your doctor about trials that may be enrolling, where they are being performed, and the inclusion criteria.

One treatment that has been shown to extend the lives of patients with IPF is **lung transplantation** (LT). It is recommended that you undergo evaluation for LT upon diagnosis. The process takes time, and because of the unpredictability of acute exacerbations, it is best to be prepared if LT becomes the best option. Patients are selected as candidates based on the urgency of their condition as well as the benefit after transplantation. Your doctor can explain the lung allocation system and whether LT fits into your management plan.

There are many different **support groups** for patients living with IPF and their families. These organizations provide current IPF information, and also assist in managing problems such as depression and preparing for end-stage pulmonary fibrosis. Although it is a very difficult and emotional topic, it is important for you to talk with your family and your health care provider about your wishes regarding end-of-life issues. These include instructions for life-support measures as well as your will. Both your health care provider and support organizations can help you and your family through this difficult time. A list of support groups is provided in the "Patient Resources" section.

Good **eating habits** can make a big difference for your breathing. Limiting your intake of fatty foods and eating smaller meals may make breathing easier.

Your **emotional health** is a critical component that frequently suffers with serious diseases such as IPF. Recognize that you are at risk for stress, anxiety, and depression; but also that your health care provider can offer help in coping with these conditions. Sufficient sleep and good relaxation are good practices that can help you feel better.

## Conclusions

IPF is a serious, life-threatening condition. However, there are positive steps that you can take to maximize your daily functioning and your enjoyment of life. Work with your doctor or health care provider to design a management plan that fits your goals.

## Web resources:

**PILOT**
http://www.pilotforipf.org

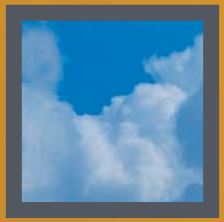
**Pulmonary Fibrosis Foundation**
http://www.pulmonaryfibrosis.org

**Coalition for Pulmonary Fibrosis**
http://www.coalitionforpf.org

**American Lung Association**
http://www.lungusa.org

**Aging with Dignity**
http://www.agingwithdignity.org

**National Hospice and Palliative Care Organization**
http://www.nhpco.org



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