

Idiopathic Pulmonary Fibrosis (IPF)

Pulmonary fibrosis (PF) describes a group of lung diseases in which thickening of the walls of the air sacs (called alveoli), caused by scarring, can result in cough, shortness of breath, fatigue and low blood oxygen levels. Pulmonary fibrosis can be caused by an identifiable irritation to the lungs, but in many cases the cause is unknown. In cases when the cause of PF is unknown, the diagnosis is *idiopathic pulmonary fibrosis* (IPF). Idiopathic means there is no known cause at this time.

What Are The Symptoms of IPF?

When you have IPF, you may find yourself becoming more short of breath or having a dry cough. You may notice you cannot do your usual activities without shortness of breath. You may feel the need to slow down or stop and rest when walking, or it may become difficult for you to walk up inclines or climb stairs because of shortness of breath. Your health care provider may notice the oxygen levels in your blood drop when you walk. Lower than normal blood oxygen levels can cause high blood pressure in your lungs (called pulmonary hypertension). This puts an added strain on your heart and can lead to heart failure if low blood oxygen levels are left untreated.

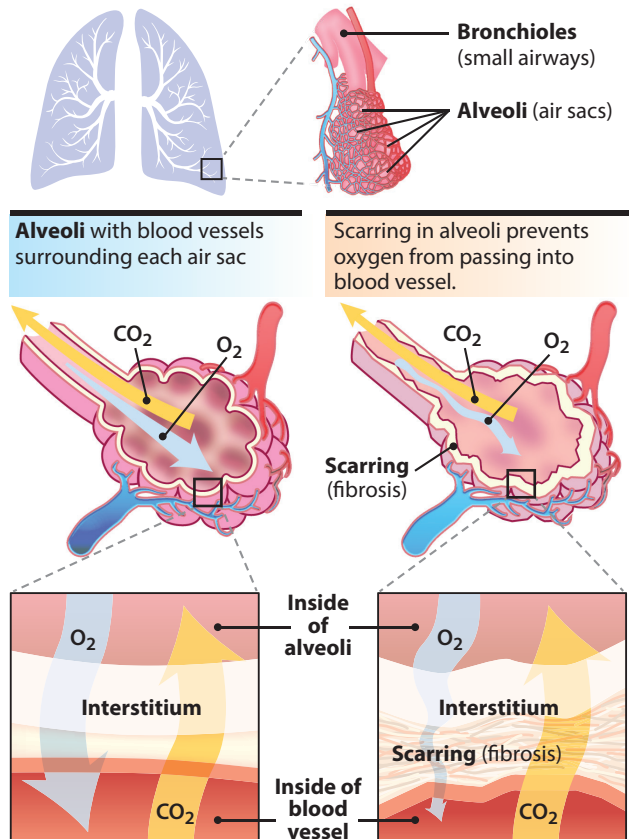
Who Develops IPF?

Most people with IPF develop symptoms of cough and shortness of breath between the ages of 50 and 70 years. IPF is not common under the age of 50 years. Historically, more men have been diagnosed with IPF than women, but IPF in women appears to be on the rise. Occasionally, IPF occurs in members of the same family. When this happens, the disease is called **Familial Pulmonary Fibrosis**. The fact that PF runs in certain families has led many experts to believe that having certain genes (genetics) may be why some people get PF.

How Does IPF Affect My Breathing?

Air flows through your nose and mouth, deep into the lungs through large airways (bronchi) and small airways (bronchioles). These breathing tubes lead air into millions of tiny air sacs called alveoli. Within the walls of the air sacs is tissue called the *interstitium*. Much like a net, a collection of tiny blood vessels called *capillaries* surround each air sac (see Figure). The oxygen in the air we breathe moves through the walls of the alveoli (including the interstitium) and into these surrounding capillaries, where the oxygen is picked up by red blood cells and carried to the heart. Carbon dioxide (CO₂) moves in the opposite direction from O₂ and is exhaled.

In IPF, the interstitium is thickened with scarring, making it difficult for oxygen to reach the capillaries. The scarring also makes the lungs “stiff” and difficult to inflate. Stiff lungs literally hold less air than normal lungs; that



is, they have less filling capacity. This low lung capacity, along with the problem with oxygen passing through the walls of the air sacs, causes shortness of breath. The amount of scarring can increase with time, making the lung even stiffer, further limiting its filling capacity, and limiting the ability of oxygen to pass through air sac walls. This scarring is generally not reversible. How fast the scarring increases, varies from person to person. Some people may remain stable (with no decline in filling capacity) for several years, while for others, their breathing can worsen more rapidly.

How Does My Health Care Provider Know I Have IPF?

Your health care provider may suspect you have IPF, based on your symptoms of a dry cough or shortness of breath. Abnormal breath sounds, called crackles can be heard by your health care provider when they have you take a deep breath. Crackles sound like Velcro® being pulled apart. You or your health care provider may also notice that the ends of your fingers and/or toes have changed shape (called clubbing). These signs will likely result in you being referred to a lung specialist (pulmonologist). The pulmonologist will do a physical examination and may order several tests, such as a chest x-ray, breathing tests or measurement of your blood oxygen level. Other tests that may be needed

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include blood tests, a high resolution CAT scan (HRCT) of your chest, echocardiogram (ultrasound or sonogram of the heart), and in some instances, a lung biopsy.

In some cases, a diagnosis of IPF can be made when a HRCT shows a particular pattern and other possible causes of PF have been excluded. In some cases, when the HRCT does not show the “typical” pattern of IPF, a surgical lung biopsy may be necessary to confirm the diagnosis of IPF. The most common way to get a biopsy of the lung is by video assisted thoracoscopic surgery (VATS) under general anaesthesia. In this procedure, the surgeon makes (two or three) small incisions in your chest, and by using a flexible camera, they are able to look inside your chest and remove samples of lung tissue for evaluation.

How Is IPF Treated?

Once the diagnosis of IPF is made, it is important for you to be seen regularly by a pulmonologist who is an IPF expert. As new treatments develop (or better ways to treat your symptoms become available), an IPF expert will know the best recommendations to make to you. Two new drugs, nintedanib and pirfenidone, have been approved by the Food and Drug Administration (FDA) for the treatment of IPF. Prior to the approval of these two new drugs, corticosteroids (also called steroids) and immunosuppressant drugs were used to treat IPF, but they often provided no benefit and may have caused significant side effects. Other therapies may be recommended to either treat your symptoms, or conditions that may develop as a result of having IPF. Such therapies include pulmonary rehabilitation, supplemental oxygen, and treatment of pulmonary hypertension. Talk with your health care provider about which drugs or other therapies will be best for your individual needs.

What About A Lung Transplant For IPF?

At this time, lung transplantation is the only known treatment to prolong life if you have IPF. Transplantation is a major surgery and requires life-long treatment with drugs that overpower your immune system to prevent rejection of your transplanted lung. Not all patients with IPF can have a lung transplant. Ask your health care provider if you should be evaluated for a lung transplant. An evaluation for a lung transplant may take months, so your health care provider may begin discussing a transplant with you before your IPF gets severe.

What Else Can Be Done To Help Me Feel Better?

Getting involved in a pulmonary rehabilitation program and a patient support group is important to help you learn more about IPF and how to manage it. You will meet others who are faced with the same challenges and share ways for dealing with having IPF. Joining a pulmonary rehabilitation program can help you improve your energy level, reduce your shortness of breath, give you a better understanding of your IPF and oxygen use, and teach you self-management skills. Your oxygen needs may change over time, so your oxygen levels should be evaluated regularly to find out how much oxygen you need at rest, with activity and with sleep. The goal is to keep your oxygen saturation levels above 89% no matter what you are doing: sitting quietly, walking, exercising, or sleeping. If you are still smoking, it

is important that you quit smoking. Exposure to smoke will worsen your breathing problem.

Are There Special Things I Should Watch For?

Because you have a chronic lung disease, try to avoid situations where you can get a cold or the flu, and get your yearly flu vaccine. A small percentage of people with IPF develop what are called acute exacerbations. These occur when breathlessness from IPF suddenly becomes worse. No one knows why **acute exacerbations** happen or which patients are likely to develop them. If you become more short of breath at any time, contact your health care provider or seek immediate medical attention.

How Can I Enroll In A Research Study For IPF?

If you are interested in enrolling in a research study for IPF, ask your lung specialist. As new treatments are being developed, research studies are needed to test them to see how well they work. These studies can only be done if people with IPF volunteer to enroll. Depending on where you live, IPF research may be happening at a center in your area. Even if you do not want to become part of a study, it is often helpful to get your care from a center that specializes in IPF.

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For additional information:

Coalition for Pulmonary Fibrosis:

<http://www.coalitionforpf.org/> 1-888-222-8541

Pulmonary Fibrosis Foundation:

<http://pulmonaryfibrosis.org/> 1-312-587-9272 U.S.

National Institutes of Health Clinical Trials

http://www.nlm.nih.gov/health/dci/Diseases/ipf/ipf_whatis.html

<http://clinicaltrials.gov/>

United Network for Organ Sharing (for transplant information):

<http://www.unos.org/> 888-894-6361

Rx Action Steps

- ✓ If you have a dry cough or shortness of breath that does not go away, see your health care provider
- ✓ If you develop sudden worsening of symptoms, seek care immediately
- ✓ If you smoke, stop smoking and limit any exposure to smoke
- ✓ Speak with your health care provider about pulmonary rehabilitation and a support group in your area
- ✓ Ask about lung transplantation
- ✓ Ask about involvement in research

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