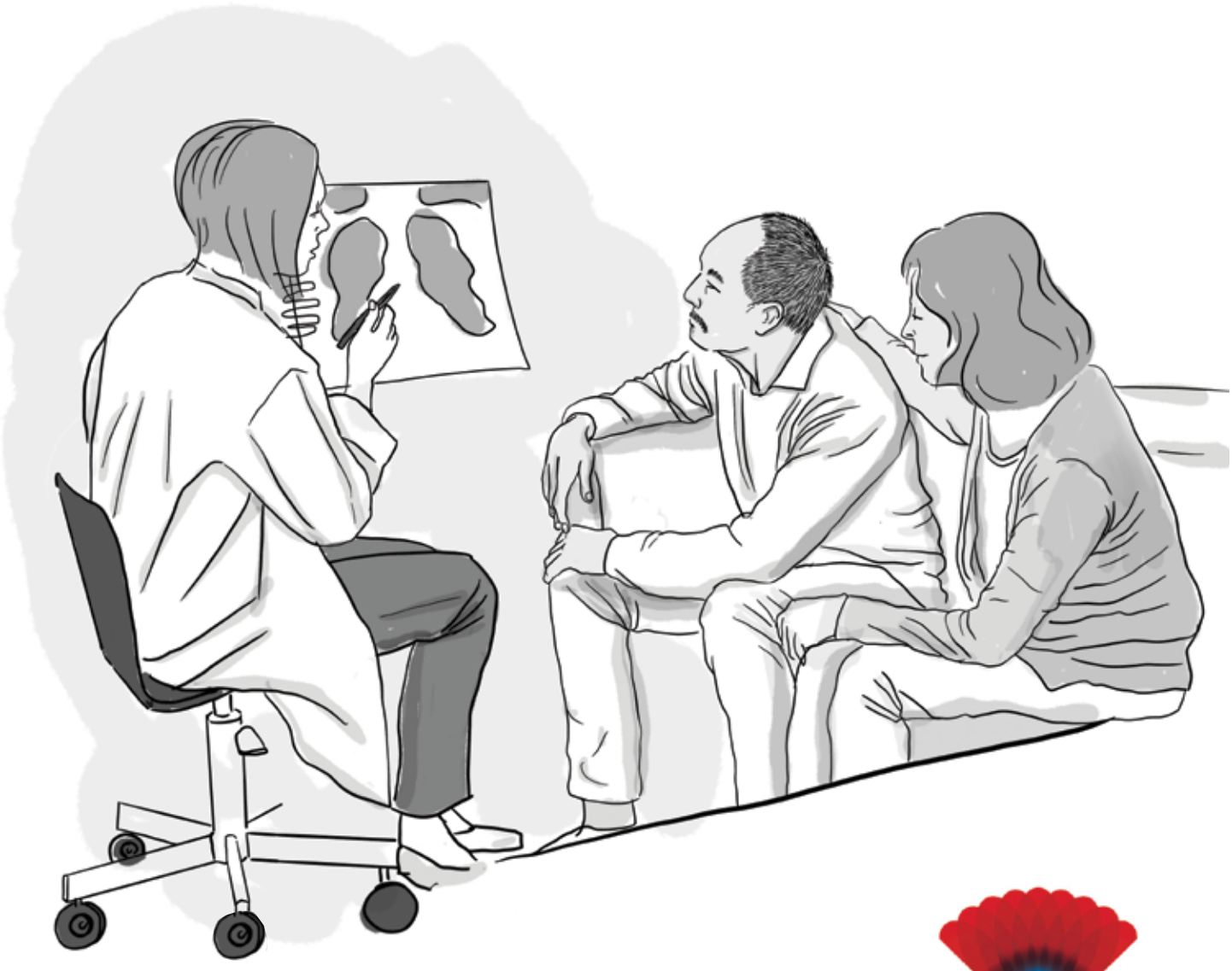
PULMONARY FIBROSIS PATIENT GUIDE



Canadian Pulmonary Fibrosis Foundation

PULMONARY FIBROSIS PATIENT GUIDE

CONTENTS

WARNING SIGNS OF A BREATHING EMERGENCY

If you are having some of the following symptoms: CALL 911

- ☐ Very rapid breathing
- ☐ Extreme tiredness/lethargy or drowsiness
- ☐ Sudden worsening of shortness of breath
- ☐ Pale, greyish skin
- ☐ Sweating
- ☐ Chest pain
- ☐ Confusion

WHEN SHOULD I CALL MY DOCTOR?

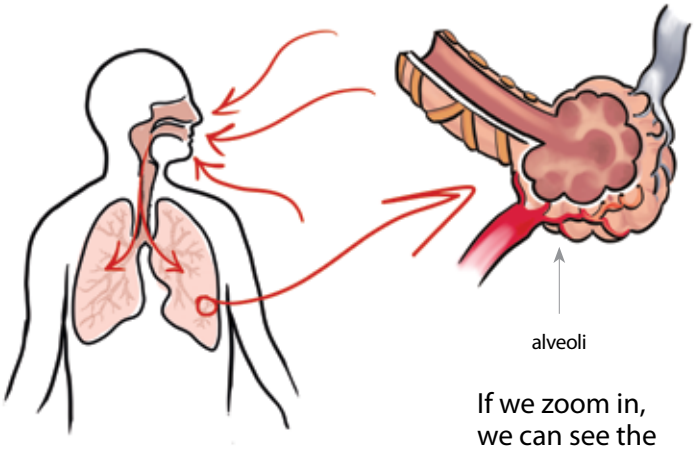
Make an appointment if you have:

- Worsening breathing / shortness of breath
- Increased cough
- Changes to your sputum production
- Fever
- Appetite changes
- Generally feeling unwell

WHAT IS PULMONARY FIBROSIS?

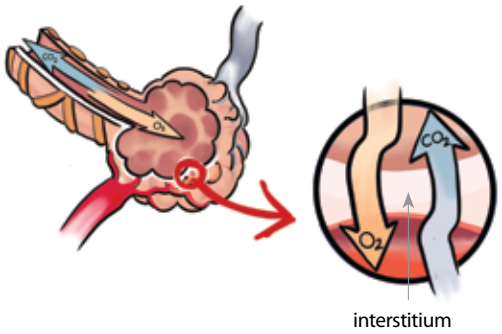
Pulmonary fibrosis (PF) is the common term used to describe a family of more than 200 different lung diseases that cause scarring (**fibrosis**) in the lung (**pulmonary**). Over time, the scar tissue can destroy the normal lung and make it hard for oxygen to get into your blood. Low oxygen levels can cause shortness of breath, especially when walking or exercising.

The PF family of lung diseases falls into a larger group of diseases called the **interstitial lung diseases (ILD)**, which includes all of the diseases that have inflammation and/or scarring in the lung. An estimated 30,000 Canadians are living with pulmonary fibrosis. About half of these have a specific kind of PF called **idiopathic pulmonary fibrosis (IPF)**. It is important to know what specific kind of pulmonary fibrosis you have, as they are all a little different from each other in how they affect breathing, how they progress, and what medications can be used to manage them.



These are your lungs. The **interstitium** is the part of the lung that holds all of the structures together–think of it as the scaffolding of your lung.

If we zoom in, we can see the airways. At the end of these airways are air sacks called **alveoli.**



In healthy lungs, the oxygen that we breathe enters the air sacks, crosses the interstitium and diffuses into our blood vessels to deliver oxygen to our body.

Carbon dioxide - our waste gas - diffuses out of the blood into the alveoli.

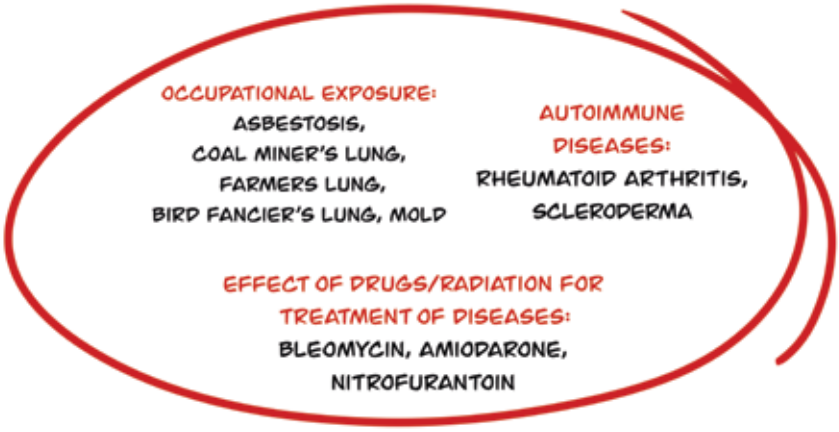
When you have interstitial lung disease (pulmonary fibrosis) there is usually **inflammation and scarring of this interstitium**. It becomes thickened and impairs the flow of oxygen into the body.

AWARENESS

There is limited awareness of PF among the public and health care professionals. It's important to shorten the time between a patient experiencing their first symptoms and being diagnosed since most medications work by delaying disease progression. **The sooner treatments can start, the more impact can be made** by stabilizing the PF patient at a less severe stage of the disease.

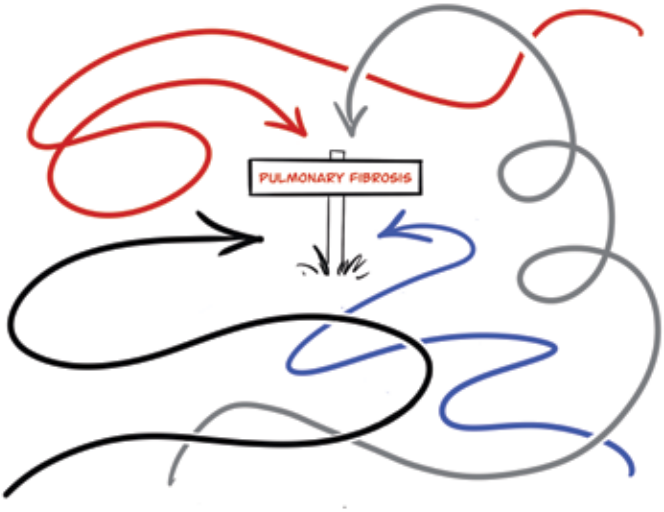
WHAT CAUSES PULMONARY FIBROSIS?

It can be challenging for doctors to figure out what causes PF. Sometimes they are able to identify one or more causes of your disease. Known causes include: occupational exposures to asbestos and inorganic dusts, organic compounds (mold, bacteria...), some medications, autoimmune diseases such as scleroderma or rheumatoid arthritis, chemo or radiation therapy. Smoking is a risk factor in many types of PF. Often, despite extensive testing, specialists are not able to identify a cause for a person's PF. When this happens, the PF is labelled idiopathic. The most common idiopathic form of PF is a specific disease called IPF.



Pulmonary fibrosis is not one disease, it is many diseases.

The cause is not always known.



LIFE EXPECTANCY

Pulmonary fibrosis is a serious disease. The prognosis varies from one patient to another depending on age, other health problems, treatments, and other factors. The average life expectancy varies from person to person. Some people live much longer than the average. Speak with your doctor about the expectations for you and your form of PF.

IDIOPATHIC PULMONARY FIBROSIS

Even after an extensive evaluation, doctors sometimes still do not know what caused your pulmonary fibrosis (PF). The most common type of PF that occurs without cause is idiopathic pulmonary fibrosis (IPF). The word "idiopathic" means a disease of unknown cause. So IPF is a scarring disease of the lungs of unknown cause.

IPF is a chronic, progressive, life-threatening disease of the lungs. IPF is usually diagnosed by:

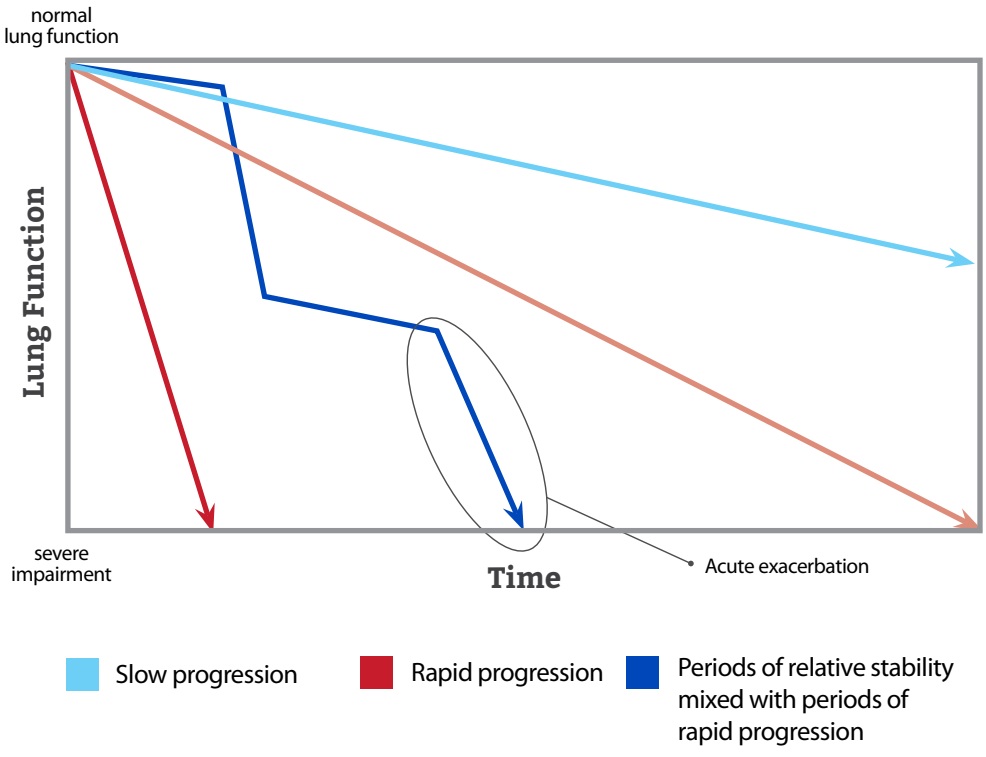
- Ruling out other causes of PF with a thorough history, physical exam and tests
- Looking at your CT scan for a specific pattern of scarring common in IPF
- Looking at lung tissue from a surgical lung biopsy for specific patterns. It is less common to need this test, often the doctor can diagnose IPF without this more invasive test

The course of IPF is unpredictable, and different for everyone. People with IPF can:

- Have symptoms that slowly worsen
- Have symptoms that worsen quickly
- Experience multiple acute exacerbations (medical term for "sudden worsening"), infections, or other complications

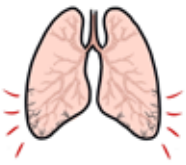
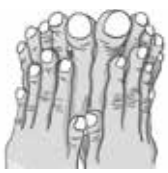
DISEASE PROGRESSION

This graph shows the different ways that IPF behaves. Idiopathic pulmonary fibrosis will worsen over time, causing more scarring in the lung, reducing lung function, and increasing shortness of breath. Some forms of IPF worsen very slowly (light blue line), some at a more intermediate rate (pink line), and some very rapidly (red line). IPF can have periods of more rapid progression (dark blue line); rapid progression can be caused by infections or by acute exacerbations of IPF.



PF SIGNS & SYMPTOMS

People can experience symptoms for a long time before learning they have pulmonary fibrosis. Early symptoms often resemble those of other conditions, and not everyone will have them all. Many of these symptoms are present in other diseases – especially lung diseases. It is important for your doctor to take a thorough history, and conduct a thorough examination. Understanding the symptoms of PF and discussing them with your doctor can help to detect the disease early.

SIGNS & SYMPTOMS	POSSIBLE SIGNS & SYMPTOMS AS PF PROGRESSES
 **SHORTNESS OF BREATH** Scar tissue makes your lungs stiff. Stiff lungs are hard to stretch, causing shortness of breath (breathlessness, dyspnea). It is NOT a normal part of aging. Usually SOB is first noticed with day-to-day activities such as showering, climbing stairs, getting dressed, and eating.	Worsening shortness of breath, even at rest.
 **COUGH** Many people with PF have a cough. It is often a dry cough, but some people may also cough up sputum or phlegm. The cough may be constant, or it might improve or get worse throughout your disease.	Cough becomes more hacking. May include bouts of uncontrollable coughing.
 **LUNG "CRACKLES"** Your doctor will listen to your lungs with a stethoscope. Often with PF, the doctor will hear "crackles" when you breathe, which sounds like Velcro® pulling apart.	
 **WEIGHT LOSS, REDUCED APPETITE, FATIGUE** Some people with PF notice they lose weight without trying and feel more tired than usual. This can be because people with PF use more energy to breathe.	Tiring quickly, even during light activities.
 **DECREASED BLOOD OXYGEN LEVELS** The doctor may do some tests to measure the oxygen in your blood *(see p.7).*	You may need supplemental oxygen therapy *(see p.10).*
 **FINGER AND TOE CLUBBING** Tips of fingers and toes becoming enlarged and the nails becoming curved can be a sign there isn't enough oxygen in your blood.	

LOW OXYGEN LEVELS

Scar tissue and inflammation in your lungs stops the oxygen you breathe from entering your bloodstream. The doctor will check your oxygen levels ("oxygen saturation", or "O2 sats") at rest and with activity, using a pulse oximeter. This is a small painless sensor placed on your finger that uses light to measure how much oxygen is in your blood.

A pulse oximeter provides important information about oxygen levels, and how well you tolerate different activities. Consider purchasing an oximeter to use at home. They are commonly available at online retailers and local pharmacies for $25-$100.

OXYGEN SATURATION LEVELS:

NORMAL	95-100%
LOW NORMAL	< 95%
SUPPLEMENTAL O2 MAY BE REQUIRED	< 90%

SIGNS OF LOW OXYGEN LEVELS

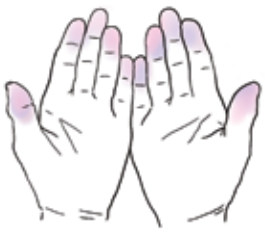
For many people with PF, oxygen levels may only be reduced a little bit while resting, but might drop much more during activity.

Shortness of breath

Fatigue, especially during activity

Weakness or less muscle function

Sleep disorders



Blueish fingertips, earlobes and lips (cyanosis)

Problems concentrating and remembering

TESTING FOR PULMONARY FIBROSIS

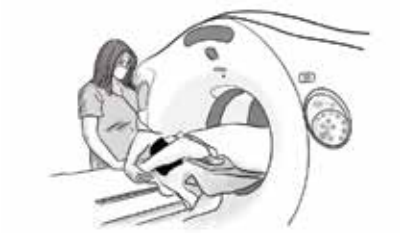
PULMONARY FUNCTION TEST

PFTs monitor how well your lungs are working. You will be asked to breathe into a mouthpiece connected to a machine. PFTs can be tiring because they involve a lot of deep breathing. A respiratory therapist (RT) will coach you through the test.

6 MINUTE WALK TEST

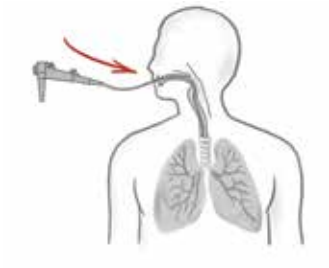
You will walk as far as you can in 6 minutes, usually in a hallway. An RT will measure your oxygen levels, blood pressure, distance walked, muscle fatigue, and how out of breath you are. This will show how your body uses oxygen with exercise, and how much activity you can tolerate.

It is common for people with PF to have low oxygen levels during the test, and to walk a shorter distance as the disease progresses.



HIGH RESOLUTION COMPUTED TOMOGRAPHY

HRCT is a special x-ray that creates a detailed image of your lungs. You will lie in a CT scanner as the pictures are taken. Your doctor will examine the scan for abnormal changes such as scar tissue or inflammation.



BRONCHOSCOPY

A procedure that lets your doctor look at your lungs and air passages. A thin tube (bronchoscope) is passed through your nose or mouth, down your throat, and into your lungs. You will be given a sedative to make you sleepy for the procedure. Not everyone with PF will need a bronchoscopy.

OTHER TESTS:

- Regular blood work
- Arterial blood gas (blood sample taken from an artery, usually in your wrist)
- Echocardiogram (ultrasound of the heart)
- Cardiopulmonary exercise test (performing exercise, usually on a stationary bike, while a doctor monitors your heart and other factors)
- Surgical lung biopsy

TREATING PULMONARY FIBROSIS

MEDICATION

There is a lot of ongoing research to study which medications can help with pulmonary fibrosis. Different medications are used for different causes of PF. It is important to know that current medications slow down or control PF progression; none of the medications stop or cure PF. All of the medications used to treat PF have side effects. It is important to follow your doctor's instructions and do regular bloodwork while taking them.

ANTIFIBROTICS

There are two antifibrotic medications approved in Canada for the treatment of PF: **nintedanib** (Ofev brand) and **pirfenidone** (Esbriet brand and generics available). They do not cure PF, but they may slow pulmonary scar tissue formation.

IMMUNOSUPPRESSANTS

Immunosuppressants suppress the activity of the immune system, which is thought to contribute to lung scarring in some types of PF. They are helpful in some types of PF, and are also used for short periods of time to treat acute exacerbations of PF (see p 4). However they can also be harmful when used long-term in other types, specifically in patients with IPF. **Prednisone** is a commonly used immunosuppressant. Other immunosuppressants include **mycophenolate mofetil**, **azathioprine**, **cyclophosphamide**, and **tacrolimus**.

PULMONARY REHABILITATION

Pulmonary rehabilitation is an important treatment for PF. It includes exercise training, breathing exercises, help with stress, anxiety, and depression, nutritional counseling, disease education and support from professionals and other patients with lung disease.

The goal of pulmonary rehab is to help you function without extreme breathlessness, and to allow you to safely exercise to improve your quality of life.

Some advantages of pulmonary rehab include:

- Helping you to walk further and strengthen your muscles so you can do more activities in your daily life
- Reducing breathlessness
- Learning about managing your disease
- Possible emotional and social support

LUNG TRANSPLANTATION

At some stage you may be considered for lung transplantation to replace one or both damaged lungs. Your doctor will assess if your lung failure is severe enough to need transplantation, if you can safely tolerate the surgery, that you have family support, and that you understand the benefits and risks (infection, rejection, medication side effects, etc).

Not everyone with PF will need a lung transplant. There are various reasons why you may not qualify for a lung transplant, or choose to decline this option. It is a serious and complicated decision you will make with your medical team.

CLINICAL TRIALS

These research studies explore whether a medical strategy, treatment, or device is safe and effective and help researchers understand how a disease works. Clinical trials produce the best data for making health care decisions, and use high standards to protect patients to make sure the results are reliable.

Clinical trials offer an opportunity to participate in important research into understanding PF and discovering new treatments.

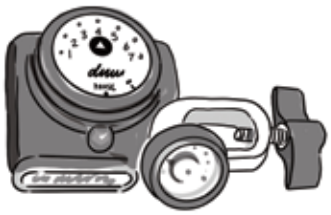
OXYGEN THERAPY

It is common for people with pulmonary fibrosis (PF) to use supplemental oxygen, especially as the disease progresses. Feeling breathless is a complicated process. Some people feel out of breath, but their oxygen levels are within the normal range. And some people won't feel out of breath when their blood oxygen levels are too low (hypoxemia). **Supplemental oxygen will only help you if your blood oxygen levels are too low.**

The **oxygen concentrator** is a machine that plugs in and uses the air in the room to create oxygen that is fed into the tubing.

PRO TIP!
A new cannula/tubing is tightly coiled and unwieldy. Soak it in hot water for about an hour before using to soften it. Dry the tubing completely so it does not get moldy.

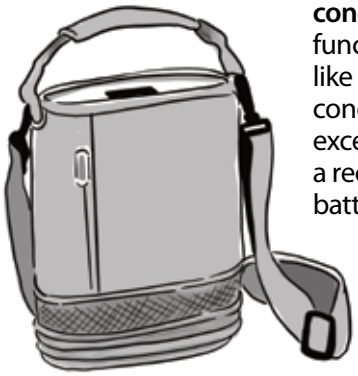
The **oxygen cannula** loops around the ears to deliver oxygen to the nose. It is sometimes attached to the **oxygen conserving device** with a long flexible tube.



Oxygen conserving devices are attached to cylinders. A dial shows when oxygen is running low.

Oxygen cylinders (compressed gas) come in different sizes/capacities.

A **liquid oxygen system** stores oxygen in liquid form and converts it to gas.



Portable concentrators function like regular concentrators, except they use a rechargeable battery.

Supplemental oxygen will not cure your PF – and it may not eliminate your breathlessness – but it can help you maintain proper blood oxygen levels. This is important for your muscles, organs, and brain to function properly. You cannot get addicted to oxygen. It is important to treat your oxygen as a prescription and use it exactly as your doctor prescribes.

It can take time to adjust to using supplemental oxygen. For example, it may be more challenging leaving the house and carrying your oxygen. On the other hand, it can also make some things easier. Since you may experience less breathlessness, you may get less tired doing activities.

LIVING WITH PULMONARY FIBROSIS

STAYING HEALTHY

Staying as healthy as possible is very important. Things you can do to stay healthy include:

- Avoid sick people, especially if they have a cold or other breathing infections
- Consider avoiding large crowds, or wearing a mask to protect yourself from infection
- Wash your hands often, and properly with soap and water for 20 seconds
- Use hand sanitizer if you are unable to wash your hands
- Avoid touching your face
- Get regular exercise
- Stop smoking

Stop smoking

Exercise

MANAGING YOUR ENERGY

It is common for people with pulmonary fibrosis (PF) to have low energy levels. This can be because of the effort to breathe, the medications you take, and the effects of your PF. Things you can do to help manage your energy include:

- Pace yourself - work at a slow, steady pace. Leave extra time to get tasks finished, and take breaks
- Eat smaller meals more often if you find eating causes breathlessness
- Adjust your expectations - you may not be able to complete the same tasks as before you were diagnosed with PF. Try to modify activities so you can safely complete them
- Let your family know your limitations, and how they can help you
- Eat a balanced, healthy diet. Proper nutrition can give you the fuel you need to do your daily activities

Pace yourself – and/or ask for help with household tasks

IMMUNIZATIONS

Talk to your doctor to make sure your immunizations (vaccines) are up to date. Some important immunizations to get include:

- COVID-19
- Influenza - get your flu shot every year in October or November
- Pneumococcal - There are two different pneumonia vaccines. Talk to your doctor about which one is right for you
- Shingles

Eat healthy

SUPPORT GROUPS

PF is a rare disease, and it can be challenging to find other people who share your experiences.

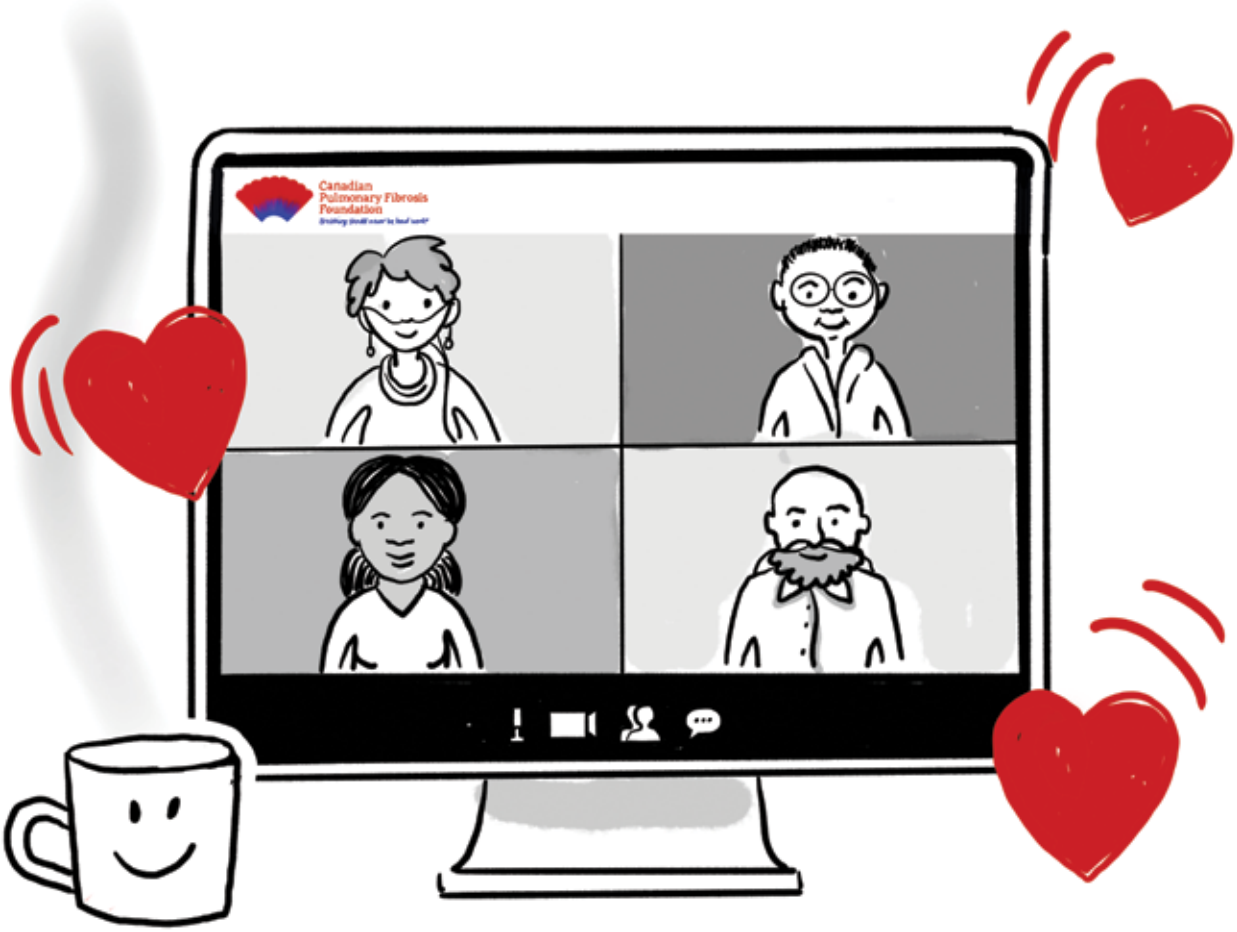
YOU ARE NOT ALONE!

There are many support groups in Canada where people with PF meet virtually or in person to discuss their fears, challenges, and share knowledge about their disease. It can be an important source of support for you and your family.

Visit

cpff.ca

for a list of support groups near you.



GLOSSARY

ACUTE EXACERBATION

A sudden, serious worsening of symptoms over the course of days or weeks. It may include increased breathlessness and cough, the need for more oxygen, general feeling of unwellness, and fever.

In some cases, there is no obvious explanation. Some people will recover to a certain degree, and others will never be the same, or continue to get worse.

ALVEOLI

Tiny air sacs at the end of the bronchioles (tiny branches of air tubes in the lungs). The alveoli are where the lungs and the blood exchange oxygen and carbon dioxide during the process of breathing in and breathing out.

CLINICAL TRIAL

Clinical trials are research studies that explore whether a medical strategy, treatment, or device is safe and effective. They also help researchers understand how a disease works. Clinical trials produce the best data for making health care decisions. Clinical trials use high standards to protect patients, and to make sure the results are reliable.

Clinical trials offer an opportunity to participate in important research into understanding PF and discovering new treatments.

IDOPATHIC PULMONARY FIBROSIS (IPF)

Pulmonary fibrosis with no known cause.

INTERSTITIAL LUNG DISEASE (ILD)

Interstitial lung disease refers to a group of over 200 chronic lung disorders characterized by inflammation and scarring that make it hard for the lungs to get enough oxygen. The scarring is called pulmonary fibrosis.

INTERSTITIUM

The interstitium is the part of the lung that holds all of the structures together like scaffolding on a building.

PULMONARY FIBROSIS

A disease where the lungs (pulmonary) become scarred (fibrosis). Over time, the scar tissue can destroy the normal lung and make it hard for oxygen to get into your blood.

NOTES

Canadian Pulmonary Fibrosis Foundation

The Canadian Pulmonary Fibrosis Foundation is a registered charity established in 2009 by Robert Davidson. The CPFF was created to raise money to research causes and treatments for PF, provide education and support for people affected by PF and their caregivers, raise awareness about PF, and represent Canadians affected by PF to governments, healthcare professionals, the media, and the public.

The Canadian Pulmonary Fibrosis Foundation works tirelessly to:

SUPPORT

EDUCATE

RESEARCH

ADVOCATE

For more information, please contact:

info@cpff.ca

or

905-294-7645

Charitable Registration # 850554858RR0001

Visit

cpff.ca

- Information about pulmonary fibrosis
- Patient and caregiver resources
- CPFF newsletter sign-up
- How to get involved

DEDICATION

Robert Davidson

The CPFF Pulmonary Fibrosis Patient Guide is dedicated to Robert Davidson, who lived with PF, and his family. Robert, who wrote the first edition of this guide, died peacefully in 2019, nine years after his double lung transplant.

THANK YOU!

Special thanks to Melissa Sulpher, and to Dr. Gokul Vidyasankar and Dr. Charlene Fell for their invaluable oversight and contributions.

Melissa Sulpher *is a Registered Nurse who has been living with pulmonary fibrosis for over 15 years.*

Gokul Vidyasankar, *B. Eng, MD FRCPC (Resp)Division Head of Respirology, Eastern Health Associate Professor of Medicine, Memorial University.*

Charlene Fell, *MD Founding Chair of the Canadian Thoracic Society's Clinical Assembly on Interstitial Lung Diseases, Director of the University of Calgary Interstitial Lung Disease Program.*

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