Living with Idiopathic Pulmonary Fibrosis

An eBook

www.LifewithPulmonaryFibrosis.com

Boehringer Ingelheim

EU-IPFF
EUROPEAN IDIOPATHIC PULMONARY FIBROSIS & RELATED DISORDERS FEDERATION

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 complements the information role of HCPs and Support Groups.

- Liam Galvin, Secretary, EU-IPFF

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About Idiopathic Pulmonary Fibrosis (IPF)

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1.1 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 quite rare. 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 factors that may increase the risk of developing IPF.

Idiopathic pulmonary fibrosis IPF

A disease where there is progressive scarring or thickening of the lungs without a known cause

Signs and **symptoms**[1,2]

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

Shortness of breath

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

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

Gradual, unintended weight loss

Tiredness and generally feeling unwell

A short dry, (hacking) cough that doesn't get better or improve over time

Rapid, shallow breathing

But what **exactly is IPF?**

IPF stands for Idiopathic Pulmonary Fibrosis. In people with IPF, the tissue inside and between the tiny air sacs (known as **alveoli**) and blood vessels of the lungs become thickened, stiff and scarred. **Fibrosis** is the medical term for this scarring.[2] 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.[2,3] This results in a shortness of breath and the organs not getting enough oxygen to function normally.[2,3]

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.

However, in most cases of **lung scarring**, an exact cause is never established and that is then what we call Idiopathic Pulmonary Fibrosis (IPF).[1]

Alveolus (plural alveoli)

Tiny air sacs in the lungs where the exchange of oxygen and carbon dioxide to and from the blood takes place

Fibrosis

Where inflamed tissue is replaced with scar tissue, making it become thickened and stiffer

Lung scarring

The tissue becomes thickened and stiff

What does 'IPF' stand for?

I	**Idiopathic**	Of unknown cause
P	**Pulmonary**	Refers to the lungs
F	**Fibrosis**	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.[2,4,5]

There is no single test that can determine an IPF diagnosis, but examination of one's medical history, a physical examination and several test results all help the pulmonologist to come to right diagnosis.

Chronic obstructive pulmonary disease (COPD)

A chronic, ongoing disease where airflow is limited to the lungs. It is usually progressive and linked to inflammation in the airways

Pulmonologist

A physician specialised in the lungs

In order to make the correct diagnosis for Peter, doctors first had to rule out all other conditions. They looked for different symptoms, asked about his medical history and performed a variety of tests so that they could reach a final diagnosis.[2,6] Because there were a range of tests to be carried out, Peter saw many different types of doctors and specialists before being diagnosed with IPF.

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 history, his family medical history, drug use, etc.[2,6]

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.[7,8] He also checked Peter's hands for widening and rounding of the fingertips – a common sign of IPF and other respiratory diseases.[5,7,8]

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 identified specific patterns in the lung tissue which indicated the presence of IPF.

Pulmonary function tests

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

Chest X-ray

A non-invasive medical test that creates images of the organs and bones inside the chest

High-Resolution Computerised Tomography (HRCT)

A type of X-ray that generates multiple, detailed images of areas inside the body

Diagnostic tests for IPF[6,9]

Chest X-ray

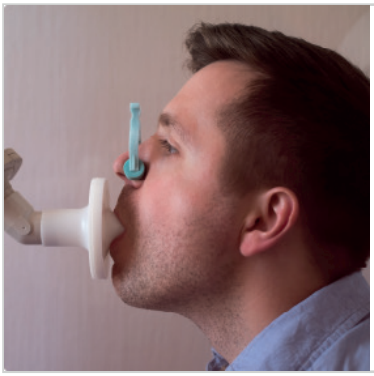
This creates a picture of your lungs, revealing shadows that suggest scarring.

Arterial blood gas test

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

Exercise testing

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



Lung function test

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

HRCT

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

Bronchoalveolar lavage

A sample of fluid is taken from the lung to be examined.

Skin test for tuberculosis

A small injection is given to check for a TB infection.

Lung biopsy

Samples of lung tissue are examined under a microscope.

Pulse oxymetry

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

1.3 How does IPF affect your **everyday life?**

Within a month, Peter recognised changes in his everyday routines. He tired more easily and daily activities that once seemed trivial to him, like walking, climbing stairs and other everyday tasks, became more and more of a challenge because of his cough and shortness of breath.[1,3] 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.**

Peter also became fearful of the reactions he received in public places. He was often embarrassed that his coughing affects those around him and that other people distance themselves from him because of it. Since Peter did not want to avoid public crowds entirely, he tried not to take these reactions to heart. When Peter noticed people nearby him shy away when he coughed, he explained that they do not have to worry because what he has is not contagious.

Some people may be able to lead a fairly 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 and improve the quality of daily life.

1.4 Lifestyle changes - Steps to help maintain your **quality of life when living with IPF**

IPF is progressive, unpredictable and may change your everyday routines. However, there are a variety of things that you could do to stay prepared for some of the challenges that accompany 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.

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.[1,9,10]

Since being diagnosed with IPF, Peter found 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.[2,9]

In addition to being active, it is also important to **relax and avoid stress**. To be physically and emotionally relaxed, Peter was told to follow a healthy sleep pattern and learn and practice relaxation techniques that may help manage stress and maintain strength.[2,11]

Like many illnesses, another important factor to reduce symptoms and improve quality of life can 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. His physician also suggested that he eat smaller, more frequent meals to prevent stomach fullness which may be accompanied by shortness of breath.[2,12]

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

How to maintain your **quality of life**[2,9,11]

Get active

Relax and avoid stress

Quit smoking

Eat a balanced, nutritious diet

Get vaccinations

1.5 Talking about IPF with your **family and friends**

Being diagnosed with IPF can be difficult, not only for you, but also for your family and friends.[13,14]

At first, Peter found it difficult to talk with his family and friends about IPF. Most people had never heard of IPF and were not aware of what he was going through.

However, being open about his condition helped Peter deal with his emotions better. It also helped his family to talk about their concerns and understand what he was going through.

As Peter learnt more about his disease he found it easier to explain it to others who could then support and help him cope with some of the negative feelings he felt. He found that changing the explanation depending on who he was talking to helped. A shorter explanation was more appropriate for younger children than for adult friends and family.

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:[2,9]

- **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.

Attending these monitoring visits is a very important part of maintaining your health.

Pulmonary function tests

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

Arterial blood gas test

A test that shows how well the lungs are working by measuring how much oxygen and carbon dioxide is in the blood

1.6.2 How does IPF progression manifest?

IPF is a progressive disease, which means that it gets worse over time. However, there is no way of predicting how fast it will progress. The rate at which IPF progresses can also change over time. Some patients may notice little change in their condition over months or years, while other patients experience a faster deterioration.[2] It is not known why some people's disease progresses faster than others.

1.6.3 Acute exacerbations

Acute exacerbations can occur at any time during the course of IPF. They can lead to faster deterioration of the disease and more time spent in hospital.[2,15]

Acute exacerbations are life-threatening events that you must take seriously.[15] That is why it is important that you work with your doctor and wider treatment team to do everything you can to look after your lungs.

What does 'acute exacerbation' stand for?

Acute	Sudden onset
Exacerbation	Considerable or rapid worsening

1.7 What other diseases are **common in a person who has been diagnosed with IPF?**

People with IPF frequently suffer from other conditions at the same time.

For quite some time, Peter woke 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 (GORD/GERD),** Peter was referred to a **gastroenterologist** for further evaluation and treatment. The specialist explained that GORD is caused by stomach acid coming up the throat and irritating the oesophagus, which can cause heartburn and other symptoms.

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

Heartburn

A burning sensation in the chest, which can spread to the throat, along with a sour taste in the mouth

Gastro-oesophageal reflux disease (GORD/GERD)

A digestive disease where stomach acid moves up out of the stomach and irritates the lining of the food pipe (oesophagus)

Gastroenterologist

A physician specialised in the management of diseases of the digestive system

Common diseases that may occur along with IPF:[16]

- **Chronic obstructive pulmonary disease (COPD):** A lung disease that causes the flow of air into and out of the lungs to be restricted. This makes it harder to breathe
- **Gastroesophageal reflux disease (GORD/GERD):** A stomach condition that causes stomach acid to come to the throat, leading to heartburn
- **Obstructive sleep apnoea:** A condition where breathing stops and starts during sleep
- **Pulmonary hypertension:** High blood pressure in the blood vessels to the lungs
- Heart conditions, such as **coronary heart disease**
- Other conditions, such as **diabetes**, **underactive thyroid** and **hiatal hernia**

- It is important to remember that every person with IPF is different and may experience other conditions too. Always be aware of any rapid changes in your condition or symptoms and discuss them with your doctor and treatment team.
- Managing other diseases can be just as important as managing IPF itself. Your doctor and treatment team will develop a treatment plan which considers all the different conditions you may have.
- Medications can also sometimes interact with one another. For this reason, if you have more than one condition, some treatments may not be suitable to treat IPF or any other diseases. Your treatment team will work with you to develop the most appropriate treatment plan.

1.8 **Emotional and physical** impact of IPF

As well as physical symptoms, IPF can affect your mental wellbeing. For example, you may start to experience a low mood if you lose personal independence, experience relationship problems or worry generally about a loss of control.[13,14]

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 future. 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.

Gradually, Peter became more and more preoccupied with the fear that he was becoming a burden on his family and friends. He also became frightened by the long-term prognosis of IPF following diagnosis. At times, he felt very down, which was unlike him. After a while he came to terms and understood that the best he could do was to accept his condition and try to enjoy his life – looking forward to getting together with his friends or playing with his grandchildren.

There is no right or wrong way to respond to the news that you have a terminal illness. The news can be shocking and difficult to accept. Some people feel numb or go silent, while others feel angry or scared for what the future holds. Everybody is different and reacts in their own way.

Give yourself the time to take in what is happening. Some people would rather be on their own, but others would rather spend time with family and friends. If you don't feel like talking straight away, you don't have to.

As hard as it can be, try not to push your emotions aside completely. At some point, it's better to express how you feel if you can manage it, even if it's uncomfortable and hard to cope with.

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 alone.

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 halved," 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 a patient support group offers an opportunity to get together with others who are facing similar experiences. It helped Peter to manage the challenges of living with his condition.[2,9,11]

As 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 online.

Rare disease

A disease that affects only a small percentage of the population

1.10 Stay motivated - Keep a **positive attitude**

Whether you have difficulties with your mental wellbeing at the moment or not, there may be times that are more difficult than others.

Below are some tips to help you maintain your mental wellbeing so that you can deal with everyday life and difficult situations, without feeling overwhelmed.

Tips to help you maintain a healthy mindset[17]

1. Talk about how you feel: talking something through and knowing that there is someone to listen and understand can make you feel much better. Friends, family or a mental health professional may be able to offer practical help or advice and give you another perspective on your disease.

2. Build healthy relationships with people: spending time with supportive family and friends can help you build a better self-image and feel more confident as you approach any challenging situations.

3. Sleep: if you have difficulty sleeping, this can have a serious impact on your mental wellbeing. Feelings of low mood and other negative emotions can be exaggerated, and you might find you have a short temper or lose confidence.

4. Diet: eating healthily can help both your physical and mental health. Eating a balanced diet at regular meal-times will help you to feel healthier and support a positive mindset.

Tips to help you maintain a healthy mindset[17]

5. Physical activity: staying active can help reduce any feelings of low mood and anxiety and boost your self-confidence. Light activities like gardening or gentle walking are a good place to start if you're not used to participating in a lot of physical activity.

6. Do something you enjoy: find time to do things you enjoy, whether it's cooking, seeing your friends or listening to music. Taking up a new hobby can also help boost your confidence and occupy your mind.

7. Relax: it's important to make time to relax, even if you don't feel under stress at the moment. This could involve spending an evening doing something you like, or even taking a short five-minute break to quietly observe your surroundings. Relaxation techniques, such as breathing exercises, yoga or meditation, can also help you relax and reduce stress.

How is IPF managed?

2.0

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 while there is no pharmacological cure for IPF yet, various therapies can help to alleviate symptoms and slow down disease **progression**. Therapeutic strategies include various medications, as well as a variety of non-pharmacological options.[2,4,9]

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.[11,16] With the help of his doctor, Peter decided which IPF therapy could work best for him, his individual situation and symptoms.

Progression

The worsening of a disease/condition over time

Comorbidities

A disease or condition that occurs simultaneously with another disease or condition

Acute exacerbations

An event characterised by sudden, severe worsening of symptoms or increases in disease severity

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.[1,8] Peter was told therapy with supplemental oxygen may be prescribed if the level of oxygen in the blood gets too low, thereby potentially causing tiredness and leading to him becoming less active.

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 needed oxygen and how often he would need to use it, the pulmonologist conducted a series of tests. 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 when exercising.

Oxygen therapy

Administration of oxygen as a medical intervention

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 improve Peter's wellbeing.[1,9,11]

Pulmonary rehabilitation programmes offer a variety of services, including:[9,11,18]

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

A team of specialists, such as doctors, nurses, physiotherapists, social workers and dieticians, generally run pulmonary rehabilitation programmes. You can receive pulmonary rehabilitation in your home, community, or your local hospital.

The goal of pulmonary rehabilitation is to provide you with education, skills, and tools to help improve management of your condition and increase your participation in social and physical activities.[18]

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

Pulmonary rehabilitation

An education and exercise programme designed to improve the quality of life for people with lung conditions

2.1.3 Lung transplantation

Another non-drug option Peter's physician informed him about was lung transplantation.

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.[9,11] A lung transplant is a major intervention and comes with its own risks including serious complications such as infections or **rejection** of the donor lungs. On the other hand, a transplant could improve the person's quality of life and help them live longer.[1,9,11]

Not everyone with IPF is eligible for a lung transplant. Some patients may have other conditions, such as high blood pressure, that make a lung transplant impossible. Many programmes have an upper age limit between 60 and 65 years. There are also only a very small number of donor organs available for transplantation.[8,11,19] Therefore, even if you are eligible, you may need to join a long waiting list for a transplant.

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.[11]

Lung transplantation

An operation to replace a patient's diseased lungs with lungs from a donor

Rejection

An immune reaction of a transplant recipient's organism to a transplanted organ or tissue

2.2 Medications

While there is no cure for IPF, medications may help to slow disease **progression.**[9] In **clinical trials,** it was shown that some medications are able to slow the decline of lung function in patients with IPF.

When Peter spoke to his doctor, he was told he would be prescribed a type of medication called an antifibrotic.

Antifibrotic medicines are one of the most recent treatment options available for IPF and can provide benefits to patients.

There are two types of antifibrotic medicines available to treat IPF: nintedanib and pirfenidone.[1,9,11]

Antifibrotics aim to prevent or slow the scarring and stiffening of lung tissue. They are effective at helping to slow down disease progression.[1,9,11]

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

People with IPF often have other diseases. These can be a significant influence on the quality of life of patients with IPF.

Different medicines can interact with one another in the body. This is why your doctor might change your medications if you are prescribed a new treatment for IPF or another disease.

Clinical trials

Medical studies that research how safe and effective a new therapy or technique is for treating a certain disease

Antifibrotics

A type of medication that aims to slow or prevent the scarring and stiffening of lungs to slow disease progression

Progression

The worsening of a disease/condition over time

2.3 Other **treatment options**

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

Palliative or supportive 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.[20,21]

As Peter's pulmonologist explained, the terms palliative care and end of life care are often confused. In reality palliative care provides relief from symptoms throughout all stages of the disease. Conversely, end of life care refers to specialist care provided at the end of somebody's life only.

Therefore, a referral to palliative care can be very effective in treating the symptoms of IPF even in the early stages of the disease.

Palliative care in IPF can involve treatments to help reduce the severity of symptoms such as breathlessness and cough. It can be used alongside other treatments to strike a balance between trying to prevent progressive lung scaring and to limit symptoms

Palliative care

Therapy that provides relief from symptoms to helps patients live more comfortably with their disease

Resources & Tips

3.0

3.1 **Helpful** web resources

If you are searching for IPF facts online, you might get an overwhelming number of results. To see a detailed overview of key topics related to IPF, please visit **www.LifewithPulmonaryFibrosis.com**

This website has been created especially for patients and caregivers who are affected by different types of pulmonary fibrosis, including 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

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 on the right 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:

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. There 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 any of the concerns relating to your IPF care.

3.2.3 Checklist for air 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 a candidate for in-flight oxygen
- ☐ Make sure you check the COVID-19 regulations of the country you are departing and the one you will be traveling to
- ☐ Ensure you know whether you will need a COVID-19 travel certificate when you travel, and how soon before departure you may need to be tested
- ☐ Investigate whether you may need to quarantine and if this may affect any plans you have
- ☐ Think about a backup plan if restrictions change when you are away

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.[22]
    - 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, will the ticket reservation 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

3.2.3 Checklist for air travel

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, as not every medication is available in every country.
- ☐ Contact the airline to confirm all necessary precautions and special requirements to be arranged on your flight.
- ☐ Contact the airport to confirm the status of your planned assistance.
- ☐ Ensure you have enough hand sanitizer and that the size complies with air travel hand luggage regulations
- ☐ Pack enough face masks or face coverings for your trip, and make sure you have one to wear on the plane, if necessary

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
- ☐ Make sure you check current COVID-19 travel restrictions, whether you're traveling at home or abroad
- ☐ Ensure you know whether you will need a COVID-19 travel certificate if you are traveling abroad, and how soon before departure you may need to be tested
- ☐ Investigate whether you may need to quarantine and if this may affect any plans
- ☐ Think about a backup plan if restrictions change when you are away

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 this before you travel
- ☐ Ensure you have enough hand sanitizer and face masks or face coverings for the duration of your trip

Glossary

4.0

4.0 Glossary

Acute exacerbations

An event characterised by sudden, severe worsening of symptoms or increases in disease severity[2]

Alveolus (plural alveoli)

Tiny air sacs in the lungs where the exchange of oxygen and carbon dioxide to and from the blood takes place

Antifibrotic

A type of medication that aims to slow or prevent the scarring and stiffening of lungs to slow disease progression[1,9,11]

Arterial Blood Gas Test

A test that shows how well the lungs are working by measuring how much oxygen and carbon dioxide is in the blood[9]

Breathing exercises

Techniques that involve breathing in a certain way to control breathlessness and strengthen your lungs

Bronchoalveolar lavage

A test where a sample of fluid is taken from the lungs to help diagnose IPF[9]

Cardiovascular

Refers to the heart, and blood vessels

Chest X-ray

A non-invasive medical test that creates images of the organs and bones inside the chest

Chronic obstructive pulmonary disease (COPD)

A chronic, ongoing disease where airflow is limited to the lungs. It is usually progressive and linked to inflammation in the airways[23]

Clinical trials

A medical study that studies how safe and effective a new therapy or technique is for treating a certain disease.

Comorbidity

A disease or condition that occurs simultaneously with another disease or condition

Complementary therapy

Therapies used alongside conventional treatment that help treat symptoms and improve overall physical and mental wellbeing

Continuous positive airway pressure (CPAP) device

A device to deliver compressed air to improve sleep in people with obstructive sleep apnoea[24]

Coronary artery/heart disease

A disease where a waxy substance (plaque) builds up inside the coronary arteries, which supply oxygen-rich blood to the heart muscle[25]

Emphysema

A lung condition where the air sacs within the lungs (alveoli) become damaged[11,23]

Fatigue

Extreme weariness resulting from exertion or illness

Fibrosis

Where inflamed tissue is replaced with scar tissue, making it become thickened and stiffer[2]

Finger clubbing

An IPF symptom where the ends of the fingers become wider and rounder[2]

Gastroenterologist

A physician specialised in the management of diseases of the digestive system

Gastro-oesophageal reflux disease (GERD/GORD)

A digestive disease where stomach acid moves up out of the stomach and irritates the lining of the food pipe (oesophagus)[26]

Heartburn

A burning sensation in the chest, which can spread to the throat, along with a sour taste in the mouth

High-Resolution Computed Tomography Scan (HRCT)

A type of X-ray that generates multiple, detailed images of areas inside the body[11]

Hyperventilation

Rapid and uncontrolled breathing

Idiopathic

Of unknown cause

Idiopathic Pulmonary Fibrosis (IPF)

A disease where there is progressive scarring or thickening of the lungs without a known cause[1,2]

Lung biopsy

The surgical removal of cells or tissue amples from the lung for examination by a pathologist[11]

Lung scarring

The lung tissue becomes thickened and stiff

Lung transplantation

An operation to replace a patient's diseased lungs with lungs from a donor

Mindfulness

A technique that helps to reduce stress and anxiety by helping to understand and manage your emotions

Nasal cannula

Small plastic tubes or prongs that fit in the nostrils to deliver supplementary oxygen[27]

Obstructive Sleep Apnoea

A sleep disorder characterised by breathing that repeatedly stops and starts during sleep[11]

Occupational therapist

A specialist who helps someone recover or live with their symptoms more easily[11]

Oxygen concentrator

A machine that removes other gases from the air to provide oxygen for oxygen therapy[9,11]

Oxygen therapy

Administration of oxygen as a medical intervention[9]

Palliative care

Therapy that provides relief from symptoms to helps patients live more comfortably with their disease[21]

Power of attorney

A formal document when someone else is given the responsibility to manage your affairs and make certain decisions on your behalf

Progression

The worsening of a disease/condition over time

Pulmonary

Refers to the lungs

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[9,11]

Pulmonary hypertension

A type of high blood pressure that affects the blood vessels to the lungs and the right side of the heart[28]

Pulmonary rehabilitation

An education and exercise programme designed to improve the quality of life for people with lung conditions[18]

Pulmonologist/respirologist

A physician specialised in the lungs

Pulse oximetry

A test used to monitor oxygen levels in a patient's blood[9,11]

Pursed lip breathing

A breathing technique to help control breathlessness and reduce anxiety[29]

Rare disease

A disease that affects only a small percentage of the population

Rejection

An immune reaction of a transplant recipient's organism to a transplanted organ or tissue

Risk factor

A variable associated with an increased risk of disease or infection

Under-active thyroid

A condition where your thyroid gland doesn't produce enough hormones

Vaccine

A product that gives protection against a specific infection

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5.0

5.0 References

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