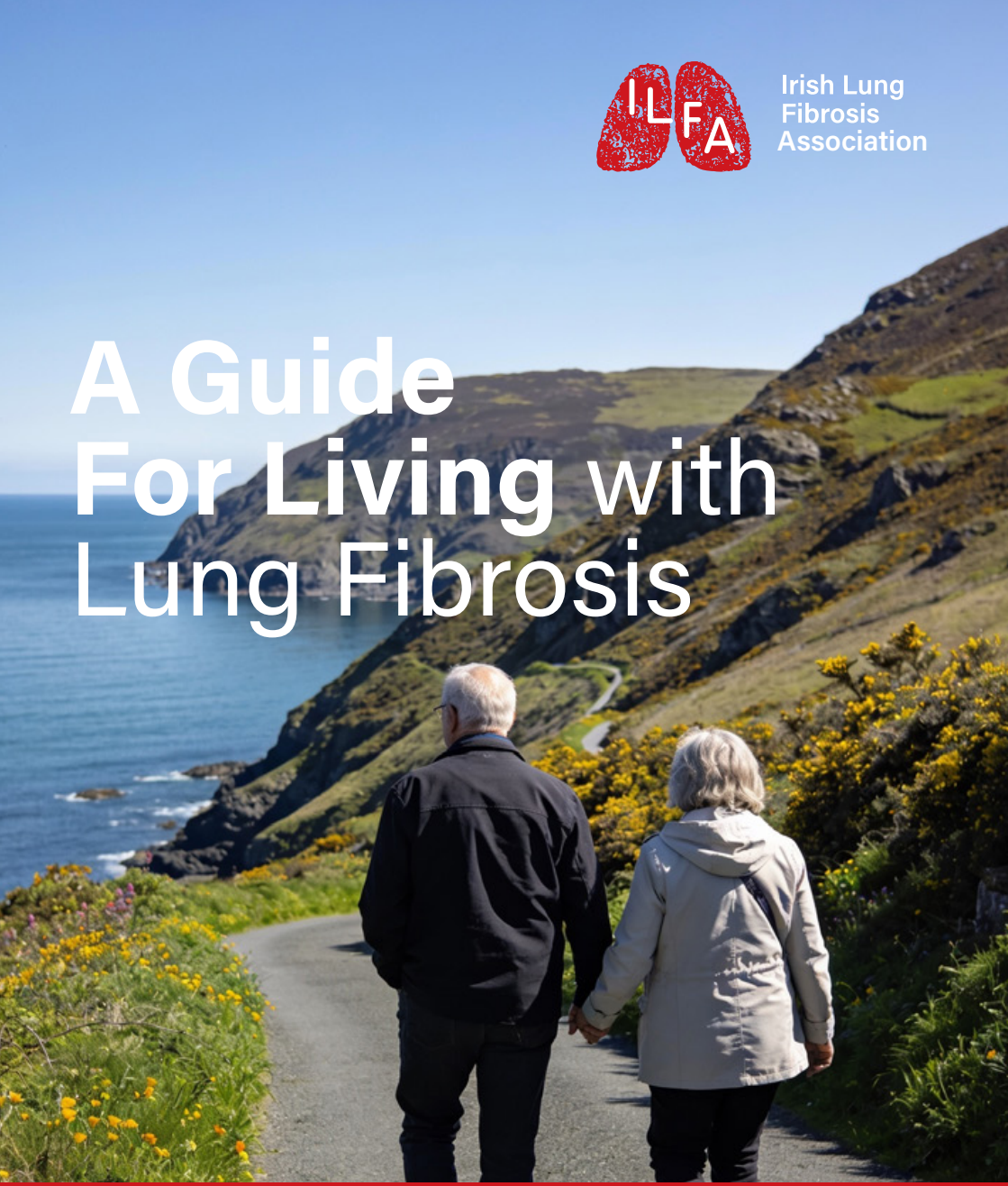
Irish Lung Fibrosis Association

A Guide For Living with Lung Fibrosis



Information for patients and carers

Learn more at ilfa.ie

About This Booklet

This booklet has been prepared to help you and your family understand lung fibrosis (otherwise known as pulmonary fibrosis or Interstitial Lung Disease).

It provides information about the cause of the disease, symptoms, diagnosis, disease progression, treatment, symptom management, maintaining your health and wellbeing and other topics. We hope you find it useful and reassuring.

If you have any questions after reading this booklet, please do not hesitate to contact us or speak with your healthcare team.

About ILFA

The Irish Lung Fibrosis Association (ILFA) is Ireland's national patient organisation for those affected by lung fibrosis. We provide education, support, advocacy and research to help patients and families affected by this condition.

Contents

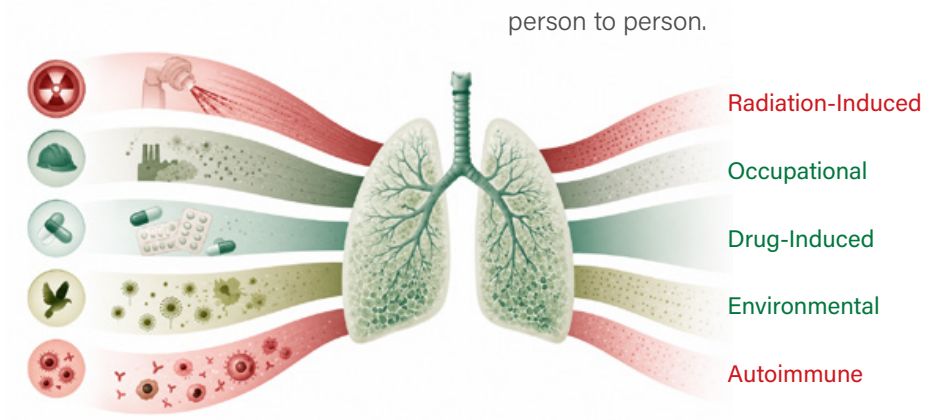
Understanding Your Condition

What is Lung Fibrosis?

Lung Fibrosis or Pulmonary Fibrosis is the name for a group of Interstitial Lung Diseases (ILD) in which scar tissue (fibrosis) develops in the lungs. Doctors call these Interstitial Lung Diseases or ILDs because the tissue damage happens in the interstitial space which is the area around the tiny air sacs in the lungs.

When we breathe in, our lungs are exposed to many types of dust particles, chemicals and toxins. Healthy lungs can repair the damage to lung tissue caused by these pollutants. However, if you have lung fibrosis, the repair process goes out of control, and too much tissue is produced causing fibrosis.

As fibrosis develops, the soft, spongy and elastic tissue in healthy lungs is replaced by hard, thick scar tissue. This makes it harder for your lungs to expand when you breathe in. Fibrosis also prevents the normal movement of oxygen from the air sacs (alveoli) in the lungs into the bloodstream, causing you to be more breathless as your blood oxygen levels fall. This can vary from person to person.



Causes

It can be challenging for doctors to know what causes lung fibrosis. It's believed that anywhere from 3 to 20% of people with lung fibrosis can have another family member with the disease, but that doesn't mean there is a genetic cause. The most common form of lung fibrosis is Idiopathic Pulmonary Fibrosis or IPF. Idiopathic means there is no known cause and a diagnosis of IPF is generally made after all other potential causes are ruled out. There are five main categories of identifiable causes of lung fibrosis.

- **Drug-Induced:** which includes medications such as amiodarone (a heart medicine), nitrofurantoin (an antibiotic), and methotrexate, bleomycin and bisulfan (all used for chemotherapy).
- **Radiation-Induced:** radiation treatment to the chest.
- **Occupational:** breathing in dust, fibres, or fumes in the workplace that contain tiny particles (such as asbestos, silica, wood, metal, or textiles).
- **Environmental:** breathing in air or dust contaminated with bacteria, viruses or animal material such as bird droppings
- **Autoimmune:** medical conditions that affect the joints and connective tissue such as arthritis, scleroderma or sarcoidosis.

Common Symptoms

The first symptom you may notice is shortness of breath especially when you are climbing the stairs, walking, exercising or taking part in an activity.

Your breathlessness can worsen as your disease progresses. Other symptoms can include:

- A dry cough.
- Low energy and fatigue.
- Changes in the shape of your fingertips. They may get bigger and more rounded. This is called 'clubbing.'
- Stomach acid and reflux (indigestion).

Diagnosis and Treatment Options

Overview

While some ILDs are stable, many ILDs like IPF are progressive which means they get worse over time. The rate of disease progression is different for everyone and can be improved with medication, other therapies, and lifestyle choices. Some patients remain stable with a gradual change in their symptoms over many years. Other patients have a steady decline in their health and their symptoms get worse. Some patients may experience a sudden and serious worsening of their symptoms. This is called an acute exacerbation.

A commonly cited statistic suggests that the average lifespan after diagnosis is three to five years however, this figure is outdated, as newer therapies have been developed that can help slow the progression of the disease. If you have concerns or questions about how your condition may develop, it's important to discuss them with your doctor.

Diagnosis

The diagnosis process starts with your doctor reviewing your medical and family history and discussing your symptoms with you. You may be asked about any continuous or repeated contact with dusts, gases, chemicals or similar substances.

Your doctor will also examine your lungs by listening with a stethoscope. During this process, they may detect sounds known as "crackles," or "rales," which resemble the noise of Velcro being pulled apart.

Your doctor may perform an additional breathing test called a pulmonary function test to measure how much air your lungs can hold. Scar tissue and inflammation can make your lung stiff which means you have to work extra hard to breathe. Your brain senses this extra work and lets you know there's a problem by triggering a feeling of breathlessness. The more scar tissue, the less air the lungs can hold.

Scar tissue also blocks the movement of oxygen from the inside of your air sacs into your bloodstream. This reduces your oxygen level, particularly during exertion or exercise. Oxygen levels can be assessed to see if they decrease after walking by using a six-minute walk test.

These tests will provide clues but if lung fibrosis is suspected then you should be referred to a specialist centre for additional imaging (X-ray or High-Resolution CT scan), lung function tests, or a biopsy.

Treatment Overview

Although there have been significant advances in recent years, including the development of new and promising medications, current treatments may help relieve symptoms and slow disease progression, but they cannot reverse lung damage or cure the condition.

There are several approaches to managing lung fibrosis, including medications, oxygen therapy, supportive strategies such as exercise and nutrition, and in some cases, lung transplantation.

The following information provides a general overview of some of the common treatments that doctors offer lung fibrosis patients. This information is not medical advice. Some treatments may be right for some people, but not for others, and no one treatment is right for everyone.

You should speak with your doctor before starting, changing or stopping any medical treatment.

Drug Therapy

Drug therapy for lung fibrosis is often specific for the type of fibrosis the patient has. Some common therapies (but not all potential therapies) include:

- **Antifibrotics** - Pirfenidone (Esbriet) and Nintedanib (Ofev) are two licensed antifibrotics used in Ireland which may help slow the worsening of lung fibrosis. The choice of which antifibrotic you need depends on your condition and the guidance of your healthcare team.

  - **Pirfenidone (Esbriet)**- Pirfenidone is taken by mouth three times a day. Common side effects are diarrhoea, nausea, vomiting, tummy pain, headache, tiredness, loss of appetite or weight, and sun sensitive skin or rash. Regular blood tests are needed to check liver function, and if you experience side effects, contact your healthcare team right away.

- **Nintedanib (OFEV)-**Nintedanib is usually taken by mouth twice a day. Common side effects include diarrhoea, nausea, vomiting, tummy pain, headache and loss of appetite or weight. Regular blood tests are needed to check liver function, and if you experience side effects, or if you notice yellowing of your skin or eyes, very dark urine, severe itching, unexplained bruising or bleeding, contact your healthcare team right away.

- **Steroids**, also called corticosteroids, are medications that help control inflammation in the lungs by suppressing or weakening the immune system.

- **Other medications**, including those for pulmonary hypertension, a common complication with lung fibrosis.

Your doctor may prescribe other medications to help you manage your symptoms and any other health problems you may have.

Oxygen Therapy

Your oxygen levels can be measured in different circumstances like at rest, while walking, or during sleep. If your oxygen levels are low, your doctor may prescribe supplementary oxygen on rest and/or ambulation.

While it can be frustrating and inconvenient to use supplementary oxygen, it can help you manage your breathlessness and improve your quality of life. Many people report that they have less breathlessness and fatigue and are better able to live an active lifestyle when using oxygen.

Your oxygen needs may change over time so tell your doctor, nurse or physiotherapist if you are more breathless. If you're having problems using supplementary oxygen, have an open conversation with your doctor about your concerns.

Pulmonary Rehabilitation

Pulmonary rehabilitation consists of classes delivered by healthcare professionals that include exercises, breathing techniques, ways to manage anxiety and stress, nutritional counselling, education, and other supportive components. A course of rehabilitation is generally 6 – 8 weeks with two classes per week. The goal of pulmonary rehabilitation is to improve your ability to function without extreme breathlessness, to increase your exercise capacity and to improve your quality of life.

To get the most out of pulmonary rehabilitation, it's best to attend the entire course. It's also important to do the exercises you learned during the course as recommended, especially once the course has finished.

Community-based pulmonary rehabilitation programmes are somewhat limited in Ireland for lung fibrosis patients, but you may have access to a rehabilitation programme through your ILD Specialist Centre, or your GP might be able to refer you to a programme in your community.

In addition, ILFA offers exercise classes several times per week and regular patient information sessions that, while not a structured pulmonary rehabilitation programme, contain many of the same elements of a structured programme.

Tip

Keep Moving

Attend your full pulmonary rehabilitation programme and continue the exercises afterwards.

Lung Transplantation

A lung transplant (surgery to remove a damaged or diseased lung and replace it with a healthy lung from a deceased donor) is the only effective treatment for people with late-stage lung fibrosis, but this is not a suitable option for everyone. It involves a major operation and depends on disease severity and your general health.

If you are being considered for a lung transplant, you will be extensively evaluated by the National Lung Transplant Centre in the Mater Misericordiae University Hospital in Dublin to determine whether you meet the candidacy requirements for lung transplant.

- To be considered a strong candidate for a lung transplant, it's important to keep your vaccinations and overall medical care up to date. If you have additional health conditions alongside your lung disease, take your medications as directed, attend all follow-up appointments, and adhere to any recommended dietary plans.

- You must also meet weight requirements. Studies demonstrate that outcomes are better when patients are at an ideal weight, as measured by Body Mass Index (BMI). Follow nutrition and exercise guidance provided by your doctor to ensure you remain at the optimal weight and BMI.

- You must not use tobacco, alcohol, or any illicit drugs to be considered for transplant. Tobacco use includes nicotine replacement therapies like patches, gum and vaping.

- You must have a strong support system with two or three caregivers who can provide continued support during the pre-and post-transplant period.

Even if you're found not to be a candidate for transplant or decide not to pursue one, your health and wellbeing will be better if you follow these guidelines.

In Ireland, lung transplant waiting times can vary, and how long a patient waits depends on the severity of their disease when they are placed on the list as well as when a suitable donor becomes available.

If your blood type is less common, you are very tall or short, or you have high levels of antibodies against tissues received from others, it may take longer to find a suitable match.

Being in the best possible physical shape at the time of your surgery will help with your recovery and transition to a more active life. If you are on the waiting list for a transplant you will receive pulmonary rehabilitation and be expected to continue with exercise programmes after your transplant.

The Mater Hospital has an excellent Transplant Resource: https://www.lungtransplant.ie/

Symptom Management

Breathlessness and cough are the major symptoms experienced by people living with lung fibrosis. You may also experience fatigue, anxiety, and depression. Contact your doctor for help managing your symptoms.

- **Breathlessness** – programmes such as pulmonary rehabilitation can enhance fitness levels and provide training in breathing techniques, along with other strategies like using fans to help ease shortness of breath. Your doctor may recommend the use of supplemental oxygen as low oxygen can be a cause of breathlessness.

- **Fatigue** – Rest is very important when you have lung fibrosis and so it's important to get 7-8 hours of good quality sleep every day if you can. Also, planning ahead and allocating plenty of time for an activity will reduce pressure and stress. It can be a good idea to split big tasks up into three or four smaller tasks with resting in between. Finally, don't be afraid to speak up and let people know when you need to walk a bit slower or when an activity is too challenging that day.

- **Cough** – There are many causes of cough in addition to lung fibrosis. Post-nasal drip from allergies, a cold, or gastroesophageal reflux (GORD) are examples that could contribute to cough. Low oxygen levels can trigger cough in some people as well. Your doctor can help identify the cause of the cough and provide treatment. There are also special exercises you can do to help manage cough. Physiotherapists with expertise in cough suppression can teach you these.

- **Medication Side Effects** - Managing medication side effects, particularly with antifibrotics like pirfenidone and nintedanib, focuses on managing gastrointestinal issues and skin sensitivity. Key strategies include taking medication with food, staying hydrated, using high-SPF sunscreen, and consulting your doctor for dose adjustments.
  - More information for nintedanib side effects: https://patient.boehringer-ingelheim.com/us/products/ofev/side-effects
  - More information for pirfenidone side effects: https://www.esbriet.com/summary.html

- **Anxiety and Depression** – it's normal for everyone to experience a range of different emotions. You might feel frightened, worried, angry or confused. You might also feel isolated or lonely at times. If you find that your mood has been low for more than a few weeks, or that anxiety is affecting your daily life and these feelings aren't going away, these could be signs that you have anxiety or depression. It's important to talk to your doctor. It might also be helpful to join a lung fibrosis support group where there are other people with similar experiences who can help. Learning some relaxation techniques and how to use mindfulness may also help you. Make sure you draw on the support of your healthcare team and family and friends. They'll want to help you.

- **Acute Exacerbation** – it's important to contact your doctor right away if you develop serious difficulty breathing, fever or flu-like symptoms, or your cough gets worse, as you may be experiencing what is called an acute exacerbation. Not everyone will experience an acute exacerbation but if you do it can be serious and you may have to go to hospital.

Tip

Track Your Symptoms

Keep a weekly symptom diary to see how your symptoms change over time.

Palliative Care

Palliative (or supportive) care is meant to help people living with life-limiting conditions have the best possible quality of life. It's focused on relieving and preventing symptoms that are bothersome or distressing. Palliative care can also help you with advanced care planning, which allows you and your family to decide your goals for care as your disease progresses.

Palliative care is provided by a multidisciplinary team (MDT) tailored to meet the needs of you and your family. The team might include a variety of medical professionals including doctors, nurses, social workers, pharmacists, occupational therapists, counsellors and others.

Palliative care can be provided in hospital, hospice or nursing home settings or at home. It is provided at three different levels depending on your illness. Early on, it can be a part of your routine check ins with your healthcare team. As your illness progresses and if your symptoms become complicated you may need more specialised care from a specialist palliative care team. You can receive any level of palliative care regardless of whether you are at home or in a care facility.

Taking advantage of palliative care soon after diagnosis can get you access to pulmonary rehabilitation, counselling, and other helpful supports.

Engaging with palliative care early in your diagnosis can have the following benefits:

- Improvement in dyspnoea management
- Greater engagement in advanced care planning
- Reduced end-of-life hospitalisation
- Greater adherence to patient wishes for care
- Improved patient and caregiver experience

Palliative care is not the same as hospice or end-of-life care which is generally focused on the last months of life. Hospice care is focused on comfort, dignity, and ensuring that the person's remaining time is as peaceful as possible. It can include treatments to control pain and other symptoms and provides bereavement support for those important to the person after death.

Palliative care is available to anyone, at any age, living with a life-limiting illness and can be accessed through your healthcare team. Many palliative care services are free for all patients and their families in Ireland. Depending on the stage of your illness or financial circumstances, you may be able to receive a medical card, which is applied for by your Consultant, GP or Social Worker to the HSE.

Tip

Visit 'Let's Talk'

Visit ILFA's YouTube Channel playlist for an excellent palliative care discussion led by Dr. Una Malloy.

Getting the Most from Medical Consultations

Visiting an ILD Specialist Centre

When you have been diagnosed with lung fibrosis you should be referred to an ILD Specialist Centre for care. There are eight specialist centres in Ireland - five in Dublin, and one in Cork, one in Galway, and one in Limerick. While the specialist centre healthcare team are experts in lung fibrosis, your lived experience with the disease also makes you an expert of your condition. The healthcare team is there to provide you with their guidance and support your care.

In addition to the specialist doctor, there are several other specialists on the multi-disciplinary healthcare team. These include nurse specialists who can offer health advice and support and physiotherapists who can advise on the best exercises to stay fit and healthy.

You may also be offered dietetic advice, mental health supports, occupational therapy, and the services of a social worker. ILD specialist services vary across the country.

It's important to take a proactive approach to your health, stay active and have a positive outlook. Keep a diary to track how you are feeling and record any changes in your physical or mental health, sickness or infections that arise. Write down the dates and reasons for visits to your GP. Keep a list of all your medicines, the dose, how often you take them and any side-effects you experience. This information can be helpful when attending your specialist centre appointments.

Medical appointments can be overwhelming and so it's good to be prepared. In addition to bringing the information noted on the previous page with you, here are some additional tips for your appointments.

- ✓ Prepare questions to ask at the appointment in advance.
- ✓ Bring someone, a family member or friend, with you.
- ✓ Take notes that you can refer to later.
- ✓ Make sure you fully understand what is said to you. If you do not understand, ask the doctor to clarify and to repeat it in non-medical terms.
- ✓ Confirm who to contact after the appointment about exacerbations or other queries.
- ✓ Ask what you should do in an emergency, for example, an acute exacerbation.

Tip

Be Prepared

Keep a diary of your symptoms, medications and questions before appointments.

Telemedicine

If you live far away from a specialist centre then you might want to have some of your appointments by phone or video, this is called telemedicine. It is your choice to use telemedicine for your appointments, your doctor should always ask for your consent.

The convenience of not having to travel is a key advantage of telemedicine, but there are disadvantages as well. Your doctor may not be able to see changes in your physical appearance that can be clues to your health such as the colour of your lips or fingernails. It can also be harder to include caregivers in the consultation and there are some tests that can't be done from home.

If you do decide to use telemedicine for an appointment, make sure you have a smartphone, tablet or laptop computer with a camera, a microphone and a good internet connection. Make sure as well that you take the appointment in a comfortable, quiet, private, and well-lit space. For video consultations, you should be facing in the direction of the light source, it should not be behind you or you'll be in shadow. You should also make sure the volume is set loud enough on the device you're using to hear what's being said.

Questions for Your Specialist

You may need to have tests carried out to either confirm your diagnosis or to monitor your ongoing health. Questions for your specialist regarding these tests might include:

What does this test involve and why do I need it?

Will I need to prepare for this test?

Are there side effects or a recovery time?

Will I need to stay in hospital?

Will I need to arrange supportive aftercare?

When and how will I get test results and what do abnormal results mean?

Maintaining Your Health - Lifestyle

Lung fibrosis can affect you physically and emotionally, but you can make positive lifestyle changes to help you cope with the challenges of living with this condition.

The most important things you can do are to learn more about your lung condition, take a proactive approach to managing your health and try to stay positive.

Lifestyle Tips:

- If you smoke, stop. Ask your doctor for help. Ask your family and friends not to smoke around you or in your home to protect you from the effects of second-hand smoke.
- Stay as active as possible. Exercise is very important for people with a lung condition and will help you to stay fit and strong. Exercise can also help improve your mood and emotional wellbeing.
- Ask your doctor to refer you to a pulmonary rehabilitation programme. If one isn't available in your area and if your doctor has cleared you for exercise, then you can join ILFA's online exercise classes.
- Ensure you're up to date on vaccinations and although it's not always possible, try to avoid exposure to respiratory infections. Wash your hands regularly and dispose of any tissues you use as soon as possible. Avoid crowded indoor places like shopping malls or visiting others in hospital. Consider carrying hand sanitizer with you, and if you know someone has a cold or chest infection, avoid close contact with them until they're feeling better.
- To help maintain a healthy body weight, follow a healthy, balanced diet. A healthy diet is one that includes a variety of foods to provide your body with all the different nutrients it needs. Healthy eating advice is outlined in the Irish Food Pyramid Guidelines, which are available online: https://assets.gov.ie/static/documents/expanded-food-pyramid-poster.pdf
- Continue to socialise and enjoy your hobbies. It is important to carry on with these activities as much as you can, just take things slowly.
- Ask your doctor for help if you are struggling physically or emotionally. If you think counselling would be helpful, ask your doctor to refer you to a counsellor or psychologist.
- Join a lung fibrosis support group to meet others and learn from their experiences.

Tip

You're Not Alone

ILFA has both regional and online support groups meeting regularly. Go to our website at ilfa.ie to find out more!

Diet and Nutrition

Note: the advice provided in this section is general nutrition advice and may not be appropriate for all individuals.

Many people living with lung fibrosis find that their body weight and other nutritional indicators can be impacted by the disease. There can be many reasons for this. It might be that breathlessness can make eating more difficult. A lack of energy can make preparing meals harder. You might have reduced appetite. As a result, losing weight and associated muscle mass is common in lung fibrosis.

It's important to eat a well-balanced diet in the management of your disease. Eating the right foods can help maintain muscle strength and support lung function. It can also help with nausea, breathlessness and acid reflux you might be experiencing. It's important to maintain a healthy weight (not being underweight or overweight) and body mass index as well. For example, losing weight unintentionally can lead to increased flare ups, increase the risk of hospitalisation, and lead to faster deterioration in lung function.

Maintaining muscle mass and strength is very important. It offers a protective effect and is associated with reductions in breathlessness and improved quality of life. The best way to do that is through diet and exercise.

For example, protein is the foundation of building muscle so it's important to eat foods high in protein like meats, chicken, fish, nuts, tofu, and beans with every meal (about 20-30 grams of protein with each meal is recommended).

Medications can cause side effects. Nausea, diarrhoea or a change in appetite can occur and this can make it harder to enjoy your food. If you experience side effects from your medication, it's important to speak to your healthcare team.

See opposite
(source: https://indi.ie/images/fact_sheets/OPNIG%20Finalised%202024%20Sarcopenia%20%20Protein.pdf)

INDI OLDER PERSONS
Irish Nutrition & Dietetic Institute

How can I get enough Protein each day?

- 3 meals containing 20 - 30g protein
- Use full fat milk or yoghurt in cooking. Drink milk with meals or snacks
- Poor appetite? Eat 3 small meals with protein containing snacks
- Add cheese/beans/lentils to soup, salad, toast, potato
- Choose products with High Protein on the label e.g. yoghurts and cereals
- Eat the protein on your plate first

Take a high protein Oral Nutrition Supplement if recommended by your doctor or dietitian

High Protein Balance Meal Ideas

Breakfast = 30g

Lunch = 28g

Dinner = 45g

Snacks

10g 10g 10g 10g

Foods containing on average 10g protein:

2 handfuls of nuts
3 slices of ham
2 eggs
33g (4 slices) chicken breast
1½ matchbox size piece of cheese
1 pot (~125g) of yoghurt with nuts
1 bowl (200g) rice pudding

Protein can be added to meals using:

Full fat milk/skimmed milk powder
Cheese/Yoghurt
Nut butters/Hummous/ground nuts

For specific dietary advice, ask your medical team to refer you to a Registered Dietitian or visit www.indi.ie to find a Registered Dietitian

INDI Older Persons Nutrition Interest Group 2024; Review Date: 2027

Here are a few strategies to help manage nutrition impact symptoms.

Symptom	Diet Strategy
Short of Breath	■ Eat when breathing feels more settled ■ Take your time eating ■ Put cutlery down between bites ■ Smaller, more frequent meals and snacks ■ Choose softer, moist foods (e.g. milk puddings, ice cream, casseroles, mashed potatoes) ■ Choose nourishing drinks (e.g. milk, smoothies) ■ Add sauces and gravy to food ■ Eat sitting upright
Fatigue	■ Try to rest before meals ■ Batch cook when energy levels are high ■ Consider on-line grocery delivery ■ Keep a stock of ready meals ■ Let family and friends help with shopping and cooking
Dry Mouth	■ Choose softer, moist foods ■ Suck on sugar-free sweets, ice lollies, and ice cubes or frozen fruit segments ■ Rinse mouth with water after using steroid inhalers ■ Avoid alcohol-based mouthwashes ■ Ensure adequate hydration ■ Consider artificial saliva sprays and mouthwashes ■ Use lip balm regularly for dry lips
Taste Changes	■ Rinse mouth with water after using steroid inhalers ■ Adopt good oral hygiene ■ Try stronger tasting foods ■ Experiment with different sauces and seasonings ■ Try herbal tea before meals

If you are experiencing nausea, try taking antifibrotic medications with food (as long as this is recommended by your doctor), avoid strong-smelling foods if you find they are a trigger, and try drinking peppermint tea or foods and drinks containing ginger.

If you are experiencing diarrhoea, taking antifibrotic medications with food may help (as long as this is recommended by your doctor). You might also want to limit your intake of sugar-free mints and gums as they can have a laxative effect. You can also try reducing your caffeine intake.

Be aware that if you are experiencing diarrhoea you can become dehydrated. It is important to drink adequate amounts of fluids to prevent this from happening. In general, women should aim for 8 x 200ml glasses of fluid per day, while men should aim for 10 x 200ml glasses. Water, milk, and sugar-free drinks all count towards your daily fluid total.

Tip

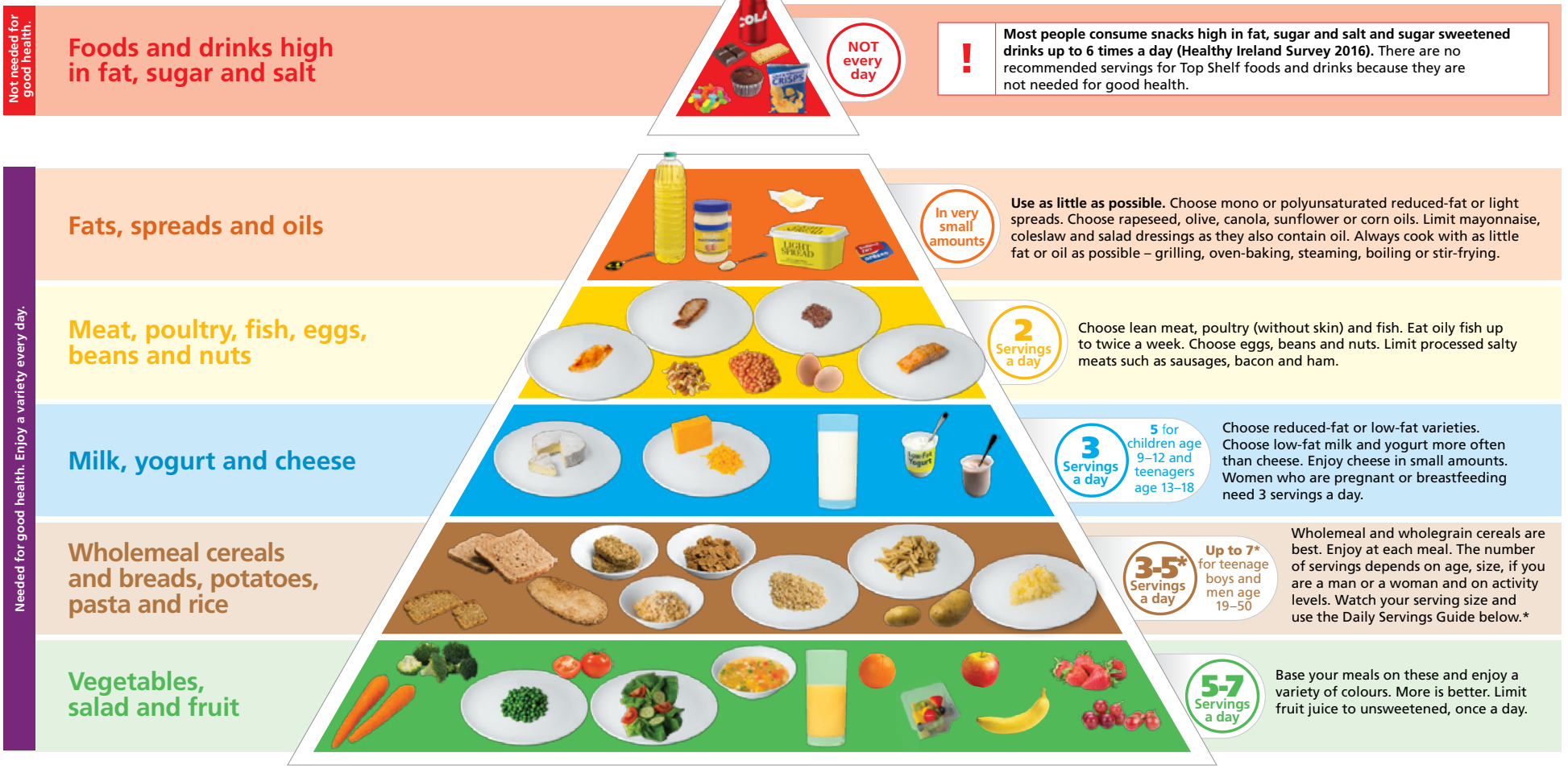
Visit 'Let's Talk'

Visit ILFA's YouTube Channel "Let's Talk" Playlist for an excellent Diet and Nutrition discussion led by Ciara Walsh.

Use the Food Pyramid to plan your daily healthy food choices everyday and watch your portion sizes.

www.healthyireland.ie

Food Pyramid for adults and children aged 5 years and over.



*Daily Servings Guide – wholemeal cereals and breads, potatoes, pasta and rice

Active	Child (5–12)	Teenager (13–18)	Adult (19–50)	Adult (51+)
♀	3–4	4	4–5	3–4
♂	3–5	5–7	5–7	4–5

Inactive	Teenager (13–18)	Adult (19–50)	Adult (51+)
♀	3	3–4	3
♂	4–5	4–6	4

There is no guideline for inactive children as it is essential that all children are active.

Average daily calorie needs for all foods and drinks for adults

♀ **Active** 2000kcal **Inactive** 1800kcal

♂ **Active** 2500kcal **Inactive** 2000kcal

Serving size guide

Cereals, cooked rice and pasta, and vegetables, salad and fruit
Use a 200ml disposable plastic cup to guide serving size.

Cheese
Use two thumbs, width and depth to guide serving size.

Meat, poultry, fish
The palm of the hand, width and depth without fingers and thumbs, shows how much you need in a day.

Reduced-fat spread
Portion packs found in cafes can guide the amount you use. One pack should be enough for two slices of bread.

Oils
Use one teaspoon of oil per person when cooking or in salads.

Drink at least 8 cups of fluid a day – water is best

Get Active!
To maintain a healthy weight adults need at least 30 minutes a day of moderate activity on 5 days a week (or 150 minutes a week); children need to be active at a moderate to vigorous level for at least 60 minutes every day.

Source: Department of Health. December 2016.

Exercise

Exercise is very important for building strength, increasing endurance, and managing breathlessness. A big problem for people with lung fibrosis is the fear of breathlessness, this can lead to immobility and disability. It is important to break out of this cycle and to manage your breathlessness, rather than let it control you.

We all get breathless doing activities and exercising because we need to take in more air (and oxygen) to give us energy. Moderate shortness of breath is acceptable and necessary and should not prevent you from going out and taking part in everyday activities and exercising. If you are breathless, stop and get your breath back when you need to, find your own strategies for doing this like stopping to look in a shop window for a while even though you are not interested in buying anything. Learn the STALL Breathing technique.

If your healthcare team has cleared you for exercise, consider this weekly schedule:

- **Aerobic Exercise:** Aim for 20 to 30 minutes of continuous or interval-based movement (e.g., gentle walking) 3 to 5 days per week.
- **Strength Training:** Perform 2 to 3 days of resistance training, focusing on high repetitions with lighter weights.
- **Flexibility:** Gentle stretches and warm-ups help prepare your muscles for exercise and prevent stiffness.

Tip

Move at Your Own Pace

ILFA offers daily exercise classes—contact info@ilfa.ie to register

Stall Breathing Technique©

S Stop what you are doing.

T Try to remain calm. Turn up your oxygen.

A Assume a comfortable position. For example, sitting or leaning.

L Let your imagination take you to a safe place. Imagine yourself there, relaxing.

L Let your breathing return to normal.

When your breathing has returned to normal, reset your oxygen to normal.

Call for medical help if your symptoms do not settle.

Your Wellbeing

Looking after your mental health is just as important as your physical health. Being diagnosed and living with lung fibrosis can be stressful.

It's common when diagnosed to feel a wide range of intense emotions and feelings. Emotions can vary but often there is a sense of shock or disbelief, fear, sadness, anger or guilt. It might seem strange, but in some cases people may experience a sense of relief at diagnosis because they now know what they're dealing with.

As the disease progresses, emotions continue to arise. For example, breathlessness is common for people with lung fibrosis. The feeling of breathlessness can lead to anxiety, low mood, negative thoughts and anger or frustration.

Looking after your mental health is an important part of your treatment. If you are experiencing poor mental health, you may be less interested in the things you previously enjoyed. You may feel more anxious, tense, or have difficulty coping. The challenges of living with lung fibrosis can be quite difficult, so you need to look after your wellbeing.

Your carers may also be struggling, so here are some suggestions for you both.

- ✓ **Connection** – keep in touch with family and friends.
- ✓ **Be Active and Healthy** - find ways to be active daily and maintain a healthy and balanced diet.
- ✓ **Live Life** – focus on the things you enjoy like listening to music, hobbies, and spending time outside in nature.
- ✓ **Rest** – aim for 7 to 8 hours of good quality sleep per night.
- ✓ **Join a Support Group** – ILFA's regional and on-line support groups are a supportive community when you are struggling.
- ✓ **Manage Breathlessness** – learn breathing and grounding techniques that can help manage the panic and anxiety that can come with breathlessness.
- ✓ **Share Your Feelings** – it can help to talk about having lung fibrosis. You may prefer to talk to family or friends, with members of your support group, try therapy, or to a counsellor.
- ✓ **Set Goals** – goals can help you focus on what you want to achieve. When developing a goal, it can help to break a big goal down into manageable pieces.

Tip

Know your Birds

The **Merlin** phone app can help you identify birds on your walks!

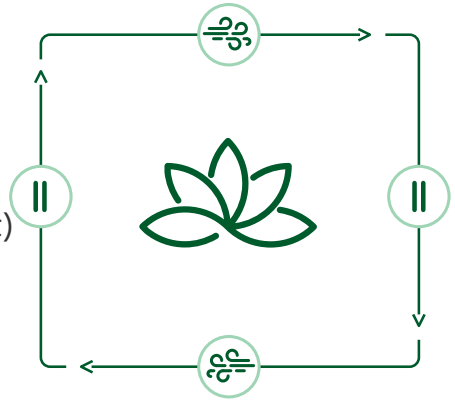
Box Breathing Technique

Box breathing is a technique that can slow your heart rate and help you feel calmer in stressful situations. It can help you manage anxiety while also strengthening the diaphragm to help you breathe easier.

Box breathing is a simple technique to calm your mind and reset your body.

Sit comfortably ▪ Follow the square ▪ Repeat for a few minutes

1. Breathe In. Count to 4 (Or your count)



2. Breathe Out.
Count to 4
(Or your count)

3. Breathe In. Count to 4 (Or your count)

4. Breath Out.
Count to 4
(Or your count)

If you can, extend the **out-breaths** to be a little longer than the in-breaths

Example: In for 4 counts, out for 5 or 6 counts

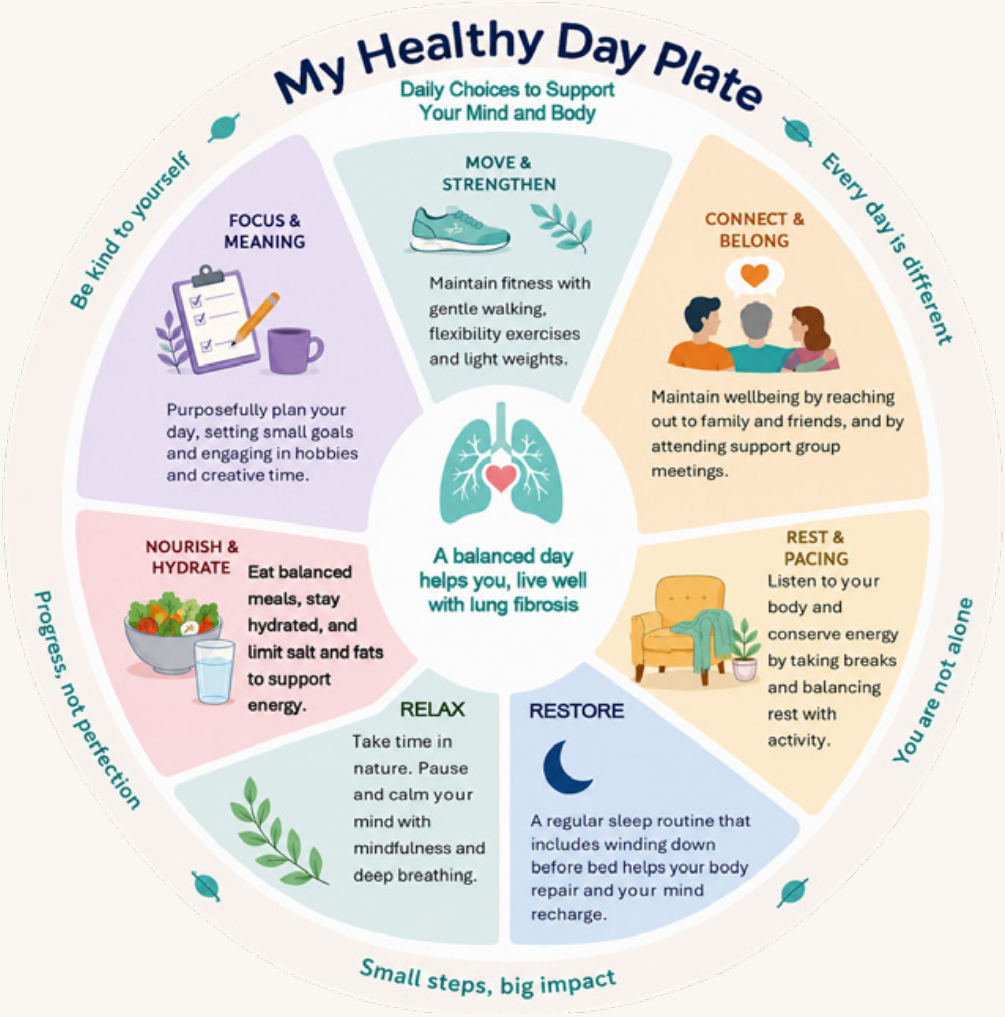
Living Well Every Day

Small daily choices that support my body, mind and lungs

It's important as well to note that mental and physical health are deeply linked. Daniel J. Siegel and David Rock created the Healthy Mind Platter which describes seven everyday activities that your mind needs to stay healthy and work well.

Think of them as "mental nutrients" your brain and relationships need to function at their best. Here is a version of their original work adapted for lung fibrosis.

Taking time to focus on each element of the platter every day helps you feel more balanced and keeps your brain working smoothly. It also strengthens the connections in your brain and helps you build better relationships with other people and the world around you.



Telling Your Family and Friends

Coming to terms with a diagnosis of lung fibrosis can be very difficult. Part of the process includes deciding who to share this news with. You may find yourself wondering whether to tell family and friends, who to tell, and when to tell them. You might worry about how people will react or feel concerned that sharing the news will upset or burden them.

There is no right or wrong way to tell people about your diagnosis and you don't need to share your diagnosis with anyone you don't want to.

The only people who need to know are the healthcare professionals involved in your care. Beyond that, the decision is entirely yours.

When deciding who to tell, it can help to think about the people you would like around you as time goes on. Many people will want to support you, but they can only do that if they understand what is happening.

You may also want to consider who is likely to cope well with the news and who may need more time or support themselves.

You do not have to tell everyone at the same time. For example, you may choose to tell close family first and speak to extended family or friends later. People may notice physical changes over time and ask questions, so it can help to think in advance about what you would feel comfortable saying.

Talking about your diagnosis can feel difficult, but many people find that sharing the news helps them feel less alone and more supported. If you are unsure how to start the conversation, these suggestions may help:

- You may prefer to tell some people individually. This can give you more space to explain things in a way that feels right for both of you.
- You may want a family member or friend to accompany you to an appointment with your healthcare team, where questions about your diagnosis and treatment can be discussed together.
- If you are part of a club, community, or social group, you might choose to tell people together rather than having many separate conversations.
- If there is someone who should know but you do not feel able to tell yourself, you could ask a trusted friend or relative to speak with them on your behalf.

People may react in different ways when they hear your news. Some may be shocked or upset. Some may not know what to say at first, while others may immediately offer support. Occasionally, people may say things that feel unhelpful, even when they mean well. These reactions can be difficult, but they are often a reflection of someone trying to process unexpected news.

People's responses may also change over time. Someone who struggles initially may become a strong source of support later on. If you are worried about upsetting someone by telling them, it may help to remember that sharing your diagnosis can also give people the opportunity to support and spend time with you.

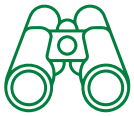
Notes:

Thinking Ahead – Care Plans

A care plan makes your wishes clear at all stages of your disease. It is a key part of an effective support system. Agreeing a care plan is a collaborative process between you, your healthcare team, and potentially your family or carers. The care plan should focus on your personal goals, your physical, mental, and social needs, and what support you require. It should primarily address your current needs but can help you make decisions about your future needs as well. Your care plan should be written down and reviewed on a regular basis.

Note, a care plan is not the same as an Advance Healthcare Directive. An Advance Healthcare Directive is a legally binding document setting out your wishes regarding medical and healthcare treatment (including treatments that you do not want) in case you are unable to make these decisions in the future.

Tip



Think Ahead, Live Well

The Irish Hospice Foundation has excellent resources to help people with life-limiting conditions (and their families) plan for the future.

Key Steps to Agree a Care Plan:

1. **Initiate the Conversation:** Talk to your GP, nurse, or care coordinator; they might also suggest it.
2. **Identify Your Needs and Goals:** Discuss your health, daily routines, emotional, social, and spiritual needs, and what you want to achieve.
3. **Include Key People:** Involve trusted family, friends, or advocates who can support you and understand your wishes.
4. **Document Your Wishes:** Record your preferences and what you *don't* want, including future wishes for legal/financial matters (refer to Advance Healthcare Directive if needed).
5. **Plan for Crisis:** Create a crisis plan with early warning signs, coping strategies, and hospital contact information.
6. **Agree & Sign:** Ensure you agree with the documented plan and receive a copy.
7. **Review & Update:** Regularly review the plan (e.g., at MDT meetings) to adjust for changes in your condition or goals.

My Care Plan

(Source: Irish Hospice Foundation)

How to Use This Plan

This helps me, my family, and the people caring for me understand what matters most and how best to support me. I can fill this in myself or with help. I can change it at any time. I can bring it with me to appointments or hospital visits. Sharing this plan helps everyone work together.

Name: **Date of Birth:**

Address:

Diagnosis:

Other important health issues:

Medications & Dosage:

Care Team:

GP:

Hospital or Specialist Doctor:

Nurse / Public Health Nurse:

Other Medical (Physio/Palliative Care, etc):

Emergency contact Details:

Symptom Tracker

Month/Year: _____ / _____

Mark one dot per symptom per week to show how you feel compared to normal.

	Week 1	Week 2	Week 3	Week 4
Cough				
Oxygen use				
Breathlessness				
Tiredness				
Diarrhoea				
Nausea				
Mental health				
Other symptoms				
				

Notes:

What makes symptoms better?

What makes symptoms worse?

Daily Living & Support

In this section, consider what you can manage by yourself and what you may need help with. Write down any equipment you use (for example, oxygen or walking aids) and support you have at home (for example, family, carers, and other services).

Emotional, Social & Spiritual Support

In this section, consider your wellbeing needs. Write down people, activities or routines that help you cope.

Planning Ahead

This section helps ensure my wishes are respected.

- Have you talked to someone about your wishes for the future?
  ☐ Yes ☐ No
- Do you have an Advance Healthcare Directive?
  ☐ Yes ☐ No ☐ Not sure
- If you cannot speak for yourself, who should speak for you?
- What matters most to you if your health gets worse:
- Where you would prefer to be cared for, if possible:
- Who to contact first (name and number):
- When to call emergency services:

Reviewing My Plan

This plan started on:

Last review date:

Next review date:

Information for Carers

It can be hard to recognise yourself as a carer, you might feel like you're simply supporting someone important to you. But understanding your role can help you get the right support.

Entitlements

Most carers are unpaid, but some may receive a benefit called Carer's Allowance from the government. The Carer's Allowance is means-tested and so to qualify your household income must be below a certain threshold. If you get Carer's Allowance then you automatically get Carer's Support Grant in June of each year. You might also be eligible for:

- GP Visit Card
- Free Travel
- Household Benefits Package
- Fuel Allowance

More information about the Carer's Allowance can be found on the Citizen's Information website.

Your Role in Patient Care

Everyone's experience of caring is different. Some people find that they become a carer gradually over time, while for others it happens suddenly. Becoming a carer is important, gratifying, and sometimes challenging. It means caring for the physical health and emotional welfare of another person which can be tiring, demanding and sometimes even frightening. Caring can be challenging and it's OK if you decide at any point you want a break or need to stop. It's important to take care of your own health and wellbeing so you can give the best support to the person you're caring for.

As a carer for someone with lung fibrosis you can advocate for them. You can also support their independence, providing assistance where needed in a nurturing, patient, and respectful manner. You can help them maintain a sense of control by ensuring they are included in all decisions related to their care.

You can also listen to their concerns and provide them with help when they request it. You can also encourage them to have a positive outlook and stay active. Consider exercising with them to keep them motivated and focused on keeping well.

It's important to be understanding, keeping in mind that anyone with a progressive disease may be frustrated, sad, or anxious at times.

Here are a few carer tips:

✓ **Stay connected to each other** – while you may now be a carer, the role you played in the patient's life before their diagnosis is still important. You are still a son, a daughter, a spouse, or a friend. It's important to continue to relate to each other in that broader context.

✓ **Mind yourself** – caring can be stressful, and It's difficult to care for others if you're feeling drained or isolated. Tend to your own emotional and physical needs. Take time to meet up with family and friends, to exercise, and to continue hobbies and other activities that support your wellbeing.

✓ **Get help** – asking family or friends to pitch in can help you better manage your loved one's care. It can be as simple as asking for help with the weekly shopping or other errands, organising appointments and helping with transport, preparing meals, dropping in for a visit, or assisting with chores. Speaking with your manager and colleagues can also be supportive if you work while being a carer.

✓ **Everyone feels grief** - it's perfectly normal for you and the person you are caring for to feel a range of emotions including grief. You probably can't avoid these emotions but there are a few techniques you can use that may help when you're feeling low. Consider taking a little time for yourself, sleep, go for a walk, or just sit quietly and reflect. You can also reach out to someone close to you to talk to. Keep to a routine of healthy eating, exercise, and regular sleep as it will help you maintain your physical and mental strength. And if you're feeling overwhelmed by your emotions, counselling might help.

How ILFA Can Help

The Irish Lung Fibrosis Association supports patients and the lung fibrosis community with education, advocacy, research, and a range of services.

Here are just a few of the services we provide to those impacted by this disease.

- Informational webinars
- Patient information and exercise packs
- Daily exercise classes
- Regular mindfulness and resilience courses
- Regional and online support groups for patients and their carers
- Research (ex. virtual pulmonary rehabilitation pilot)
- Nursing advice line
- Grants for psychological counselling
- Pulmonary fibrosis awareness month campaigns
- Training bursaries for medical professionals and patients

If you would like to know more about our services or if you would like to sign up for one of the services we provide, visit our website at **ilfa.ie** to learn more or contact us by email at **info@ilfa.ie** or phone at **+353 (0)1 592 2501**.

Resources Mentioned In this Booklet

- **ILFA Resources:** https://ilfa.ie/information-resources/
- **ILFA YouTube Channel:** https://youtube.com/@irishlungfibrosisassociati1875?si=FLM4rXSwjawUAr4k
- **Mater Hospital Lung Transplant Site:** https://www.lungtransplant.ie/
- **More information on Nintedanib:** https://actionpf.org/portals/0/page-content/documents/resources/68712eea33630ec9d73418fb_Nintedanib%20English%20DIGITAL.pdf
- **More information on pirfenidone:** https://actionpf.org/portals/0/page-content/documents/resources/68712f24ef891fe49b4c09cb_Pirfenidone%20English%20DIGITAL.pdf
- **ILFA's Let Talk Nutrition with Ciara Walsh:** https://www.youtube.com/watch?v=v6m0cv_X03E&t=13s
- **Irish Heart Foundation Food Diary Booklet:** https://irishheart.ie/wp-content/uploads/2018/01/IHF-Food-Diary-2018-A5-V3.pdf
- **The HSE's Living Well Programme** (a six-week HSE self-management programme that helps people live well with their health condition). The programme is delivered by the regional centres. https://www2.hse.ie/living-well/
- **Merlin phone app** (developed by Cornell Lab Ornithology Department) https://merlin.allaboutbirds.org/
- **Family Carers Ireland Emergency Care Plan:** https://www.familycarers.ie/media/r1apwoyj/family-carers-ireland-emergency-plan.pdf
- **Advance Care Directive:** https://www.decisionsupportservice.ie/services/advance-healthcare-directives/making-advance-healthcare-directive
- **Irish Hospice Foundation Care Plan:** https://hospicefoundation.ie/wp-content/uploads/2022/08/Think.Ahead_.Personal.Wishes.and_.Care_.Plan_.pdf
- **Carer's Allowance:** https://www.citizensinformation.ie/en/social-welfare/carers/carers-allowance/
- **Irish Hospice Association Palliative Care Booklet:** https://hospicefoundation.ie/wp-content/uploads/2025/05/Palliative-Care-Booklet-Irish-Hospice-Foundation.pdf
- **ILFA's Let's Talk Palliative Care and Hospice Care Video:** https://youtu.be/WxU4uGd0Hd4

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Contact Us

We are here to help. If you have any questions or would like more information, please get in touch with us.

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Important Note

This booklet is intended for general information purposes only and does not replace the advice of your healthcare team. Always consult your doctor or specialist if you have concerns about your health or treatment.

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Irish Lung Fibrosis Association