

LIVING WELL WITH PULMONARY FIBROSIS BY RESPIPLUS™

A Learning Tool for Patients and Their Families



DEALING WITH A DIAGNOSIS OF PULMONARY FIBROSIS

This guide belongs to:

Healthcare Professional:

Institution:

1ST EDITION JUNE 2016

THE SKILLS YOU NEED TO MANAGE YOUR PULMONARY FIBROSIS

Pulmonary Fibrosis (also known as interstitial lung disease) is a disease that can affect every part of your life. As the disease progresses, so will its impact on your life and your daily activities. The exacerbations of the disease, or period of worsening symptoms can greatly affect your quality of life.

There are things that you can do to help you live well with this disease. This module is part of “Living well with Pulmonary Fibrosis”, a self-management program, created for those affected by the disease by medical experts in collaboration with patients.

HOW CAN THIS PROGRAM HELP ME?

With “Living well with Pulmonary Fibrosis” you will learn skills to adopt healthy new lifestyle behaviours. This module is your guide. It was designed for you to be able to review at your own pace. Take it from the top or skip to the topics that are most meaningful to you. Share it with people close to you so that they can understand what you are going through.

We wish you well as you go through this program. Do not hesitate to discuss whatever thoughts and feelings you have with your resource person, professionals in your healthcare team and your physician.

On behalf of Team RESPIPLUS



WELCOME TO THE MODULE "DEALING WITH A DIAGNOSIS OF PULMONARY FIBROSIS"

Identify, with the help of your resource person, the subjects on which you need additional information to better manage your Pulmonary Fibrosis. Check off each subject that interests you or is important for you at this time.

Dealing with a diagnosis of Pulmonary Fibrosis

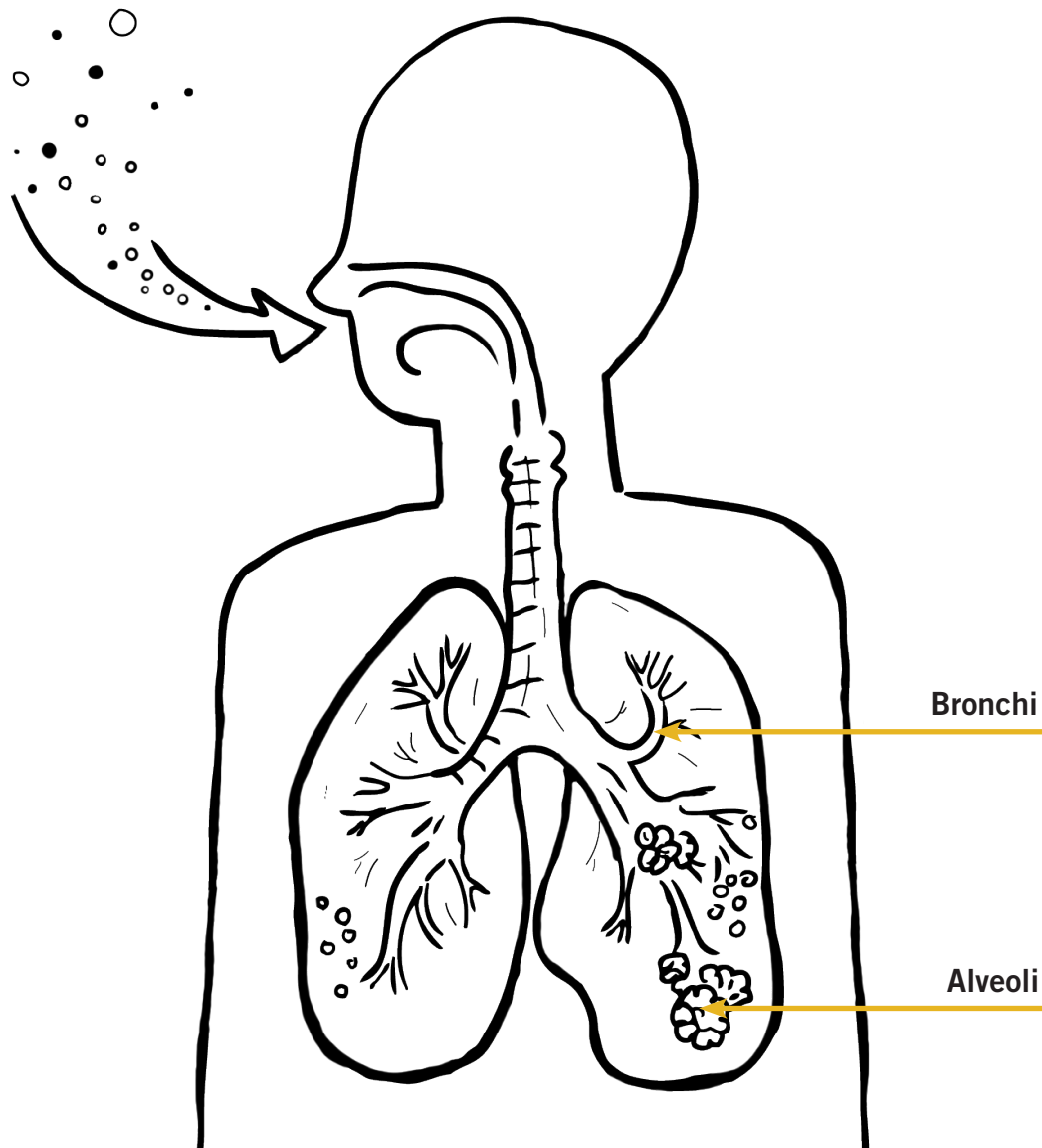
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Living well with Pulmonary Fibrosis means adopting and maintaining healthy life habits and behaviours in order to improve your quality of life.



DEALING WITH
A DIAGNOSIS OF
PULMONARY
FIBROSIS

NORMAL LUNG



1

Every time you breathe, air is drawn into your lungs – along with dust and pollutants.

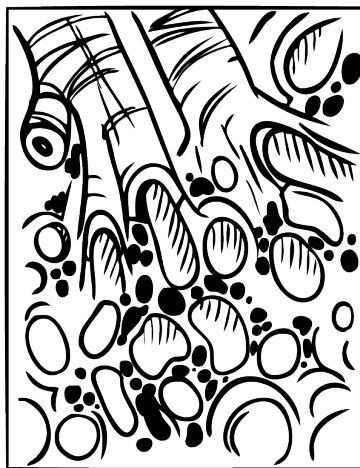
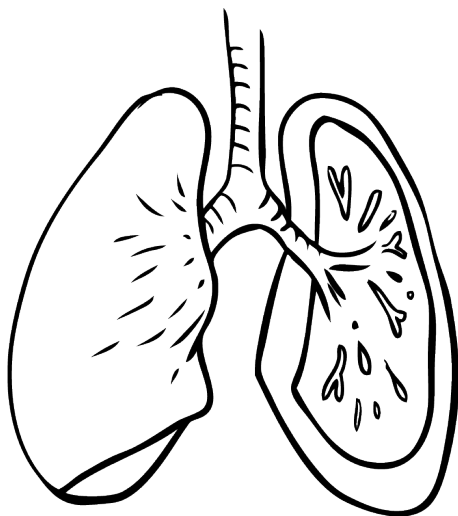
2

Air will go in through the bronchi and reach the alveoli where gas exchange will occur. There, oxygen crosses the lung tissue and enters the bloodstream through the interstitium where it can now reach all the organs in the body.

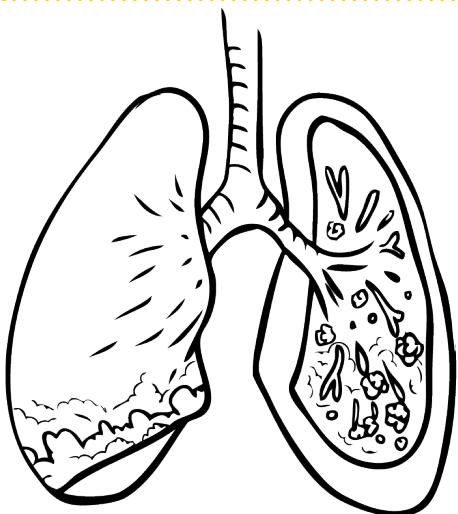
PULMONARY FIBROSIS

Pulmonary fibrosis is a disease characterized by progressive scarring in the lungs. Scar formation causes the lung interstitium (part of the lung where gas exchange occurs) to become thicker and stiffer. After you breathe in, oxygen needs to diffuse through the interstitium to reach the blood circulation. In pulmonary fibrosis, the ***thickened interstitium*** makes it harder for the oxygen to reach the blood circulation.

HEALTHY LUNG



FIBROTIC LUNG



Interstitium

PULMONARY FIBROSIS:

ITS SIGNS AND SYMPTOMS

WHAT ARE THE SIGNS AND SYMPTOMS OF PULMONARY FIBROSIS?

- Shortness of breath, particularly when exercising and during daily activities
- Frequent non productive cough
- Fatigue
- Loss of appetite
- Rounding and enlargement of the fingernails (clubbing) in advanced disease
- Crackle sounds (similar to velcro sounds) may be heard by your doctor when listening to your lungs with a stethoscope



SHORTNESS OF BREATH IN PULMONARY FIBROSIS

When you have pulmonary fibrosis the simple act of breathing can become an effort. Not being able to breathe easily can affect every part of your life. It can affect your body by making you tire out quickly. It can also affect your emotions by making you feel tense and anxious.

WHY DO YOU FEEL SHORT OF BREATH WHEN YOU HAVE PULMONARY FIBROSIS?

When you have pulmonary fibrosis, oxygen has trouble reaching the blood circulation. The interstitium (part of the lung where gas exchange occurs) becomes stiffer and thicker because of the scarring caused by the disease.

YOU FEEL SHORT OF BREATH

Since you have difficulty to move air in your lungs and absorb oxygen, you feel short of breath.

Living better with pulmonary fibrosis means being more in control by saving your energy and relaxing, dealing with anxiety and shortness of breath while continuing to exercise regularly.

In this series of modules, you will learn a number of strategies to do so, which will help you feel less tired and less anxious.

TYPES AND CAUSES OF PULMONARY FIBROSIS

TYPES OF PULMONARY FIBROSIS

Pulmonary fibrosis is a category of major breathing diseases that causes lungs to become thick and stiff over time. There are many different types and causes of pulmonary fibrosis.

- Sometimes the doctors will find that you have a type of fibrosis without any known causes: those will be called ***idiopathic fibrosis***. Examples include ***idiopathic pulmonary fibrosis (IPF)*** and ***idiopathic non-specific interstitial pneumonia (NSIP)***. ***For more information see pages 11 to 12.***
- Sometimes, for many different reasons, the doctors can't clearly identify what type of fibrosis you have. They will say you have an ***unclassifiable pulmonary fibrosis***. ***For more information see page 12.***
- Some fibroses are caused by exposure to different agents. ***Hypersensitivity pneumonitis*** can occur after a repetitive exposure to an antigen. Antigens are inhaled foreign substances such as mold, dust or animal protein (bird exposure for example). ***For more information see page 13.***
- Different ***rheumatologic conditions*** and diseases of the immune system such as rheumatoid arthritis, scleroderma, polymyositis, dermatomyositis, Sjogren's syndrome and mixed connective tissue disease can cause inflammation and fibrosis in the lungs. ***For more information see page 14.***

IDIOPATHIC PULMONARY FIBROSIS (IPF):

WHY DOES IT OCCUR?

The exact mechanism responsible for IPF is not well understood. The word “**Idiopathic**” means the cause of the disease is unknown. Worldwide many people are currently doing research on this topic to try to get a better understanding of the disease.

IPF more commonly affects people from 50 to 70 years old.

Here are some potential risk factors that may cause people to be more susceptible to IPF:

Age	The majority of patients with IPF are diagnosed after the age of 50 years.
Smoking	Majority of IPF patients are smokers or former smokers. If you are smoking, please refer to the module <i>Adopting and Maintaining a Healthy and Fulfilling Lifestyle</i> .
Family history	A genetic component to IPF is suspected. Some families will have multiple members with a diagnosis of IPF.
Acid reflux	Many patients with IPF have acid reflux or heartburn (which your physician may refer to as GERD – Gastroesophageal Reflux Disease). Current research is underway to better understand the link between both these diseases.

The course of the disease can vary dramatically between patients. For example, some people will have rapidly progressive fibrosis while others will experience a slower progression of the disease. Many patients live only about 3 to 5 years after the diagnosis (while some are impacted more quickly and others may survive beyond the 5 year window). There is currently no cure for IPF. New available medications are now used to slow the disease progression in patients with IPF.

IDIOPATHIC NON-SPECIFIC INTERSTITIAL PNEUMONIA (NSIP)

Idiopathic NSIP is a rare type of lung disease with no clear known cause. It affects predominantly women who never smoked. The majority of patients are diagnosed in their sixth decade of life.

The prognosis of patients with idiopathic NSIP is usually good with a low mortality rate.

Treatment of NSIP depends on disease severity and evolution in time. It may be treated with immunosuppressant medication like prednisone or mycophenolate mofetil.

UNCLASSIFIABLE PULMONARY FIBROSIS

Sometimes doctors are unable to identify what type of pulmonary fibrosis you have, they will then say that you have unclassifiable pulmonary fibrosis. Potential reasons explaining why you are being told you have unclassifiable disease include the inability to undergo a surgical lung biopsy (because of too advanced lung disease or too high surgical risk), the inability to make a diagnosis because of insufficient tissue on the lung biopsy or conflicting results between different diagnosis tests.

Refer to the module "Treating Pulmonary Fibrosis" for more information on medications.

HYPERSENSITIVITY PNEUMONITIS (HP)

HP is a type of PF related to an immune reaction within the lung to an inhaled agent. Over 300 different agents, most often organic in origin, have been reported to cause **HP**. The most common exposures (agents) include protein on bird feathers, mold from various sources and sometimes microorganisms such as different bacteria or mycobacterium avium complex (MAC).

The causative exposures cannot be identified in up to 50% of cases.

The central treatment of HP is antigen avoidance.

Treatment with steroids appears to accelerate the initial recovery from some cases of HP, however long-term outcomes may not be affected by corticosteroid treatment. In many cases of HP, we often add an immunosuppressant drug, such as mycophenolate mofetil or azathioprine to help minimize the exposure to steroids.

Refer to the module "Treating Pulmonary Fibrosis" for more information on medications.

CONNECTIVE-TISSUE DISEASE RELATED PULMONARY FIBROSIS

Some type of **autoimmune or connective tissue disease (CTD)** like rheumatoid arthritis, scleroderma, Sjogren's syndrome, systemic lupus erythematosus or myositis can affect the lungs and cause pulmonary fibrosis. CTD can cause lung inflammation or scarring, or both. The exact reason why some patients with CTD develop lung disease and others will not is still unclear.

In some patients the lung disease will be the first sign of the CTD, while others will develop it later in the evolution of the disease.

Treatment of CTD related pulmonary fibrosis depends on disease severity and evolution in time. It is usually treated with immunosuppressant medication. The most commonly used medications are prednisone, mycophenolate mofetil, azathioprine, cyclophosphamide or rituximab. Some of these names may be familiar to you as they can also be used to treat the non-pulmonary manifestations of CTD.

Refer to the module "Treating Pulmonary Fibrosis" for more information on medications.

HOW IS THE DIAGNOSIS OF PULMONARY FIBROSIS MADE?

Diagnosing pulmonary fibrosis and identifying the precise type of fibrosis is a challenging process. This disease may be difficult to diagnose and you may need to see many doctors before knowing what type of pulmonary fibrosis you have.

Your doctor will start by asking you many questions regarding....

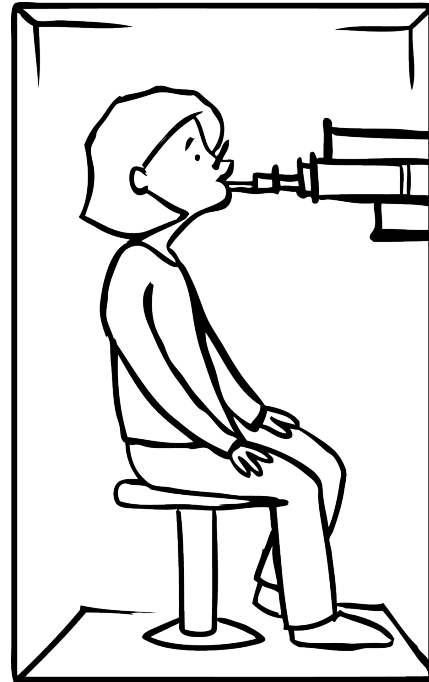
MEDICAL HISTORY AND PHYSICAL EXAM

Your doctor will start by doing a complete medical history and physical exam. You will be asked many questions regarding your medical history, medications, symptoms, your smoking history and exposure to different agents associated with lung fibrosis.

YOUR DOCTOR MAY ORDER SOME OF THESE TESTS TO MAKE THE DIAGNOSIS

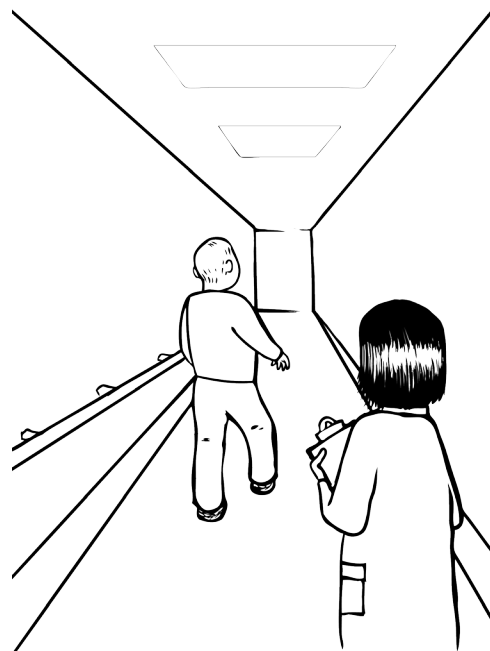
PULMONARY FUNCTION TESTS

These are a series of breathing tests that allow measurements of the volume of air in your lungs, airflow and how well your lungs exchange oxygen. The results of these tests give information about the severity and evolution of the disease.



WALK TEST

During this test you will be asked to walk the longest distance possible in 6 minutes. The distance you walk and your oxygen saturation (the amount of oxygen in your blood) will be measured. A base point will be established so that future walk tests can measure changes over a period of months.



YOUR DOCTOR MAY ORDER SOME OF THESE TESTS TO MAKE THE DIAGNOSIS

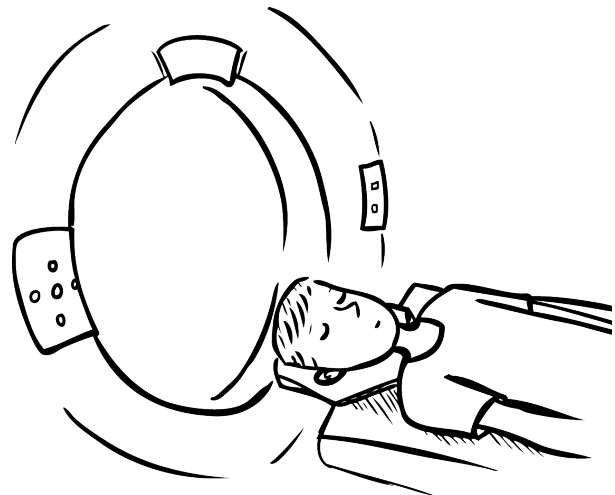
BLOOD TESTS

Your doctor may order blood tests to look for the presence of rheumatologic conditions. These conditions sometimes provide insight concerning the cause or type of Pulmonary Fibrosis.



HIGH RESOLUTION COMPUTED TOMOGRAPHY (HRCT)

This is a type of CT scan that allows your doctor to see clear images from inside your lungs. Some images might be taken while you are lying on your back, on your stomach or while breathing air out. This test is done in only a few minutes and provides your doctor with helpful information regarding the type and severity of the disease.



YOUR DOCTOR MAY ORDER SOME OF THESE TESTS TO MAKE THE DIAGNOSIS

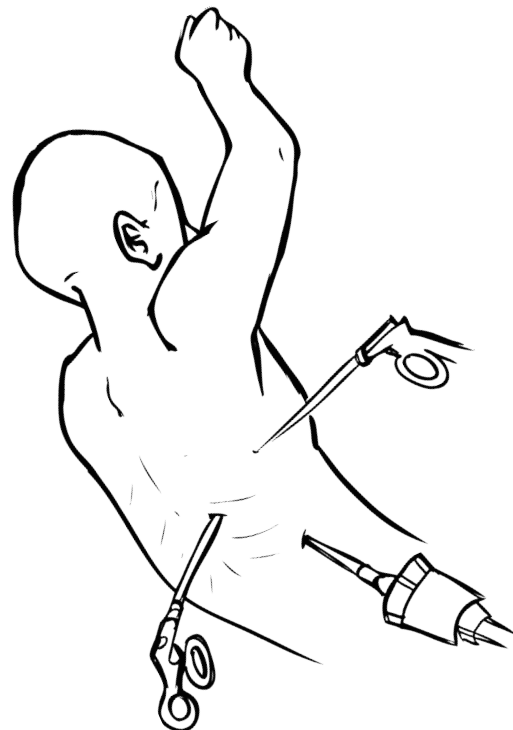
BRONCHOSCOPY

In this test, your doctor introduces, through your nose or mouth, a small flexible tube with a camera at the end. This camera allows visualization of your airways. Fluid and tissue sample may be collected during the procedure.



SURGICAL LUNG BIOPSY

Some patients will need to undergo a surgical lung biopsy to establish a diagnosis. Thoracic surgeons perform this procedure. Patients need to stay in the hospital for a few days after the biopsy. This test is only performed when the type of pulmonary fibrosis cannot be determined by the previously described tests.



WHAT TO EXPECT FOR DISEASE FOLLOW-UP?

Once you get a diagnosis of pulmonary fibrosis you will be scheduled for regular medical visits with your doctor, who will monitor your symptoms.

You may also have access to other healthcare professionals or a care team in which a resource person will be assigned to you. This person will be responsible for coordinating your care and guiding you in the learning of how to self-manage your disease and will be your first contact person when you need help.

You can expect to have repeated pulmonary function tests every couple of months.

Your doctor may also repeat walking tests or CT scans if a re-evaluation of your lung disease is needed.

YOUR DOCTOR HAS SAID YOU HAVE PULMONARY FIBROSIS

What worries you most about the idea of having this diagnosis?

Do you know someone with Pulmonary Fibrosis? If so, have you spoken to that person about it?

It might also be helpful for you to meet other people with pulmonary fibrosis. You could discuss how they rose to the challenge. Please see the resource section on page 26.

Talk to your doctor or your resource person about wanting to meet other patients or contact some pulmonary fibrosis patient associations in your region.

ADJUSTING TO A DIAGNOSIS OF PULMONARY FIBROSIS

A diagnosis of pulmonary fibrosis can change the way you see yourself. You may feel trapped, isolated, of no use to your family and friends. You may feel hopeless. These are common reactions.

We are going to look at the concerns often felt by people who are diagnosed with pulmonary fibrosis:

- **You can discuss them with your doctor or resource person.**
- **Support groups are also a resource for gaining understanding of how to deal with the disease.**
- **You can also develop a plan for improving the situation such as trying to get additional information, cultivating a positive attitude or asking for help.**

ADJUSTING TO A DIAGNOSIS OF PULMONARY FIBROSIS

Emotional Consequences

(perception of the disease progression and awareness of death, feeling of dependency, loss of self-esteem...)

My concerns:

My plan:

Family-Related and Social Consequences

(loss of job, limited recreational activities and mobility, isolation...)

My concerns:

My plan:

Disease-Related Consequences

(symptoms, costs, etc....)

My concerns:

My plan:

DON'T FORGET!

Don't keep your concerns and fears to yourself. Discuss your feelings with your doctor, your resource person, your family and friends.

INFORMING YOUR FAMILY AND FRIENDS OF YOUR DIAGNOSIS OF PULMONARY FIBROSIS

BEFORE:

How will you tell your friends and family about your diagnosis?

How do you plan to address their concerns?

You need to tell your family and friends that even though you have pulmonary fibrosis, you are ready to do as much as you can to continue to be active and to enjoy life within your limits.

Tell them that you need their support to help you cope, manage and live with the disease. Their support starts with being able to discuss with them concerns and fears related to the disease. If possible, clearly inform your carers of how best they can help you.

AFTER:

How did they react to the news?

What were their concerns?

How can your family and friends best help you?

Ask your family and friends to express their concerns and speak to your doctor or resource person about them.

HERE IS AN OPPORTUNITY TO PLAN AHEAD!

You have learned new skills to better manage your Pulmonary Fibrosis. You need to integrate these new habits into your daily life, and maintain them on a long term basis, if you would like to see positive results on your health and on your quality of life. It is important to establish a plan by writing down your objectives (the small steps are as important as the big ones!) and the time to achieve them.

My objectives in order to live better with my Pulmonary Fibrosis for the next month:

1. _____
2. _____
3. _____

Are you confident that you can attain your objectives for the next month?

1	2	3	4	5	6	7	8	9	10
not at all confident					very confident				

My objectives in order to live better with my Pulmonary Fibrosis for the next ____ months:

1. _____
2. _____
3. _____

Are you confident that you can attain your objectives for the next ____ months?

1	2	3	4	5	6	7	8	9	10
not at all confident					very confident				

Notes: _____

Patient's signature

Date: _____

Healthcare professional's signature

Date: _____

THE INTEGRATION AND MAINTENANCE OF YOUR PROGRAM

**TAKE THE TIME TO REVIEW YOUR OBJECTIVES AND
YOUR PLAN.**

1. Which objectives have you been able to attain?

2. How did you manage to attain these objectives?

3. Are there any objectives that you have not yet attained? Which ones?

4. What could help you attain these objectives?

Notes:

HELPFUL RESOURCES



Canadian Lung Association

<http://www.lung.ca/>

Alberta & NWT

<http://www.ab.lung.ca>

Telephone: 780-488-6819

British Columbia

<http://www.bc.lung.ca>

Telephone: 604-731-LUNG (5864)

Manitoba

<http://www.mb.lung.ca/index.php>

Winnipeg: 204-774-5501

Westman Region: (204) 725-4230

New Brunswick

<https://nb.lung.ca>

Telephone: 506-455-8961

Newfoundland & Labrador

<http://www.nf.lung.ca>

Telephone: 709-726-4664

Nova Scotia

<http://www.ns.lung.ca/site/>

Telephone: 1-902-443-8141

Ontario

<http://www.on.lung.ca>

Telephone: 416-864-9911

Prince Edward Island

<http://www.pei.lung.ca>

Telephone: 902-892-5957

Quebec

<http://www.pq.lung.ca>

Telephone: 514-287-7400

Saskatchewan

<https://sk.lung.ca>

Toll Free: 1-888-566-LUNG (5864)



RQAM

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DE L'ASTHME ET DE LA MPOC

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Regroupement Québécois d'asthme et MPOC

<http://www.rqam.ca/>

Telephone: 418-650-9500



Canadian Pulmonary Fibrosis Foundation

<http://www.canadianpulmonaryfibrosis.ca/>

Telephone: 905-294-7645



Canadian Network for Respiratory Care

<http://cnrchome.net/>

Telephone: 905-880-1092

Toll Free: 1-855-355-4672

For a full updated list, you can visit
our website
livingwellwithpulmonaryfibrosis.com

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ACKNOWLEDGEMENTS

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www.LivingWellWithPulmonaryFibrosis.com



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