LIVING WELL WITH PULMONARY FIBROSIS

BY RESPIPLUS™

A Learning Tool for Patients and Their Families

TREATING 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

RESPIPLUS

WELCOME TO THE MODULE "TREATING 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.

Treating Pulmonary Fibrosis

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

TREATING PULMONARY FIBROSIS

MEDICATIONS FOR PULMONARY FIBROSIS AND THE IMPORTANCE OF TAKING THEM PROPERLY

Another very important part of managing your pulmonary fibrosis is to take your medication as prescribed.

Medications do not cure pulmonary fibrosis – but they may help reduce the progression of the disease if taken as directed by your doctor.

YOU SHOULD KNOW THE FOLLOWING ABOUT YOUR MEDICATION:

- the name of each medication you are taking
- what your medication does
- the dose you should be taking
- when you should be taking your medication
- how to take your medication properly
- medication precautions and side effects
- management of side effects

ANTI-FIBROTICS (IPF)

ANTI-FIBROTICS FOR IPF AND THE IMPORTANCE OF TAKING THEM PROPERLY

ANTI-FIBROTICS

This class of medication is recommended for patients with a diagnosis of IPF

Goal of therapy:
To reduce the progression of idiopathic pulmonary fibrosis with the intent of maintaining the respiratory capacity.

- **The results of anti-fibrotic therapies may widely vary from one individual to another**
- **The medication may slow the progression of the disease but will not change your symptoms**
- **It is essential that you take the medication as directed by your doctor and pharmacist**

The exact mechanisms of medications used in the treatment of IPF are also not fully understood. It is important to discuss with your doctor as your treatment progresses to make sure that the medications you are taking are still right for you.

Anti-fibrotics are aimed at slowing the progression of your IPF - these medications will not repair the damage that has already been done to your lungs. They can only attempt to prevent further damage from occurring, or cause it to occur slower.

ANTI-FIBROTICS[1] – GENERAL INFORMATION

Anti-fibrotics are important medications in the management of your idiopathic pulmonary fibrosis.

1. The medication may or may not cause some side effects. These side effects may vary widely from one individual to the next and can be dealt with as they arise with the help of your doctor who may choose to adjust the dosing of medication accordingly.

2. These medications are known to incur some side effects which can be reduced when taken with food.

3. These medications can be affected by tobacco use and must be taken with out smoking. If you are an active smoker, now is a perfect time to begin your journey towards a healthy life tobacco free. Please refer to the module ***"Adopting and Maintaining a Healthy and Fulfilling Lifestyle"*** for more information on this topic, and always let your doctors know if you are actively smoking as this may affect your treatment.

4. These medications may also cause some damage to your liver. Your doctor will usually take bloodwork to make sure your liver is okay before starting the treatment, and may take bloodwork regularly afterwards to make sure you are tolerating your treatment well. This is a completely normal precaution.

5. Any change that you notice after starting the medication should be signaled to your pharmacist, nurse or prescribing doctor.

6. Any time you feel that you are getting short of breath more easily than usual you should always call your pharmacist, nurse or prescribing doctor - never ignore your breathing!

---

[1] At the time of publication, there are only 2 anti-fibrotics available for IPF (Pirfenidone and Nintedanib).

PIRFENIDONE (ESBRIET)

ESBRIET® (pirfenidone) is a treatment for mild to moderate idiopathic pulmonary fibrosis (IPF) in adults. Esbriet is taken orally 3 times daily with food.

POSSIBLE SIDE EFFECTS:

a) **Nausea, loss of appetite and gastric reflux**
b) **Dizziness and fatigue**
c) **Skin rash (especially after being in the sun)**
d) **Diarrhea and bloating**
e) **Damage to your liver**

THEREFORE IT IS IMPORTANT TO:

- Take Esbriet with or immediately after a good sized meal every time.

- Space meals out by at least 4 hours to allow time to digest the food and medication.

- Pay attention to your weight and signal to your doctor or your resource person (if you experience decreased appetite and weight loss).

- Always wear sunscreen to protect from the sun. SPF50 with UV-a and UV-b protection is the minimum, applied twice daily and re-applied as needed. It is also important to wear protective clothing to cover sun exposed areas, including a hat and/or long sleeves.

- Inform your health care provider if you notice any rash forming on your skin, as they may want to temporarily reduce the dosing of the medication to help the rash heal.

- **Your doctor will take a blood sample every month for the first 6 months of treatment to make sure your liver is doing alright, and every 3 months thereafter.**

NINTEDANIB (OFEV™)

OFEV™ (nintedanib) is a prescription medicine used to treat people with IPF. Ofev is taken orally twice daily with food. Your doctor will tell you how much OFEV to take and when to take it.

Taking your medication as prescribed is one of the steps towards maintaining a healthy lifestyle.

POSSIBLE SIDE EFFECTS:

a) **Liver problems**
b) **Diarrhea**
c) **Nausea, and vomiting**

THEREFORE IT IS IMPORTANT TO:

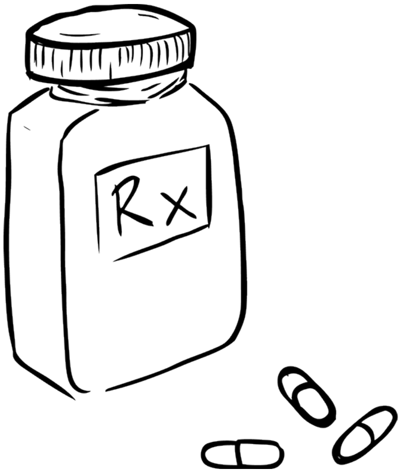
- Take Ofev after a meal with a beverage every time – do not open the capsule or crush it. Take it whole as indicated on the packet.

- Call your doctor right away if you have unexplained symptoms such as yellowing of your skin or the white part of your eyes (jaundice), dark or brown (tea colored) urine, pain on the upper right side of your stomach area (abdomen), bleeding or bruising more easily than normal, or feeling tired.

- Tell your doctor if you have diarrhea, nausea or vomitting or if these symptoms do not go away or become worse. Tell your doctor if you are taking over-the-counter laxatives, stool softeners and other medicines or dietery supplements that can cause diarrhea. Your doctor may recommend that you drink more fluid or take medecine like Loperamide (Imodium®) to treat diarrhea.

TEST YOUR KNOWLEDGE ON ANTI-FIBROTICS

Why are anti-fibrotics for Pulmonary Fibrosis important?		
They reduce shortness of breath	☐ True	☐ False
They may reduce the progression of fibrosis	☐ True	☐ False
They reduce cough	☐ True	☐ False
They may cause side effects	☐ True	☐ False
Following recommendations may reduce side effects	☐ True	☐ False
It is normal to have blood tests regularly	☐ True	☐ False
Anti-Fibrotics should be taken without food	☐ True	☐ False



Review your answers with your doctor or your resource person in order to evaluate your learning needs on medications treating your disease.

IMMUNOSUPRESSANTS

MEDICATION – IMMUNOSUPPRESSANTS

The goal of this category of medication is to suppress the immune system, reduce the inflammation in your lungs and stabilize your lung function.

All of the immunosuppressive medication can decrease your ability to fight an infection.

Wash your hands frequently and try to stay away from people who are sick. Make sure your vaccinations are up to date.

PREDNISONE

Prednisone is often the first line of treatment for different types of pulmonary fibrosis (Hypersensitivity pneumonitis, pulmonary fibrosis related to connective tissue disease or idiopathic NSIP for example). It has a fast onset of action but can be associated with important side effects. Your doctor may want to add another immunosuppressant drug (steroid sparing) to be able to lower the prednisone dose you need.

POSSIBLE SIDE EFFECTS:

a) Changes in mood
b) Increased appetite and weight gain
c) Thin, fragile skin
d) Sleep disturbance
e) Increased in blood pressure
f) Increased in blood sugar
g) Swelling
h) Osteoporosis (weakening of the bones)
i) Increased risk of infection
j) Cataracts and glaucoma

Talk to your doctor if you have any problems while taking the medication.

MONITORING

Your doctor may ask you to monitor your blood pressure and/or blood sugars to make sure it remains within the normal ranges

It is important to remain active and have a good alimentation while on prednisone therapy to try and minimize the potential side effects. Consult our module ***"Get moving... Breathe Easy with Pulmonary Fibrosis"***.

MYCOPHENOLATE MOFETIL (CELLCEPT®)

Mycophenolate is part of the class of drug called immunosuppressants.
It can be recommended for the treatment of your pulmonary fibrosis.

This medication is often taken twice a day.

POSSIBLE SIDE EFFECTS:

a) **Upset stomach, nausea, vomiting**
b) **Diarrhea – constipation**
c) **Low number of white blood cells (leukopenia)**
d) **Increased risk of infections**

Talk to your doctor if you have any problems while taking the medication.

MONITORING

Your doctor will order regular blood tests to monitor the level of your white blood cells.

AZATHIOPRINE (PrIMURAN)

Azathioprine is part of the class of drug called immunosuppressants.
It can be recommended for the treatment of your pulmonary fibrosis.

POSSIBLE SIDE EFFECTS:

a) Upset stomach, nausea, vomiting
b) Diarrhea
c) Low number of white blood cells (leukopenia)
d) Liver damage
e) Increased risk of infections

Talk to your doctor if you have any problems while taking the medication.

MONITORING

Your doctor will order regular blood tests to monitor the level of your white blood cells and liver enzymes.

CYCLOPHOSPHAMIDE (^{Pr}PROCYTOX)

Cyclophosphamide is part of the class of drug called immunosuppressants. It can be recommended for the treatment of your pulmonary fibrosis.

This medication can be given either intravenously every couple of weeks or everyday by mouth.

POSSIBLE SIDE EFFECTS:

a) **Upset stomach, nausea, vomiting**
b) **Fatigue**
c) **Weight loss or lost of appetite**
d) **Damage to your kidneys or urinary tract**
e) **Low number of white blood cells (leukopenia)**
f) **Increased risk of infections**

Talk to your doctor if you have any problems while taking the medication.

MONITORING

Your doctor will order regular blood tests to monitor the level of your white blood cells, liver and renal functions. Serial urine analysis will also be done.

RITUXIMAB (RITUXAN®)

Rituximab is part of the class of drug called immunosuppressants.
It can be recommended for the treatment of your pulmonary fibrosis.

This medication is administered intravenously usually every couple of months.

POSSIBLE SIDE EFFECTS:

a) **Upset stomach, nausea, vomiting**
b) **Fatigue**
c) **Fever and chills occurring during the intravenous treatment (infusion reactions)**
d) **Painful sores or ulcers on lips or mouth**
e) **Low number of white blood cells (leukopenia)**
f) **Increased risk of infections**

Talk to your doctor if you have any problems while taking the medication.

MONITORING

Your doctor will order regular blood tests to monitor the level of your white blood cells, liver and renal functions.

TEST YOUR KNOWLEDGE ON IMMUNOSUPPRESSANT MEDICATION

Why are immunosuppressants for Pulmonary Fibrosis important?		
They suppress the immune system and reduce the inflammation in your lungs	☐ True	☐ False
They may increase your risk of infections	☐ True	☐ False
They can be associated with many side effects	☐ True	☐ False
Many require regular blood tests for toxicity monitoring	☐ True	☐ False

Review your answers with your doctor or your resource person in order to evaluate your learning needs on medications treating your disease.

OTHER MEDICATIONS

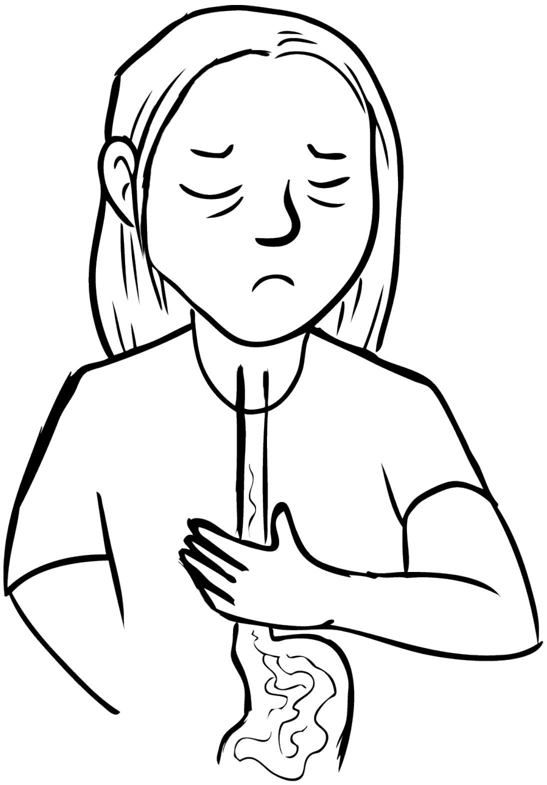
OTHER MEDICATIONS

GASTROESOPHAGEAL REFLUX DISEASE (GERD)

GERD is a condition where acid from your stomach produced for the digestion of food backs up into your throat. It is thought that some individuals with GERD may breathe in tiny drops of acid from their stomachs, which may even cause injury to the lungs. Although the role of GERD in relation to pulmonary fibrosis is still poorly understood, many individuals with pulmonary fibrosis also suffer from this problem.

Often, to treat the symptoms of GERD as well as its potential harm to the lungs, your doctor may use medication to reduce the acidity in your stomach.

These medications are usually in a class named “Proton Pump Inhibitors” (PPIs) and reduce the production of acid in the stomach to relieve GERD.



OTHER MEDICATIONS

It is recommended that all pulmonary fibrosis patients receive a pneumonia vaccine and get their annual flu shots

EXACERBATIONS

An exacerbation for patients with pulmonary fibrosis can be described as a sudden deterioration of lung function without any identifiable cause. For patients, this means a sudden increase in shortness of breath or coughing as well as a decrease in ability to tolerate effort. Although the exact criteria for exacerbations can vary, it is widely accepted that a worsening of pulmonary function within a 30 day period which is not caused by an infection, a heart problem or another type of lung injury can be called an exacerbation. Pulmonary fibrosis exacerbations often leave permanent damage to the lung and contribute to a decrease in lung function which cannot be reversed.

MEDICATIONS TO TREAT EXACERBATIONS

Antibiotics	Oral Anti-Inflammatory (Prednisone)
Are useful in treating respiratory infections (sinusitis, infected bronchitis, pneumonia, etc.)	Can be prescribed for a short period when your respiratory symptoms get worse (exacerbations)
Come in a variety of types	
The treatment has to be taken exactly as prescribed	

IS YOUR MEDICATION WORKING?

PULMONARY FUNCTION TESTS

Once you start taking medication for your pulmonary fibrosis it may be difficult to know exactly how well it is working. Unlike an infection which is cured by antibiotics, pulmonary fibrosis cannot be cured by medication.

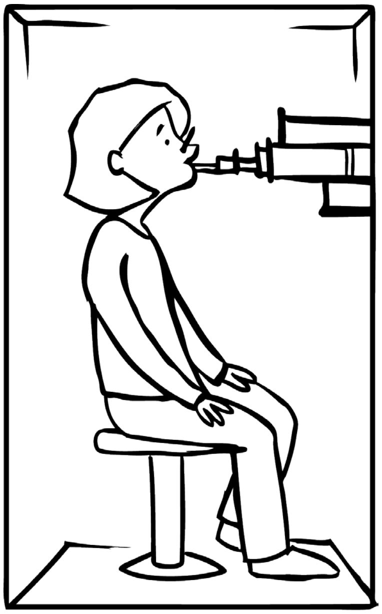
In order to assess the evolution of your disease your doctor may order pulmonary function tests and six minute walk tests at regular intervals.

When you undergo pulmonary function tests, respiratory therapists will make you breathe in different ways to be able to obtain different measurements like forced expiratory flows, lung volumes and the gas exchange capabity of your lungs. Of those, your doctors will be mainly interested in looking at the Forced vital capacity (FVC) and the diffusion capacity of the lung (DLCO).

FVC	DLCO
This represents the maximum amount of air you can breathe out at expiration after you completely fill your lungs during a maximal inspiration.	This measures how well your lungs exchange gases between the air and your circulatory system when you breathe.

Your doctor will look at the evolution of these tests over time to assess if your lung disease is stable or has progressed.

These tests can be physically challenging and it is recommended to be well rested when preparing to undergo them.



NON-PHARMACOLOGICAL TREATMENT

OXYGEN

At the later stages of pulmonary fibrosis, the lungs may have an increased difficulty getting the oxygen from the air into the bloodstream. This usually causes an increase in shortness of breath as well as a decrease in the ability to tolerate activities and effort.

In an effort to curb this loss of lung function, supplemental oxygen may be beneficial in your plan of action. The oxygen supplied in this way may help the lungs get the amount needed to properly supply the body, providing an increase in tolerance for activity and a decrease in shortness of breath.

For more information on oxygen therapy, please refer to the module ***"Integrating a long-term oxygen therapy into your life"*** of this series.

PHYSICAL ACTIVITY AND EXERCISE

The regular practice of exercise can be helpful in increasing your ability to tolerate effort and may increase your endurance. Exercise may also strengthen important muscles involved in breathing, making every breath count for just a little bit more. When dealing with pulmonary fibrosis, it is important to practice **within your limits.**

An exercise regimen can be designed with the help of a physiotherapist or exercise therapist. You may also consult our module ***"Get moving… breathe easy with Pulmonary Fibrosis"*** which promotes the importance of physical activity in general.

If you are an active smoker, quitting will greatly help with physical activity and exercise. For more information, please refer to the module ***"Adopting and Maintaining a Healthy and Fulfilling Lifestyle"*** of this series.

LUNG TRANSPLANT

Lung transplant is a treatment option for a small number of patients with advanced lung disease. Patients need to fulfill precise criteria to be considered a good candidate for transplant. If you have not done so already, you will want to ask your doctor in advance to see if you are a potential candidate for lung transplant. If your doctor considers you may be a good candidate for transplant, he will refer you to a lung transplant clinic where you will meet the transplant team and be evaluated.

CHECK YOUR CONFIDENCE LEVEL IN TAKING YOUR MEDICATIONS

On a scale of 1 to 10, circle the number that best represents the confidence that you have in your ability to apply the strategies and techniques taught in this section.

1. Can you list the medications you are currently taking for your respiratory condition?

1 2 3 4 5 6 7 8 9 10

not at all confident very confident

2. Do you think you can take your medications as prescribed?

1 2 3 4 5 6 7 8 9 10

not at all confident very confident

3. Do you think taking your medications as prescribed can reduce the severity of its side effects?

1 2 3 4 5 6 7 8 9 10

not at all confident very confident

4. Do you think taking your medication as prescribed can slow the progression of your disease?

1 2 3 4 5 6 7 8 9 10

not at all confident very confident

CHECK YOUR CONFIDENCE LEVEL IN TAKING YOUR MEDICATIONS

If you feel confident that you can take your medication as prescribed, continue to apply the strategies suggested in this section.

If you don’t feel confident that you can take your medication as prescribed, what are the reasons?

____

____

What could you do to feel more comfortable to take your medication as prescribed?

____

____

Can you list the medications you are currently taking for your respiratory condition?

____

____

Can you list other medications you are taking?

____

____

Don’t hesitate to discuss your concerns about your medications and how they should be properly taken with your doctor or your resource person.

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

THE LUNG ASSOCIATION™
L'ASSOCIATION PULMONAIRE

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
RÉSEAU QUÉBÉCOIS
DE L'ASTHME ET DE LA MPOC
La référence des professionnels en santé respiratoire

Regroupement Québecois d'asthme et MPOC
http://www.rqam.ca/
Telephone: 418-650-9500

Réseau canadien pour les soins respiratoires
Canadian Network for Respiratory Care

Canadian Network for Respiratory Care
http://cnrchome.net/
Telephone: 905-880-1092
Toll Free: 1-855-355-4672

CANADIAN PULMONARY FIBROSIS FOUNDATION

Canadian Pulmonary Fibrosis Foundation
http://www.canadianpulmonaryfibrosis.ca/
Telephone: 905-294-7645

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

Notes:

Notes:

Notes:

Notes:

Notes:

ACKNOWLEDGEMENTS

Authors

Julie Morisset, MD • Diane Nault, RN, MSc • Alexandre Savard, RN, BScN • Maria Sedeno, BEng, MM

Collaborators

Deborah Assayag, MD • Jean Bourbeau, MD, MSc, FRCP • Bruno-Pierre Dubé, MD • Patricia Côté, RN, M.A., directrice exécutive et conseillère principale, RQESR • Geneviève Tremblay, Directrice programmes de santé, APQ • Dominique Massie, Directrice générale, APQ • Graphic Design: Cassandra Giaccari, BFA

Participating Institutions

- The Research Institute of the McGill University Health Center
- University of California, San Francisco
- Centre Hospitalier de l’Université de Montréal
- RQESR - Réseau québécois d’éducation en santé respiratoire
- APQ - Association pulmonaire du Québec

Living Well With Pulmonary Fibrosis has also been updated in consultation with groups of healthcare professionals and Pulmonary Fibrosis patients participating in a pilot study at the University of California, San Francisco and Centre Hospitalier de l’Université de Montréal.

The development of this program has been made possible thanks to a non-restrictive educational grant offered by:

Boehringer Ingelheim

Roche

LIVING WELL WITH
PULMONARY FIBROSIS
BY RESPIPLUS™

www.LivingWellWithPulmonaryFibrosis.com

RESPIPLUS