



Idiopathic Pulmonary Fibrosis (IPF)

A GUIDE TO LIVING WITH IDIOPATHIC PULMONARY FIBROSIS

THANK YOU

A special thank you to those who made this guide possible.

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DEDICATION

For those people who have had their breath taken away from Idiopathic Pulmonary Fibrosis. This guide is for you and your loved ones.

LUNG SASKATCHEWAN

At Lung Saskatchewan we work to deliver on our mission: to improve lung health one breath at a time. Our commitment to improving lung health in Saskatchewan began in 1911 during the tuberculosis epidemic that swept across the province and Canada. Today, we continue to fight for the 1 in 5 people in Canada affected by lung diseases.

Thanks to our work, Saskatchewan has seen tremendous strides in lung health education, research, health promotion, and advocacy. It is through these four priorities we are able to offer best-in-class, timely, and relevant opportunities to all of our stakeholders.



CHAPTER 1: IDIOPATHIC PULMONARY FIBROSIS (IPF)

We often don't think about our breathing until something takes our breath away. Idiopathic Pulmonary Fibrosis (IPF) may be doing just that for you or someone you love. There is currently no cure for IPF. However, this book has been created to help you have a better understanding of the disease, to understand possible treatment options, to know what you can do to manage your symptoms and to assist you in preparing for your future.

What is IPF?

Pulmonary fibrosis is a long-term disease where the lungs (*pulmonary*) become scarred (*fibrosis*). The scarring causes the part of the lung where oxygen is moved to the blood (*interstitial*) to become thick and stiff. If the reason or cause of the disease is unknown it is called *idiopathic*. Some people refer to pulmonary fibrosis as a *restrictive disease* because it is hard to fully expand the lungs when taking a breath in.

I – Idiopathic (unknown cause)

P – Pulmonary (lungs)

F – Fibrosis (scar tissue)

Pulmonary fibrosis and idiopathic pulmonary fibrosis are a subgroup of *interstitial lung diseases* called *interstitial pneumonia*. Among the many interstitial lung diseases, Idiopathic Pulmonary Fibrosis (IPF) is the most common. It is more common in males over the age of 50.



Moe Martin, Double Lung Transplant Recipient, IPF

Signs and Symptoms

When the lungs become scarred, it is hard to get oxygen to the organs and body. The lack of oxygen can cause different signs and symptoms. Knowing what to look for is important so you and your health care provider can come up with a plan to diagnose the problem.

Do you have any of these signs and symptoms?

- ❑ **Shortness of breath** - Shortness of breath is not a normal part of aging. It is one of the most common symptoms of IPF. Many people first notice it when exercising or climbing the stairs leaves them more breathless than usual. As IPF gets worse, people become more short of breath.
- ❑ **Dry cough** - Most people with IPF have a dry and frequent cough. If you have a cough that lasts longer than 3 weeks, please see a health care provider.
- ❑ **Inspiratory crackles** - This is a 'velcro-like' crackling sound in the lungs upon taking a breath in. It is heard with a stethoscope.
- ❑ **Digit/finger clubbing** - About half of people with IPF have rounding and widening of the fingers and toes. This may occur when there isn't enough oxygen in the blood.
- ❑ **Weight loss and feeling unwell** - Some people with IPF notice they lose weight without trying, have a general feeling of being unwell and feel more tired than usual. This is because the disease requires people to use more energy to breathe.
- ❑ **Pedal edema** - As the disease gets worse, a slow development of leg swelling (edema), might be seen. This swelling happens when the heart pumps less blood throughout the body, which causes the body to hold onto more fluid. The extra fluid builds up in the lowest area of the body, the legs.

These signs and symptoms do not mean you have IPF. They may also be as a result of other conditions.

If you have any of these signs and symptoms, tell your health care provider today.

Pulmonary fibrosis is often divided in two categories.

- 1. Known Cause:** Certain environmental exposures (things from where we live or work), medications for inflammation (anti-inflammatory), cancer (chemotherapy), infection (antibiotics), the heart (cardiac), the stomach (gastrointestinal) and the brain (neurological) can cause scarring in the lungs. Radiation, certain inhaled drugs or chemicals (cocaine, zinc chloride, ammonia) and connective tissue disease (rheumatoid arthritis, polymyositis, and scleroderma) are also linked to pulmonary fibrosis.
- 2. Unknown Cause:** As the name (*idiopathic*) suggests, we do not know what causes IPF. There are different factors that may increase a person's chances of getting the disease.

Possible IPF Risk Factors

- **Smoking** – Our lungs are meant to inhale oxygen. Anything that is burnt is harmful to our breathing. Smoking is strongly linked to causing many lung diseases including IPF.
- **Environmental Exposures** – A variety of environmental exposures have been shown to increase the risk of getting IPF. Certain jobs and occupations such as farming, raising birds, hair dressing, and stone cutting/polishing may be connected. Contact with livestock and different kinds of dust (metal, wood, vegetable and animal) are also risk factors.
- **Viruses** – Researchers have been looking at the possibility of viral infections being associated with IPF. There has been a focus on *Epstein-Barr* (a virus in the herpes family that is best known to cause infectious mononucleosis often referred to as mono) and *hepatitis C* (a viral infection that can cause liver damage) specifically. Despite the likelihood of an association between IPF and these viruses, research has not been able to say for sure that they cause IPF.
- **Gastroesophageal Reflux Disease (GERD)** - Gastroesophageal reflux disease (GERD) is a chronic digestive disease. GERD occurs when stomach acid or content flows back into your food pipe (esophagus). The backwash (reflux) irritates the lining of your esophagus and causes GERD. This may also be related to IPF, however, more knowledge and research is still needed in this area.

- **Genetics** - Genetics can play a role in IPF. A very small percentage of people with IPF have reported two or more family members with the disease.
- **Gender** – IPF is more common in males.
- **Age** - IPF is more common in people 50 years and older. The risk of getting IPF for both women and men increases with age.

Disease Course

An early and accurate diagnosis of IPF is key. Being diagnosed at an earlier stage of the disease may allow you to have more treatment options and improve your quality of life.

Learning you have IPF is very difficult. Understanding the disease, treatment options, and ways to manage your symptoms will help you plan your future. Many people are told they have IPF after their disease is quite advanced and they are already very sick. Everyone's journey is different. The average survival rate is 2-3 years from the time of diagnosis. There are people with IPF who live much longer.

Some risk factors, such as smoking, can also make the disease get worse at a faster rate. The course of IPF is unpredictable.

People with IPF can:

- Have symptoms that slowly get worse over time.
- Have symptoms that get worse very quickly.
- Experience various *acute exacerbations*, which consist of sudden worsening in symptoms, and there is an abrupt decline in how the lungs function. After an *acute exacerbation*, some people will recover to a certain degree and others will never be the same or continue to get worse. People with IPF can also suffer from infection, or from unknown complications.

How fast and how much a person's lung function declines helps health care providers predict how advanced the disease is, however no two people will have the exact same experience.

Chapter 1 Summary

- As the name suggests, Idiopathic Pulmonary Fibrosis (IPF), is a lung disease with unknown cause.
- IPF belongs to a group of lung conditions called Interstitial Lung Diseases (ILD) which cause scarring of the tissues in the lungs. The scarred tissues in the lungs cause the area in which oxygen is moved into the blood to become stiff and thick. This leads to reduced oxygen in the blood.
- IPF is a long-lasting (chronic) disease that gets worse over time. Everyone with the disease will progress differently.
- IPF is the most common of the Interstitial Lung Diseases (ILDs).
- The most common IPF symptoms are: shortness of breath, frequent dry cough, inspiratory crackles, clubbing of the fingers and toes, and pedal edema.
- Although there is no known cause for IPF, different factors are connected to IPF such as: smoking, contact with livestock and various kinds of dust. Farming, raising birds, chemicals from hairdressing, and stone cutting/polishing viruses, Gastroesophageal Reflux Disease (GERD), age (50+), and male gender are also risk factors.

“From our first breath to our last breath, the air that we breathe in and out is our greatest physical interaction with the world around us.”

Dr. Brian Graham

Past President and CEO
The Lung Association of Saskatchewan (now Lung Saskatchewan)

CHAPTER 2: DIAGNOSIS

How do I find out if I have IPF?

There are many different tests available that your doctor may recommend to help you find the right diagnosis. If you have any symptoms or you think you may be at risk for IPF, be sure to talk to your doctor. The correct diagnosis and finding out as early as possible is important, as it may have an impact on the type of treatment you receive. You may meet with different doctors and specialists to help determine an accurate diagnosis.

Because there is not one particular test to diagnose IPF, it may take some time to find out if you have the disease. You will likely need to have many different tests. The tests listed below will help your doctor make the right diagnosis for you. Sometimes doctors have to rule out other possible diseases first. They often will start with the tests that are the least invasive and only order more if needed. These tests will help you and your doctor get a better understanding of your condition.

Tests to Find Out if You have IPF

- **Medical History:** Your health care provider will first ask you about your health and your family's health. They will want to know any symptoms you may be having (example: coughing or shortness of breath during physical activity) and if you have any other medical conditions. Letting them know about any medications you currently take or have taken in the past is also important. Be sure to also tell your doctor where you have lived and worked, and if you have ever smoked. The more details, the better.
- **Physical Examination:** A physical examination is the most common test. Your health care provider will check for any visible signs and symptoms that could indicate IPF. They will likely look for rounding and widening of your fingers and toes (clubbing), listen for a dry cough, and using a stethoscope, try to hear any crackling sounds in your lungs.



- **Pulse Oximetry:** This measures the amount of oxygen that is carried in the blood (oxygen saturation). This method uses a small sensor device which is usually placed on one of your fingers to determine how much oxygen is in your blood. The oxygen level will be given a percentage. This can help your doctor decide if you will need supplemental oxygen.
- **Arterial Blood Gas (ABG):** This test also measures the amount of oxygen that is carried in your blood. It is very accurate but more invasive than pulse oximetry. A blood sample is usually in your wrist with a needle from an artery and then analyzed to see how much oxygen is in your blood.
- **Pulmonary Function Test (PFT):** A PFT is a series of breathing tests used to find out how well you move air in and out of your lungs and how well oxygen enters your bloodstream from your lungs.
- **Six Minute Walk Test:** Shortness of breath when exercising or doing physical activity is usually the first symptom people notice with IPF. Because of this, the six minute walk test is often used if IPF is suspected. For this test you will be asked to walk for six minutes at your own pace. While walking the tester will measure the amount of oxygen in your blood using a pulse oximeter. This test measures how far you can walk in six minutes, how difficult it was for you to complete the test, and if your oxygen levels in your blood dropped with exercise.

- **Blood Test:** Although a blood test cannot confirm IPF, it can be performed to help rule out other lung-related conditions. This is an essential part of determining the correct diagnosis.
- **Chest radiography (x-ray):** A chest radiograph or x-ray is a test that uses electromagnetic waves to create pictures of the structures in and around your chest. This will often provide some useful information such as the size of your lungs, help rule out other lung diseases and show possible distribution of disease in the lungs.



- **Bronchoscopy:** A bronchoscopy is a test that allows the doctor to look through your nose or mouth into the trachea (windpipe) and down to the bronchi (large airways) in your lungs. This is done with a thin, flexible tube with a light and camera on the end called a bronchoscope. Your doctor can take tissue or fluid samples if needed, through a channel in the bronchoscope. The bronchoscopy takes about 30-60 minutes but the appointment will likely take a few hours. You will have an intravenous (IV) line inserted to allow you to receive sedation (medicine to make you sleepy). You will be sleepy, but not completely asleep during the test. The doctor will also spray liquid freezing (anesthetic) into your mouth and throat to make them numb. It is normal to cough during the procedure. A bronchoscopy might be uncomfortable but is not painful. Your oxygen level and blood pressure will be monitored throughout the bronchoscopy. You will be given oxygen to breathe through your nose during the test.
- **High Resolution Computed Tomography (HRCT):** This test is also called a CAT or CT scan. CT scanners take many detailed x-rays that are blended together by a computer. You lie down in the long tube of the CT scanner and stay very still

as the x-rays are being taken. You will be monitored by a health care provider from a separate room. This process usually takes 15-30 minutes and is very important in confirming IPF.

- **Lung Biopsy:** If your diagnosis is still unclear with the other tests listed above, you may have a lung biopsy. This test involves small incisions into the side of the chest to remove a small part of the lung tissue and examine it under a microscope. Prior to the procedure, the chest is cleaned and a needle is administered to numb the area. If you have this test done, you will need to stay in the hospital for a few days. Although there are risks involved with this surgical procedure, it can provide a more definitive confirmation if needed.

Preparing for Your Tests

Like any test, you may be given specific instructions to follow to help you get ready for the exam. Be sure to ask your health care provider if you should eat, drink or take your regular medications before your tests. Let them know if you are experiencing any changes in your health since they last saw you (fever, worsening of symptoms, new symptoms etc.). Also find out what you should wear and bring to your test so you are best prepared.

Diagnosing IPF

Some of the tests you may have done might be conducted to rule out other conditions, or to help monitor the development of IPF. In order to diagnose you with IPF doctors must:

1. Rule out other known causes of interstitial lung disease (toxins, environmental or occupational exposures).
2. See a specific pattern on the chest x-ray, referred to as 'usual interstitial pneumonia pattern', which shows scarring and other abnormalities in the lung tissues.
3. See a specific pattern in the lung tissue from the CT scan or surgical lung biopsy.



Talk about it: Talk about your worries and fears. Reach out to family and friends, your health care providers, a counsellor or someone that may give you spiritual guidance. Some people like to talk to people who are in a similar situation. There are support groups and communities of people living with lung disease.

Learn more

Understanding the disease and knowing what to expect is important. It will likely help you better manage your symptoms and inform you about the different ways to prepare for your future. Reading this guide is a great way to educate yourself about IPF.

Finding out you have IPF

There is no right or wrong way to feel when you are given a diagnosis. It is normal to feel many emotions. You may feel shocked, angry, scared, sad, resentful, helpless, or frustrated. Some people say they felt alone when they were told they had IPF even though they were surrounded by family or friends. It is important to know, you are not alone.

Ask questions

You will have questions and concerns. Write them down and ask your team of health care providers. They will help answer your questions and link you to other resources in your community.



Chapter 2 Summary

- Talk to your doctor if others in your family have IPF or if you have any signs and symptoms common to IPF.
- To help determine a diagnosis or what tests you may need, tell your doctor about any other conditions you have, if you currently smoke or have smoked and provide them with a list of your current and past medications. Don't forget to mention where you have lived and worked. Certain things in your environment may have put you at risk for IPF.
- There is not one particular test to find out if you have IPF. You may need to have many different tests.
- Finding out you have IPF is difficult. Remember you are not alone and there are people to help you.
- Talk about IPF, learn more and don't be scared to ask questions.



Nicole Nelson, Lung Ambassador, Diagnosed with pulmonary fibrosis at 18 years old

CHAPTER 3: COMORBIDITIES, MULTI-MORBIDITIES AND IPF

Having IPF may increase your risk of developing other illnesses, and having an additional illness may cause your IPF to become worse.

If you have IPF along with another long-term illness, it is called a *comorbidity*. If you have two or more illnesses at the same time, it is referred to as a *multi-morbidity*.

Comorbidities are Common to those with IPF

- **Pulmonary infection** is an infection in the lung.

It is the most common comorbidity among people with IPF and can be caused by viruses, bacteria, or fungi. A pulmonary infection can also cause an acute exacerbation (sudden worsening of breathing symptoms).

- **Emphysema** occurs when the air sacs in your lungs are damaged, making it harder for you to breathe out completely. Emphysema is part of a lung disease called chronic obstructive pulmonary disease (COPD). Smoking is a key risk factor for both IPF and emphysema.

- **Pulmonary hypertension** is high blood pressure in the arteries of the right side of the heart. This condition is associated with acute exacerbations (sudden worsening of breathing) and can cause IPF to worsen quickly. Pulmonary hypertension in IPF is likely from many factors. The narrowing of the blood vessels and arteries may be from low oxygen levels that are usually seen in IPF.

- **Lung cancer** is a type of cancer where cancer cells grow out of control, taking over normal cells and organs in the body. Lung cancer starts in the lungs.

The two major types of lung cancer are;

1. Small cell lung cancer
2. Non-small cell lung cancer.

Of the two types of lung cancer, non-small cell lung cancer is the most common. It is slower to spread and grow compared to small cell lung cancer.

The risk of developing lung cancer is about seven times higher in people with IPF. The most common risk factor for IPF and lung cancer is smoking. Further research is needed to know for sure if aggressive treatments are appropriate and beneficial for people with both IPF and lung cancer. Most people with IPF and lung

cancer who are not treated surgically have chemotherapy. However, because only a few studies have examined the use of chemotherapy for people who suffer from lung cancer and IPF, the best treatment plan is still not known. More research is needed to explore the best treatment options for people with lung cancer and IPF.

- **Gastroesophageal Reflux Disease (GERD)**

is a long term disease which can irritate the digestive system. This occurs when acid or other contents from the stomach flow back into your esophagus (your food pipe). The acid/stomach content can irritate the lining of your esophagus, causing a burning sensation in your chest, or chest pain. It can also make swallowing difficult, your throat to become sore or hoarse, food to come back up, and/or a feeling of a lump in your throat. People with IPF are more likely to develop GERD. Antacid therapy is usually recommended. Treating GERD may improve survival and slow down the progression of IPF. However, the effect of anti-acid therapy to help manage GERD in patients with IPF requires further investigation.

- **Cardiovascular disease** is commonly referred to as “heart disease.” Cardiovascular disease usually includes conditions that involve constricted or blocked blood vessels which can lead to a heart attack, chest pain, or a stroke. People with IPF are more likely to have cardiovascular disease. Although the exact reason for this is unclear, some researchers think lung injury and repair may affect the way the blood clots, which can cause a narrowing of the arteries. Another way in which cardiovascular disease and IPF may be linked is the lack of oxygen due to the scarring of the lungs. People with IPF are at least twice as likely to have coronary artery disease. Coronary artery disease is when plaque builds up in the coronary arteries. Further studies are needed on what the best treatment is for those with both IPF and cardiovascular disease. Unfortunately, some of the treatment options for cardiovascular disease can cause an even greater risk to the person with IPF.

- **Diabetes mellitus** is a disease in which the body either cannot produce insulin or cannot properly use the insulin it does produce. This leads to high levels of glucose (sugar) in the

blood, which can damage organs, blood vessels and nerves. You have a greater risk of developing diabetes if you have IPF. Having both these diseases may make your IPF worse sooner.

- **Obstructive sleep apnea**, also known as OSA or obstructive sleep apnea-hypopnea syndrome, means you have apneas (short pauses in your breathing when you sleep). These apneas may last for 10 to 30 seconds or even longer. People with obstructive sleep apnea often snore and can stop breathing dozens or hundreds of times each night. This can lead to low levels of oxygen and sleep disruption. Untreated sleep apnea can cause people to not only feel tired, but it can also affect their memory, and put them at risk for heart disease or a stroke.

More and more people with IPF are being diagnosed with obstructive sleep apnea. The increased risk seems to be related to a lower lung capacity and severe restriction of the lungs.

The best treatment for obstructive sleep apnea is continuous positive airway pressure (CPAP) produced by a flow generator, also called a CPAP machine. This pressure is delivered to your airway through tubing and a mask which is held in place by headgear. This creates a 'splint' that supports the tissues at the back of the throat, holds the airway open, and prevents the airway from collapsing. Once your airway is open, the pauses in your breathing and snoring are stopped. If you stop using the CPAP machine, your symptoms will return. CPAP treatment in patients with both IPF and obstructive sleep apnea (moderate to severe) have resulted in significant improvement in their daily living activities, and in both the quality of sleep and life. A recent study found that correct usage of the CPAP machine in people with IPF and obstructive sleep apnea also appeared to allow people to live longer.

- **Depression** is a medical illness that negatively affects how you feel, the way you think, and how you act. Depression causes feelings of sadness and may cause a loss of interest in activities previously enjoyed.

It is upsetting, scary, and challenging to be diagnosed with IPF. Many people feel depressed when diagnosed, or over time as their symptoms begin to worsen. Recognizing the symptoms of depression and asking for help is important.

Symptoms of depression can vary from mild to severe and can include:

- Feeling sad or having a depressed mood
- Loss of interest or pleasure in activities previously enjoyed
- Changes in appetite — weight loss or gain not related to dieting
- Trouble sleeping or sleeping too much
- Loss of energy or feeling more tired than usual
- Increase in purposeless physical activity (e.g., hand-wringing or pacing) or slowed movements and speech (actions observable by others)
- Feeling worthless or guilty
- Difficulty thinking, concentrating or making decisions
- Thoughts of death or suicide

Being diagnosed with IPF is difficult. Many people may also have another condition or disease (comorbidity) that accompanies IPF. Receiving an early diagnosis and the right treatment of the comorbidities is very important. Despite the fact that recent studies have provided health care professionals guidance for the management of comorbidities, most treatment recommendations are based on studies done with people who do not have IPF.

More research is needed to better understand how these different diseases impact people with IPF, how they are linked, and how they should best be treated. Talk to your doctor about these comorbidities to learn how they can screen for them. Having more than one illness may affect how fast your IPF worsens, how your disease is treated, and your overall quality of life.

IPF is complicated and when people have more than one illness, it becomes even more difficult to treat and understand. Often people with IPF work with a team of health care providers, referred to as a *multidisciplinary team* who specialize in different diseases and areas of health. If you have more than one illness, you may have even more people on your multidisciplinary team to ensure you are getting the specialized care you need.



Your multidisciplinary team may include but are not limited to the following:

- **Primary physician** - a doctor that is generally in charge of your overall care.
- **Respirologist** - a specialist that is responsible for treatment decisions and assessments of lung function.
- **Psychologist** - a professional that will provide you with counselling services and coping strategies.
- **Pathologist** - a scientist or medical doctor who specialises in pathology, which is the diagnosing of disease using organs, tissue, and body fluids.
- **Dietitian** - a health care professional that will help you develop a plan that will meet your dietary and nutritional needs.
- **Occupational therapist (OT)** - a professional that will provide services to restore self-care (personal care, mobility), leisure (social activities, sports) and productivity (play, school, employment, homemaking work and leisure skills). Occupation refers to the activities and tasks of daily life that have value and meaning to a person.
- **Respiratory Therapist (RT)** - a health care professional that cares for patients by evaluating, treating, and maintaining cardiopulmonary (heart and lung) function.
- **Social Worker** - A professional concerned with helping you and your family enhance your individual

and collective wellbeing. They aim to help you develop skills and abilities to cope with your disease, as well as link you to resources in the community that can support and assist you with your needs.

- **Physical Therapist (PT) and Exercise Therapist (ET)** - professionals that are responsible for developing an individualized exercise program and helping you manage your disease and overall health.
- **Radiologist** - a health care professional that will conduct chest x-rays and high-resolution computed tomography (CT) scans.
- **Pharmacist** - a health care professional that will dispense your medications.
- **Respiratory Nurse (RN)** - a health professional that supports you in your care plan.

A multidisciplinary team must have effective communication, as some of your health care professionals may not work in the same place. It is important that your primary physician is aware of everyone that is helping you as well as all of the medications (both prescribed, over the counter, and herbal/natural supplements) that you are taking. Even though your conditions may not have a cure, your multidisciplinary team can show you various ways to best manage your symptoms and improve your quality of life.

Chapter 3 Summary

- Many people with IPF may also have another long-term illness which is called a *comorbidity*.
- Two or more illnesses at the same time is referred to as a *multi-morbidity*.
- Pulmonary infection, emphysema, pulmonary hypertension, and lung cancer are some of the common comorbidities associated with IPF.
- Gastroesophageal Reflux Disease (GERD) and cardiovascular disease are also seen more often in people with IPF.
- People with IPF also have a greater risk of developing diabetes. Having both IPF and diabetes can make IPF worsen more rapidly.
- More people with IPF are also being treated for obstructive sleep apnea (OSA). OSA means you have apneas, which are short pauses in your breathing when you sleep.
- It is common for people with IPF to experience depression. It is important to recognize the warning signs of depression and ask for help.
- A *multidisciplinary* team is a team of health care professionals who specialize in different areas of health. Most people with IPF will have several health care professionals helping them with their care.

“With a vision of healthy lungs for everyone, our mission is to improve lung health one breath at a time.”

Lung Saskatchewan

CHAPTER 4: MANAGEMENT

IPF is a chronic or long-term disease, which means other than a lung transplant, it cannot be cured. There are however, ways to help manage IPF.

Medications

Currently no medications exist that can cure IPF or decrease the amount of scarring that is already in the lungs. Previously, there have been very few medications available for people with IPF. Today there are medications available that help to slow the progression of the disease. Recent research discoveries have made new medications available and clinical trials are showing promise for more options in the future.

Everyone is different and treatment options will vary from person to person. The decision to use any medication for IPF should be approached carefully. Medications for IPF may have mild to severe side effects. It is important to talk to your doctor about different treatment options and learn about potential medication side effects.



Medications Commonly Used to Treat IPF

- **Pirfenidone (Esbriet®):** an anti-scarring (anti-fibrotic) medication that slows the progression of IPF. Some people taking Pirfenidone have had side effects, most commonly stomach upset and skin rash, particularly with exposure to sun. Pirfenidone has been approved by Health Canada for the treatment of mild to moderate IPF.
- **Nintedanib (OFEV®):** an anti-scarring (anti-fibrotic) medication that slows development of IPF. The most common side effect for this medication is diarrhea.



- **Corticosteroid pills (Prednisone®):** an anti-inflammatory medication which can reduce the swelling in the airways by lowering the immune system. Corticosteroids are only used when a person with IPF has an acute exacerbation (sudden worsening in symptoms, and a decline in lung function). Corticosteroids can be harmful for people with IPF that have scarring that is not getting worse or is getting worse slowly.

Some medications such as Warfarin®, Imatinib®, Ambrisentan®, cyclophosphamide®, and a combination of prednisone®, azathioprine® and N-acetylcysteine® together are no longer recommended as a general guideline for IPF therapy. Recent studies of these medications have demonstrated a lack of benefit and have shown potential harm in people with IPF. Often the negative side effects and costs to the drugs outweighed any possible benefit. However, people who have another and/or known reason to be treated with anticoagulation (medication that stop blood clots from forming in conditions such as venous thromboembolic disease or atrial fibrillation), should follow treatment guidelines for these conditions independent of their IPF.

Quitting smoking, the best thing you can do for your health!

Stopping smoking will not only prevent lung disease, it will help you manage any existing lung disease such as IPF. If you smoke, remember that you are not alone. There are many options to help you become tobacco-free.

Did you know talking about quitting is proven to help people quit?

Understanding why you smoke and what makes you want to smoke is important in order to help come up with a quit plan that works best for you. Talking with health care providers about quitting, along with the use of medication has been shown to have the most success.

Methods to Help You Quit

In the past, the majority of people quit smoking using the *cold turkey* method, a quit method where no medications are used. Luckily, now there are many more ways available to help you quit.

- **The Cold Turkey Method:** Quitting cold turkey means that you stop smoking immediately, without the use of any medications. Quitting *cold turkey* is free, and does not require a prescription. However, when you quit suddenly, the amount of nicotine in your bloodstream declines quickly. As a result, you may have some symptoms of withdrawal. How strong these symptoms are and how long they last vary, but for most people they generally improve in a week or two.
- **Nicotine Replacement Therapies (NRTs):** The goal of nicotine replacement therapies (NRTs) is to replace the nicotine you would normally get from smoking. Nicotine is the addictive part of tobacco. NRTs are recommended by Health Canada as a safe and effective way to quit smoking. The amount of nicotine in NRTs may be less than the amount you would normally get from smoking. However, dosages can be adjusted and NRTs can be combined. (E.g., use of the NRT patch and NRT gum etc.). NRTs are recommended for short term use, however, longer term use is still safer than the use of tobacco products. To ensure safety and best results, follow the treatment plan outlined by your health care provider. In Canada, NRT products that are currently available over the counter at pharmacies

include the patch, gum, inhaler, lozenge and nasal spray. Some work place health plans may cover the cost of NRTs even though a prescription is not needed to purchase them.

- **Prescription Drugs:** Champix® (Varenicline) and Zyban® (Bupropion) are two of the most commonly used prescription drugs to help people quit smoking. Some of the costs may be recovered depending on the type of coverage you have if you have a health insurance plan. Champix® and Zyban® are taken in a pill form but act very differently in the body. These medications require a prescription from a prescribing health care provider. It is important to discuss with your health care provider any pre-existing health conditions that you have as well as any medications you are currently taking before using these medications.

We can help you be tobacco-free!

Join the Lung Saskatchewan's Sask Quits online support group or visit LungSask.ca/tobacco to find educational materials, resources and services that are available to help you become tobacco-free.



Oxygen

Supplemental oxygen is a treatment option that can be prescribed by a health care provider. It is not a cure for IPF and it will not take away your shortness of breath. When IPF worsens, the fibrosis (scarring) in the lungs allow less oxygen to enter the blood. Oxygen in the blood is needed for all organs, including your brain to repair and function.

Having low oxygen levels in the blood can cause people to have:

- more fatigue, especially during physical activity,
- problems concentrating and remembering,
- reduced quality of life,
- less muscle function,
- sleep disorders,
- swelling (*edema*) in their legs,
- a blue colour on their fingertips, earlobes or lips (*cyanosis*),
- other conditions including heart disease,
- a worsening of existing health problems, and
- an increased risk of death.

Supplemental Oxygen Myths vs Facts

Myth: Supplemental oxygen is prescribed if you are short of breath.

Fact: Supplemental oxygen is only provided if the oxygen in your blood is low. If you do not have low levels of oxygen in your blood, oxygen will not help you.

Myth: Supplemental oxygen is addictive.

Fact: Supplemental oxygen is not addictive. Your body needs a certain amount of oxygen to maintain normal body functions.

Myth: If you are given supplemental oxygen you will be cured and you will no longer need your other medications.

Fact: Supplemental oxygen will not cure your IPF. If your doctor has prescribed other medications you should continue to take them unless they tell you otherwise.



Charlotte L'Oste Brown, Photo taken when she was living with IPF before her lung transplant

Common Questions about Supplemental Oxygen

Does everyone with IPF need supplemental oxygen?

No. Only people who have low levels of oxygen in their blood will be prescribed oxygen. A low blood oxygen level is called *hypoxemia*.

How will I know if I need supplemental oxygen?

Tests (oximetry and/or arterial blood gas) are done to measure the amount of oxygen in your blood. Using oximetry, (a small sensor device which is usually placed on one of your fingers to determine how much oxygen is in your blood), your health care provider can find out the oxygen levels in your blood when you are at rest, sleeping, and/or doing physical activity. An arterial blood gas test uses blood taken from the arterial artery, usually in your wrist. Treatment guidelines recommend people with IPF who have low levels of oxygen in their blood receive supplemental oxygen. For people who have low levels of oxygen in their blood and have other long-term lung disease, supplemental oxygen has proven to be helpful. There remains a degree of uncertainty about the

benefits oxygen therapy has for people with IPF. Cost, inconvenience, and health impact for those with IPF and on oxygen are still being studied.

If I am prescribed supplemental oxygen, do I need it all the time?

It depends.

- Your doctor may prescribe oxygen:
- All day and night, all the time (*continuous*).
- Only at night while you sleep (*nocturnal oxygen*).
- Only while you exercise or do physical activity (*ambulatory/exertional oxygen*).

How much oxygen will I need?

Everyone's prescription for oxygen will be different. You will be told how much oxygen you need (*flow rate*) and how long you need to use it (*duration*). Your doctor may tell you to increase your flow rate when you are exercising or sleeping.

My doctor gave me an oxygen prescription, now what?

Once you have an oxygen prescription, you will need to contact an oxygen supplier. The supplier will show you what kind of equipment you need, how to use the equipment and go over some important safety tips. They can also help explain possible funding options.

Oxygen Delivery Devices

Oxygen can be delivered using different systems and devices. Your oxygen supplier along with your doctor, can help you pick the one that is right for your lifestyle and budget.

1. **Nasal Prongs:** All systems have a tube to deliver oxygen into your nose.



2. **Concentrator:** The air we breathe is a mixture of odorless gases made up of nitrogen, oxygen and other gases. An oxygen concentrator is a medical device that takes in air, removes nitrogen and the other gases and delivers pure oxygen.



If you have a home oxygen concentrator, you will need a backup oxygen supply (e.g., a tank of compressed oxygen). The backup is used if the power goes out or if the concentrator stops working.

Advantages:

- ✓ You do not need to have your tanks filled.
- ✓ It can save you some money.
- ✓ There is little maintenance.
- ✓ It is easy to use.

Things to Consider:

- You will need a source of electricity. A concentrator must be plugged into an outlet at all times.
- You will need portable oxygen tanks that you can take with you when you leave your home.
- You must have an open storage space for the concentrator.
- The concentrator is heavy and therefore not very easy to move around or with you.

3. **Portable Oxygen Concentrator:** Another type of concentrator that is smaller and easily moved.

Advantages:

- ✓ This has all the advantages of a concentrator but can also be taken outside of the home and moved around with you. It can be used with an adapter, battery, or electricity.



Things to Consider:

- Some portable oxygen concentrators deliver oxygen primarily when you breathe in through your nose only.

4. **Compressed Gaseous Cylinders:**

High purity oxygen in a gaseous form is compressed and stored in an aluminum or steel tank.



Advantages:

- ✓ High flow rates are possible.
- ✓ The oxygen is not wasted because the gas does not evaporate.
- ✓ It is quiet.
- ✓ Aluminum tanks are light.
- ✓ No electricity is needed. It does not have to be plugged in.
- ✓ Smaller cylinders may be used when going places or for a backup with other systems.

Things to Consider:

- The tanks only last for a certain amount of time before the oxygen will run out.
- Steel cylinders are heavy.
- There is some skill needed to adjust and change the flow regulator.
- Special storage is required.

5. **Portable Liquid Oxygen:**

High purity oxygen in a liquid form is stored in an insulated container.



Advantages:

- ✓ You can store a high volume of oxygen.
- ✓ High flow rates are possible.
- ✓ It is quiet.
- ✓ You can fill the portable oxygen units.
- ✓ It is light and easy to take with you where you go.

Things to Consider:

- The containers require filling.
- The oxygen evaporates whether it is used or not.
- The oxygen is very cold in the liquid state.
- Knowledge and skill is needed to fill the tanks.
- It is more expensive.

6. Oxygen Conserving Device:

Oxygen is delivered intermittently as you breathe. When the oxygen is delivered will vary.

The oxygen may be delivered:

- for a portion of your breath in,
- during the entire time you breathe in,
- every breath, or
- every second breath.

An extra dose of oxygen may also be delivered from time to time as well.

Advantages:

- ✓ The intermittent oxygen delivery saves oxygen and allows the oxygen supply to last longer. Because of this you can go out with your oxygen longer. It also allows for smaller and lighter portable units to be used.
- ✓ There is less drying and irritation of the inside of your nose.
- ✓ It can be used with cylinders.

Things to Consider:

- This device is usually more expensive.
- It may be difficult to match the flow delivery to your breathing pattern.
- Many of these devices need a battery for power.
- You cannot use humidification with this device.
- Some patients may not receive the oxygen they require with this system.



Tips and Facts about Oxygen Therapy

1. **Ask how to use the equipment:** Have the oxygen supplier show you how to use the equipment before you go home. Don't be afraid to ask for clarification. Take notes and for additional support, have a family member, caregiver or friend come with you to learn about your equipment.
2. **Use your oxygen as prescribed to you by your health care provider:** Know how much oxygen to use (*dose/flow rate*) and how long to use it (*duration*). Understand if your flow rate needs to change when you exercise, take part in physical activity or when you are sleeping.

3. **If you are prescribed continuous oxygen try to use your oxygen all the time.** People prescribed continuous oxygen are encouraged to use their oxygen 24 hours of the day.
4. **Keep your equipment clean:** Clean equipment works better and helps prevent infections. The oxygen supplier will teach you when to replace your oxygen tubing and nasal prongs.
5. **Find out the contact information (phone number, email, location etc.) of your oxygen supplier in case you have problems with your oxygen equipment or questions about oxygen therapy.**
6. **Ask about funding:** Oxygen therapy can be expensive especially over the long term. If you meet certain criteria, you may qualify for government funding for oxygen therapy – ask your doctor or your oxygen supply company if you are eligible. Funding varies from province to province and may only cover some of your supplemental oxygen needs.
7. **If you smoke, consider quitting. DO NOT SMOKE WHILE USING OXYGEN.** No one should smoke within 10 feet or 3 meters of oxygen equipment because it is a dangerous fire hazard. We can help you quit! Contact Lung Saskatchewan to find out about the educational materials, resources and services that are available to help you become tobacco-free. LungSask.ca/tobacco
8. **Plan before you travel:** Travelling with oxygen is very possible however it will require you to plan ahead. Before you decide to travel, contact your doctor and your home oxygen supplier. Your doctor can help you decide if you are able to take the type of trip you have planned. The home oxygen supplier will help coordinate the equipment you need while you are travelling. They will also help you arrange for equipment when you arrive at your destination.

You may want to consider arranging for an oxygen concentrator at your destination. It is important to ensure that your oxygen supply and delivery system(s) is appropriate for all parts of your trip (en route and at your final destination). Make sure to plan for an adequate supply of oxygen during your travels. Your options may include portable supplies that can be plugged into a car battery. Check to make sure you do not need any plug in adapters

for your trip. Always make sure your equipment can be safely and securely stowed while traveling. Don't forget to contact your airline or travel agent to advise them that you will be travelling with oxygen and find out if there are special requirements for your oxygen.

My Oxygen Care Plan

Work with your oxygen supplier to answer these questions to develop your personalized oxygen care plan.

- How much oxygen do I need? _____
- What is my flow rate at rest? _____
- What is my flow rate with activity? _____
- What is my flow rate when I am sleeping? _____
- How long do I have to use my oxygen for? _____
- What kind of equipment do I need? _____
- How do I keep my equipment clean? _____
- How often do I replace my oxygen-source tubing and nasal prongs? _____
- What is the number I can call if I have problems with my equipment or questions about my oxygen? _____
- What do I need to do to see if I qualify for government funding for oxygen therapy? _____
- Other: _____

Move to Breathe Easier with Pulmonary Rehabilitation

Pulmonary rehabilitation is an exercise and educational program designed for people with lung disease. The program often includes, but is not limited to aerobic training (walking, biking etc.), resistance or strength training (lifting weights

or bands), flexibility exercises (stretches) and educational sessions about ways to best manage your disease. The programs usually include information on healthy living and diet. Pulmonary rehabilitation is also a way for people to meet and feel supported by others who have lung disease.

Most pulmonary rehabilitation programs are currently designed for people with chronic obstructive pulmonary disease (COPD), however many programs accept and welcome people with IPF. Some locations may even have pulmonary fibrosis rehabilitation programs. It might not seem logical to take part in exercise especially if it makes you breathless, however staying active is a recommended part of managing IPF. A recent update to the treatment guidelines for IPF recommends that the majority of patients with IPF should take part in pulmonary rehabilitation. Studies specific to IPF are currently looking at understanding the best types of exercises and how often they should be done.

Why Take Part in Pulmonary Rehabilitation

- Exercise training has been proven to be a safe and effective way to prevent and to help manage chronic (long term) conditions including IPF.
- It can allow you to walk further, do more physical activity, improve or maintain muscle strength and increase your ability to do everyday tasks.
- You may have less shortness of breath.
- You will learn more about managing IPF.
- It will provide you with emotional and social support.
- It could improve your quality of life and help you to feel better all around.

Types of Pulmonary Rehabilitation

1. **Supervised Pulmonary Rehabilitation:** This is a structured and supervised exercise program for people with lung disease. In general, it seems that supervised exercise training programs provide greater benefit for people with IPF. More and more locations have supervised pulmonary rehabilitation. Some of the exercise programs are self-referral and others require your doctor to refer you. Programs may be based out of a hospital, a health/rehabilitation center, or in the community.

Find a pulmonary rehabilitation program near you!

To find out if there is a pulmonary rehabilitation program near you, speak to your health care provider, lung specialist, or contact **Lung Saskatchewan at 1-833-385-LUNG (5864) or visit our website at LungSask.ca.**

2. Self-directed Home-Based Pulmonary

Rehabilitation: There are not as many studies that examine the impact home-based pulmonary rehabilitation programs have on people with IPF. Some smaller studies found that home-based pulmonary rehabilitation may reduce shortness of breath and fatigue, while improving the amount of exercise people can do and overall health-related quality of life. If you live in a town where no formal lung exercise programs are available, you can still gain many of the benefits of pulmonary rehabilitation with your own self-directed home-based exercise program. Some locations have technologies in place to help deliver health services, such a pulmonary rehabilitation, to individuals, particularly those living in an isolated area or with limited access to transportation. This can be done using cell phones, monitored home exercise training programs, videoconferencing, telemonitoring and/or Telehealth.

Before you begin any new exercise program in the community or at home, speak to your health care provider. You should be assessed first with some sort of exercise test (E.g., 6 minute walk test) and your oxygen levels will likely be monitored with an oximeter while you exercise. The assessment will help prescribe what kind of exercise is best for you, help determine the best way to start exercising and will ensure your safety. It may be suggested that you use oxygen for exercise or if you are already using oxygen at rest, that you may need to increase the flow of your oxygen while you take part in physical activity. Together with your health care provider, a plan can be developed that is right for you.

Self-Care: Maintaining a Healthy Lifestyle

Taking care of yourself and preventing colds and flus is also very important in managing your IPF.

Health Tips:

- Ask your doctor about getting a flu (*influenza*) shot every year.
- Ask your doctor about getting pneumonia (*pneumococcal*) shot every 5 years.
- Wash your hands often and properly with soap.
- Try to avoid touching your mouth, nose or eyes.
- Try to stay clear of people who are ill with chest infections or colds.
- Stay as fit as you can. (*Find out about pulmonary rehabilitation*).
- Contact Lung Saskatchewan at 1-833-385- LUNG (5864) or visit our website at LungSask.ca.





General Tips if You Get Short of Breath when Eating

- Eat small frequent meals throughout the day (5-6 small meals instead of 3 big meals).
- Rest before eating.
- Eat slowly and chew foods well.
- Cut your food into small, bite-size pieces.
- Eat foods that are nutritious and easy to chew and swallow.
- Choose foods that will maximize nutrition in a small amount.

Everyone is different! You may need a personalized dietary plan. Ask your doctor about a referral to a dietitian that specializes in chronic disease management nutrition.

Eat Well - Breathe Easier

Your body needs a lot of energy. People with IPF use more energy breathing.

If you are underweight, you may lack energy. A health care provider may advise you to obtain more calories through a specific diet.

If you are overweight, your muscles will have to work harder and this can worsen your shortness of breath. Excess weight around your waistline can make breathing even more difficult because it places more pressure on your diaphragm – which assists your lungs with breathing. A balanced and healthy diet can help you achieve a healthy weight.

General Tips for Healthy Eating

- Enjoy a variety of foods.
- Choose lean meats.
- Limit salt, sugar, alcohol and caffeine.
- Eat plenty of fruits and vegetables.
- To save time and energy, find recipes that are quick and easy but also nutritious.
- When possible, prepare more than one meal at a time and freeze what you do not eat so you can still eat healthy when time is limited or when you are feeling unwell.
- Drink plenty of water.



Charlotte L'Oste Brown, Double Lung Transplant Recipient

Acute Exacerbations of IPF

Some people with IPF have *acute exacerbations* (a sudden worsening of symptoms), where there is an abrupt decline in how the lungs function. After an acute exacerbation, some people will recover to a certain degree and others will never be the same or continue to get worse. People with IPF can have episodes or worsening from a lung infection (*pneumonia*), a clot in the lung (*pulmonary embolism*), a collapsed lung (*pneumothorax*), heart (*cardiac*) failure or from unknown complications. When the cause cannot be identified the term acute exacerbation for IPF is often used. An acute exacerbation can happen at any time if you have IPF. There are no known risk factors for an acute exacerbation. However some studies have found that a sudden worsening of symptoms is more common in people with advanced disease.

Signs and Symptoms of an Acute Exacerbation of IPF

- A worsening in cough, shortness of breath, fatigue,
- Fever,
- Phlegm (*sputum production*),
- General feeling of being unwell.

IPF Acute Exacerbations are serious.

- IPF Acute Exacerbations may;
- require you to stay in the hospital,
 - require technology and machines to help you breathe,
 - make your disease worse, and
 - cause death.

Treatment of IPF Acute Exacerbations

There is limited research on proven and effective therapies for acute exacerbations of IPF. Many doctors will prescribe anti-inflammatory medication (*systemic corticosteroids*). Most of the research supports treating the worsening of symptoms and low oxygen in the blood (*hypoxemia*) with supplemental oxygen. Equipment to help people breathe (*mechanical ventilation*) may be considered to treat lung (*respiratory*) failure.

Some IPF therapies (Nintedanib and Pirfenidone) may help prevent the development of an acute exacerbation of IPF.

Monitoring IPF

It is important for you and your doctor to monitor your health. Setting up regular check-ups with your doctor is key. Depending on how your disease is progressing, your doctor may recommend different treatment options.

When should I seek medical help?

If you have a sudden worsening of breathing, cough, sputum production, fever, loss of appetite or a general feeling of being unwell, make an appointment to see your doctor right away.

Warning Signs of a Breathing Emergency:

- Very rapid breathing
- Sucking in skin above the breastbone and between the ribs
- Being really tired/lethargic (because of the work of breathing)
- Finding it hard to speak - you can't finish a sentence
- Nostrils flaring out
- Pale, grey, sweating
- Chest pain
- Blue lips or fingers
- Confusion
- Drowsiness
- Extreme shortness of breath

If you or someone you love experiences a breathing emergency, call 911 and go to the nearest emergency department right away! The person having the breathing emergency should not drive.

“To breathe freely is a beautiful gift and should never be taken for granted. Take good care of your lungs – they are precious.”

Jill Hubick
Registered Nurse, Lung Saskatchewan

Chapter 4 Summary

- IPF is a chronic or long-term disease that has no cure other than a lung transplant. There are however ways to help manage IPF.
- No medications currently exist that can cure IPF or decrease the amount of scarring that is already in the lungs. However, there are medications available that help to slow the progression of the disease. Recent research discoveries have made some newer medications available, and clinical trials are showing promise for more options in the future. The most common medications used to manage IPF are Pirfenidone (Esbriet®), Nintendanib (OFEV®), Corticosteroid pills (Prednisone®) – only for acute exacerbations.
- Stopping smoking will not only prevent lung disease, but help you manage any existing lung disease such as IPF.
- Being prescribed oxygen is not a cure for IPF and it will not take away your shortness of breath. When IPF gets worse, the fibrosis (scarring) in the lungs allows less oxygen to enter the blood. Oxygen in the blood is needed for organs including your brain to repair and function. A low oxygen level in the blood is called *hypoxemia*.
- An assessment or test (oximetry or arterial blood gas) will be done to see if you need supplemental oxygen. If you require supplemental oxygen you may need it all the time (*continuous*), only with physical activity (*exertional*) or when you are sleeping. Work with an oxygen supplier to determine the right type of oxygen equipment/device for you.
- Use the *My Oxygen Action Care Plan* in this chapter to work with your doctor and oxygen supplier to make sure you know when and how to use your oxygen equipment. When learning about how to use your oxygen, consider bringing along a family member, caregiver or support person.
- Use your oxygen as directed by your doctor.
- If you are prescribed continuous oxygen try to use your oxygen all the time. People prescribed continuous oxygen are encouraged to use their oxygen 24 hours of the day.
- Keep your supplemental oxygen equipment clean.
- Find out the contact information (phone number, email, location etc.) of your oxygen supplier in case you have problems with your equipment or questions about oxygen therapy.
- Ask about supplemental oxygen funding.
- If you smoke, consider quitting. **DO NOT SMOKE WHILE USING OXYGEN.** No one should smoke within 10 feet or 3 meters of oxygen equipment because it is a dangerous fire hazard.
- Lung Saskatchewan Association can help you quit smoking! Join the Lung Saskatchewan's Sask Quits online support group or visit LungSask.ca/tobacco to find educational materials, resources and services that are available to help you become tobacco-free.
- Plan before you travel with your supplemental oxygen.
- Pulmonary rehabilitation is an exercise and educational program designed for people with lung disease. Treatment guidelines recommend most patients with IPF should take part in pulmonary rehabilitation.
- Taking care of yourself and preventing colds and flus is also very important in managing your IPF.
- People with IPF use more energy breathing compared to people who do not have a lung disease. Maintaining a healthy weight can help with your breathing and energy needs. Nutrition plays an important role in achieving this.
- Some people with IPF have *acute exacerbations*, which consist of sudden worsening in symptoms, and there is an abrupt decline in how the lungs function. After an *acute exacerbation*, some people will recover to a certain degree and others will never be the same or continue to get worse.
- IPF acute exacerbations are serious and can be deadly.
- If you have a sudden worsening of breathing, cough, sputum production, fever, loss of appetite or a general feeling of being unwell make an appointment to see your doctor right away.
- Know the warning signs of a breathing emergency. If you or someone you love experiences a breathing emergency, call 911 or go to the nearest emergency department right away! The person having the breathing emergency should not drive.

CHAPTER 5: ADJUSTING AND COPING WITH YOUR DIAGNOSIS

Receiving a diagnosis of IPF can be very difficult. There is no right or wrong way to feel. Everyone will cope differently. You might feel very little at first or find yourself in shock and disbelief. With time, many people experience a range of emotions including anger, fear, frustration, helplessness, resentment, denial, or sadness. Some people grieve the life they once led. While others find relief in knowing some answers as to why they have been so ill and feel a sense of acceptance.

Being Told You Have IPF – You are Not Alone!

Most people know very little about IPF before their diagnosis. At first many will have a sense of relief that it isn't cancer but are quickly shocked by the poor prognosis IPF has. You may not remember everything your doctor told you when you first found out you had IPF. This is normal. Being told you have IPF can be overwhelming. You may find it helpful to make a follow-up appointment with your doctor and bring along a support person who can also learn more about the disease. Don't be scared to write your questions or concerns down and bring them to your next appointment.

IPF and its Impact

IPF will not only impact your physical health, but also your social and emotional wellbeing. It will also affect your loved ones, work life and overall quality of life.

Talk about it!

Whatever feelings you are having, you do not have to go through it alone. It can help to talk about your worries and fears. You may find comfort in talking to your family, partner, or friends. However, sometimes it is easier to speak to a doctor, nurse, counsellor, social worker, or spiritual leader. Several people try to figure out why this is happening to them and feel as if no one knows what they are going through. You may prefer to talk to other people with IPF. Many places have support groups for people with breathing problems or specifically for those with pulmonary fibrosis. With technology being more and more part of our daily lives, some support groups are online.

Join A Support Group

Visit LungSask.ca/pulmonaryfibrosis to learn more.

Managing Your Life at Home

As the disease gets worse, there is a decline in physical function. The majority of people first notice they are more short of breath than usual when they are exercising. However, eventually people with IPF find it is challenging to walk upstairs, shower, do house work, cook, clean, or get dressed. Putting a plan and supports in place to assist with these daily activities of living is key. This could include adjusting how you do things to save energy, working with local programs that can help bring you meals, have home care come into your home to help with some of your personal care, moving to a place that has assisted living or asking for help from a support person.

Ask your health care provider about client/patient access services. There may be programs in your province/community that can help you with meals, personal care, transportation and/or support you in other ways.

Work and Careers

You may need to make adjustments or changes to your work life/career. When you are very short of breath, focusing on work and managing fatigue can be difficult or no longer possible. You might not be able to work as long and part time work may be an option. Depending on the type of work you do, you may need to change jobs. You may also need to consider leaving the workforce all together.



Adjusting to Life with IPF

Educating family and friends

Having your friends and family understand IPF is important. It can be difficult for others to realize the debilitating and enormous impact the disease has on you. People without lung disease don't know what it feels like to be extremely short of breath and fatigued all the time. There is often a general misconception that people who have supplemental oxygen are no longer short of breath.

The disease can affect your social life and your ability to interact with friends and family like you are used to. This can cause feelings of isolation and loneliness. Many people with IPF struggle to go out in public, even if they are feeling well enough to do so, because they are self-conscious of the reactions they receive from others when they cough. Some people may assume you are contagious. Many people with IPF feel embarrassed by their oxygen tank or that their coughing is disturbing others.

If your friends and family have a good understanding of what IPF is and how it affects you, they can find ways to be supportive and helpful. Talking to your loved ones about IPF as soon as possible can allow everyone to be on the same page and reduce any assumptions or unrealistic expectations both parties may have. Remember you don't have to go through this alone. Working with the people in your life to find ways to adjust is beneficial for everyone.

Activities - Managing your Energy

Our hobbies and activities of daily living bring us both independence and joy. They are also a large part of our social lives granting us the ability to interact with others. Modifications can usually be made so you can keep doing the activities that are important to you longer.

Tips to Manage your Energy

1. **Pace yourself** - Work at a slow, steady pace. Leave yourself extra time to get things done. Try things like putting your clothes out the night before if you have an early-morning appointment. Pace your meals. Eat small meals often and rest after eating.
2. **Set priorities** - How can you shorten your 'must - do' list? Learn to 'say no' to the things you don't want to do or can't do. Ask yourself what your priorities are. Try to modify and still take part in activities that bring you happiness.
3. **Plan ahead** - Before leaving the house think about the places you will be going. Can you park near the door? Are there benches to sit on so you can rest? Write the items on your grocery list in the same order you would find them in the grocery store. If you start in the vegetable aisle, put lettuce and peppers at the top of your list. That way, you won't have to walk as far. Spread out your activities throughout the week, balancing work and rest each day. Do your best to keep, your schedule flexible and put off your plans on days you have less energy.
4. **Adjust activities** - Find easier, less tiring ways of doing chores. These changes may help you stay active, rather than avoiding the activities altogether.

Examples of Modifying Activities

- Shower while sitting on a bath stool.
- After a bath/shower, slip on a terry cloth robe instead of drying yourself with a towel.
- If you like to golf, rent a golf cart so you are not using the energy walking from one hole to the next.
- When cooking or baking, try to find a stool that you can sit on while chopping or mixing in the kitchen. Sitting takes less energy than standing.
- While making meals, make extra and freeze to use later.
- Make bed-making easier by replacing heavy covers with lightweight electric blanket or synthetic duvet.
- To make meals easier, try using the microwave or slow-cooker.
- Consider applying for a special parking permit for your car, so you can park closer to the doors.

5. **Choose clothing carefully** - Dress for comfort. Try wearing loose fitting clothing and under garments. Anything that adds further restriction especially to the upper body can make you more uncomfortable. Clothing made with some spandex and stretch can also be helpful.

6. **Utilize helpful tools** - Carrying objects takes extra energy and can leave you more breathless. Try using a trolley or wagon for taking in and out groceries or to clear the dishes at the table. This will also reduce the number of trips you need to complete one task.

Items that can make everyday tasks easier:

- Long handled shoehorns
- Long-handled dustpans
- Bath seat
- Grab bars for the bathtub
- Hand-held shower head
- Shower seat
- Raised toilet seat
- Folding gardening stool

7. **Ask for help** - Asking for help doesn't mean losing your independence, it's a way of saving your energy for other, perhaps more important things. Ask for help with tasks that cause you to be the most short of breath and tired. Consider hiring help for household chores and yard work like lawn and garden maintenance, snow shoveling, and fall clean-up. Consider programs that deliver meals and/or groceries.

8. **Think about your posture** - Working with your back bent or shoulders slumped can cause muscle strain and increase the likelihood that you will hold your breath. Try to avoid bending and lifting. Push, pull or slide instead. If you must lift or carry, lift with your legs, use both hands, and hold the object close to your body. Choose work surfaces that allow you to sit or stand straight. Change positions often.

9. **Use relaxation and stress management techniques** - Being short of breath can make anyone feel anxious and may cause you to panic. This may only make you even more short of breath. It can be a vicious cycle. Added tension and worry takes energy and strength as well.

There are several relaxation and stress management techniques that are proven to reduce anxiety,

decrease blood pressure and heart rate, help with pain management and improve overall quality of life.

Relaxation and Stress Management

Techniques:

• **Progressive Muscle Relaxation:** Progressive muscle relaxation is a technique where you purposefully tense and relax different muscle groups in your body. The technique teaches you to become aware of how tension and then relaxation feels in different parts of the body. It is designed to help you recognize signs of muscle tension caused by stress. With this technique you can counteract your stress by relaxing your muscles.

Here's how to give progressive muscle relaxation a try.

1. Get comfy. Loosen tight clothing, take off your shoes and get into a relaxing position. You can lie down or sit in a chair that supports your head.
2. Curl both fists, tightening your lower and upper arms. Tense your muscles for 5-7 seconds. Then relax.
3. Scrunch up your forehead, squint your eyes, purse your lips, press your tongue to the roof of your mouth and lift up your shoulders for 5-7 seconds. Then relax.
4. Arch your back by pressing out your stomach for 5-7 seconds. Then relax.
5. Pull your feet and toes back towards your face, tightening your shins for 5-7 seconds. Now relax all your muscles.

• **Visualization** - Day dreaming, memories, and self-talk are all types of visualization. Visualization can be very effective with stress and anxiety, and can calm or brighten moods.

Here are some tips for visualization.

1. Find a quiet place where you can relax. Loosen tight clothing, take off your shoes and get into a comfortable position. You can lie down or sit in a chair that supports your head. Close your eyes.
2. Mentally scan your body and release any muscle tension.
3. Imagine a scene or image that brings you happiness and calmness using all of your senses. Think about how it looks, sounds, smells, feels and tastes.

For example: Imagine the sights of a sandy beach, surrounded by turquoise water, tall

green pine trees, and a clear blue sky above. Hear the waves from the water reach the shores edge and listen to the birds singing in the trees. Feel the warm sun against your face and imagine sinking your toes into the soft cool, wet sand.

Other scenarios may include watching the sun set or rise, being in a garden, your own backyard or in a safe, comfortable place.

- **Meditation:** This technique is used to promote a natural shift of awareness to a wakeful but deeply restful state.

Here is an example of meditation.

1. Sit in a quiet place. Make yourself comfortable.
 2. While sitting, close your eyes and repeat a 'mantra,' a meaningless sequence of sounds that are unique to you or even a calming phrase or word. Example: repeat the sound 'om,' or 'relax'. Meditation is recommended for 20 minutes twice daily.
- **Deep Breathing:** Deep breathing can help with relaxation. Focusing on the breath may also be helpful when completing everyday tasks. Try thinking about breathing in deep through your nose and out through your mouth when making a meal, getting dressed, etc.

Chapter 5 Summary:

- Receiving a diagnosis of IPF can be very difficult. There is no right or wrong way to feel. Everyone will cope differently.
- IPF will not only impact your physical health, but also your social and emotional wellbeing. It will also affect your loved ones, work life and overall quality of life.
- Talking about being diagnosed with IPF may help you cope. Consider speaking with your doctor, a nurse, counsellor, social worker or spiritual leader. You may prefer to talk to people with IPF. Many places have support groups for people with breathing problems or specifically for those with pulmonary fibrosis.
- Putting a plan and supports in place to assist with these daily activities of living is key. This could include adjusting how you do things to save energy, working with local programs that can help bring you meals, have home care come into your home to help with some of your personal care, moving to a place that has assisted living or asking for help from a support person.
- Educate your family and friends about IPF so they can find ways to be supportive and helpful.
- Consider ways to modify activities to save energy.
- Don't be scared to ask for help.
- Being short of breath can make anyone feel anxious and may cause you to panic. This may only make you even more short of breath. It can be a vicious cycle. Added tension and worry takes energy and strength as well.
- Consider trying relaxation and stress management techniques such as progressive muscle relaxation, visualization, meditation and deep breathing.

“At Lung Saskatchewan we want disease to take a back seat to living and breathing well.”

Erin Kuan

President & CEO, Lung Saskatchewan

CHAPTER 6: LUNG TRANSPLANTATION

Much of this chapter was adapted from The Lung Association, Alberta and NWT Lung Transplant Information for Alberta & NWT Residents Handbook.

IPF is a severe lung disease. For people who have severe lung disease, a lung transplant may be their last treatment option to survive. A lung transplant can offer hope, but it can be a complex and scary process. A lung transplant can also be expensive, and it comes with health risks. It is a lifelong commitment. You will need medication and treatment for the rest of your life.

There are five hospitals in Canada that perform lung transplants. The world's first successful lung transplant was done in 1983 in Toronto, Ontario. Today over 200 lung transplant surgeries are performed each year in Canada. The number of transplants done each yearly, depends on the number of donor lungs that become available.

What is a lung transplant?

When you have a lung transplant, your diseased or damaged lung(s) are removed and replaced with a healthy lung that is donated from another body (donor lung). One or both lungs can be replaced.

Where do most donated lungs come from?

Most donor lungs come from the body of someone whose brain has lost function but the rest of the organs are healthy. This is referred to as 'brain death.' Brain death can happen with a serious trauma or sudden bleed inside the brain.

What makes a donor lung a good match?

1. Matching blood type
2. Size of the donated lung(s) – often related to height
3. Where the donor is geographically located
4. Antibodies you may have against the certain donors' blood and tissues (antibodies are proteins in blood and tissue that play a key role in your immune system)

Lungs that are too large can sometimes be trimmed down. Age, gender and race are not important.



Nicole Nelson, Double Lung Transplant Recipient

Who may need a lung transplant?

- People with a severe lung disease who despite all available treatments and medications are continuing to worsen.
- When your life expectancy is one to two years without a lung transplant.
- When a lung transplant could result in a better quality of life.

Most of the lung transplants done in Canada are a double lung transplant (both lungs are removed and replaced with donor lungs). The decision, however, will depend on the number of donors available and the medical needs of the patient.

Who might not be able to have a lung transplant?

People who have:

- Cancer (except some skin) – it is best to be cancer-free for at least five years
- Heart, liver or kidney diseases that can't be treated
- Non-curable chronic infections (example: Hepatitis B, Burkholderia, Mycobacterium Abscessus)
- Significant problems with the shape of the chest wall or spine
- The inability to follow medical therapy
- A lack of support system
- An addiction to tobacco, drugs, or alcohol

If you smoke, and would like to be considered for a lung transplant, speak to your health care provider. You can also join the Lung Saskatchewan's Sask Quits online support group or visit LungSask.ca/tobacco to find educational materials, resources and services that are available to help you become tobacco-free.

When may a lung transplant be too risky?

A lung transplant might be too risky if you:

- Have a critical illness (Example: on a breathing machine (mechanical ventilator) and/or are needing powerful medications to support your blood pressure)
- Are severely overweight or underweight
- Have very brittle bones (osteoporosis)
- Have chronic lung infections
- Have other medical conditions (Example: diabetes) that could result in a more difficult recovery from a

lung transplant

How will I know if I can be put on the lung transplant waiting list?

You need to be evaluated by health care professionals. A team of medical professionals will inquire about medical problems and screen for various cancers. You will also have several tests such as a CT scan, (high resolution picture of your lungs and perhaps other organs), pulmonary (lung) function tests (breathing tests), an echocardiogram (ultrasound of the heart) and possibly other tests as well. These tests often take about a week to do. You will require a support person to be with you during these tests. You will also need to have a strong social support system of friends or family that will be able to take care of you after your transplant. If it is determined that you are a suitable candidate for a lung transplant, you will be asked to attend a physiotherapy program. Sometime during this program you will be added to the lung transplantation wait list. This program is done at a transplant center which may not be where you live. It might require you and your support person to travel and find accommodations in the transplant center city.

The transplant team may also ask you to make certain lifestyle changes before being placed on the transplant waiting list. Examples include dental work or optimizing weight.

Why do I need a support person?

A lung transplant is a major surgery that affects all parts of your life, not just your body. The entire process of a lung transplant can be very stressful and is usually full of ups and downs. Your support person should be someone you can talk to and someone that can help you stay positive. They will also need to help you get to appointments, get groceries, do the laundry, and assist with personal care before and after the surgery.

Who should be my support person?

Being a support person is very demanding and a full-time job before the lung transplant and afterwards during recovery. A support person is often your spouse/partner or a close family member or friend. One or more people can make up a support 'team' so that they can share the work, costs, and if needed travel.

A support person should be okay with spending lots

of time in hospitals as they will be accompanying you to tests, appointments and will be with you during your recovery. If working, they will have to be able to leave their job temporarily and commit to being there for you during this very stressful time.

How long does my support person have to stay?

Your support person will need to go with you to all your medical appointments and tests before the surgery. They will need to stay with you at all times while you are an inpatient at the hospital (usually 1 month) and also while you are an outpatient (usually 2 months). An outpatient is a person who has medical treatment at the transplant center but is not admitted overnight. As an outpatient you will be required to stay close to the transplant center. On average your support person will be with you for at least 3 months after your lung transplant surgery. This time will vary for everyone. A support person is essential to the success of a transplant. Speak to the transplant team if this is a concern for you.

What should I do while I am waiting for a lung transplant surgery?

Use this time to exercise as tolerated to ensure your body is in the best shape possible for surgery. Your health care providers may recommend you work with a pulmonary rehabilitation program. The better shape you are in before the surgery, the easier and quicker you will recover. Ask your health care provider team about advice on nutrition and diet. While you are on the transplant waiting list you will have appointments often with the medical team to monitor your condition. It is very important to let your health care providers know if your health changes.

How close do you have to be to the transplant centre when waiting?

You must be living 2.5 hours travelling distance (by land or by air) of the transplant center. It is important that you can get to the hospital quickly if you receive your call that there are donor lungs available for you.

How long is the waiting time for a transplant?

The time you have to wait for a lung transplant is different for everyone. It will also be different for the various transplant centers. Available donor lungs will be given to the best match to someone on the lung

transplant list. It will also depend on how sick the person waiting for new lungs is.

Some people will wait a few days after being added to the lung transplant list, others wait over two years.

Disease severity is assessed on the amount of scarring seen on your CT scan, your pulmonary function tests, shortness of breath, oxygen levels, how far you can walk, and if you have pulmonary hypertension in addition to IPF.

There is no set system to determine the exact severity of IPF. Unfortunately, some people will die while waiting for a lung transplant. Some people may also have to withdraw from the transplant waiting list due to other medical conditions that arise while waiting.

How will I be notified when suitable organs become available?

You will receive a phone call telling you to come to the transplant center. Your medical team will have already determined if you need to drive or travel by air to the transplant center. You will need to have a cell phone with you at all times in anticipation for 'the lung transplant call'. You will also need a bag packed so you are ready to get to the hospital immediately.

How long is the surgery?

A lung transplant surgery usually takes 6-8 hours. This could take longer if there are complications. You will be asleep for the entire surgery. The incision will be located below the lower part of your chest. When you wake up, you will have chest tubes in your sides and a breathing tube in your mouth. You will also be given medication to help with the pain and discomfort from the surgery until you recover.

How long is the recovery from surgery?

Recovery will vary. Everyone heals differently. Some people may have complications. Some more and some less than others. Most people who have a lung transplant are in the Intensive Care Unit (ICU) in the hospital for a few days to a week. Usually they then remain in the hospital for three or more weeks after that. During your hospital stay the doctors will determine the right medications and recovery plan for you. After being released from the hospital, you will have to remain near the transplant center for at least two more months. (A minimum three months total from the time of surgery). If you live far away



Charlotte L'Oste Brown, Double Lung Transplant Recipient

from the transplant center, you will need to plan to move during this time along with a support person to help you (often a friend or family member). During your stay you will continue rehabilitation and physiotherapy. The pulmonary rehabilitation will be continued after you are discharged from the hospital on an outpatient basis. You will also have blood tests, chest x-rays, and spirometry (breathing tests) after your transplant. Some people may also need to have a bronchoscopy as well. Your breathing tests will improve for the next year after your transplant as long as your new lungs remain healthy. You will be required to monitor your breathing using a portable micro-spirometer (a device for measuring the amount of air you can blow out). A micro-spirometer will help detect problems early, even before you have symptoms. Regular blood tests are also needed to be sure your immune system is adequately suppressed so your new lungs are not rejected. The blood tests will also help monitor other organs (kidneys and liver). This is important as other organs can be affected by medications needed after a transplant.

What are the most common medical problems people face after a lung transplant?

1. Rejection
2. Infection
3. Increased risk of skin cancer and other cancers

What is rejection?

Your immune system protects you against germs, and other cells that don't belong in your body. It will recognize your new lungs as new or 'foreign'. It will try to attack and damage them. This is called rejection. After the transplant, you will be given anti-rejection medications (or immunosuppressants), to prevent rejection. Anti-rejection medications must be taken forever, or rejection will occur.

What do anti-rejection medications do?

Anti-rejection medications lower your body's immune responses. This helps to prevent your body from attacking the new lung(s). A combination of anti-rejection medications is needed to prevent rejection. Most lung transplant patients have to take 3 types of anti-rejection medications. Anti-rejection medications have to be taken exactly as instructed.

Anti-rejection medications are very effective at lowering your immune system. This is great for preventing rejection. However, it also means that you will be at a higher risk of becoming, sick or getting an infection. With time, the amount of anti-rejection drugs you will need to take will be reduced. Anti-rejection medications should only be adjusted by the transplant team.

Anti-rejection medications can also have other side effects. Some examples include kidney damage, high blood pressure, increased cholesterol levels, or increased blood sugar. Some people have to take other medications to help with the side effects of the anti-rejection medications. You may also need to take antibiotics or anti-viral medications to help prevent lung infections. Anti-rejection medications also increase the risk of cancers.

What is acute rejection?

Just over a third of all lung transplant recipients will develop acute rejection within the first year after their transplant. It can, however, happen at any time. This is temporary and can be fixed in hospital with medication.

What is chronic rejection (bronchiolitis obliterans syndrome)?

When the body rejects the donated lung over a long period of time, the smallest airways inside the lungs (bronchioles), become swollen and scarred causing the airways to become very narrow. This makes it difficult to breathe. Chronic rejection is difficult to treat. It is the most common reason for death in those with a lung transplant. In very rare cases, some may receive another lung transplant. This is usually identified by specific changes in your pulmonary function tests (lung breathing tests). If these changes happen, you may need to have a bronchoscopy (a tube-like instrument inserted into your airways to view your breathing passages) with a biopsy (small

sample of tissue removed and examined) of your transplanted lungs before treatments for chronic rejection can begin.

What can cause rejection?

The most common reasons why rejection can occur include the following:

- Infections that involve the lungs
- Injury to the lungs that happens during and immediately after transplant surgery (primary graft dysfunction)
- Not taking immunosuppressive medicines regularly and following your treatment plan after transplant
- Gastroesophageal Reflux Disease (GERD) - Gastroesophageal reflux disease (GERD) is a chronic digestive disease. GERD occurs when stomach acid or content flows back into your food pipe (esophagus). The backwash (reflux) irritates the lining of your esophagus and causes GERD.

What are the signs that may indicate rejection?

- Fever
- Flu-like symptoms
- Chest congestion
- Coughing
- Shortness of breath
- New pain around the lung
- Generally feeling unwell
- Fatigue

Talk to you transplant provider right away if you have any new or worsening of symptoms.

Your health care provider team will also monitor you for rejection with breathing tests and bronchoscopy. (See chapter 2 for more on breathing tests and bronchoscopy).

How is rejection treated?

Mild rejection is usually treated with a high dose of corticosteroids (anti-inflammatory medications), which are given first by intravenous (IV) and then by pill. More severe acute rejection is treated with additional medication that will slow down or stop your immune cells even more.

What can I do to help prevent infection and/or illness?

- ✓ Wash your hands with water and soap properly and often
- ✓ Take care of your teeth and gums
- ✓ Protect your skin from scratches and sores
- ✓ Speak to your health care provider about getting a flu vaccination and a pneumonia vaccination
- ✓ Try to avoid large crowds and people with colds and flus

*Note: Some other vaccines are unsafe for people who have had an organ transplant. Check with your transplant team before receiving any other vaccines.

Why will I be more at risk of developing skin cancer and other types of cancers if I have a lung transplant?

The anti-rejection medicines increase your risk of cancer. Regular visits with your transplant health care team, will help you to watch out for and manage complications. Prevention measures such as wearing sunscreen, light weight long sleeves and pants as well as a hat when outdoors will also help lower your risk.

The Cost of a Lung Transplant

Having a lung transplant is a very expensive process. Although the cost of the surgery itself is covered, there are many other costs associated with the process. It is important to consider what your plan might be for funding these related costs.

For residents of Canada, some provinces will pay for all the tests done in hospital, appointments with health care professionals, the surgery itself, and the stay in hospital after the surgery. This usually also includes the cost of immunosuppression medications for those that have received a lung transplant.

What costs are often not covered?

Cell Phone or Pager: You will need to have a cell phone or pager with you at all times while you are on the transplant waiting list so that you can be notified if a suitable donor has been found. This is not usually covered.

Travel and Accommodations: Travel and accommodations are rarely paid for by provincial/territorial government. Methods of travel will vary depending on where you live, your medical condition,

the length of stay, and your financial situation. Once you have arrived in the city of the transplant center, will also require transportation to and from the hospital. Parking is also an additional cost. You and your support person will need to stay in the city where the transplant center is located for the six week transplant assessment phase and usually for about 3 months during the recovery phase after the lung transplant. Accommodations may include staying with family or friends, hotels or renting near the transplant center. You may also need to eat out more often during the weeks of testing.

As a lung transplant candidate, you will have to be very careful that you do not get any lung infections before or after the surgery. The risk of infection is higher once the surgery is complete due to the anti-rejection medications. For this reason, the transplant center specialists do not want you living in a place where you will be sharing space with a lot of people. If possible, consider other transportation options other than public transportation (bus, streetcar, subway) during this time for the same reason.

People that live outside of where the transplant center is located usually rent an apartment that they will share only with their support person.

Lost Wages: If you are working, you and your support persons lost wages will not be compensated by the transplant program.

Medical Equipment:

- **Oxygen:** If you require home oxygen before the surgery that is covered by a private health insurance plan or government plan, it is important to ensure that your provider will be able to accommodate you if you need to travel for your transplant (especially if this is out of province).

You will be required to purchase specific medical equipment to monitor your health status after your lung transplant.

- **Micro-spirometer:** a device used to monitor your lung function (breathing) every day at home after the transplant. (~\$300-\$600)
- **Alert Bracelet:** (~ \$100 for the first year and \$40/year after that)
- **Digital Thermometer:** to monitor temperature – a common sign of infection (~\$20)
- **Digital Weigh Scale:** (~\$40)
- **Digital Blood Pressure Cuff:** (~\$100)

You may also be required to purchase other medical equipment.

Certain Medications: Medications needed to treat side effects of the anti-rejection/suppressant drugs or other conditions will need to be paid for by either your private insurance or by yourself. Contact your insurance company to determine which medication will be covered.

Are there assistance programs available?

Lung Saskatchewan: Lung Saskatchewan offers the Caring Breaths Financial Assistance Program to reimburse essential support programs, services and medical equipment to improve lung health for patients and their primary caregivers. Visit LungSask.ca/Caring-Breaths for more information.

Service Clubs and Community/Charitable

Groups: Speak to your transplant team social worker to find out about community service clubs and community groups that fundraise for medical needs of residents that are not covered. (Some examples that are not limited to and will vary depending on your province/territory include: The Lion's Club, Rotary Clubs, Kinsmen Club, Shriners, Knights of Columbus, Kiwanis Club, and Royal Canadian Legions).

Accommodations: Some provinces have housing available at a lower rate that is designated for people who need to stay close to the hospital for medical procedures such as a lung transplant. Speak to your transplant team social worker to find out what is available in the city where your transplant center is located.

Also make sure to talk to a travel agent or search the internet for a hotel/motel/bed and breakfast close to the hospital. Hotels can be very expensive. Call the hotel directly and ask if they have any special rates. Some hotels close to the transplant center may offer a discount to people travelling for medical reasons.

Travel: Speak to your transplant team social worker to find out if there are transportation programs/services that are available for this reason specifically and if you can have access to them.

Hope Air is a charitable organization with a mission "to help Canadians in financial need fly to necessary medical treatment." 1-877-346-4673 www.hopeair.org

Friends and Family: Don't be scared to ask for help. Your friends and family may organize a fundraising event or they may be able to help you financially. They might have rewards points they can use for travel tickets for you and your support person, groceries, or hotel accommodations. (Examples of some "loyalty reward points:" Air Miles®, Aeroplan®, President's Choice PC® Points, Via Rail Preference Points™).

Canada Health Act: Under the Canada Health Act, the Federal government has the responsibility for funding health care services for certain groups including: the Canadian Armed Forces (CAF), the Royal Canadian Mounted Police (RCMP), First Nations and Inuit, and inmates of federal prisons. People in one of these categories may be eligible for medical travel benefits and medical coverage that is not provided in the provincial/territorial health plan.

Current members of the Canadian Armed Forces (CAF): Contact your human resources department.

Former members of the CAF and RCMP: Contact Veterans Affairs: 1-866-522-2122 or www.vac-acc.gc.ca

Employment Insurance Compassionate Care

Benefits for Caregivers: A person may be eligible for Employment Insurance Compassionate Care Benefits if they are away from work to provide care or support to a very ill family member. Service Canada: 1-800-206-7218 www.servicecanada.gc.ca

Canada Pension Disability: The Canada Pension Disability program's primary role is to replace portion of income for CPP contributors who cannot work because of a disability that is both severe and prolonged. www.servicecanada.gc.ca 1-800-277-9914

Income Tax Tips

Certain medical expenses related to organ transplants may be deductible from your income tax. You may also qualify for the Disability Tax Credit. Everyone's tax situation is different, contact an accountant or professional tax advisor to discuss your options. You may need to gather or save specific receipts or paperwork. Ask about medical costs and prescriptions not covered, premiums paid under a Health Service Plan, temporary housing costs, parking at the hospital, food, non-prescription drugs prescribed by a doctor, renovations/alterations to the home because of your health.

You may also qualify for the Disability Tax Credit during your recovery from your transplant surgery.

Canada Revenue Agency (CRA): www.cra-arc.gc.ca

Canada Pension Plan Disability Benefit: You may qualify to receive funds from the Canada Pension Plan Disability Benefit. www.canada.ca/en/services/benefits/publicpensions/cpp/cpp-disability-benefit/eligibility.html

Private Health Insurance Plans: Private Health Insurance Plans offer different coverage for medications, travel and other services depending on the plan itself and the insurance provider/company. Group health insurance plans may be available through your employer or bank.

Bank or Credit Union: You might have savings, RRSPs, or other investments that can be cashed in. Some people get a line of credit or take out a second mortgage. Speak to your bank/credit union, an accountant or a financial advisor to see what your options are. It is important to understand the consequences of cashing in any investments or insurance policies before you do this.

Your home: You and your support person may need to temporarily relocate for a few months closer to the transplant center. You may also not be able to afford the home you currently have with the loss of wages of you and possibly your support person. Although renting out your home while you are away can be stressful, it may help you financially. It may make financial sense to downsize your home or move to a more affordable area as well.

Building a Budget: Funding a lung transplant is not easy for most people. You may have some difficult financial choices to make. Ask advice before your run into money problems.

Speak to the social worker on your transplant team about any financial concerns you have.

How long can people live after a lung transplant?

The survival rates will vary for everyone. This depends on rejections, infections, side effects, age and other medical conditions. The survival rate and success of transplants continues to improve.

Lung Transplant Planning Checklist

If you would like to be considered for a lung transplant, the checklist below is a tool to help guide you through the process.

Lung transplant inquiries

- Make an appointment with my doctor or lung specialists to ask if I could be considered for a lung transplant
- Write down my questions before any appointments to make it easier to remember them.
- Discuss any concerns or questions I have with family, doctor, respirologist (lung specialist), and/or members of the transplant team.
- Ask my health care team if I need to lose weight or make any lifestyle changes.
- Have a dental check-up and get any needed dental work done.
- Discuss with close family and friends what it means to be placed on the lung transplant list.
- Decide if I want to speak to someone who has already had a lung transplant (Speak to your transplant team social worker or your provincial/territory Lung Association to connect you with someone who has already gone through a lung transplant).

Support Person or Team

- Decide who my support person or team of people will be.
- Ask my support person if they can afford to leave their job or relocate. (If you live outside of the city the transplant center is located in).
- Share with my support person that they may qualify for financial supports such as Employee Insurance or Compassionate Care Benefits.

Health Insurance:

Do I have a health insurance plan? Yes ___ No ___

If I have a health insurance plan check my coverage for:

- Anti-rejection medications
- Non-prescription drugs
- Other medications (medications to treat side effects of anti-rejection medications)
- Medical Travel Expenses
- Oxygen (if needed)
- Micro-spirometer

- Medical Alert Bracelet
- Digital Thermometer
- Digital Weigh Scale
- Digital Blood Pressure Cuff
- Is coverage valid outside of the province and for how long? (* If I would need to travel outside of my province for a lung transplant).
- What is the co-payment or deductible for my health insurance plan?

If I don't have a health insurance plan:

- Can I get one with my "pre-existing" lung condition?
- Would a plan cover treatments and medications for my current lung condition?
- What are the premiums and co-payment or deductible?
- Let provincial/territorial health care plan know if I move out of province for a lung transplant.

Travel and Accommodations Costs (if applicable)

- Gather information on places to live close to the transplant center and costs that may be covered or discounted
- Add up costs to travel between my home and transplant center
 - Initial assessment week
 - Regular planned and scheduled check-ups: (usually this is done every 3 months during the first year of transplant, every 6 months during the second year and then once a year). – Confirm this schedule with your transplant team.
- Add up costs for accommodations for:
 - Initial assessment (one week of tests)
 - 2-3 days for each check-up after transplant
 - Apartment rental while on wait list (time unknown).* Some people need to live closer to the transplant center while on the transplant waitlist.
- Add up the approximate costs for food and other living expenses per month while living away from home
- Add up the approximate costs for phone, cable electricity and hook-up fees
- Add up transportation costs to get to the transplant center

- If I use home oxygen – contact my supplier to arrange for setup in my apartment in my new location and during travel for transplant

Funding for Expenses

- Write down all the sources of income and savings or investments I have.
- Am I eligible for other benefits?
- Can I get disability benefits (Canada Pension Plan, Union, Government Programs)
- Decide if I need to sell my house, downsize, or move in with family or friends.
- Discuss all my options for funding with my accountant, lawyer, real estate agent, banking representative
- Look into redeeming loyalty reward points (Ex. Travel Rewards etc.)
- Think about having a fundraising event. (Ask service clubs or other organizations to help).

Taxes

- Discuss my tax situation with an accountant or tax advisor.
- Keep receipts for renovations or equipment needed because of any disability.
- Keep a log of mileage and expenses for travel.
- Keep receipts for any expenses I pay that are related to the transplant.
- Find out about disability tax credit.

Other Notes _____

Organ donation can give the gift of breath.

Speak to your families about organ and tissue donation today!

Chapter 6 Summary

- For those suffering from severe lung disease, a lung transplant may be their last treatment option to survive. A lung transplant also comes with health risks and it is a lifelong commitment as you will need medication and treatment for the rest of your life.
- When you have a lung transplant, your diseased or damaged lung(s) are removed and replaced with a healthy lung that is donated from another body (donor lung). One or both lungs can be replaced.
- There is a list of criteria that identify who can have a lung transplant. In order to qualify for surgery, you need to be evaluated by transplant health care professionals. A team of medical staff will inquire about medical problems, complete several medical tests and screen for various cancers.
- If you are a suitable candidate for a lung transplant, you will be asked to attend a physiotherapy program and sometime during this program you will be added to the lung transplantation wait list. This program is done at your transplant center, which may not be where you live which might require you and your support person to travel and find accommodations.
- Everyone who is on the transplant list will need to have a support person. A support person should be okay with spending lots of time in hospitals as they will be accompanying you to tests, appointments and with you during your recovery.
- Recovery from a lung transplant will vary for everyone. Most people who have a lung transplant are in the Intensive Care Unit (ICU) in the hospital for a few days to a week. Usually they then remain in the hospital for three or more weeks after that. After being released from the hospital, you will have to remain near the transplant center for at least two more months. (A minimum three months total from the time of surgery).
- The most common medical problems people face after a lung transplant are rejection, infection, and an increased risk of skin cancer and other cancers.
- Having a lung transplant is a very expensive process. Although the cost of the surgery itself is covered, there are many other costs associated with the process. It is important to consider what your plan might be for funding these related costs.

- The survival rates for a lung transplant will vary for everyone. This depends on rejections, infections, side effects, age and other medical conditions. The survival rate and success of transplants continues to improve.



“I wasn’t waiting for someone to die, but rather a woman who had no other choice but to wait for someone to give the gift of life.”

Charlotte-L’Oste-Brown
Double Lung Transplant Recipient

CHAPTER 7: END OF LIFE CARE/ADVANCED CARE PLANNING

Other than a possible lung transplant, there is no cure for IPF, and even a lung transplant has significant risks as well. For most people with IPF, with time their health will decline further. There will be more symptoms and acute exacerbations (a sudden worsening of breathing due to complications such as infection or causes that are not known) that are associated with an increased risk of dying. Death is a natural stage in the cycle of human life. Discussing advance directives, goals of care, palliative care, and hospice care, are important conversations in the end of life care in IPF. Because the disease course of IPF is unpredictable, end of life care should be discussed as early as possible.

What is end of life care?

End of life care is an active, caring approach that treats, comforts, and supports those who are living with or dying from chronic life threatening conditions.

End of life care is sensitive to personal, cultural and spiritual values, beliefs and practices and includes support from families and friends. It also includes the period of bereavement.

What is palliative care?

Palliative care is a special kind of healthcare for the both the individual (and their family), who is living with a life threatening illness that is usually at an advanced stage.

Goals of Palliative Care

To provide:

- comfort,
- dignity,
- the best quality of life for both the person dying and their family, and
- relief of pain and other symptoms as much as possible.

Palliative care does not only focus on your physical needs but also your mental, emotional and spiritual well-being. It will not speed up or postpone death.

* Note: End of Life care and Palliative care definitions are from Canadian Advocacy for Canadians, 2000).



Common fears of dying:

- losing independence
- becoming a burden
- losing control
- not being able to breathe (suffocation)
- not being able to communicate
- pain
- being easily forgotten
- losing self and dignity
- dying alone or without anyone caring

Talking about death and dying can be difficult for some people. Many people try to avoid it because they feel they are giving up hope. However, making sure your wishes are known and having a plan in place is beneficial and important.

What is advance care planning?

Advance care planning is understanding and considering options for your future health care decisions as well as identifying your wishes. An advance care plan may be a set of verbal or written instructions describing what kind of care you would want (or not want) if you were no longer able to speak for yourself. It may also include who you would like to make your decisions for you if you can no longer do so.



Common requests when dying:

- To be free of pain,
- To live with the highest possible level of functioning,
- To resolve long standing conflicts,
- To satisfy dying wishes,
- To hand over control of their care to a significant other.

What are advance directives?

Advance directives are directions given as to what kind of care you would like to receive and who is to make these decisions should you not be able to any longer. This also referred to as a 'living will.'

What is a proxy directive?

A proxy directive specifies who you want to make decisions for you when you are no longer able to make the decisions yourself. The terms for this across provinces and territories may vary and may be referred to as:

- Medical Proxy
- Health Representative or Agent
- Power of Attorney for Personal Care
- Power of Attorney for Healthcare

What is Hospice Care?

Hospice services are often for those with severe disability and/or symptoms and may include services within the home or at a dedicated 'hospice' bed or unit in hospital or long-term care home.

What is medical assistance in dying?

On June 17, 2016, the federal government introduced legislation that allows eligible adults to request

medical assistance in dying. The following information is from the Government of Canada's website on Medical Assistance in Dying.

There are two types of medical assisted dying available in Canada.

The doctor or nurse practitioner will:

1. Administer a substance that causes death
2. Prescribe a drug for the person who is eligible for medical assisted dying to take themselves to bring about their own death.

Who is eligible for medical assistance in dying?

In order to be eligible for medical assistance in dying, you must meet all of the following criteria.

You must:

- be eligible for health services funded by the federal government, or a province or territory (generally visitors to Canada are not eligible for medical assistance in dying).
- be at least 18 years old and mentally competent. This means being capable of making health care decisions for yourself.
- have a grievous and irremediable medical condition which includes:
 - a serious illness, disease or disability
 - be in an advanced state of decline that cannot be reversed
 - experience unbearable physical or mental suffering from your illness, disease, disability or state of decline that cannot be relieved under conditions that you consider acceptable
 - be at a point where your natural death has become reasonable foreseeable (this takes into account all of your medical circumstances and does not require a specific prognosis as to how long you have left to live)
 - You do not need to have a fatal or terminal condition to be eligible for medical assistance in dying
- make a voluntary request for medical assistance in dying that is not the result of outside pressure or influence.
- give informed consent to receive medical assistance in dying
 - Informed consent means you have consented (given permission) to medical assistance in dying

after you have received all of the information you need to make your decision including:

- ◆ Your medical diagnosis
 - ◆ Available forms of treatment
 - ◆ Available options to relieve suffering, including palliative care.
- you must be able to give informed consent both: at the time of your request and immediately before medical assistance in dying is provided.
 - you can withdraw your consent at any time and in any manner.

How do I request medical assistance in dying?

You can ask a health care provider for information about medical assistance in dying at any time.

The process for requesting medical assistance in dying requires you to complete the following steps.

1. **Talk to your doctor or nurse practitioner about end of life care options in relation to your medical condition or circumstance.**
2. **Submit a written request:** The written request must state that you want to have a medically assisted death. Some provinces and territories may require that you complete a specific form. This form may be provided by your health care provider or available on a provincial or territorial website. If you are unable to write, another adult can sign the request on your behalf under clear direction. This adult must be at least 18 years of age, understand what it means to request medical assistance in dying and not benefit from your death (example: they must not be an heir to your estate). The written request must be signed and dated before two independent witnesses, who must also sign and date the request. An independent witness must be 18 years of age and understand what it means to request medical assistance in dying as well. To be considered independent means that the witnesses cannot benefit from your death, be an owner or operator of a health care facility where you live or are receiving care and cannot be directly involved in providing you with health or personal care.
3. **Undergo medical assessments:** Your doctor or nurse practitioner must make sure that you are eligible to receive medical assistance in dying according to all of the listed criteria. A second

doctor or nurse practitioner must also provide a written opinion confirming that you are eligible. You must also be informed that you have the right to withdraw your request at any time. The physician or nurse practitioner providing the original assessment and the one giving the second opinion must be independent. They must not hold a position of authority over the other, could knowingly benefit from your death, and they cannot be connected to the other or to you in a way that could affect their objectivity.

4. **Wait a 10-day reflection period to receive the service.** You must wait a period of at least 10 clear days after signing your written request before the service can be provided so that you have time to consider your request. If you do decide to proceed after the 10 days, your practitioner can have confidence in your true desire to receive the service. An exception may be made to the 10 clear day reflection period if both you and your first and second medical doctor or nurse practitioner agree that your death is fast approaching, and/or you might soon lose your capacity to provide informed consent.

Can I withdraw after I have requested medical assistance in dying?

Yes. You may choose to withdraw your request at any time in the process and in any manner.

For more information on medical assistance in dying visit the Government of Canada's website: www.canada.ca/en/health-canada/services/medical-assistance-dying.html

What do you want your end of life care to look like?

- Take time to reminisce and reflect on your life.
- Express your true feelings, fears, expectations, wishes and hopes.
- Ask all of your questions.
- Talk about it.

End of life care questions to consider

- What are your end of life goals?
- Do you wish to be cured?
- Do you wish to live longer?
- Do you wish to improve or maintain function/ quality of life/independence?
- Do you wish to be pain free and comfortable?
- Do you wish to maintain control?
- What does a good death look like to you?
- Who would you like to support you?
- Have you previously had an experience with serious illness, or has someone close to you had an experience with serious illness or death? If so, if you were in this situation again, what would you hope for?
- What worries you the most?
- Do you think your symptoms and illness will eventually be worse than death?
- If you can no longer make decisions for yourself, who would you like to make them for you?
- What if, based on changes in your health, the doctors recommend something different from what you have told your love(d) ones or the people making decisions on your behalf?
- Will you give your loved one(s) permission to work with your doctors to make the best decision possible for you, even if it may differ from what you said you wanted in the past?
- Are there certain decisions about your health that you would never want your loved one(s) to change under any circumstances?
- Where do you want to die (at home, or in care)?
- Do you have any spiritual or cultural requests when you die?

For more information on advance care planning for you or your loved ones including work books and guides visit Speak Up's website

www.advancereplanning.ca

Chapter 7 Summary

- End of life care is an active, compassionate approach that treats, comforts and supports those who are living with or dying from chronic life threatening conditions.
- Palliative care is a special kind of healthcare for the both the individual (and their family), who is living with a life threatening illness that is usually at an advanced stage.
- Advanced care planning is understanding and considering options for your future health care decisions as well as identifying your wishes.
- Advance directives are directions given as to what kind of care you would like to receive and who is to make these decisions should you not be able to any longer. This also referred to as a 'living will.'
- A proxy directive specifies who you want to make decisions for you when you are no longer are able to make the decisions yourself.
- Hospice services are often for those with severe disability and/or symptoms and may provide services within the home or a dedicated 'hospice' bed or unit in hospital or long-term care home.
- On June 17, 2016, the federal government introduced legislation that allows eligible adults to request medical assistance in dying. There is specific information as to who would be considered eligible from the Government of Canada's website. There are two types of medical assisted dying available in Canada.

The doctor or nurse practitioner will:

1. Administer a substance that causes death
 2. Prescribe a drug for the person who is eligible for medical assisted dying to take themselves to bring about their own death.
- For more information on advance care planning for you or your loved ones including work books and guides visit Speak Up's website www.advacecareplanning.ca

“Have you ever watched an infant sleep, or sat at the side of a grandparent as they quietly rest? There is something remarkable about every breath taken. Breathing allows you to feel the beauty of being alive and present.”

Lori Nagy
Professional Education Associate
Lung Saskatchewan

CHAPTER 8: ALTERNATIVE THERAPIES AND CLINICAL TRIALS

Because IPF has no cure (other than a lung transplant), and treatments are still rather limited, some people choose to try alternative therapies such as naturopathy (the use of natural agents such as water, air, herbs and tissue manipulation and/or electrotherapy), acupuncture (a Chinese medical practice or procedure that treats illness or provides local anesthesia by the insertion of needles at specific sites of the body), hypnosis (the treatment of a symptom, disease by means of hypnotism), salt room therapies (inhaling of low concentrations of salt deep into the lungs) as well as others.

Alternative therapies are not scientifically proven to be both valid and reliable treatments. If you choose to try any form of alternative therapy, be informed about your decision and any possible risks it may have. Be sure to let your health care provider team know, as it may negatively interact with other treatment or medications you are taking.

Stem Cell Therapy

Stem cell therapies are gaining more and more interest from both the medical community and from the public. Stem cells are in many tissues in our body and serve as a sort of internal repair system. When a stem cell divides each new cell has the potential to either remain a stem cell or become another type of cell with a more specialized function such as a muscle cell, a brain cell, or a red blood cell etc. Given stem cells unique regenerative abilities, stem cells offer new potential for the treatment of disease. However, much more work and research is still needed.

Stem cell therapies are still in their early stages of clinical trials and further research is needed to determine the benefits and/or risks that they may have on lung conditions such as IPF. Such therapies will not be clinically available to patients until the safety and the effectiveness have been rigorously documented and scientifically proven.

Currently, the only option for patients to access stem cell therapy as well as other experimental therapies is through participation in approved clinical trials, in which patients are closely monitored and the potential benefits and harm can be objectively assessed. All treatment therapies should be

discussed with your doctor prior to participation. There are risks involved in participating in clinical trials including unpleasant or serious side-effects and there is no guarantee of a benefit.

Clinical Trials

Clinical trials may allow you to play an active role in your own healthcare, gain access to new and potentially beneficial treatments, obtain expert medical care at leading healthcare facilities, and help others with IPF by contributing to medical research. If you are interested in a clinical trial for IPF, ask your doctors whether there are any trials that you may be able to join.

Information about the current Clinical Trials in Canada for IPF can be found on the Health Canada Website at the following link: www.clinicaltrials.gov

Chapter 8 Summary

- Alternative therapies are not scientifically proven to be both valid and reliable treatments. If you choose to try any form of alternative therapy, be informed about your decision and any possible risks it may have. Be sure to let your health care provider team know, as it may negatively interact with other treatment or medications you are taking.
- Stem cell therapies are still in their early stages of clinical trials and further research is needed to determine the benefits and/or risks that they may have on lung conditions such as IPF.
- Currently, the only option for patients to access stem cell therapy as well as other experimental therapies is through participation in approved clinical trials, in which patients are closely monitored and the potential benefits and harm can be objectively assessed.
- If you are interested in a clinical trial for IPF, ask your doctors whether there are any trials that you may be able to join.
- Information about the current Clinical Trials in Canada for IPF can be found on the Health Canada Website at the following link: www.clinicaltrials.gov

Other Resources:

- American Thoracic Society Patient Education:
www.thoracic.org
- Canadian Association for Retired Persons:
www.carp.ca
- Canadian Association of Transplantation:
www.transplant.ca
- Canadian Blood Services:
www.bloodservices.ca
- Canadian Health Network:
www.canadian-health-network.ca
- Canadian Lung Association:
www.lung.ca
- Canadian Life and Health Insurance Assoc. Inc.:
www.clhia.ca
- Canadian Medical Alert™ Foundation:
www.medicalalert.ca
- Canadian Organ Replacement Register (CORR):
www.cihi/corr
- Canadian Pulmonary Fibrosis Foundation:
www.cpff.ca
- Canadian Society of Transplantation:
www.cst-transplant.ca
- Government of Canada
(Medical Assistance in Dying):
www.canada.ca/en/health-canada/services/medical-assistance-dying.html
- Health Canada Clinical Trials:
www.clinicaltrials.gov
- Living Well with Pulmonary Fibrosis:
www.livingwellwithpmonaryfibrosis.com
- Speak Up:
www.advancecareplanning.ca
- Trafford Publishing (books on lung transplantation):
www.trafford.com
- TransWeb: Transplantation and donation information:
www.transweb.org



LUNG AMBASSADOR

Charlotte L'Oste Brown

Double Lung Transplant Recipient, IPF

Charlotte was diagnosed with pulmonary fibrosis in 2003. In the summer of 2014, a lung infection and thick forest fire smoke caused Charlotte's oxygen saturation to drop to 38%. A normal level is 98%. Charlotte's condition was serious and she was hospitalized. She had two choices: she could die or wait for a lung transplant. Feeling like she had way too much to live for and wanting to spend more time with her daughters, Charlotte chose the latter. On December 1, 2015, she was placed on the transplant wait list.

While waiting for that life changing call, Charlotte became a Lung Ambassador for Lung Saskatchewan. She encouraged everyone to talk to their family about their wishes to become an organ donor. She said, "people should talk about it today because people waiting may not have tomorrow."

Charlotte received a double lung transplant on Mother's Day in 2017. Today she continues to advocate for lung health, research and organ donation as Lung Ambassador with Lung Saskatchewan.



LUNG AMBASSADOR

Nicole Nelson, Double Lung Transplant Recipient, Pulmonary Fibrosis

Nicole Nelson was diagnosed with pulmonary fibrosis at just 18 years old. She had a long family history of an inherited form of pulmonary fibrosis. The disease had already taken 16 of her family members, so when she was told she had it too, although devastating, it didn't come as a big surprise.

In 2013 she noticed a rapid decline in her health. In February of that year she caught a cold that she couldn't seem to get over. She had a dry cough and was very short of breath. She found herself avoiding stairs and parking close to doors because walking far distances and doing what once seemed effortless became impossible. By January 2014 she was placed on the lung transplantation waiting list.

In July 2014, Nicole received her life saving call. After her transplant, Nicole says, "the very, first thing I wanted to do was take the biggest breath I could."

Today, Nicole is back at work full time and enjoys spending time with her husband and pets. Nicole continues to share her story and advocate for improving lung health as a Lung Saskatchewan Lung Ambassador.



LUNG AMBASSADOR

Moe Martin, Double Lung Transplant Recipient, IPF

For those that know Moe Martin, they describe the 69 year old husband and father of two as an optimistic man who has dedicated much of his life to his family, career and community. With over 25 years in the workforce, Moe was excited about retiring. He looked forward to having more time to spend with his family and the ability do the things he dreamed of.

In June 2015, all of Moe's plans were no longer. Pulmonary fibrosis not only threatened to take his retirement dreams away, but also his breath. After undergoing many tests, Moe qualified to be put on the lung transplant list. After months of waiting and hoping, Moe received his life-saving call in the winter of 2017, and a chance to live out his dreams.

Since the transplant Moe is doing exactly what he set out to do in his retirement. He also focuses on maintaining healthy lifestyle with diet and exercise. Moe is a Lung Ambassador for Lung Saskatchewan and continues to advocate for change in lung health with his inspirational story.



LUNG AMBASSADORS

Ken and Bea Ambrose, Their journey with IPF

Ken and Bea Ambrose were married for almost 60 years. Together they had two sons, six grandchildren and four great grandchildren. They describe their family as caring and always there for them.

In 2000, Bea developed a cough and ended up in the hospital with pneumonia. Over the next few years she developed pneumonia several times, and was finally diagnosed with IPF in 2007. Bea was prescribed medication to try and slow the progression of the disease, however the side effects were significant and she was forced to stop taking them.

Bea's diagnosis progressed over time and required her to be on oxygen and uses a walker or wheel chair to get around. Bea enrolled in a pulmonary rehabilitation program where she learned about ways to better manage her IPF including helpful exercises. She wished she had been referred to pulmonary rehabilitation sooner and that there had been more information on IPF when she was first diagnosed.

The couple shared their journey with IPF in this information guide as their hope is this guide will be a valued resource to those diagnosed and their families. "Our wish for the future is that there will be more resources dedicated towards IPF to help find a cure for this terrible disease. We also would like to see more medications that have less side effects to become available to help slow the progression of the disease.

Sadly Bea passed away at the age of 80 in 2019. We are grateful for Ken and Bea's courage to share their story to help others."

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