Understanding Your Body

Idiopathic Pulmonary Fibrosis (IPF)
Dear patients, family members and caregivers,

The goal of this book is to help you to better understand your disease. We will describe what happens when you have idiopathic pulmonary fibrosis (IPF) and what is known about IPF at this time.

We will explain medical tests that you may need. In addition, this book will give you tips to help you stay as healthy as possible.

At the end of the book, there is space for you to write down any questions you may have. Please take a moment to go over this book with your doctor or health care provider and discuss any questions or concerns.
What does IPF stand for?

Idiopathic pulmonary fibrosis (IPF) uses three medical terms to describe one disease:

“**Idiopathic**”
When the cause of a disease is not known, it is called idiopathic.

“**Pulmonary**”
This is the medical term for lungs. This book will describe where the lungs are located and how they work.

“**Fibrosis**”
This means tough or leathery because scar tissue has formed. We will describe how lungs with fibrosis are different from healthy lungs.
How common is IPF?

IPF is a rare disease. Only about 14 to 43 people out of 100,000 have IPF.

The disease occurs throughout the world and affects people of all races.

IPF is much more common in older people but can occur at younger age.

Researchers all over the world are working on better understanding the disease.
**Where are the lungs?**

Lungs are spongy air-filled organs located on either side of the chest. The heart sits between the two lungs.

As we breathe air in through our nose and mouth, our lungs inflate like a balloon. The lungs get bigger as air fills all the deep spaces in the lungs. When we breathe out, the lungs become small again.

The trachea (also called the windpipe) moves air into the lungs through a pair of tubes called bronchi (each tube is called a bronchus).

The main bronchi branch into smaller bronchi and then into even smaller tubes called bronchioles, which end in very small air sacs called alveoli.
How do the lungs work?

When you breathe in, your lungs fill with air. Oxygen (O2) from the air can then enter the blood. This happens in the alveoli, which are located next to blood vessels. The blood carries oxygen through the body, which needs oxygen to function.

The lungs also remove carbon dioxide (CO2) from the body. Carbon dioxide leaves the blood vessels and enters the alveoli where it can be breathed out. Too much carbon dioxide in the body is not healthy.

The air we breathe is sometimes polluted. Our lungs work to remove dirt and germs from the air. This helps us to stay healthy.
What happens in IPF?

The small picture inset shows how oxygen and carbon dioxide move between the blood vessels and alveoli of the lungs. In a healthy lung this is a very short distance to travel. This makes it easy for oxygen to enter the body and carbon dioxide to leave the body.

A lung with IPF does not expand as well as a healthy lung. The tough, leathery tissue in the lung does not move easily. This makes it harder for a patient to breathe.

In addition, the leathery tissue located between the alveoli and blood vessels is thicker, making it harder for oxygen to enter the body and carbon dioxide to leave the body.
What are the symptoms of IPF?

At first, IPF usually causes no symptoms or only mild symptoms. As the disease becomes worse, you may experience:

- Dry cough
- Shortness of breath and tiredness
- More frequent colds and lung infections
- A bluish skin color (called cyanosis)
- Changes in the shape of your finger nails (called clubbing)
- Poor appetite and weight loss

Over time you may also develop heart disease and blood clots.
How is IPF diagnosed?

Your doctor will examine you and take your medical history (asking questions about your health and the health of family members). You may also need some special tests.

Computerized axial tomography (usually called a CT scan or CAT scan) can examine the organs inside your body. A CT scan uses x-rays and a computer to get a detailed picture of your lungs.

The CT scanner is a large machine shaped like a donut. It takes x-rays as your body moves through the opening. This usually takes just a few seconds and is painless.

A CT scan can detect changes in your lungs that suggest IPF.
What is a lung biopsy?

For most patients IPF can be diagnosed based on medical history, physical exam, and the CT scan.

For some patients it is necessary to take a small piece of lung tissue. A specialist will look at this tissue under a microscope to diagnose the problem.

Taking a piece of lung tissue is called a surgical lung biopsy. Before the biopsy, you will take medications to put you to sleep so that you do not feel any pain.

The surgeon will insert a tube with a tiny camera into the chest, so the lung can be seen on a video monitor.

A small piece of lung tissue can then be taken with a special device called a stapler, which cuts and seals the lung at the same time.
How can the function of the lungs be tested?

To understand how well your lungs are working, you will take a pulmonary function test using a breathing machine called spirometer.

During the pulmonary function test, you take a deep breath in and then blow out as hard and fast as you can into a tube. This test also measures lung volume (how much air is in your lungs when you take a deep breath). People with IPF have a harder time filling their lungs and usually have smaller lung volumes.

The amount of oxygen in your blood can be measured with a finger sensor. If your oxygen level is below 88%, you may need extra oxygen.

The oxygen in the blood can also be measured with a blood sample, usually taken from a blood vessel in the wrist. This is called an arterial blood gas test.
How is IPF treated?

Since the lung scarring (fibrosis) in IPF is permanent, the disease cannot be cured. Your doctor may refer you to a specialist, who may choose one or more of the following treatments:

- Medical treatment
- Pulmonary rehabilitation to help you learn to manage your symptoms (this might include exercise training, nutrition counseling, or group support)
- Supplemental oxygen to help keep your oxygen levels at a healthy level
- Lung transplantation
- In rare cases, steroids may be used
Will my lungs get worse?

It used to be thought that all patients with IPF get worse very quickly. However, recent research shows that in some patients the disease worsens slowly. Some patients will even stay the same over time.

Unfortunately we cannot tell which patients will get worse and which patients will stay the same. This is an important area of research, because understanding why some patients get worse might help us find better treatments for this disease.

New medications that are currently being tested might help slow the disease and keep symptoms from getting worse.
What should I do to keep my IPF from getting worse?

Symptoms generally become more severe over time and worsen when you get a lung infection. Please make sure to:

- Use all your medications as prescribed
- Stay active
- Get vaccines for flu and pneumonia, because lung infections often cause IPF to worsen
- If you still smoke, you should ask your doctor about ways to quit. You should also ask people around you to stop smoking
- See a doctor who specializes in lung diseases (pulmonologist), and if possible seek care at an IPF specialty center
When should I seek medical help?

Call your doctor if you notice any changes or have any concerns that you wish to discuss. In addition, please seek medical help right away if you have any of these warning signs:

- Unusual trouble breathing or shortness of breath
- Pain or pressure in your chest
- A cough you cannot control
- Coughing up blood
- High fever
- Diarrhea, nausea or vomiting
- Unusual weakness
Questions for My Doctor:

For more information and other available books go to www.pocketdoktor.com or email to post@pocketdoktor.com

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1. Edition 2013
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Authors: Erica Herzog, M.D.; PhD.; Felix Knauf, M.D.; Philipp Kirchhoff, M.D.
Design: Patrick Lane
Printed in Germany

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