

Idiopathic pulmonary fibrosis (IPF)



UHN

Information for patients

What is Idiopathic pulmonary fibrosis (IPF)?

Idiopathic pulmonary fibrosis (IPF) is a condition that causes progressive scarring of the lungs. IPF causes the lungs to become stiff, making it hard to breathe. The lungs are then unable to provide the body with enough oxygen.

The cause of IPF is unknown.

Some facts about IPF are:

- IPF probably affects over 10,000 people in Canada
- 3,000 - 4,000 new cases of IPF are found each year
- IPF is found mostly in people between the ages of 50 and 70
- IPF affects more men than women
- Most people with IPF are former smokers
- There are no proven risk factors for IPF, but 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 of IPF often come on slowly and include:

- Shortness of breath with activity
- Chronic, dry hacking cough
- “Crackles” in the lungs heard by your doctor through a stethoscope
- Rounding of the fingernails - a condition called “clubbing”

Please visit the UHN Patient Education website for more health information:

www.uhnpatienteducation.ca

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Since the signs and symptoms of IPF can be just like the symptoms of other scarring lung diseases, your doctor will need to make sure that you do not have any other conditions. You may need to visit your doctor several times so that he or she can finalize your IPF diagnosis and treatment.

How will my doctor know if I have IPF?

The diagnosis of IPF requires input from lung specialists, radiologists, and in many cases, pathologists experienced in the evaluation of patients with interstitial lung disease. In many cases, a face-to-face discussion among these different disciplines is necessary for an accurate diagnosis.

To find out if you have IPF, your doctor will start by talking to you about your medical history and, he or she will also do a physical examination. This will include talking with you about problems you have that could be related to lung scarring, your medications, any occupational and environmental exposures that might cause lung disease, listening to your chest with a stethoscope, and examining your skin and joints.

In addition to a careful medical history and physical examination, you may need to have the following tests to help your doctor make an accurate diagnosis:

Pulmonary Function Testing (PFT) – This test involves a series of breathing tests that measure your lungs’ airflows, the volume of air in your lungs, and the ability of your lungs to extract oxygen from the air.

High Resolution Computed Tomography (HRCT) – This is a special type of CT scan that provides your doctor with high- resolution images of your lungs. Images may be taken in several different ways: while you are lying on your back, lying on your chest, and possibly, while having you breathe air out of your lungs. The HRCT is an extremely valuable test to help your doctor find out whether or not you have IPF.

- Having a HRCT is no different than having a regular CT scan but requires a special protocol; they both are performed on an open-air table and take only a few minutes.

Blood Tests – Blood tests (serology) that will look for various antibodies may be needed to see if you have a connective tissue disease, such as Rheumatoid arthritis or Scleroderma.

- Auto-antibodies are antibodies (immune factors normally produced by your body to fight infection) that react to and, damage your own body tissues. Some patients with connective tissue diseases develop lung problems before other parts of the body are affected.

Six Minute Walk Testing – This test looks at distance you can walk and the oxygen saturations (amount of oxygen in your body tissues measured by finger or ear probe) you have while walking.

Bronchoscopy – During this test, a flexible fiberoptic scope, about the size of a pencil, is passed down your trachea or “breathing tube” into your lungs to get fluid and tissue samples. Your doctor will “numb” the back of your throat and give you medication, if needed, to make you comfortable during this procedure. This test will help your doctor find out if you have IPF.

- Bronchoscopy is an outpatient procedure, so you will not need to stay overnight stay in the hospital. This test is not always needed in cases of IPF, but may be done by your doctor in certain situations.

Surgical Lung Biopsy –Some patients will need to have a lung biopsy to be sure that they have IPF.

- A lung biopsy is a surgical procedure usually done with small tools and cameras through one inch-long incisions in the side of the chest. A surgical lung biopsy is performed by a chest surgeon under general anesthetic (you will be asleep during the biopsy). You will need to stay a few days in the hospital after the procedure.

What is the treatment for IPF?

Unfortunately, there is no cure for IPF. As well, there is no treatment available that is proven to be effective.

Recent research data suggests a potential role for several therapies that require further study:

- **N-Acetylcysteine (NAC)** – One research study found that adding N-acetylcysteine (NAC) to traditional prednisone-containing therapies (see below) may be helpful, but it is not clear how helpful this drug is when used on its own. N-acetylcysteine (NAC) is available over the counter at many pharmacies and nutritional stores and has very few side effects.
- **Prednisone plus Azathioprine** – using two medications, such as Prednisone and, a second drug such as Azathioprine (or Mycophenolate and Cyclophosphamide) has been used for the treatment of IPF.
 - So far, it has been found that this treatment is not usually effective. If used to treat IPF, research has shown that these therapies work better if combined with N-acetylcysteine (NAC).
- **Pirfenidone** – Three large research studies of Pirfenidone have suggested that this drug may have a small benefit in treating IPF. Pirfenidone may slow the rate of scarring and stiffening of the lungs, but only by a very small amount. This drug has not been approved for use in the United States or Canada.
- **Other medications under study** – There are ongoing research studies of other medications for IPF. At this time, some are showing that they may be helpful to treat IPF in the future, but more research will be needed before these medications are approved for widespread use in people with IPF.

Your doctor will talk to you about which, if any, of these treatments may be right for you.

What other things can be done for people with IPF?

- **Pulmonary rehabilitation** is an important and effective treatment for patients with IPF. Pulmonary rehabilitation is an exercise and educational program designed just for patients with lung disease.

- A **Lung transplant** may be an effective treatment option for some patients. You should know, though, that a lung transplant is not always right for everyone. Your doctor will talk to you about this, if needed.
- Careful attention to certain associated medical problems such as **gastroesophageal reflux disease (GERD)** and **pulmonary hypertension (PHTN)** may also be important.

Where can I find more information about IPF?

If you have questions about IPF, please talk to your doctor or refer to the websites below.

Please be aware that there is a lot of information about IPF available online, but many websites have information which may not be accurate. If you are searching for information, we recommend that you only refer to reputable websites, such as the ones listed below.

Online Resources for IPF	
www.lung.ca/diseases-maladies/a-z/pfibrosis-fibrosep/index_e.php	
www.nlm.nih.gov/medlineplus	→ search for idiopathic pulmonary fibrosis
www.patients.uptodate.com	→ search for idiopathic pulmonary fibrosis
http://cpff.ca	→ website of the Canadian Pulmonary Fibrosis Foundation
www.pulmonaryfibrosis.org	→ website of the American Pulmonary Fibrosis Foundation
www.coalitionforpf.org/	→ website of the Coalition for Pulmonary Fibrosis