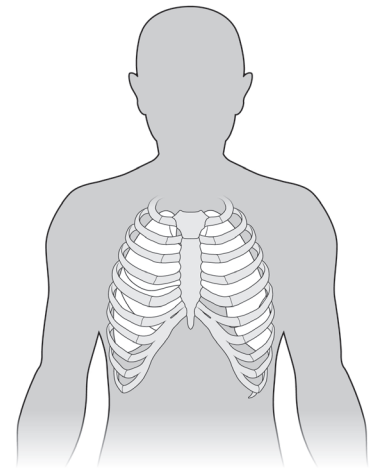


# Idiopathic Pulmonary Fibrosis (IPF)

## Information for patients and families

Read this handout to learn about:

- What idiopathic pulmonary fibrosis (IPF) is
- Signs and symptoms
- How your doctor will know if you have it
- Tests you may need
- Treatments
- Where to get more information



## What is idiopathic pulmonary fibrosis (IPF)?

Idiopathic pulmonary fibrosis (IPF) is a condition that causes scars to form in your lungs. More and more scars form over time. IPF causes your lungs to become stiff. This makes it hard to breathe. Over time, your lungs will not be able to give your body enough oxygen.

We do not know what causes IPF.



## **Some facts about IPF are:**

- IPF may affect over 10,000 people in Canada
- There are 3,000 to 4,000 new cases of IPF each year
- Most people who get IPF are between the ages of 50 and 70
- More men have IPF than women
- Most people with IPF used to smoke
- A small number of patients have a family history of lung scarring

## **What are the signs and symptoms of IPF?**

Most people find that the signs and symptoms come on slowly. They include:

- Getting out of breath when you are active
- A dry hacking cough that does not go away
- “Crackles” in the lungs that your doctor can hear with a stethoscope
- Rounding of the fingernails - a condition called “clubbing”

The signs and symptoms of IPF can be the same as other scarring lung diseases. This means your doctor will need to make sure that you do not have any other conditions. You may need to visit your doctor several times to find out if you have IPF. You will also need to decide on your treatment together.

## **How will my doctor know if I have IPF?**

IPF is hard to diagnose. You may need to have many tests. Then a team of specialists will talk about the results of the tests. Your team may include Respiriologists (lung specialists), Radiologists (x-ray specialists), Rheumatologists (joint specialists) and Pathologists (microscope specialists). Together, they will decide if you have IPF.

First your doctor will talk to you about your medical history and do a physical exam. At this visit, your doctor will:

- talk with you about problems that may relate to lung scarring
- ask about any medicine you are taking
- talk about what you have been exposed to at work and in the environment that might cause lung disease
- listen to your chest with a stethoscope
- check your skin and joints

The next step is to do some tests.

## **What tests will I need?**

Your doctor will talk to you about which tests are right for you. Here are some tests that help to show if a patient has IPF:

### **Pulmonary Function Testing (PFT)**

This breathing test will measure:

- the flow of air in your lungs
- the volume of air in your lungs
- how well your lungs take oxygen from the air

### **High Resolution Computed Tomography (HRCT)**

This is a special type of CT scan that makes detailed pictures of your lungs. The scan can take pictures while you lie on your back and while you lie on your chest. It can also take pictures while you breathe air out of your lungs. The HRCT is a very valuable test to help your doctor find out whether or not you have IPF.

Having an HRCT is the same as having a regular CT scan. Both scans have you lie on an open-air table and only take a few minutes. Only the steps for doing a HRCT are different.

### **Blood Tests**

Blood tests will show if you have other diseases. They will check for antibodies. These will show whether you have a connective tissue disease, such as Rheumatoid arthritis or Scleroderma. Some people with these diseases have lung problems before they get any other symptoms.

### **Six Minute Walk Testing**

This test looks at how far you can walk. It also measures the amount of oxygen in your body while you are walking. The test uses a probe on your finger or in your ear.

### **Bronchoscopy**

Some patients will need to have a bronchoscopy to be sure that they have IPF. This test collects samples of fluid and tissue from your lungs. First your doctor will numb the back of your throat. They will give you medicine to make you more comfortable. Then the doctor will use a flexible fiberoptic scope, which is about the size of a pencil. The scope passes down your trachea or “breathing tube” into your lungs. Then the doctor collects the samples. This test will help your doctor find out if you have IPF.

Bronchoscopy is an outpatient procedure. This means you do not need to stay overnight in the hospital.

### **Surgical Lung Biopsy**

Some patients will need to have a lung biopsy to be sure that they have IPF. A lung biopsy is a type of surgery to take samples of lung tissue for testing. You will have the biopsy under general anesthetic (you will be asleep).

- A chest surgeon will do the biopsy. They will make cuts in the side of your chest that are 1 inch (2.5 cm) long. Then the surgeon will use small tools and cameras to collect samples from your lungs.
- You will need to stay in the hospital for a few days after the biopsy.

## What is the treatment for IPF?

Unfortunately, there is no cure for IPF.

There are 3 types of treatment. But, no treatment is proven to cure IPF. Your doctor may recommend that you try one of these.

The 3 types of treatment are:

1. Medicine
2. Pulmonary rehabilitation
3. Lung transplant

Read about each of these treatments in the sections below.

### 1. Medicine

Recent research suggests some drug therapies may be helpful, while others may not work for certain types of patients. Many of these drugs need more study.

#### a. Pirfenidone (Esbriet)

The CAPACITY and ASCEND trials have shown that this drug slows down scarring in patients with mild to moderate cases of IPF. It does not undo the damage that has already been done to the lungs. But once you start taking this drug, it slows down the decline in how well your lungs function.

- You need to take Pirfenidone (Esbriet) 3 times a day.
- Common side effects are nausea, heart burn, diarrhea, skin rash and sun sensitivity.
- Pirfenidone (Esbriet) is available in Canada.

#### b. Nintedanib (OFEV™)

The INPULSIS trials have shown that this drug also slows down scarring in patients with mild to moderate cases of IPF. It does not undo the damage that has already been done to the lungs. But once you start taking this drug, it slows down the decline in how well your lungs function.

- You need to take nintedanib 2 times a day.
- Common side effects are diarrhea, nausea and decreased appetite.
- Nintedanib (OFEV) is available in Canada.

### **c. Treatment of GERD**

Gastroesophageal reflux (gas-tro-ee-sof-a-gee-ul ree- fluhk-s) disease (GERD) is also known as acid reflux. It happens when stomach contents flows back into your foodpipe (esophagus). This is the tube that leads from your mouth to your stomach. It can be due to weakening of the point where the esophagus meets the stomach (the lower esophageal sphincter), or problems with the squeezing of the muscles of the esophagus, or stomach. It is thought that there is a strong link between GERD and IPF. The reason for this is not known. Most patients with IPF are not able to feel if they have GERD.

There is growing evidence that treating patients with IPF for GERD (even if they do not have symptoms) can help slow down the decline in how well your lungs function. Studies in this area are still going on.

- Most patients with IPF should be treated with a prescription antacid from a class called Proton Pump Inhibitors (PPI)
- These medications reduce the amount of acid being made in the stomach
- PPI medications are generally safe and well tolerated, but if taken for a long time, they may slightly increase the risk of pneumonia (lung infections), gastrointestinal infections or bone fracture.
- Examples of PPI medications are: dexlansoprazole (Dexilant), esomeprazole (Nexium), lansoprazole (Prevacid), omeprazole (Losec), pantoprazole (Pantoloc and Tecta) and rabeprazole (Pariet)

### **d. N-Acetylcysteine (NAC)**

Although there was previously some research that suggested adding N-acetylcysteine (NAC) to therapies using prednisone is helpful (see below), the ACETYLCYSTEINE trial found that NAC has no effect in IPF. Generally, this medication is no longer used to treat IPF.

### **e. Prednisone plus Azathioprine plus NAC**

This therapy uses 3 medicines together. This combination has often been used with patients with IPF. However, a large placebo controlled trial called the PANTHER trial showed that this treatment does not improve outcomes for patients with IPF. In fact, results are worse for patients with a definite diagnosis of IPF. These patients are more likely to end up in hospital, have side effects, or even die, than patients who got a placebo. This treatment is still important and useful for similar diseases. However, patients who definitely have IPF should avoid it.

### **f. Other medicines**

Research studies continue on other drugs to treat IPF. Some studies are showing that these drugs may be helpful to treat IPF in the future. However, there will need to be more research to approve these drugs for people with IPF.

Ask your doctor if there are other medicines that may be right for you.

## **2. Pulmonary rehabilitation**

Pulmonary rehabilitation is a special exercise program for patients with lung disease. These exercises do not make your lungs stronger, but they do make your arms and legs stronger. When you get stronger and into better shape, you will feel better and be able to do more. These programs also teach you how to deal with all the stress that comes with having lung disease. They teach patients how to manage their daily chores and use their Oxygen. Most patients find these programs help improve their physical and psychological well-being.

## **3. Lung transplant**

A lung transplant may be the right choice for some patients. But, it is not right for everyone. You can talk to your doctor about this.

## Where can I find more information about IPF?

If you have questions about IPF, please talk to your doctor. You can also find out more on the websites below.

There is a lot of information about IPF available online. However, some websites have information that is not correct.

Only use websites that have a good reputation, such as the websites below.

Name **Canadian Pulmonary Fibrosis Foundation**

Website <http://cpff.ca>

Name **American Pulmonary Fibrosis Foundation**

Website [www.pulmonaryfibrosis.org](http://www.pulmonaryfibrosis.org)

Name **American Thoracic Society – IPF patient information**

Website <http://www.thoracic.org/patients/patient-resources/resources/idiopathic-pulmonary-fibrosis.pdf>

Name **The Lung Association – IPF patient information**

Website <http://www.lung.ca/lung-health/lung-disease/idiopathic-pulmonary-fibrosis>

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