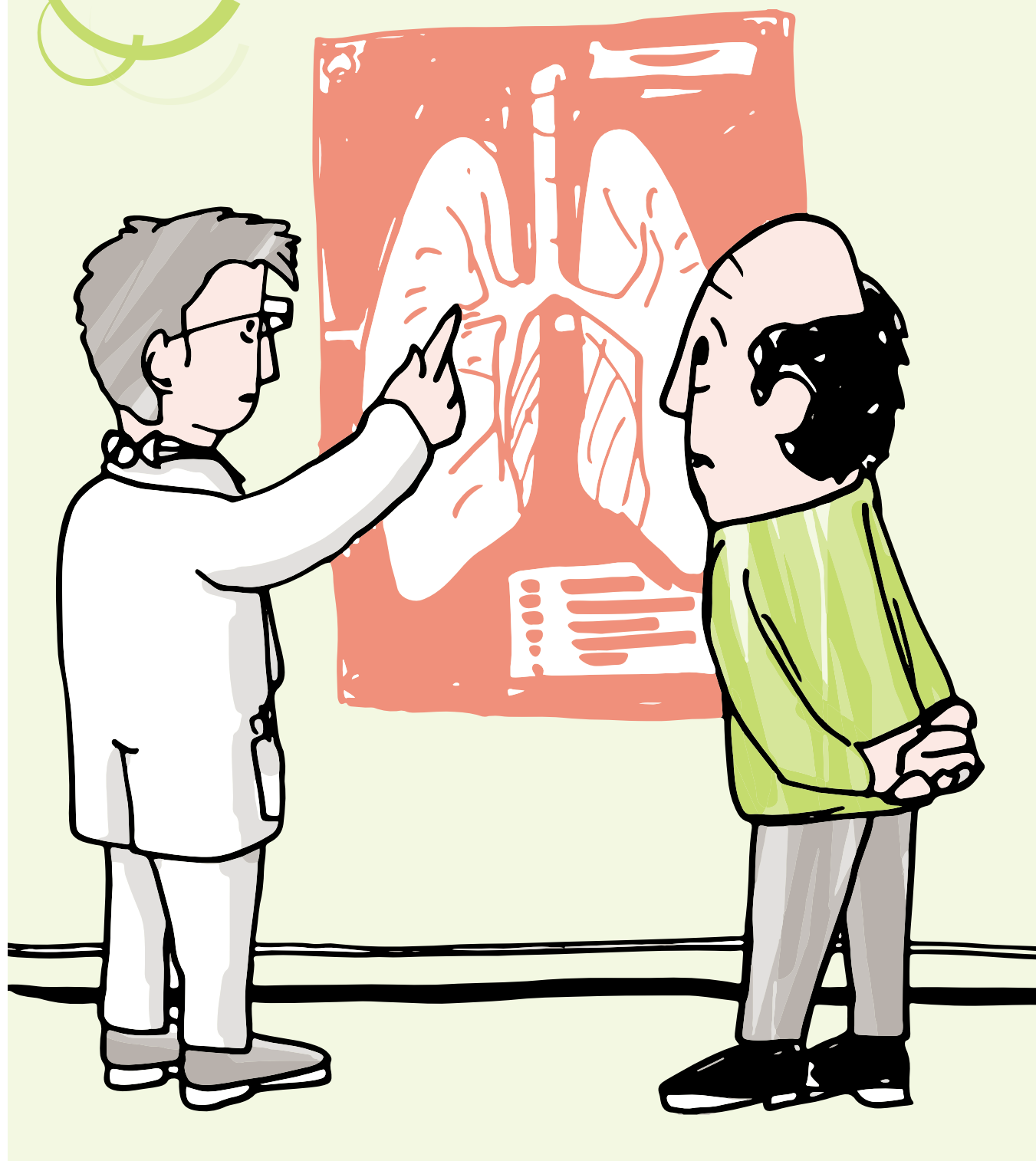


# Life with IPF



## Life with IPF – an eBook

# Preamble

**This eBook is endorsed by the European Idiopathic Pulmonary Fibrosis & Related Disorders Federation (EU-IPFF)**

*“Upon Diagnosis and at different stages on a patient’s IPF journey, accurate unbiased and easily understood information is a vital need for patients and carers.*

*I believe a resource like this compliments the information role of HCPs and Support Groups”*

**Liam Galvin, Secretary, EU-IPFF**



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# 1.0 About Idiopathic Pulmonary Fibrosis (IPF)

## 1.1 IPF – What's behind all this?

Let's meet a typical person with IPF. We will name him Peter. Peter was diagnosed with **Idiopathic Pulmonary Fibrosis (IPF)**, a serious lung disease that is not very frequent. He is often breathless, especially during exercise, and has a dry, hacking cough that doesn't get better accompanied by unintended weight loss, tiredness, and a widening and rounding of the tips of his fingers and toes. As a man over the age of 60 and a smoker, Peter has some antecedents that may increase the risk of developing IPF.



## Signs and Symptoms<sup>1</sup>

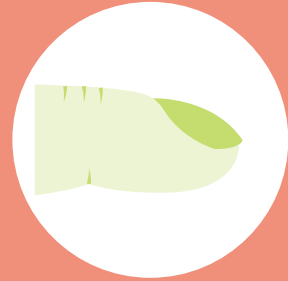
Some of the most common signs and symptoms of IPF include:



Shortness of breath



Dry, hacking cough that doesn't get better



Clubbing (widening and rounding) of the tips of the fingers or toes



Gradual, unintended weight loss



Tiredness and generally feeling unwell



Aching muscles and joints



Rapid, shallow breathing



"Velcro-like" crackles heard by the physician during lung auscultation

## Risk factors for IPF<sup>2</sup>

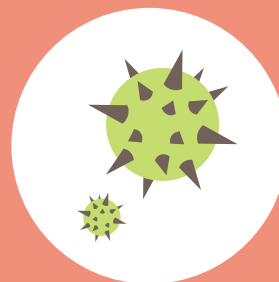
In addition to age above 40 years and male gender, other factors that are thought to increase the risk of developing IPF include:



Cigarette smoking



Environmental exposures



Viral lung infections



Gastro-oesophageal reflux disease (GERD)



Diabetes



Genetic link

## But what exactly is IPF?

IPF stands for Idiopathic Pulmonary Fibrosis, a rare condition that affects 14 – 43 people per 100 000, with a total of approximately 3 million patients worldwide.<sup>3</sup> In people with IPF, the tissue inside and between the tiny air sacs (known as **alveoli**) and blood vessels of the lungs becomes thickened, stiff and scarred. **Fibrosis** is the medical term for this scarring.<sup>2</sup> IPF becomes worse over time and as the scarring gets thicker and more widespread, the lungs lose their ability to transfer oxygen into the bloodstream. This results in a shortness of breath and the organs not getting enough oxygen to function normally.<sup>3,4</sup>

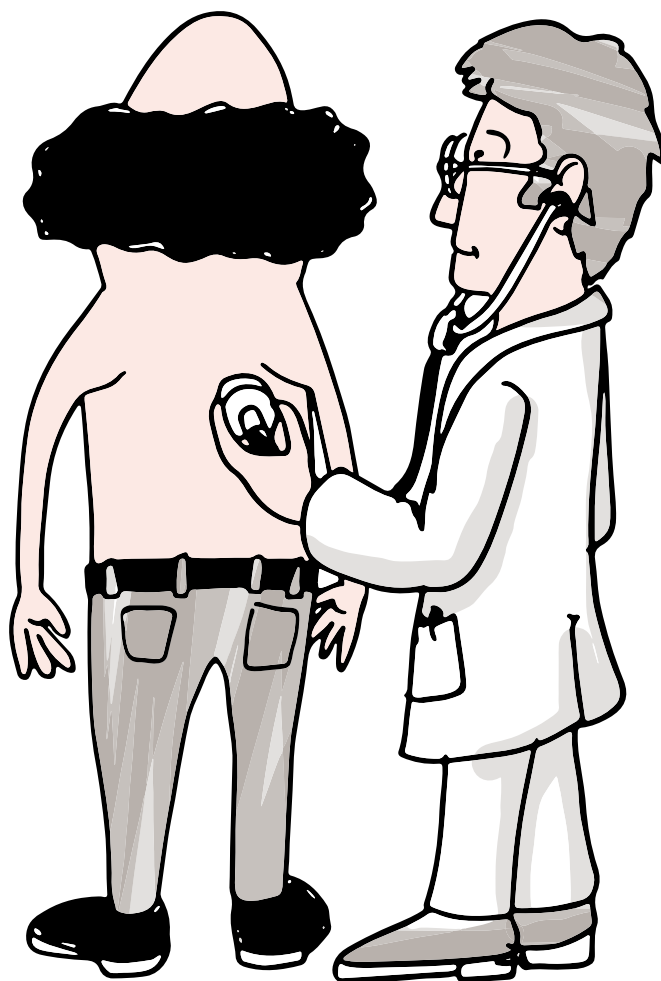
Sometimes doctors can find out what is causing **lung scarring**. For example, exposure to environmental pollutants and certain medicines can cause fibrosis in the lungs.<sup>1</sup> However, in most cases of lung scarring, an exact cause is never established – and that is then what we call Idiopathic Pulmonary Fibrosis (IPF).<sup>1</sup>

## What does IPF stand for?

<b>I</b>	<b>Idiopathic</b>	Of unknown cause
<b>P</b>	<b>Pulmonary</b>	Refers to the lungs
<b>F</b>	<b>Fibrosis</b>	Formation of scar tissue

## 1.2 How is IPF diagnosed?

When Peter initially visited his doctor with breathing problems, his physician attributed his symptoms to coronary heart disease or **chronic obstructive pulmonary disease (COPD)**. As his symptoms did not get better with COPD treatment, he went to see a **pulmonologist** who finally diagnosed Peter with IPF. Peter felt it took a while to get an accurate diagnosis, but as he learned from other people diagnosed with IPF, it can take much longer until a correct diagnosis is established. IPF can be hard to diagnose because it causes the same kind of symptoms as some other lung diseases.<sup>2</sup> There is no single test that can determine an IPF diagnosis, but a medical history, physical examination and several test results all help the pulmonologists to come to the right diagnosis.



In order to achieve the correct diagnosis for Peter, the pulmonologist first had to obtain a **differential diagnosis** and exclude a number of other lung diseases, such as COPD, sarcoidosis or infections. Lung diseases associated with environmental factors, such as exposure to asbestos, side effects of medications or a systemic disease that may affect the lungs indirectly, also needed to be ruled out.

To exclude illnesses that cause symptoms similar to IPF, the pulmonologist conducted a complete and detailed **medical history** to find out about whether Peter has ever smoked, his work, his family medical history, drug use, etc.<sup>2</sup>

A **physical examination** gave the doctor a better understanding of Peter's lung health. When listening to the lungs with a stethoscope, the physician detected a distinctive, Velcro-like crackling sound, which is present in more than 80% of patients with IPF.<sup>5</sup> He also checked Peter's hands for widening and rounding of the fingertips – a common sign of IPF and other respiratory diseases.<sup>1</sup>

In order to conclusively diagnose IPF, Peter's pulmonologist carried out a number of special tests:

To understand how well Peter's lungs are working, Peter had to undergo **pulmonary function tests**, which determined how well his lungs take in and release air.

A routine **chest X-ray** created a picture of Peter's lungs, revealing some lung abnormalities and shadows that suggest scarring of the lung tissue.

However, many patients with significant scarring have a normal chest X-ray, so IPF cannot be ruled out from a chest X-ray alone.

Peter's pulmonologist needed to perform an **HRCT** (High-resolution computerised tomography) of the chest to get a more detailed image of his lungs and thereby identifying specific patterns in the lung tissue which indicate the presence of IPF.

## What is an HRCT scan and how is it done?

An HRCT (high-resolution computerised tomography) scan uses x-rays and a computer to provide very sharp pictures which show cross sections through the organs inside the body, e.g. the heart and the lungs. In contrast to a traditional chest X-ray, an HRCT scan allows physicians to see a lot of small details of the organs, which can be helpful in making a specific diagnosis.

An HRCT scan is generally taken and interpreted by a team of specialists consisting of radiologists, nurses and technologists.

The HRCT scanner is a large, tunnel-like machine which is open at the back and the front. During the scanning procedure, the patient is positioned on a moveable examination table that moves slowly backwards and forwards through the hole in the centre of the scanner.

As the table slides through the opening, an X-ray beam rotates around the patient's body to create a series of pictures of the inside of the body.

To ensure a high quality of the images, the patient is asked to hold very still and at times to hold his breath for a while. Depending on the individual situation, the whole scanning procedure takes between 15 and 30 minutes and is absolutely painless for the patient.

After the exam, the images will be analysed by an imaging physician who specialises in the interpretation of HRCT scans. Shortly after that, they will be submitted to the attending physician, who will then discuss and explain the results to the patient.



HRCT scanner



Chest HRCT image

In about two thirds of the cases, IPF can be diagnosed based on medical history, physical exam, and CT scan. However, for some patients it is also necessary to undergo a so-called surgical **lung biopsy**. In this procedure, a surgeon removes a small lung tissue sample for microscopic analysis to achieve an ultimate diagnosis.

### What is a lung biopsy and how is it done?

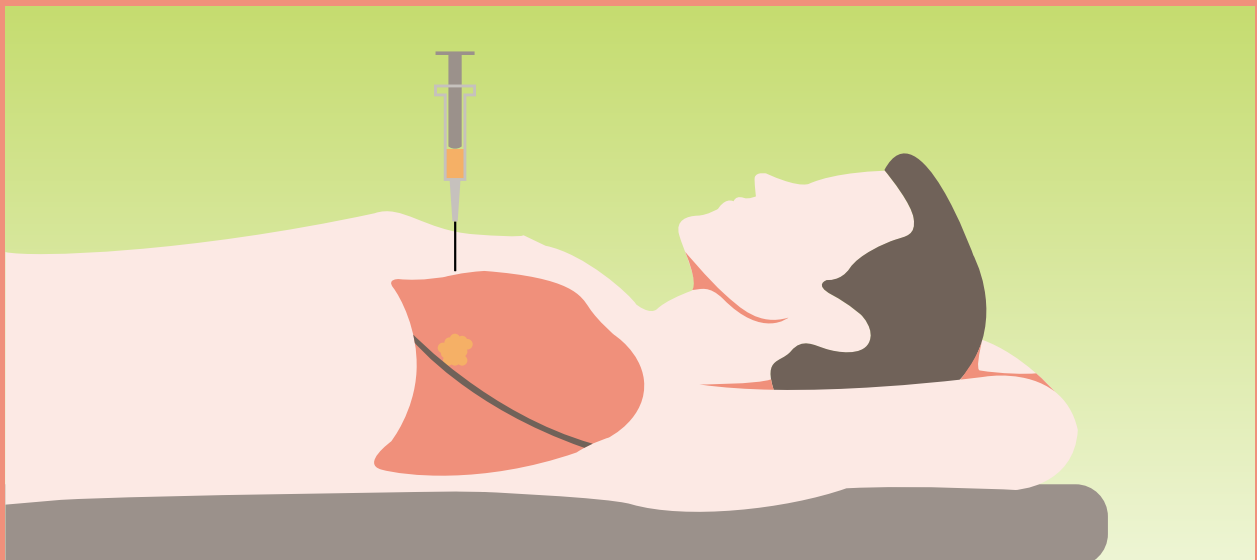
A lung biopsy is a surgical procedure performed by a chest surgeon and a surgical team in which a small piece of lung tissue is removed and analysed. Before the procedure, the patient is put under general anaesthesia.

To look inside the lungs, the surgeon makes a small incision and inserts a tube-like viewing instrument with a miniature video camera into the chest, so a picture of the inner lungs is transmitted to a video monitor.

The physician then identifies the area that appears altered due to disease and extracts a small tissue sample for laboratory analysis. The lung tissue is cut and sealed at the same time by a special device called a stapler.

Once the sample is removed, it is immediately sent to a pathologist who will then examine the tissue under a microscope to diagnose the problem. The final diagnosis usually takes five to seven days.

To ensure a sufficient follow-up care after the surgical intervention, the patient usually needs to stay in hospital for a few days to fully recover from the lung biopsy.



A surgical lung biopsy

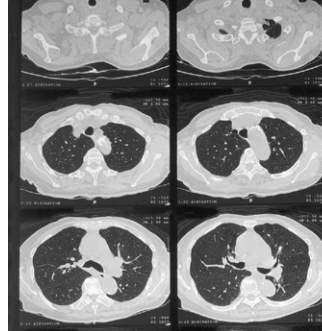
### Diagnostic tests for IPF<sup>6</sup>

#### Chest X-Ray



This creates a picture of your lungs, revealing shadows that suggest scarring.

#### HRCT



A type of X-ray that provides sharper and more detailed pictures than a standard chest X-ray.

#### Lung function tests



Tests such as spirometry measure how much air you can blow out of your lungs after taking a deep breath.

#### Pulse oximetry



This test uses light to estimate how much oxygen is in your blood.

#### Arterial blood gas test



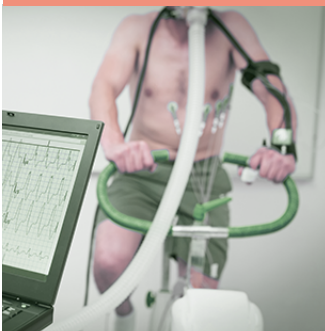
Oxygen and carbon dioxide levels are measured in blood taken from an artery.

#### Skin test for tuberculosis



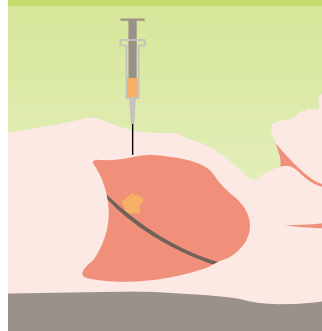
Done to rule out TB.

#### Exercise testing



Measures how well your lungs move oxygen and carbon dioxide in and out of your bloodstream when you're active.

#### Lung biopsy



Samples of lung tissue are examined under a microscope.

## 1.3 How does IPF affect your everyday life?

Already in the last month, Peter has recognised changes in his every day routines. He tired more easily and daily activities that once seemed trivial to him like walking or climbing stairs, showering, doing the housework, and other every-



day tasks became more and more a challenge for Peter because of his cough and shortness of breath.<sup>1</sup> More frequently he found himself gasping for air and stopping to take a break. The difficulty with physical activity also forced him to give up some hobbies he once loved to do. Hiking, fishing, traveling and other leisure activities suddenly looked out of reach. But after Peter learned more about IPF, he was able to cope and to keep up with some of his favourite activities without letting his illness get in the way.

Find out more about staying motivated in chapter **1.10 – Stay motivated - Keep a positive attitude.**

Since a few month ago, Peter feels fear of the reactions he gets in public places. He's often embarrassed that his coughing affects those around him and that other people distance themselves from him because of it. Since Peter does not want to avoid public crowds entirely, he tries to not take these reactions to heart. When Peter notices people nearby him shy away when he coughs, as if they were afraid of what he has, he tries to explain that they do not have to worry because what he has is not contagious.

Some people may be able to lead an almost normal life in spite of being diagnosed with IPF. Others may find everyday life more difficult. Coping with an IPF diagnosis can be challenging. Every person may have good and bad days. However, there are ways to lessen the burden of IPF on daily life and improve the quality of life.

## 1.4 Lifestyle Changes – Steps to help maintain your quality of life while living with IPF

IPF is progressive, unpredictable and may change your every day routines. However, there are a variety of things that you could do to stay prepared for some of the challenges accompanied with this disease.

When Peter visited his pulmonologist for a regular check-up, he was told to consider making some lifestyle changes that may help to reduce his symptoms and improve his quality of life.

Due to Peter still being a smoker, the first and most important thing his pulmonologist recommended to do was to **quit smoking** as soon as possible to prevent further damage to the lungs. He also told him to ask his family and friends to avoid smoking around him as second-hand smoke can be just as harmful. Peter was assigned to a support programme and his doctor recommended other beneficial methods to help him stop using tobacco.<sup>6-8</sup>

Since being diagnosed with IPF, Peter finds it easier to sit back and limit his physical activities in an attempt to avoid shortness of breath. To prevent developing a completely inactive lifestyle, his pulmonologist told him to **get active** and stay in shape but also learn to understand his limits and get enough rest. Regular, moderate exercise strengthens the muscles and helps keep the body working as efficiently as possible to manage shortness of breath.<sup>7,8</sup>

In addition to being active, it is also important to **relax and avoid stress**. To be physically and emotionally relaxed, Peter was recommended to follow a healthy sleep pattern and learn and practice relaxation techniques that may help manage stress and maintain strength.<sup>7,8</sup>

Like many illnesses, another important factor to reduce symptoms and improve quality of life may be to **eat a balanced, nutritious diet**. In order to maintain a healthy body weight, Peter was advised to eat more fruits, vegetables, whole grain products, as well as lean meats and low-fat dairy products and to avoid saturated fat, sodium (salt) and added sugar. Along with that, his physician told him to eat smaller, more frequent meals to prevent stomach fullness which may be accompanied by shortness of breath.<sup>7,8</sup>

For staying as healthy as possible, another step the physician recommended to Peter was to **get vaccinations and avoid catching seasonal colds/flu** or other secondary illnesses because these may cause IPF to worsen.

### Steps to help maintain your quality of life<sup>6-8</sup>



Quit smoking



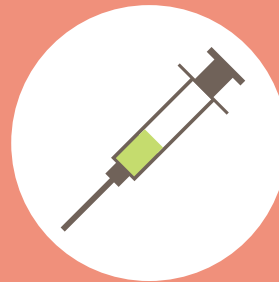
Get active



Relax and avoid stress



Eat a balanced, nutritious diet

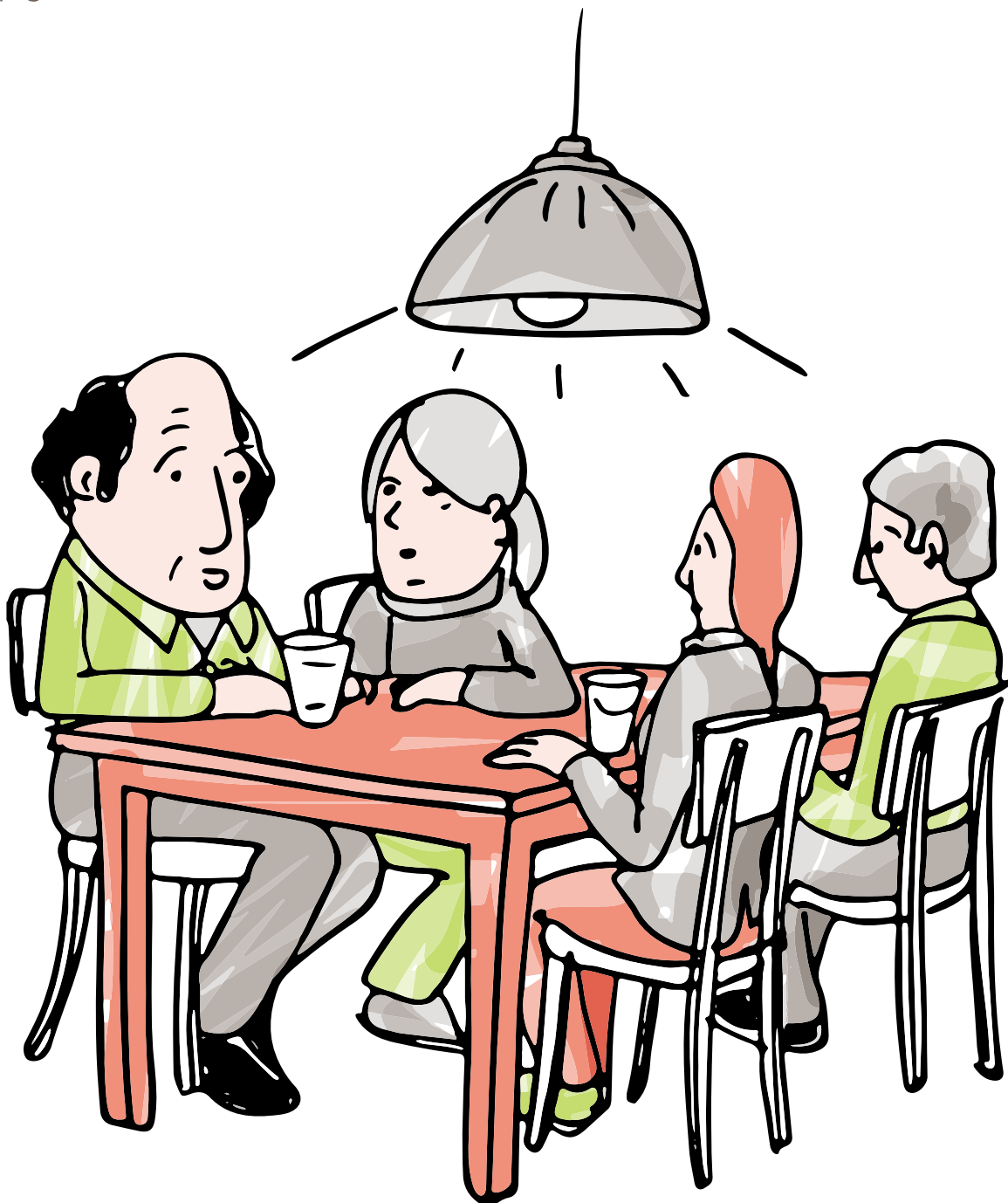


Get vaccinations

## 1.5 Talking about IPF with your family and friends

Being diagnosed with IPF can be devastating, not only for you, but also for your family and friends.<sup>9</sup> It might be difficult, but talking about the disease may help one feel less alone and ease the burden.

When Peter was diagnosed with IPF, he was worried about telling his family and friends and sharing his diagnosis with others. He wondered whom to tell and how to initiate a conversation about IPF. Finally, he started by creating a list of close people that he wanted to talk to in person and took a few notes to help get the conversation started.



Peter shared his diagnosis with his wife and children first, then he told other family members and his closest friends. Before talking to them, Peter thought about how much information he wanted to share with each person because all of these circumstances required a different level of detail.

To initiate a conversation about IPF, Peter tried to find a quiet and appropriate setting – breaking the news in a mild and gradual manner. He initially explained that the condition he is suffering from is called Idiopathic Pulmonary Fibrosis (IPF), a rare lung disease that usually gets worse over time. Peter told his loved ones that there is no cure for IPF yet, but that there are treatments to slow the disease **progression** in addition to the other things his physician recommended to do that may make him feel better.

The news caused many kinds of reactions, from supportive, to sobering and even speechless. When they learned that Peter was suffering from a disease with an unpredictable course, some of Peter's loved ones felt sad and uncomfortable or even wanted to avoid the topic altogether, while others were frightened about the possibility of losing him. After a while, Peter realised that talking about his condition or just sitting together and holding hands is often the best way to overcome these fears and difficulties.

As Peter talked to family and friends, a range of different questions about his disease came up, which he was not able to answer alone. For Peter, it was helpful to write these questions down and to discuss them with his pulmonologist.

Keeping those closest to him involved and informed about his illness, helped Peter to cope with his IPF diagnosis and feel less alone.

## 1.6 What should you expect after the diagnosis of IPF?

### 1.6.1 How is the monitoring of IPF performed?

In order to ensure that the disease is properly managed, Peter was scheduled for regular medical appointments every 3 to 6 months.

During these visits, his pulmonologist conducted a variety of tests to help assess the disease<sup>2</sup>:

- **Pulmonary function tests** reveal any change in lung function since the last examination.
- **Arterial blood gas tests** determine the levels of oxygen and carbon dioxide in blood taken from an artery.
- **Surveys and questionnaires** evaluate changes in symptoms, quality of life, psychological well-being and activity levels.

The attendance to this monitoring visits is a very important part of maintaining your health.



### 1.6.2 How does IPF progression manifest?

IPF is a condition which gets worse over time, but disease **progression** can be slow or rapid and varies from patient to patient.<sup>2,10,11</sup> In some patients, symptoms quickly become more severe, while in others they remain stable over a long period of time.<sup>2,10,11</sup> The individual course of the disease is hard to predict.<sup>2</sup>

A while after Peter visited his pulmonologist again, he gradually felt more breathless and fatigued and some activities were becoming more and more exhausting. Since his physician had told him to call the office right away if there were any changes in his symptoms, Peter didn't hesitate to make an appointment soon after he noticed a difference.

Peter's pulmonologist did a complete physical examination and explained that IPF steadily progresses over time. As the lung tissue becomes more and more scarred it is increasingly difficult for oxygen to get into the blood-stream through the lungs. This usually correlates to people feeling more and more breathless during everyday tasks and to the patient's organs not getting enough oxygen to function properly.<sup>3,4</sup>

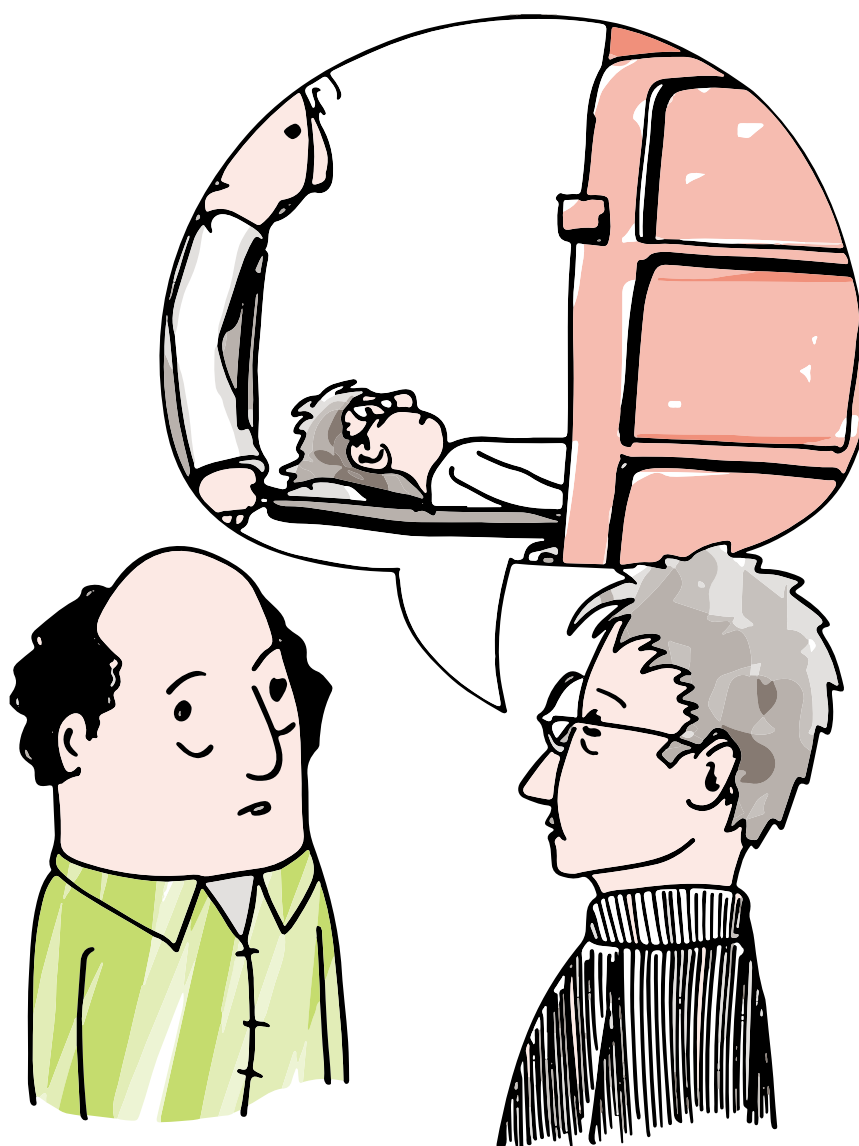
### 1.6.3 Acute exacerbations

The daily life of a person with IPF can be interrupted by events called **acute exacerbations**, where the person experiences a dramatic worsening in disease symptoms due to an unknown cause over a short period of time, usually a few weeks.<sup>2</sup>

#### What does acute exacerbation stand for?

<b>Acute</b>	Sudden onset
<b>Exacerbation</b>	Dramatic worsening in disease symptoms due to an unknown cause

James, Peter's friend from a patient support group, has already experienced one acute exacerbation and shared his impressions: When James experienced this acute exacerbation, he suffered from severe difficulty in breathing, a worsened cough accompanied with fever and flu-like body aches.<sup>12</sup> This sudden, severe worsening of the disease occurred over a period of one month without any known cause. When James went to see his physician, the doctor performed several tests and a **high-resolution computerised tomography (HRCT)** to diagnose an acute exacerbation.<sup>12</sup> He told him that these events are very severe and may even be life-threatening complications that, unfortunately, often cannot be predicted.<sup>13</sup> In order to monitor and manage his acute exacerbation, James was admitted to hospital, where he was given pharmaceutical therapy to relieve his symptoms.<sup>2</sup> After discharge, James was instructed to get in touch with the doctor's office right away if any acute deterioration of symptoms occur again anytime in the near future.



## 1.7 What other diseases are common in a person that has been diagnosed with IPF?

Persons with IPF frequently suffer from other conditions, or **comorbidities**, at the same time.

For quite some time, Peter wakes in the night with **heartburn** and a sour taste in his mouth. The next time Peter went to see his pulmonologist, he told him about these symptoms and sleep disturbances. In order to diagnose the cause of his symptoms, the pulmonologist conducted some additional tests.



The results of the tests suggested the presence of a condition called **gastro-oesophageal reflux disease (GERD)**. Peter was referred to a **gastroenterologist** for further evaluation and treatment. The specialist explained that GERD is caused by stomach acid coming up the throat and irritating the esophagus, which can cause heartburn and other symptoms. Aspiration of stomach acid into the lung can ultimately contribute to the damage of the lung tissue. In order to control his symptoms, Peter received medications that neutralise stomach acid and provide quick relief.

Along with GERD, IPF is associated with a variety of other respiratory and non-respiratory diseases.

## **Common other diseases that may occur along with IPF: <sup>2,6</sup>**

- Pulmonary hypertension (high blood pressure in the blood vessels that supply the lungs),
- Gastro-oesophageal reflux disease (GERD; chronic acid reflux),
- Chronic obstructive pulmonary disease (COPD; chronic inflammatory lung disease that causes obstructed airflow from the lungs and gets worse over time),
- Obstructive sleep apnoea (repeated pauses in breathing during sleep).

Due to the fact that comorbid conditions associated with IPF may negatively impact the quality of life, it is important to comment to the physician any other symptoms you may have, so the adequate diagnosis and treatment for these comorbidities could be provided, to improve your overall well-being.<sup>14</sup>

## 1.8 Emotional and physical impact of IPF

Changes in daily routines and physical health, a lack of independence as well as a poor prognosis may affect your feelings and leave you with strong emotions – both positive and negative.<sup>15</sup>



Before encountering IPF symptoms, Peter generally felt satisfied with his life. Initially, he did not fully understand the severity of his disease and the burden it would cause on his life, both physically and emotionally. As Peter learned more about his diagnosis, he experienced many different feelings. Aside from wondering “Why me?”, he felt sad, angry and afraid about his upcoming life. He wished doctors had known what was causing his symptoms earlier, and he was feeling powerless as he assumed there was nearly nothing he could do.

Another challenge Peter experienced with IPF was his persistent IPF-related cough. Many times, he felt isolated and embarrassed due to not being able to control it, making it difficult to interact and communicate with other people, thereby limiting his social life, leisure activities and hobbies.

Gradually, Peter became more and more preoccupied with the fear that he was becoming a burden on his family and friends. Along with that, the poor prognosis of IPF and the possibility of death within three to five years after diagnosis frightened him. At times, he was depressed, which was very unlike him. After a while he came to terms and understood that the best he could do was to accept his disease and try to enjoy his life – looking forward to getting together with his friends or playing with his grandchildren.

Emotional health often suffers with serious diseases such as IPF. It's natural to feel both angry and curious and experience a significant emotional burden when dealing with IPF. For many people, the road to their diagnosis is long and frustrating – seeing a number of different doctors and experiencing considerable delays in receiving an accurate diagnosis. Following the diagnosis of IPF, many persons miss the freedom they enjoyed before they knew about their disease. Learning that they will have to cope with pain, breathlessness, and physical distress can result in stress, anxiety, and a sense of being powerless against it.

The first step to free oneself of the intrusive and burdensome effects of IPF, is to acknowledge feelings – both positive and negative – as they are a normal part of the process of accepting the diagnosis and further to cope with the diagnosis one step at a time. In the process of coping with the diagnosis, people go through several stages of mental transition until they reach the point of acceptance.

If emotional stress becomes overwhelming, you should not be ashamed to reach out for professional help and support to share feelings, needs, and concerns. There are definitely helpful resources available.

## 1.9 Where to get support?

After trying to cope with his physically and emotionally exhausting IPF diagnosis on his own, Peter had to admit he couldn't handle his disease and the accompanying rollercoaster of emotions all alone anymore.

Asking for help is not always easy, but Peter recognised that once his disease progresses, he would need his loved ones more and more. Heeding the advice "A problem shared is a problem solved," he reached out for support from his family and friends. His family members and close friends tried to help Peter through this intense time by offering practical and emotional support. This helped Peter very much, however he also had the desire to share his thoughts and feelings with other people dealing with the diagnosis of IPF who were also experiencing all he was going through. In order to find people in the same situation, Peter talked to his pulmonologist who then recommended him to join an IPF support group associated with the hospital.



Participating in this specific IPF support group helped Peter better manage the challenges of living with IPF. Getting together, creating a sense of community, sharing experiences and feeling understood all decreased his sense of isolation and strengthened him during his IPF journey.

Due to the reason that IPF is a **rare disease**, there is not always a specific support group in every area. Alongside face-to-face meetings with other people with the same diagnosis, some IPF organisations offer support groups that are available by telephone or as an online group. Another possibility for concerned patients is to consider acting as a volunteer and form a patient support group on their own to provide relief to other people in the same situation in their area.

Along with support groups, there are many other chances to reach out for support. Receiving professional counselling and psychosocial support provided by social workers, professionally trained therapists and psychologists, is an excellent way to get the support people with IPF may need. Religious or spiritual counselling may also be an option for people with IPF seeking comfort and support.

## 1.10 Stay motivated – Keep a positive attitude



Living with a serious, life-threatening condition like IPF is not easy. However, there are a variety of steps that you can take to stay motivated, keep a positive attitude and maximise enjoyment of life.

Since Peter joined the IPF support group he learned from them that it is important to become actively involved in his treatment and care for his health by regularly talking to his doctors and nurses. Peter now tries to be prepared for his medical visits by keeping a journal of questions to be answered by his pulmonologist during the visit, taking notes at his appointments or bringing a friend or family member with him. Peter focuses his energy on getting the best treatment for his condition, thereby feeling he has regained some control over what is happening to him.

In order to support this feeling of control, to foster hopefulness, and to stay motivated, Peter tries to be mindful of the present moment – concentrating on today's known facts rather than the uncertainties of the future or the **progression** of his disease.

Even though Peter has received a life-changing diagnosis, he tries to continue doing things he enjoys and to keep up with some of his favourite activities without letting his illness get in the way. If he feels up to going for a morning walk or doing some gardening, he does as much as he can. Even if he wanted to visit his family and friends on the other side of the country or wanted to go on a vacation, IPF wouldn't automatically cut his holiday hopes short. Traveling with IPF may mean that Peter will encounter some extra challenges, but with a good organisation and planning, there is no reason to stay at home.

**Read more about "Travelling with IPF" on [www.LifewithIPF.com](http://www.LifewithIPF.com)**

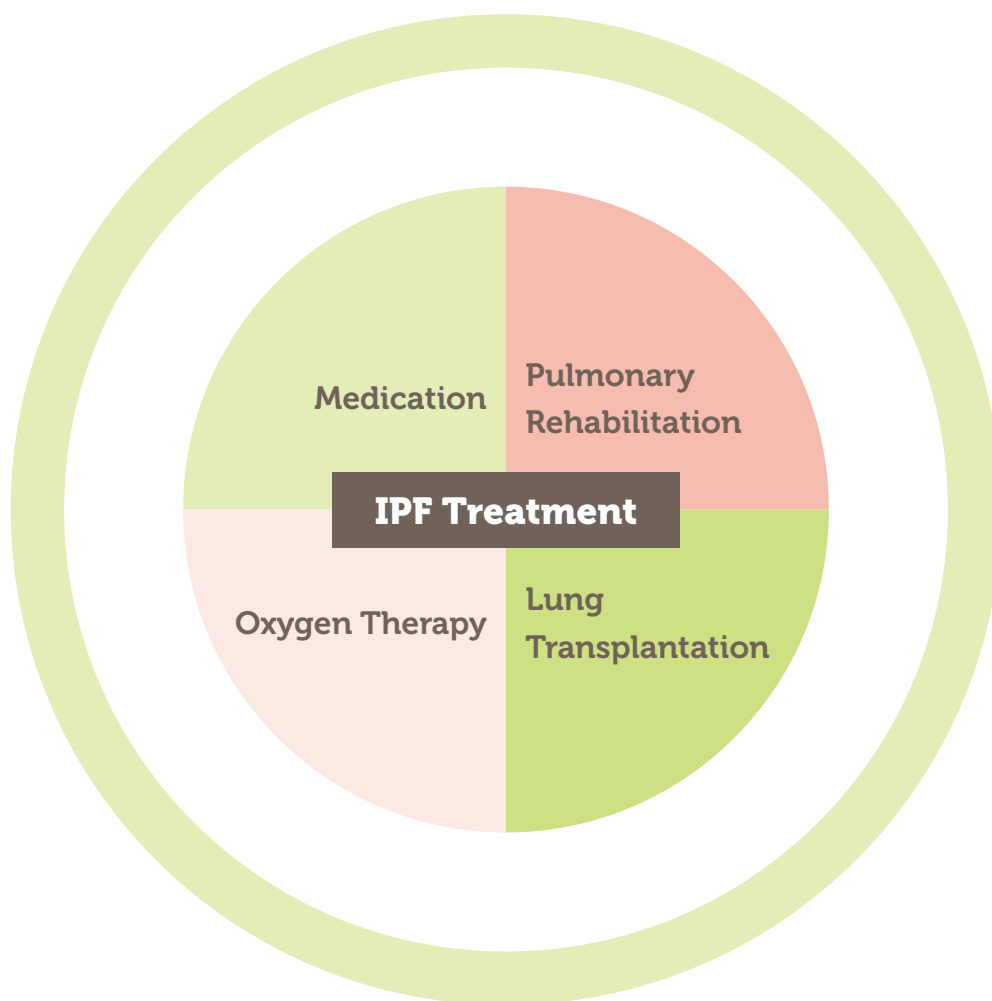
Generally, a positive attitude can help patients, as well as their family and friends, cope with the disease and do better over time. Positive thinking can be helpful and healing, and may get you through the most difficult days.

## 2.0 How is IPF managed?

When Peter was newly diagnosed with IPF, he tried to find out as much as possible about the disease and his therapy options from his doctor and treatment team to better understand this condition.

His pulmonologist told him, that, whilst there is no pharmacological cure for IPF yet, various therapies can help to alleviate symptoms and slow down disease **progression**. Therapeutic strategies that may stabilise the disease include various medications, as well as a variety of non-pharmacological options.<sup>6</sup>

Treatment decisions for IPF are highly personal for each person and may be affected by additional factors such as the presence of other diseases (**comorbidities**), the possible adverse events of the treatments and the risk of **acute exacerbations** of IPF.<sup>2</sup> With the help of his doctor, Peter decided which IPF therapy could work best for him, his individual situation and symptoms.



## 2.1 Non-pharmacological therapies

Alongside medicinal treatment, non-pharmacological therapies may stabilise the disease and improve symptoms.

### 2.1.1 Oxygen therapy

The first non-pharmacological intervention Peter's physician informed him about was **supplemental oxygen**, which can help reduce shortness of breath and improve patients ability to perform everyday tasks.<sup>2,16</sup> Peter was told therapy

with supplemental oxygen may be prescribed if the level of oxygen in the blood gets too low, thereby potentially causing symptoms of **fatigue**, irritability, and lack of concentration.

At first, supplemental oxygen may only be needed during exercise and sleep, but as the disease progresses and the lungs increasingly lose their ability to oxygenate the blood, it may be required all the time to keep oxygen levels in the blood at a healthy level.

To check whether Peter needs oxygen and how often he would need to use it, the pulmonologist conducted a series of tests to determine Peter's oxygen saturation and arterial blood gases. The tests confirmed he is a candidate for supplemental **oxygen therapy** and should use it during exertion. Accordingly, Peter was prescribed supplemental oxygen given through nasal prongs to be worn during moderate exertion.



### 2.1.2 Pulmonary rehabilitation

Since it is a standard intervention for people with chronic lung disease, **pulmonary rehabilitation** was another non-pharmacological therapy the pulmonologist highly recommended to Peter in order to improve his well-being.<sup>2,16,17</sup>

Pulmonary rehabilitation programmes offer a variety of services, including:<sup>2,17</sup>

- Physical conditioning
- Exercise training and breathing exercises
- Anxiety, stress, and depression management
- Advice and support to improve nutrition
- Education on the disease

The pulmonary rehabilitation programmes are generally run by a multi-disciplinary team consisting of doctors, nurses, physiotherapists, social workers, dieticians, and other healthcare specialists and can be carried out in the home, community or hospital settings. The goal of pulmonary rehabilitation is to provide skills, tools and tricks that help you manage the disease and take control of your individual symptoms.



In order to get expert guidance, Peter was advised to take part in a pulmonary rehabilitation programme over a period of a few months.

Along with a personalised exercise programme, Peter learned breathing techniques and was provided with nutritional counselling and emotional support.

The pulmonary rehabilitation programme helped him to feel better, improve his energy, strength and endurance, and have a better outlook on his disease.

### 2.1.3 Lung transplantation

Another non-drug option Peter was informed about by his physician was **lung transplantation** as a therapy to treat IPF.

In a lung transplantation, one or both damaged lungs are replaced with the lungs from a donor. This choice of treatment may be required if the IPF is quickly worsening or very severe.<sup>17</sup> A lung transplant is a major intervention and comes with its own risks including serious complications as infections or **rejection** of the donor lungs. On the other hand, a transplant could improve the patient's quality of life and help them live longer.<sup>1,2,17</sup> Not everyone with IPF is a lung-transplant candidate - some patients may have **comorbidities** that make a lung transplant not possible. Also, many programmes have an upper age limit of 60 or 65 years.<sup>1,2,17</sup> Another limitation is the very small number of donor organs available for transplantation, which means that in reality, less than 1% of patients with IPF will ever be able to get a transplant. For that reason, lung transplantation is not a suitable treatment for everybody with IPF. However, it can be a very successful treatment for a small proportion of people.

Since lung transplantation is a potential option for all IPF patients, soon after he was diagnosed, Peter underwent an evaluation for a lung transplantation. Although the first evaluation showed that he was not currently a candidate for a lung transplant, Peter will be reassessed regularly to see if he may be considered as a candidate for a lung transplant in a more advanced stage of the disease.<sup>2,18</sup>

Usually, the selection of candidates for lung transplantation is dependent on the urgency of their condition as well as their predicted individual outcomes and benefits after the transplant.

## 2.2 Medications

Whilst there is no cure for IPF, medications may help to slow disease **progression**.<sup>2,19</sup> In **clinical trials**, it was shown that some medications are able to slow the decline of lung function in patients with IPF.

When Peter talked to his physician about which treatment is the best for his individual situation, the physician informed him, that, fortunately, in their country two medications are available for the treatment of IPF, while in other countries, only one medication or no medications are available yet.

The medications the pulmonologist informed Peter about are:

- OFEV® (active substance: nintedanib)
- Esbriet® (active substance: pirfenidone)

Both drugs are **anti-fibrotic** drugs which means that they can help reduce new scarring and stiffening of the lungs in patients with IPF.

Peter had a conversation with his physician regarding the characteristics of each drug, to select which treatment may suit him best.



### 2.3 Other treatment options

When talking about treatment options of IPF, the physician informed Peter that, along with medicinal and non-pharmacological treatments, **palliative care** is another aspect to consider.

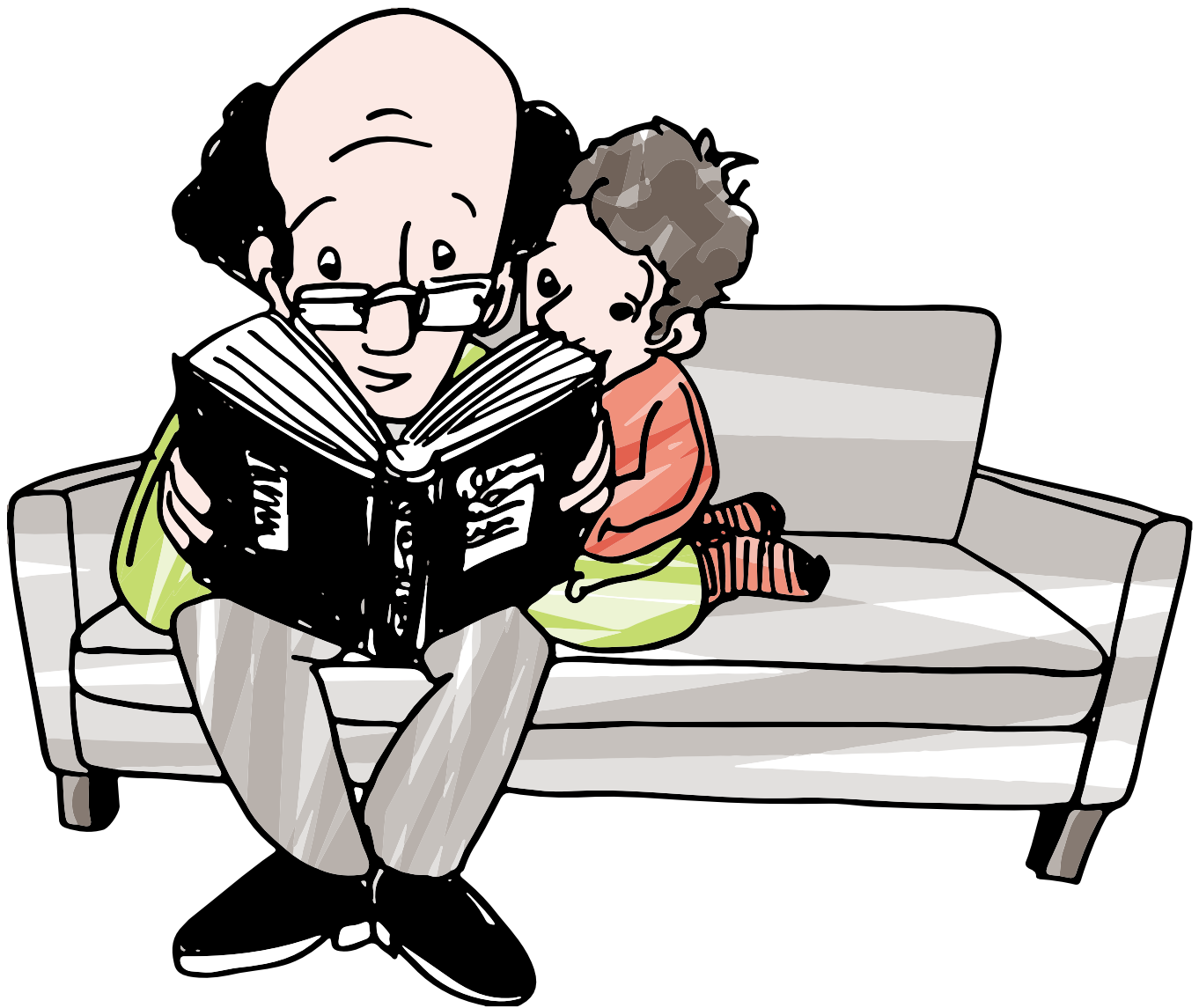
Palliative or symptomatic care is a central part of the treatment of IPF. It is designed to relieve physical and emotional suffering and improve the patient's quality of life, through social, psychological and spiritual means.<sup>2</sup>

As Peter's pulmonologist reported, traditionally, people think about palliative care as only being applied late in the disease and toward the end of somebody's life. When in reality, a referral to palliative care can be very effective in treating the symptoms rather than treating the underlying cause even in the early stages of the disease.

Looking at IPF, palliative care can involve treatments to help reduce the symptoms of breathlessness and cough. It can be used alongside other treatments to strike a balance between trying to prevent progressive lung scarring and to limit symptoms.

## 2.4 Coming to terms with IPF

After Peter was attuned to the IPF therapy that works best for him, he was doing much better than he originally expected. He takes his medications as indicated and attends the IPF patient support group to share his experiences and feelings with people in the same situation. Peter tries to continue doing things he enjoys without letting his illness get in the way, looking forward for simple social events like getting together with friends or playing with his grandchildren. Peter understood that the best thing he could do was to accept his disease, be mindful of the present moment and try to enjoy life as much as possible.



# 3.0 Resources & Tips

## 3.1 Helpful web resources

If you are searching for IPF facts online, you might get a huge and overwhelming number of results. To get a detailed overview of all the hot topics related to IPF, please visit [www.LifewithIPF.com](http://www.LifewithIPF.com)

LifewithIPF.com is a website created especially for patients and caregivers who are affected by IPF. It offers disease information, guidance and support, answers to frequently asked questions, patient stories and a variety of downloadable resources.



**Patient organisations** serve as a patient advocate and help by providing a forum to connect with others who face similar challenges to obtain information about living with the disease, and to receive support in various ways.

The following search terms could be useful in order to find an appropriate IPF patient organisation:

- "IPF Patient Organisation",
- "Idiopathic Pulmonary Fibrosis Patients",
- "IPF Patients Foundation",
- "IPF support group",
- "IPF help"

## 3.2 Checklists and tips for download

### 3.2.1 Important contacts for your care

You may meet many different medical professionals during the treatment of IPF. Having an up-to-date and complete list of key people, caregivers, physicians, pharmacies, hospitals and support groups to contact in an emergency makes it faster and easier to get appropriate help.

- Fill in the information below and keep it somewhere handy where you can find it quickly even in the confusion of a crisis.
- It could be helpful to make a few copies – you might want one for the refrigerator, one to carry with you when you are out of the house, one to keep in the car and one for your desk at work.
- You could also share this list with anyone who might be called upon to act in your absence.
- It is important to regularly review and revise your list to make sure the names and numbers are up to date.

**IPF doctor (Pulmonologist)**

Name/number:

**Pharmacist**

Name/number:

**Hospital**

Name/number:

**Support group**

Name/number:

**Other care providers**

Name/number:

Name/number:

Name/number:

Name/number:

### 3.2.2 Notes and questions for your next doctor's appointment

IPF is a challenging and unpredictable disease. Therefore, it is important to be well informed about the disease and the options associated with it.

Always keep in mind that your physicians, nurses and healthcare providers are partners in your treatment. Make sure you discuss any questions and concerns you may have about IPF to get a firm understanding of the disease and how you should care for yourself.

To be prepared for your appointments, it may be helpful to consider and write down all the questions you have as they come to mind, and share them with your doctor and healthcare team at your next visit. Below are some questions you may find helpful to ask your physician.

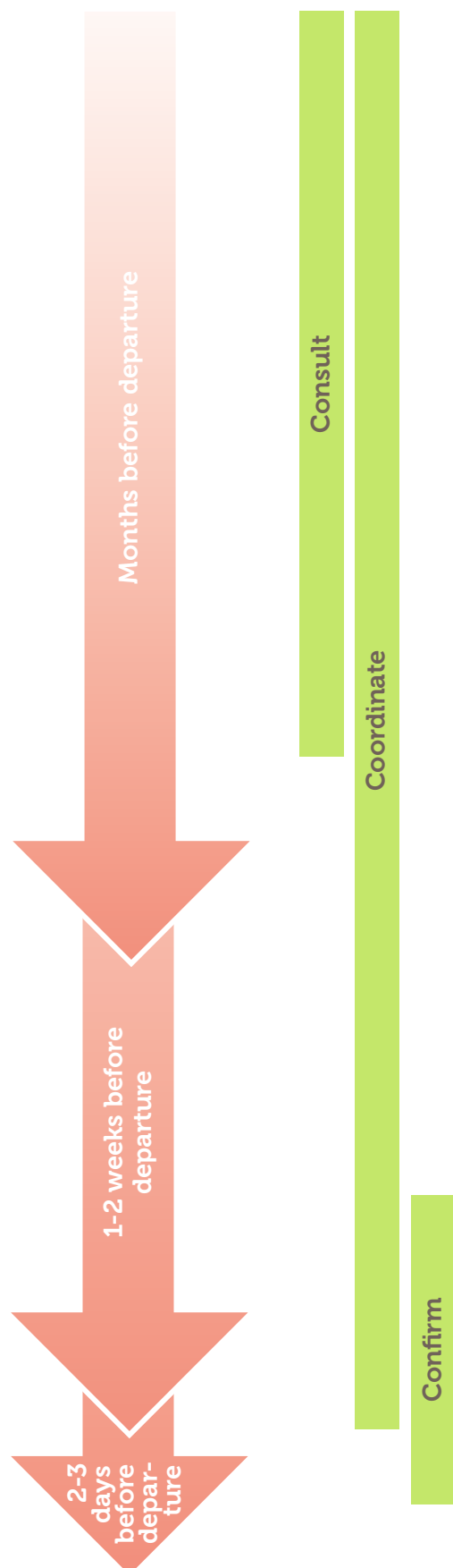
#### Possible questions to ask your pulmonologist:

- How can I help myself?
- What are my personal treatment options?
- How will my symptoms be monitored and how frequently will I be tested?
- What can I do to better cope with the diagnosis of IPF?
- How can I find local support groups?
- Can I benefit from supplemental oxygen?
- Would a lung transplantation be an option for me?

Feel free to use the space below to write down your own questions or take notes on all of the concerns relating to your IPF care.

#### Notes and questions for your doctor:

## 3.2.3 Checklist for plane travel

**Before planning your trip...**

- ☐ Talk to your doctor about your travel plans and any concerns you may have
- ☐ If you are not currently on oxygen therapy, your doctor will need to determine whether you are candidate for in-flight oxygen

**Before booking your airline tickets...**

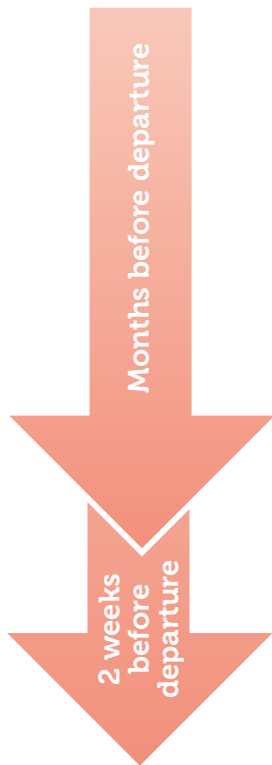
If you need in-flight oxygen, check the airline's oxygen policy and "fit-to-fly" requirements

- ☐ Does the airline supply in-flight oxygen, or are you required/allowed to bring your own portable oxygen supply?
- ☐ What type of oxygen delivery system does the airline offer on board?
- ☐ Available flow rate settings?
- ☐ Mask or nasal cannula?
- ☐ What are the costs?
- ☐ Information regarding battery supply/in-flight electrical supply
- ☐ What are the policies regarding use of oxygen during take-off and landing?
- ☐ Complete and submit the airline's MEDIF form.<sup>3</sup>
  - One portion will be completed by you, the other must be completed by your doctor
  - Only after the airline reviews the completed form and determines you are eligible to fly, the ticket reservation will be finalised
- ☐ Contact the airport(s) you will be using to arrange for assistance to/from the gate
- ☐ Check with your insurance provider about your coverage while travelling – additional traveller's insurance may be beneficial or necessary

**Before enjoying your get-away...**

- ☐ Make sure you have an adequate supply of all prescription medicines as well as new prescriptions for refills. Please note that you need to check the local situation since not every medication is available in every country.
- ☐ Contact the airline to confirm all necessary precautions and special arrangements will be realised on your flight.
- ☐ Contact the airport to confirm the status of your planned assistance.

### 3.2.4 Checklist for car, bus or train travel



#### Before planning your trip...

- ☐ Talk to your doctor about your travel plans and any concerns you may have regarding
  - Climate
  - Air quality
  - Terrain
  - Altitudes
  - Mode of transportation

#### Before booking your journey...

- ☐ Check with your insurance provider about your coverage while travelling – additional traveller's insurance may be beneficial or necessary

#### Before enjoying your get-away...

- ☐ Make sure you have an adequate supply of all prescription medicines as well as new prescriptions for refills. Please note, that not all medication is available in all countries around the world. Check out before!

# 4.0 Glossary

**Acute exacerbations:** Episodes characterised by sudden, severe worsening of symptoms or increases in disease severity that occur without any known cause and often lead to hospitalisation<sup>20</sup>

**Alveolus (pl. alveoli):** Very small air sacs found in the lungs where the exchange of oxygen and carbon dioxide takes place<sup>21</sup>

**Anti-fibrotic:** Acting to block or reduce tissue scarring<sup>22</sup>

**Arterial Blood Gas Test:** Provides information on the levels of oxygen and carbon dioxide found in the blood from an artery to see how well the lungs are working<sup>23</sup>

**Asthma:** A chronic respiratory disease characterised by symptoms such as wheezing, shortness of breath, chest tightness and cough that vary over time in their occurrence, frequency and intensity.<sup>24</sup>

**Cardiovascular:** Refers to the heart, and blood vessels<sup>25</sup>

**Chest X-ray:** A non-invasive medical test that creates images of the organs and bones inside the chest<sup>23</sup>

**Clinical trials:** Research studies that test how well new therapies work in humans<sup>21</sup>

**Comorbidity:** A disease or condition that occurs simultaneously with another disease or condition<sup>22</sup>

**Chronic obstructive pulmonary disease (COPD):** A common preventable and treatable disease, characterized by persistent airflow limitation that is usually progressive and associated with an enhanced chronic inflammatory response in the airways and the lung to noxious particles or gases.<sup>26</sup>

**Coronary artery disease:** A disease in which a waxy substance (plaque) builds up inside the coronary arteries, which supply oxygen-rich blood to the heart muscle.<sup>23</sup>

**Differential diagnosis:** The distinguishing of a particular disease or condition from potential alternative diagnoses which presents itself with similar signs and symptoms<sup>21</sup>

**Fatigue:** Extreme weariness resulting from exertion or illness<sup>21</sup>

**Fibrosis:** The repair and replacement of inflamed tissues or organs by fibrous connective tissue; eventually results in replacement of normal tissue with scar tissue<sup>21</sup>

**Gastroenterologist:** A physician specialising in the management of diseases of the digestive system

**Gastro-oesophageal reflux disease (GERD):** A chronic digestive disease, which occurs when stomach acid or content flows back into the food pipe, irritating the lining of the oesophagus<sup>27</sup>

**Heartburn:** A burning sensation in the chest, which can spread to the throat, along with a sour taste in the mouth<sup>27</sup>

## 4.0 Glossary

**High-Resolution Computed Tomography Scan (HRCT):** A type of X-ray that generates multiple, detailed images of areas inside the body<sup>23</sup>

**Idiopathic:** Of unknown cause<sup>21</sup>

**Idiopathic Pulmonary Fibrosis (IPF):** Progressive scarring or thickening of the lungs without a known cause<sup>23</sup>

**Lung biopsy:** The surgical removal of cells or tissue samples from the lung for examination by a pathologist<sup>23</sup>

**Lung scarring:** The lung tissue becomes thickened and stiff

**Lung transplantation:** Surgical replacement of a patient's diseased lungs with lungs from a donor<sup>23</sup>

**Obstructive Sleep Apnoea:** A potentially serious sleep disorder characterised by breathing which repeatedly stops and starts during sleep.<sup>27</sup>

**Oxygen therapy:** Administration of oxygen as a medical intervention<sup>23</sup>

**Palliative care:** Non-curative therapy to reduce symptoms and provide comfort to patients suffering from a serious or life-threatening disease<sup>21</sup>

**Progression:** The worsening of a disease/condition over time

**Pulmonary:** Refers to the lungs<sup>21</sup>

**Pulmonary function test:** A group of tests used to check how well the lungs take in and release air and how well they supply oxygen to the rest of the body<sup>21</sup>

**Pulmonary hypertension:** A type of high blood pressure that affects the arteries in the lungs and the right side of the heart<sup>27</sup>

**Pulmonary rehabilitation:** A broad therapeutic concept with the aim of improving the quality of life for the patient<sup>23</sup>

**Pulmonologist:** A physician specialising in the lungs

**Pulse oximetry:** A test used to monitor a patient's blood oxygen saturation<sup>21</sup>

**Rare disease:** A disease that affects only a small percentage of the population<sup>28</sup>

**Rejection:** An immune reaction of a transplant recipient's organism to a transplanted organ or tissue<sup>21</sup>

**Risk factor:** A variable associated with an increased risk of disease or infection<sup>21</sup>

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