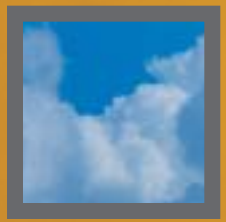

PILOT™

Pulmonary Fibrosis Identification:
Lessons for Optimizing Treatment

IDIOPATHIC PULMONARY FIBROSIS: PATIENT INFORMATION BROCHURE

Supported by an unrestricted educational grant from

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PATIENT INFORMATION

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UNDERSTANDING IPF

What is Pulmonary Fibrosis?

Pulmonary fibrosis is the progressive scarring of the lungs that occurs when air sacs known as alveoli (see figure 1) gradually become replaced by fibrotic tissue or 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 IPF?

Idiopathic pulmonary fibrosis (IPF), a specific type of pulmonary fibrosis, is a progressive and debilitating lung disease. It is characterized by scarring of the lung that worsens over time, causing a stiffness of the lungs, which makes it difficult to breathe. In some patients, the development of scarring occurs quickly, while in others it happens over a longer period of time. 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.

What causes IPF?

Idiopathic means "unknown," and although the exact cause of IPF is not known, it is understood that IPF involves a change in the lung's normal healing process. It is believed that recurrent injuries to the lungs of the patients with IPF result in an exaggerated healing cycle in which extra scar tissue is produced. Over time, the scar tissue causes hardening in the lungs, which makes it difficult to breathe. Potential risk factors associated with the disease have also been identified.

The Main Parts of Your Lungs

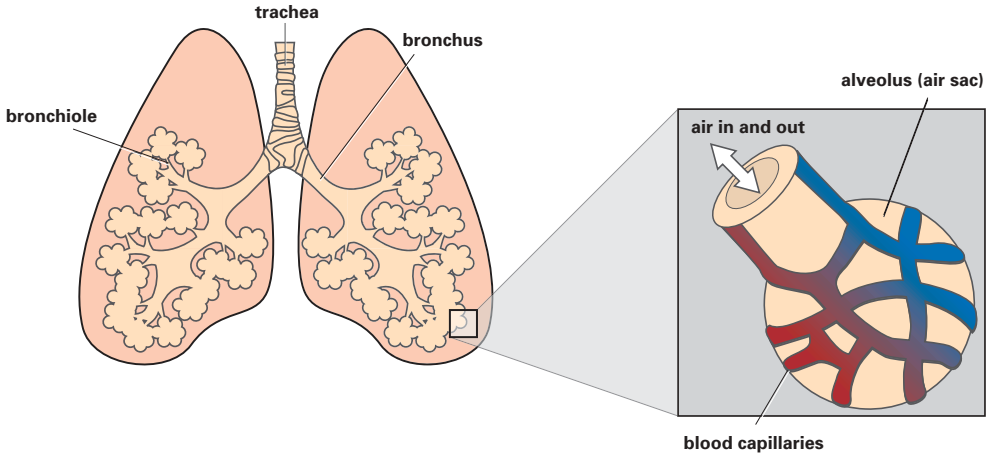


Figure 1. **With pulmonary fibrosis, the air sacs become thicker and stiffer, which in turn makes it difficult to breathe**

UNDERSTANDING IPF

Some of the risk factors associated with IPF include:

- Genetics—10% to 15% of the cases occur in patients who have family members with 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 GERD (gastroesophageal reflux disease)

What are the early and late symptoms of IPF?

The early symptoms of IPF typically include:

- Cough: usually dry and nonproductive, meaning that no phlegm or mucus is produced
- Shortness of breath occurs while performing physical activities
- Other symptoms may include 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, which is an enlargement of their 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.

UNDERSTANDING IPF

Why is IPF difficult to diagnose?

Since the cause of IPF is unknown, it can be difficult to diagnose. An accurate diagnosis of IPF is achieved by ruling out other diseases. This process may be time consuming because the symptoms of IPF, such as cough and shortness of breath, are common symptoms associated with a number of other diseases, including asthma, COPD, and congestive heart failure. In addition, these symptoms can be mild and progress slowly; therefore, patients may not contact their doctor right away.

How is IPF diagnosed?

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

- Blood tests
- Pulmonary function tests
- Chest x-ray and CT scan
- Lung biopsy (VATS) or bronchoscopy

Blood tests will help your doctor to rule out other diseases. **Pulmonary function tests** are done to measure your breathing capacity, determine the "stiffness" of your lungs, and calculate the amount of oxygen that travels from your lungs to your bloodstream. You may also have an **x-ray** and a **CT "CAT" scan** of your chest done. These studies will help to identify the pattern of scarring in the lungs. A **lung biopsy** may also be needed. The chest surgeon will do this in an operating room while you are under general anesthesia. The procedure is called **Video-Assisted Thoracoscopic Surgery (VATS)**. Small incisions are made on the side of your chest, and then small

samples of tissue are taken. These samples are then sent to a laboratory and analyzed under a microscope. Sometimes a smaller biopsy will be done by a procedure called **bronchoscopy**. First, your doctor will give you medicine to make you relaxed and numb your airway. Then a small tube will be placed through your nose or mouth, into your lungs in order to perform the biopsy.

How is IPF treated?

There are several medications that IPF patients can try. Despite all of the studies being done, no one knows which treatment is the best. Your doctor will help you decide which medication is most appropriate for you.

Oral steroids (eg, prednisone, prednisolone) are often prescribed for patients with IPF. While there have been no studies showing them to be effective in the treatment of IPF, steroids may help to minimize inflammation in the lung that may occur as a result of the disease. Oral steroids do cause side effects, such as increased appetite, weight gain, and insomnia (inability to sleep). They can also make you feel more emotional. If you use oral steroids for a long period of time, you may develop brittle bones, and your body may not be able to fight off infections as well.

Immunosuppressants or medications that decrease your body's immune system, (eg, azathioprine and cyclophosphamide) may also be used. These medications can also cause side effects, including nausea, vomiting, fever, and skin rash. **Colchicine** is another treatment your doctor may recommend, and side effects may include nausea, vomiting, diarrhea, and hair loss.

Interferon gamma-1b is an injection that your doctor may recommend. Recent studies suggest that this medication may improve the lung's ability to respond to injuries that might otherwise result in acute exacerbations (rapid

UNDERSTANDING IPF

Medication	Method of Delivery	Benefits	Common Side Effects
Oral Steroids	Oral	May help decrease inflammation in the lungs	• Increased appetite • Insomnia • Long term use may lead to brittle bones
Immunosuppressants • Azathioprine • Cyclophosphamide	Oral	May help decrease inflammation and "scarring" of the lungs	• Nausea/vomiting • Fever • Skin rash
Colchicine	Oral	May help to slow fibrosis "scarring" in the lungs	• Nausea/vomiting • Diarrhea • Hair loss
Interferon Gamma-1b	Injection	May improve lung's ability to respond to injuries that might otherwise result in rapid worsening of IPF	• Flu-like symptoms, such as fever, chills, and body aches

worsening) of IPF. The most common side effects associated with interferon gamma-1b are flu-like symptoms, including fever, chills, and body aches. These side effects usually diminish within the first few months of therapy.

In addition to these medications, there are several potential therapies that are currently being investigated in ongoing clinical trials. You may want to speak with your doctor to determine if you are a candidate for one of these trials.

Non-drug treatments

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 may be considered for lung transplantation, but there are many factors that need to be considered before that is recommended. Your doctor can discuss these options with you.

Living with IPF

In addition to taking the medication your physician prescribes, there are a number of things you can do to help manage the symptoms of IPF. As with all diseases, it is important to live a healthy lifestyle. Quitting smoking is a very important lifestyle change to improve your health—smokers are advised to quit. You should eat a healthy diet, maintain a proper weight, and get enough rest. If you are short of breath after eating a meal, try eating smaller portions more frequently—up to six or eight times a day. Exercise helps to keep your body working as efficiently as possible and improve your energy and muscle tone. Your physician can also

recommend starting in a pulmonary rehabilitation program. It offers patients an exercise program that is structured and monitored by health care professionals. These professionals provide nutritional advice, breathing techniques, information about your disease, and emotional support.

Is there a cure for IPF?

Unfortunately, there is currently no cure for IPF. However, there are a number of different therapies being studied, and the results are promising. If you are interested in being enrolled into one of these treatment trials, you should speak with your doctor. Your doctor can help you determine which trial may be right for you.

Are there support groups for IPF patients and their families?

Yes, there are many different support groups for patients living with IPF and their families. These organizations provide you and your family with current IPF information, and also assist you in managing difficult things, such as dealing with 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 life-support measures. Both your health care provider and support organizations are there to help you and your family through this difficult time. You should speak with your health care provider about resources available in your area. In addition, a list of support group Web sites is provided in the "Patient Resources" section of these materials.

PATIENT RESOURCES

For answers to questions regarding idiopathic pulmonary fibrosis (IPF), please visit the following Web sites.

Coalition for Pulmonary Fibrosis
www.coalitionforpf.org

PILOT™
www.pilotforipf.org

Pulmonary Fibrosis Foundation
www.pulmonaryfibrosis.org

American Lung Association
www.lungusa.org/diseases/pulmfibrosis.html

National Institutes of Health
www.nlm.nih.gov/medlineplus/pulmonaryfibrosis.html

National Heart, Lung, and Blood Institute
www.nhlbi.nih.gov

Canadian Lung Association
www.lung.ca/diseases/pulmonary_fibrosis.html

For information on patient and family support organizations and end-of-life care, please refer to the following sites:

Aging With Dignity
www.agingwithdignity.org

National Hospice and Palliative Care Organization
www.nhpco.org

Caregiving.com
www.caregiving.com

Well Spouse Foundation
www.wellspouse.org

PATIENT RESOURCES

Please note that there are many other Web sites listing IPF facts, but information acquired from the Internet may not always be accurate. When using the World Wide Web, remember the following tips:

- Check the source—Information should come from a reliable source, such as a national organization or large university hospital
- Check the date—Make sure that the information is up to date
- Confirm the information—Check that the information is referenced to scientific sources, and always ask your doctor to help clarify any issues or questions

WHAT YOU NEED TO KNOW ABOUT HOME OXYGEN THERAPY AND PULMONARY REHABILITATION

Patients with IPF often require home oxygen therapy to help make breathing easier and more comfortable in order to continue being as active as possible in daily life. Oxygen therapy is often used together with pulmonary rehabilitation, which offers patients exercise training, breathing techniques, medical education, nutrition advice, and emotional support from a team of health care professionals. Patients also learn how to use and adjust their oxygen levels while in pulmonary rehabilitation. The following are some important things you should know about oxygen therapy and pulmonary rehabilitation.

Pulmonary rehabilitation and oxygen therapy are two important resources for IPF patients. Both are intended to help prolong life, reduce disability, and increase the level of functioning.

Tips for Oxygen Therapy and Pulmonary Rehabilitation

- You will need a prescription for both oxygen therapy and pulmonary rehabilitation. So be sure to speak with your health care provider
- In most cases, both oxygen therapy and pulmonary rehabilitation will be covered by your insurance
- If you are started on oxygen therapy, it is important to use it as directed by your health care provider
- Choose your oxygen supplier carefully—make sure your supplier delivers and installs the equipment, provides 24-hour service, 7 days a week, offers information on the use and cleaning of the equipment, and accepts payment from your insurance company

Tips for Oxygen Therapy and Pulmonary Rehabilitation

- Only adjust the flow settings on the oxygen tank if specifically instructed to do so by your health care professional. For example, you may need to adjust your oxygen flow rate before exercising
- Make sure that you understand your home oxygen equipment and that you have detailed instructions on how to care for this equipment
- Remember to always avoid smoking and have your visitors avoid smoking while you are using your oxygen
- Always request to be seated in the "no smoking" section at restaurants
- Do not use any flammable products while wearing your oxygen
- Remember to remain at least 5 feet from open flames and sources of heat, such as gas stoves, lighted fireplaces, and candles
- Avoid substances and medications that will decrease your breathing, such as alcohol and narcotics (codeine, Vicodin®, Percocet®)
- The use of cotton gauze and water soluble lubricants such as K-Y Jelly® can decrease skin irritation from oxygen tubing and nasal prongs
- Some patients worry that they will become "addicted" to oxygen therapy, but please remember this is not true

WHAT YOU NEED TO KNOW ABOUT HOME OXYGEN THERAPY AND PULMONARY REHABILITATION

- Do not hesitate to contact your physician if you experience:
  - Difficulty breathing
  - Frequent headaches
  - Blue lips or fingertips
  - Drowsiness
  - Dizziness

For more information on home oxygen therapy and pulmonary rehabilitation, visit the following Web sites:

American Association for Respiratory Care
www.aarc.org

American Association of Cardiovascular and Pulmonary Rehabilitation
www.aacvpr.org

Breathin' Easy
www.oxygen4travel.com

National Home Oxygen Patients Association
www.homeoxygen.org

TIPS FOR A HEALTHY LIFESTYLE

Living a healthy lifestyle may help in slowing the progress of your chronic lung disease. Stop smoking, eat a well-balanced diet, and follow a regular exercise program—these actions are critical elements of a healthy lifestyle.

Stop Smoking

- Make a personal decision to quit smoking and select a specific date and time to begin
- Develop a list of reasons to quit smoking
- Speak with your health care provider about your plan and options that can help you
- Always seek the support of your family and friends
- Carry a list of reasons why you quit smoking to help you through difficult moments
- Seek distractions when the urge to smoke occurs
- Do not allow a "slip" to stop you. Pick a new date and remain positive
- Reward yourself for not smoking

Remember, there are benefits to quitting smoking, such as improved taste and smell, increased oxygen levels in your blood, lower blood pressure and heart rate, warmer body temperature, and a reduced risk of cancer and heart attack.

For more information and resources about quitting smoking, visit the US Public Health Service Web site:
http://www.surgeongeneral.gov/tobacco/welcome.htm

TIPS FOR A HEALTHY LIFESTYLE

Healthy and Well-Balanced Diet

- Reduce your intake of saturated fats, such as meat, eggs, and butter
- Eat the recommended five portions of fruit and vegetables each day
- Choose good sources of protein, such as chicken and fish
- Smaller and more frequent meals may make breathing easier

Regular Exercise

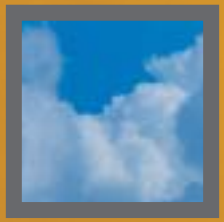
- Add regular exercise as prescribed by your health care provider to your daily routine. Exercise is recommended to keep breathing capacity up and to maintain muscle strength
- Consider enrolling in a pulmonary rehabilitation program to learn breathing exercises, increase strength, and enhance your support network
- Remember you can continue to exercise while on oxygen therapy by learning to adjust your oxygen flow rates appropriately. You should speak with your health care provider to learn how to adjust your oxygen flow rate
- You should speak with your health care provider for more information on starting in a pulmonary rehabilitation program

TIPS FOR A HEALTHY LIFESTYLE

Emotional Health

- Seek support systems, such as family, friends, health care providers, and support groups
- Recognize the importance of finding help to manage stress, anxiety, and depression
- Speak with your health care provider if you feel that you need medication to help manage your stress, anxiety, or depression
- Prepare for the changes in your life realistically, but stay hopeful and keep a positive attitude
- Recognize the need for you and your family to go through the grieving process
- Remember that sleep is important for both your mind and body
- Incorporate relaxation into your daily life

It is important to remember that the course of IPF varies greatly from person to person. Some patients advance quickly while others remain stable for longer periods of time. You can help yourself by following the same sensible health measures that everyone should observe. By keeping in tune with your disease, you will become less intimidated by treatment, and you may increase treatment success, while lowering risks of complications. As a well-informed patient, you will be in a better position to discuss all of your health care with your physician.



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