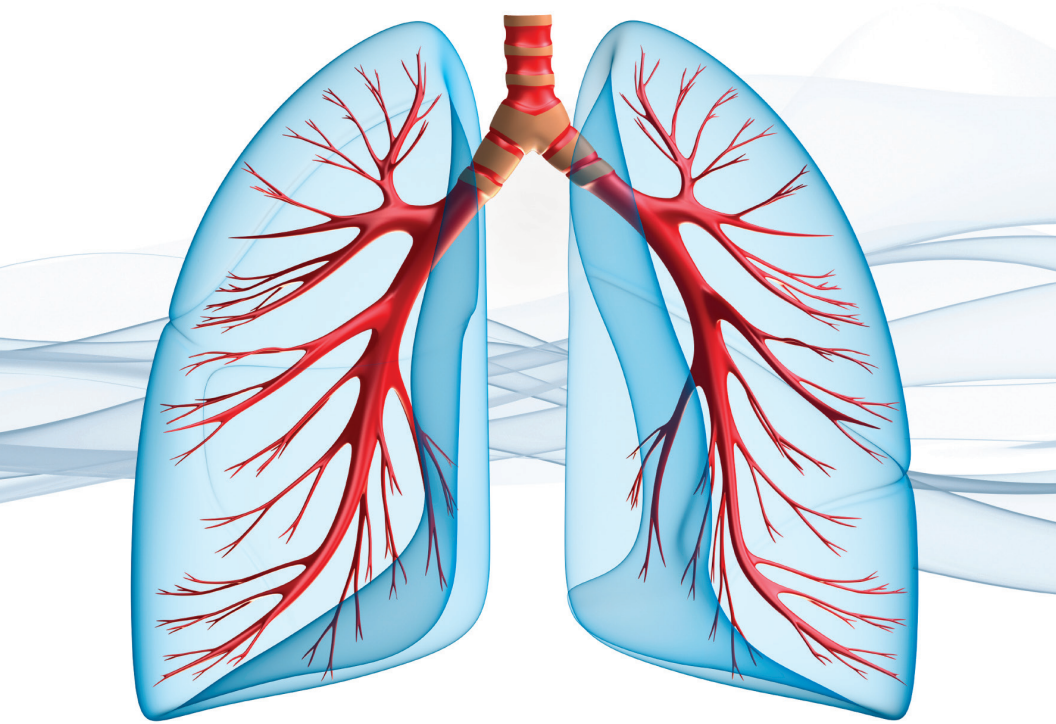
CANADIAN PULMONARY FIBROSIS FOUNDATION

BREATHING SHOULD NEVER BE HARD WORK

IDIOPATHIC PULMONARY FIBROSIS

PATIENT INFORMATION GUIDE



Written by Robert Davidson, Director, President and Founder of the Canadian Pulmonary Fibrosis Foundation,
based on his personal experiences as an IPF patient and double lung transplant recipient.
Some of your experiences may be different.

“NEVER SURRENDER”

Winston Churchill used these words in 1939
when confronted with the threat of invasion by foreign forces.

These were the words I stood by after my diagnosis of
Idiopathic Pulmonary Fibrosis (IPF) in October, 2007.
I believe they helped me through those most difficult days.
I never gave up hope. To give in is to let the disease win.
In my case, it lost. I WON.
I received my new lungs on January 30, 2010, with grateful thanks
to my donor and his or her family. I urge everyone who receives this
frightening diagnosis to “NEVER SURRENDER.”

Robert Davidson

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grants from Hoffmann La Roche Canada Inc. and Boehringer Ingelheim (Canada) Ltd..
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WHAT'S IN THIS GUIDE FOR YOU

When a patient is diagnosed with pulmonary fibrosis (PF), they may not have heard of it. It is a relatively unknown and rare disease with up to an estimated 30,000 sufferers in Canada.

Once they start looking into it, however, they soon find out it is quite frightening, with a poor prognosis – death within three to five years of diagnosis. Their physicians may not have the necessary time or resources to fully explain all the details of the disease, and what they can expect as it progresses.

You, as the patient, must take charge of your own health care and find out what questions need to be asked and answered.

The Canadian Pulmonary Fibrosis Foundation was established by Robert Davidson who had IPF and survived through the gift of new lungs on January 30, 2010. He has a lot of the answers to those non-medical questions frequently asked by people suffering from IPF.

This guide provides Robert's advice on how patients should manage and advocate for their proper care. It will also answer many of the questions that patients and their caregivers may have, and set them in the right direction to get the answers to other questions.

It is not intended as a substitute for professional medical advice and patients should always consult their physicians with any questions they may have regarding specific medical conditions and symptoms.

The Canadian Pulmonary Fibrosis Foundation is here to help. If you have any questions, or just want to chat about the disease and how it is affecting you, please contact us at Robert@cpff.ca or 905-294-7645.

WHAT IS PULMONARY FIBROSIS?

Pulmonary Fibrosis is one of more than 200 related diseases of the lung known as Interstitial Lung Diseases (IID). IPF is the most common form and has no known cause. Up to 30,000 Canadians are believed to be affected by all forms of PF, with an estimated 5,000 deaths each year. They are part of a subgroup of ILDs called interstitial pneumonia.

In patients with PF and IPF, the lung (pulmonary) tissue becomes scarred (fibrosis) and over time, as the scarring becomes thicker and more widespread, the lungs lose their ability to transfer oxygen into the bloodstream. As a result, patients become short of breath and the brain and vital organs are deprived of the oxygen necessary for survival. Some IPF patients may notice some memory loss (I found my memory returned after my lung transplant), and some may find their hands and feet get cold.

WHAT CAUSES PULMONARY FIBROSIS?

Some other diseases such as scleroderma and rheumatoid arthritis may cause PF and it is important to carefully examine each patient to see if there is a related cause which may be treatable, resulting in improved life expectancy.

There are certain environmental exposures that are believed to be associated with PF, including:

- **Exposure to contaminants** such as asbestos, coal dust, silica, as well as animal proteins
- **Lung infections**
- **Certain prescription drugs** - always check for side effects of drugs you are prescribed
- **Gastroesophageal Reflux Disease** (GERD)
- **Smoking**. If you smoke you should try and stop and help those around you stop

GENETICS

There is a lot of research being undertaken into the effects of the genes on a person contracting PF or supporting other causes to develop into PF. gene variants may predispose some people to contracting PF. Also, mutations in certain familial genes show that the disease may be genetic in some families, particularly if more than one person in the family has contracted PF. Other genetic research indicates that in some patients with specific genetic traits, the disease will progress at a greater rate than those without it.

It is suggested at this time that patients do not go out and get genetic testing as this may only cause unnecessary alarm. Not all

of the genetic variances mean that you will get PF. Suffice to say, there is considerable research being done in the field which may result in future cures or preventative gene work. This is something that PF patients should discuss carefully with their doctor.

HOW MANY PEOPLE HAVE IT?

Finding out how many people have PF and IPF, and how many die from it, was very difficult until now, and even now it is not easy.

Statistics Canada does not have PF as a recognized cause of death; rather it is buried into one of the 200 related diseases. It is only in the past few years that the medical community has started to set up Interstitial lung Disease units and track the incidence of the disease.

It is estimated that up to 0.09 per cent of Canadians (30,000) suffer from all forms of pulmonary fibrosis, and that 5,000 die from it each year. There are about 6,000 new cases each year in Canada. This number is expected to increase as the population ages. While this is tragic for those people, it may push governments to take the disease more seriously and properly fund research and new treatment options.

IPF is more common in men than women – about 60 per cent men to 40 per cent women. It is very rare in patients under age 50. Younger people can certainly get PF, but it tends to be due to other causes such as rheumatoid arthritis, scleroderma or other exposures. It affects all races.

WHAT ARE THE SYMPTOMS?

You may not notice symptoms immediately. It could take months, or years, before there are any noticeable effects. Once you start to notice the symptoms, the disease is established and it becomes essential to determine if there is any cause that may be treatable.

The most common symptom of PF is shortness of breath, known as dyspnea. This symptom can be difficult to notice, especially in older people who may attribute this shortness of breath to aging, or may suspect a heart problem. As the disease progresses, the shortness of breath becomes more noticeable, particularly when carrying out normal daily activities, such as showering or going up stairs. Some people find it difficult to speak on the phone, running noticeably short of breath while talking.

As the disease advances the patient may develop a dry, hacking cough with no phlegm being produced. This cough can become very troubling, sometimes causing the patient to vomit. This should be reported to your doctor if it gets too bad, as there are prescription drugs that may ease the cough reflex. Watch out for drugs that may have a side effect of constipation. This is very stressful on IPF patients and causes an alarming drop in oxygen saturation when using the washroom.

Other symptoms you may encounter include:

- **Loss of appetite**
- **Rapid weight loss,** not necessarily related to the loss of appetite
- **Extreme tiredness and loss of energy**
- **Chest discomfort**
- **Coldness in the extremities** (hands and feet)
- **Memory loss**

As the disease progresses, getting up and down stairs will become more difficult, as will everyday tasks of getting up and showering in the

morning. Oxygen may be necessary to get around. Refer to the section on oxygen later in this guide for more information.

WHAT'S INVOLVED IN THE DIAGNOSIS?

Once you have been referred by your family doctor to a respirologist, probably after an initial x-ray, you will be subjected to a number of tests. It is very important to receive the correct diagnosis. If you are misdiagnosed, treatment options may be missed and IPF is easy to misdiagnose.

The doctors will listen to your lungs and may hear crackles, a bit like velcro being opened. He or she will also check your hands for clubbing of the fingertips, a common sign of IPF. These signs will help determine other test requirements.

You will be sent for a High Resolution CT scan which clearly shows the scarring pattern. the position of the scarring on the lungs tells the specialist a lot and will be instrumental in making the diagnosis.

Other tests include a **pulmonary function test** which is done in the hospital and involves various tests of breathing, both hard and soft,

into a machine testing lung volume output capacity, and gas exchange (the inhalation of oxygen and exhalation of carbon dioxide). It will measure how effective your lungs are at getting oxygen into the bloodstream, and getting the old stuff out. The scarring affects the alveoli, the little sacks that do this job.

You will probably be sent for a **six minute walk test** which, while very simple, is very important as it measures the oxygen saturation when you walk for six minutes without stopping. Your medical team also tracks how far you are able to walk in that time. A reduction in distance tells them a lot.

One of the more invasive tests is the **bronchoscopy** where a tube is inserted into the bronchial tubes and fluids injected into the lungs and retrieved so that the sample may be tested in the lab. At the same time a small piece may be taken from each lung for analysis. While these tests are a little uncomfortable, they do tell the doctors so much.

The doctor may ask for a **surgical lung biopsy**. This is another invasive procedure and they will usually ask for this only if they are unable to confirm the diagnosis through all the other tests. 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.

Of course you will have lots of **blood tests**, but you may as well get used to them as you will be having them very frequently after your lung transplant.

For a more detailed description of the diagnostic tests, please refer to Dr. Shane

Shapera's paper on Idiopathic Pulmonary Fibrosis, a link to which may be found on our web site at www.cpff.ca

TREATMENTS

There have been exciting developments in treating IPF since 2012 that slow down the progression of the disease. This gives us immediate hope.

Esbriet™ (pirfenidone) is an anti -fibrotic/anti –inflammatory agent approved by health Canada for the treatment of mild to moderate idiopathic pulmonary fibrosis in adults. Another treatment called Ofev™ (nintedanib) was also recently approved by Health Canada for the treatment of IPF. Talk to your doctor about which of these treatments may be suitable for you. Do not delay as early diagnosis and treatment is very important for you. Clinical trials demonstrated that both these treatments were effective in treating mild to moderate IPF in patients.

Make sure you get your pneumonia and flu shots. You don't want to get these illnesses while you have IPF or PF as they can cause exacerbations of the disease which can result in more irreversible damage to your lungs.

Your doctor may also prescribe some drugs that can help with some of the symptoms of IPF, particularly the dry hacking cough. Sometimes the side effects of these drugs can outweigh the benefit of easing symptoms, so don't be too upset if your doctor does not prescribe them for you. If you are prescribed such drugs, it is very important that you follow the doctor's instructions to the letter to avoid serious repercussions.

Pulmonary rehabilitation can be very effective in keeping the lungs supple and helps your breathing. Everyone who has pulmonary

fibrosis should try to get as much exercise as possible without over-exerting themselves. Of course, if your doctor specifically instructs you to avoid exercise, you should listen to that advice. It wouldn't hurt to ask why though.

It is best if you can find a lung rehab program close by your home to get expert guidance in the best pulmonary therapy program for you. Even if you cannot find such a facility, try and get exercise without over-exerting yourself.

In Toronto, we have an excellent pulmonary rehab facility at West Park Healthcare Centre. This is the only inpatient program in Ontario and is staffed with excellent staff and respirologists. An inpatient program is useful if you cannot find a lung rehab centre close to your home, or are unable to travel to an outpatient program for other reasons.

For those of you who cannot locate a rehab facility close to home the Foundation has produced an exercise video designed by PF specialists specifically for PF patients. This is freely available on our web site at www.cpff.ca

OXYGEN THERAPY

At some point you may be prescribed oxygen. Although you will feel breathless while you have PF and feel you need oxygen, it still may not be the appropriate time for oxygen therapy. Only a series of tests will confirm if you qualify.

You will be tested for your oxygen saturation; how much of the hemoglobin contains oxygen. It should be greater than 90 per cent at all times. They will also test your arterial blood gases by taking a small amount of blood from an artery, usually the wrist. You may be tested while at rest and while walking. These tests will determine whether you qualify for oxygen.

Once you are given oxygen, you might consider acquiring an oxymeter which measures your oxygen saturation. If it goes below 89 per cent, then you should be using your oxygen. If you measure in the 90 per cent or above range, you are getting enough oxygen.

While taking oxygen, you may start having some side effects such as headaches, confusion or increased sleepiness. This may mean you are getting too much and should consult your provider about the flow you are on.

Oxygyen should not unduly affect your lifestyle, and your oxygen provider will work with you to provide the best equipment to do the job and meet your lifestyle needs.

Everyone, including animals, need oxygen to survive. Without oxygen the internal organs such as the heart, liver, kidneys, brain etc are affected. You cannot become addicted to oxygen – you already are.

As you probably know, oxygen supports combustion. It is one of the elements needed for fire and can be explosive. It is essential that you not be around any flames or high heat sources such as candles, cigarettes, heating pads, etc. You must not smoke while using your oxygen. Also, make sure it is stored at least six feet away from any

source of combustion. Watch out for simple things you may forget about, such as pilot lights or hair dryers. If you cannot manage to dry your hair without using oxygen, have someone else dry it for you while you store the oxygen a safe distance away. You'll be getting a little bit spoiled at the same time, so enjoy it.

Travelling has become much easier with the development of portable oxygen concentrators. These devices run on arechargeable battery pack and can be plugged in to a car lighter or electrical outlet. They are also safe to take on airplanes.

Before flying or travelling to high altitudes, you should consult your doctor. You may be sent for an altitude test which tests your altitude tolerance to airplane pressure of 8,000 feet. You should also discuss your oxygen needs with the airline before booking the flight. Many airlines charge extra for oxygen on board, and some do not allow you to bring your own supply on board. They may also require a doctor's letter confirming your requirements. You might want to get some extra battery packs. Oxygen qualifies as a medical expense for income tax purposes.

LUNG TRANSPLANT

At some stage you may be considered as a candidate for a lung transplant. This is currently the only intervention known to significantly prolong life in IPF patients.

The first successful single lung transplant was done at Toronto General Hospital in 1983. The first double lung transplant was done at the same hospital in 1986. Toronto is one of the most experienced centres for lung transplant in North America, and provides care that is on par with the best programs in the world.

If you are referred there for a transplant, indeed to any Canadian Transplant Unit, you can be sure of first-class treatment.

Forty five percent of lung transplants in Canada go to patients with pulmonary fibrosis.

The criteria for receiving a lung transplant are constantly changing. It used to be that people over the age of 55 were considered too old, now people as old as 74 are receiving new lungs and the operation success rate is very high. New anti-rejection drugs are becoming available that significantly reduce the risk of rejection.

Although there are risks with a lung transplant, the quality of life after a transplant can be vastly improved (the quality of my life certainly did.)

There are, of course, risks, but these risks are being addressed constantly by excellent researchers around the world.

You should know that in Canada, apart from the very limited availability of donor lungs, the only criteria for receiving a lung transplant are:

- **Your blood type must match** with the donor lungs
- **The lung size must be right for you** – you can't put 7 litre lungs in a 4 litre space, or vice versa
- **Your general health** (except for the IPF) and whether you can take an 8 hour operation

With the right patient at the right time, a lung transplant, while being a very risky and major operation, can indeed be a miracle, extending the patient's life expectancy and improving their quality of life.

CLINICAL TRIALS

Ask your doctors whether there are any clinical trials going on that you may be able to join. This has two benefits: firstly you will have the opportunity to try new drugs that, hopefully, will come on the market and which may help you; secondly, you are contributing to important research and that will make you feel good.

WHAT YOU CAN DO

The first and most important thing is to **take charge of your own health care**. Every person is different and this disease is still relatively new to the medical community and the community at large. As mentioned earlier, it is only in the last few years that hospitals across the country have set up Interstitial lung Disease units. There are still a number of physicians, including respirologists, who may not have the most up-to-date information about the disease or the treatments available.

Upon diagnosis, you should ask to be referred to your nearest ILD facility and Lung Transplant Centre. These are the people who are constantly updating their knowledge of IPF. They don't like surprises, such as seeing a patient for the first time when they are in end stage IPF.

HERE ARE SOME OTHER THINGS YOU CAN DO:

Stay in shape. Patients with respiratory issues often find it easier to sit back and not make the effort on their lungs, since it is easier and less painful. But, as mentioned earlier, pulmonary rehab will help you improve the quality of your everyday life. It will also help get you ready for that lung transplant. If you are listed for a transplant, you will be required to attend pulmonary exercise therapy three times a week anyway, so you may as well start now.

Quit smoking. It may go without saying, but it is a hard thing to do, so start as soon as possible. Also, ask your friends and family to avoid smoking around you.

Try to avoid strong smells. Perfumes and cooking smells, in particular, can irritate the lungs and start a painful coughing fit.

Relax, if you can. This will help you cope and ease your mind. It will also help you get enough sleep to maintain your strength.

Try and get your own personal oxymeter. These are very useful to help you keep track of your oxygen saturation so you don't over exert yourself. Talk to your doctor about safe saturation levels.

Let your family and friends help. You will need them more and more as the disease progresses so don't be afraid to ask them. You can always repay them after your transplant and they will feel privileged to be able to help.

Join or form a support group and help others. It will help you to take your mind off your problems. As they say, a problem shared is

a problem solved. The Canadian Pulmonary Fibrosis Foundation is always looking for volunteers to help out by providing support to other sufferers in your area just by talking to them, either in meetings or by telephone. You can all help each other.

Learn to be your own advocate. It is important to learn how to ask questions of your health care providers and be able to tell them what you need in a very focused and clear way. When you go to see your doctor, bring a list of questions or concerns that you may have about your disease and insist that they be answered in a way that you understand. Taking a loved one with you to these visits can also help ensure that your concerns are addressed, as they too can be advocates on your behalf.

Encourage people to sign up to be organ donors. You don't want them to end up being donors before their time, but they will feel happy to support you in this way. Visit Transplant.ca.

Most of all,
NEVER SURRENDER.

NOTES

CANADIAN PULMONARY FIBROSIS FOUNDATION

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