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### What is pulmonary fibrosis?

1

### What symptoms do patients with pulmonary fibrosis notice?

1
- Breathlessness
- Cough
- Clubbing
- Anxiety and Depression
- Hypoxia (Low oxygen)

2

### How is Pulmonary Fibrosis diagnosed?

3

### What treatment is available?

4
- Best Supportive Care
- Oxygen Treatment
- Pulmonary Rehabilitation
- Influenza and Pneumonia Vaccinations
- Anxiety and Depression

4

### What will happen in the future?

6

### What happens if it gets worse?

7

### Why have I been referred to palliative care?

7

### How can I help myself?

8
- Diet
- Exercise
- Smoking
- Breathing Techniques and Relaxation
- Home Adaptations and Equipment
- Welfare Benefits

8

### What about getting out and about?

11
- Mobility Aids
- Motoring Benefits
- Going on Holiday
- Travel Insurance
- Travel with Oxygen
- Travel by Car
- Travel by Train
- Travel by Air

11

### What Support is available?

15
- Specialist Nurse
- Support Groups
- Breathe Easy

15
What is pulmonary fibrosis?

Pulmonary fibrosis means scarring of the lungs. In the outer parts of the lungs are very tiny airsacs, called alveoli. These airsacs are the part of the lungs where oxygen is absorbed. In pulmonary fibrosis the airsacs become steadily more damaged through scarring, so that the lungs are less able to absorb the oxygen that the body needs.

Pulmonary fibrosis can be caused by several things. For example, some people who have arthritic conditions; like rheumatoid arthritis or systemic sclerosis may develop pulmonary fibrosis as part of their condition. Some people who have worked in certain occupations, such as with high levels of asbestos dust may develop pulmonary fibrosis; this particular type of pulmonary fibrosis is called asbestosis. Sometimes certain drugs and even birds can also cause other types of pulmonary fibrosis. However, there is a group of people who develop pulmonary fibrosis for who there is no clear cause that can be identified. The word used to describe this type is “idiopathic” and people may therefore be given a diagnosis of idiopathic pulmonary fibrosis (IPF) in these circumstances.

What symptoms do patients with pulmonary fibrosis notice?

Breathlessness
The most common symptom is breathlessness. People with pulmonary fibrosis usually describe steadily worsening breathlessness over time. They may notice that it becomes more difficult to rush or exercise at the start and then find it harder to walk up an incline or climb the stairs. In the early stages many people attribute this to just getting older or being out of shape. If the condition worsens then the level of breathlessness is also likely to get worse and for some people this can mean it is difficult to do everyday things like getting dressed and showering without feeling breathless.

Cough
Seventy five percent of people with pulmonary fibrosis also experience a dry cough. It may feel as though there is sputum (phlegm) to cough up but it is a struggle to do so. Usually, there is no additional sputum present but the nerves in the lung have become over-sensitive and so cause the urge to cough. This cough can be irritating and distressing.

Clubbing
Approximately half of people with pulmonary fibrosis will develop a change in the shape of their finger and toe nails. The ends of their fingers and toes become more rounded and can become more prominent. This is called clubbing. It is not usually painful or uncomfortable but in extreme cases may make it difficult to perform some tasks such as doing up small buttons.

Anxiety and Depression
Some people with pulmonary fibrosis will experience feelings of anxiety and may also become low in mood. For many people there can be uncertainty about what the condition means and what to expect in the future. These uncertainties and their symptoms, which may be worsening, can all contribute towards making anxiety and low mood worse.

Hypoxia (Low oxygen)
If the scarring in the lungs starts to cause the body’s oxygen levels to drop, this can make the work of the heart harder. Over time the heart may develop some strain as a result. If this happens people may notice their ankles swelling and they may also feel more tired than previously.
How is Pulmonary Fibrosis diagnosed?

Your doctor may suspect that there is pulmonary fibrosis present if he or she hears "crackles" when listening to your chest. These tend to be more easily heard at the back and bottom areas of the lungs. Through a stethoscope they sound like velcro or like milk being poured on rice crispies.

It is likely that you will be asked to have a chest X-ray. Usually this shows some signs of scarring but does not give enough detail for the type of pulmonary fibrosis to be diagnosed.

It is very unusual to have pulmonary fibrosis and for there to be no crackles audible when your doctor listens to your chest and a normal chest X-ray. Therefore, if both these are normal this is reassuring.

However, if the chest X-ray suggests there may be pulmonary fibrosis present, then your doctor is likely to refer you to a respiratory specialist for a further examination, pulmonary function tests and earlobe blood gases to assess your breathing pattern and oxygen levels and additional tests.

One of the most useful tests for diagnosing pulmonary fibrosis is a high resolution CT scan (HRCT) of the lungs. This test gives detailed pictures of the lungs and in many cases gives enough information for the type of pulmonary fibrosis to be diagnosed.

Some of the different types of pulmonary fibrosis can have overlapping features on the HRCT scan. Therefore, the scan needs to be interpreted together with the information your specialist has obtained through talking to you, examining you and usually taking blood tests also. Therefore, the final diagnosis about the type of pulmonary fibrosis is usually made at a special multidisciplinary team meeting that includes different experts.

For some people it is not possible to make a clear diagnosis based on the HRCT scan appearance alone. The respiratory specialist may then recommend that you have a lung biopsy, so that the pattern of scarring can be looked at more closely under the microscope. The biopsy is usually performed using keyhole surgery.

Occasionally it may also be necessary to look down into the lungs using a small flexible telescope. This procedure is called a bronchoscopy and is usually done as a day procedure when you are lightly sedated.

What treatment is available?

The answer to this question depends greatly on the final type of pulmonary fibrosis that has been diagnosed. Therefore, treatment recommendations will vary depending on the pattern of scarring, the severity of scarring and whether or not there is an underlying cause. Unfortunately, some types do not have a drug treatment that has been proven to make a difference to the way the disease behaves.

Idiopathic Pulmonary Fibrosis (IPF) is the name given to pulmonary fibrosis that has no identifiable cause. For IPF, there are ongoing clinical trials of treatment for which we are still waiting for the results, so there may be effective treatments available in the future.

For other types of pulmonary fibrosis a careful decision needs to be made weighing up the risks of a treatment against the potential benefits and this should be done together with the respiratory specialist. It is also important to ask about how much benefit a given treatment can be expected to give. For example, some treatments are unable to reverse or improve the scarring process but may slow down the worsening. In such cases, there would not be a noticeable improvement but without the drug the condition could progress more quickly.

Some people with pulmonary fibrosis would benefit from having a lung transplant. There are many considerations to take into account before such a step is made and there is a very detailed assessment process that needs to be completed.

Best Supportive Care

There are some treatments that are useful for most people with pulmonary fibrosis these are often grouped under the heading best supportive care and will be discussed below:

Oxygen Treatment

The body needs oxygen to function. As the scaring in the lungs seen in pulmonary fibrosis affects the transfer of oxygen from the lungs to the blood stream the supply may be reduced in some people. This can result in breathlessness, fatigue, cyanosis (a blue tinge to your lips, fingers) and fluid retention. Giving supplementary oxygen if the oxygen in the blood is low may help with breathlessness and enable people to be more active.
But not all people who are breathless will have low oxygen. This is important because oxygen treatment will not help relieve symptoms unless the blood levels are low. By testing your oxygen saturation with a finger probe and taking an arterial or earlobe blood gas sample, your doctor is able to determine if you may benefit from supplementary oxygen and refer you for a formal oxygen assessment. Some people may require oxygen all the time (Long term oxygen therapy LTOT) while others only during activity (Ambulatory). If you need oxygen during activity, you will have an additional ambulatory test. Your oxygen saturation will be measured while you walk for 6 minutes without supplemental oxygen and then for 6 minutes with supplemental oxygen. This allows us to assess if you will benefit from wearing oxygen during activity and how much to give you. People with Pulmonary Fibrosis may benefit from ambulatory oxygen before LTOT is required. It is therefore important that you advise your doctor of any significant changes in your ability to carry out everyday activities so this can be assessed. It is also not unusual for people to require a higher flow rate when they are active than when they are resting.

At the end of the assessment, if you need oxygen, the assessor will discuss the best mode of delivery with you, demonstrate the chosen equipment, and order your oxygen to be delivered to your home. Once it is installed, they will then usually complete a safety visit to your home.

**Pulmonary Rehabilitation**

Having a lung condition can result in a gradual decrease in activity and fitness resulting in you becoming deconditioned and able to do less physically. Pulmonary rehabilitation programmes are designed specifically for people with long-term lung conditions to help you be as active and functional in your daily life as possible. It is particularly useful in helping people who find it hard to do the things they would like to because of breathlessness or tiredness in their arms and legs. The content of programmes may vary but most contain an exercise program, breathing retraining, education, nutrition, and emotional support.

The most important component is a supervised exercise programme to improve endurance and strength. Many people worry as they may not have exercised for years but after an initial assessment the programme is personalised and designed to meet your individual needs and ability. At the end of the programme, people are generally able to walk three or four times further than they could at the beginning.

Being breathless can also be frightening, people are often afraid to be active to avoid this feeling. The programme will also help you deal with anxious feelings you experience when you feel breathless that can make the sensation of breathlessness feel worse than it needs to give you more control.

**Influenza and Pneumonia Vaccinations**

Pulmonary fibrosis weakens the lungs’ defences against infections such as pneumonia and influenza. It is recommended that people with pulmonary fibrosis have their vaccinations kept up to date.

**Anxiety and Depression**

We know that people with pulmonary fibrosis can become anxious or depressed. There are effective treatments available. Your doctor can advise about the need for such treatment.

**What will happen in the future?**

For many people with pulmonary fibrosis the condition gets worse with time. The rate at which this happens can be very variable and depends upon the type of pulmonary fibrosis and the response to any treatment that has been given. Unfortunately, for some people with pulmonary fibrosis, the condition will eventually shorten their life.

At the moment, it is not possible to give people detailed and accurate information about how their pulmonary fibrosis will behave and what will happen to them specifically in the future. For some, the fibrosis stays stable, without getting worse for a long period. However, for other people, the fibrosis progresses steadily over a few months or years. This is best individually monitored by and discussed with your specialist.

Sometimes, people with pulmonary fibrosis experience a sudden worsening of their condition. This may mean worsening breathlessness or cough. This may be caused by an infection in the lungs or actual worsening of the pulmonary fibrosis. There are also other causes for such a deterioration and so medical help is usually needed. Such episodes can occur unexpectedly and are called exacerbations.
What happens if it gets worse?

Measuring your lung function and oxygen levels during visits to the hospital clinic will give more accurate information about whether the condition is stable or the pulmonary fibrosis is progressing. Most people will also notice that their breathlessness is getting worse. However, sometimes, there are other reasons for worsening breathlessness so additional tests may be needed to determine this.

A change in treatment may be required, although this will depend upon the type of pulmonary fibrosis and the cause of the worsening symptoms. This should be discussed with the hospital specialist.

Even though we may be unable to offer a curative treatment we will be able to help relieve uncomfortable symptoms such as cough and breathlessness. This may be done by your specialist alone or in conjunction with palliative care services.

Why have I been referred to palliative care?

Traditionally you may associate palliative care with the treatment of cancer and wonder why your doctor has suggested a referral to them. The palliative care team are experts in helping relieve symptoms. In addition they can also provide support to both people with pulmonary fibrosis and their carers to help them manage the changes that are taking place. They have a well established team of doctors and nurses based in the community and local hospices.

A referral may be made to help with a specific short term problem or more general ongoing support. The level of help they provide will vary according to your needs. For example their help may be useful early on when you have just been diagnosed and are trying to come to terms with the information you have been given, for symptom control such as breathlessness, providing support for patients waiting for lung transplantation and more traditionally for end of life care as pulmonary fibrosis progresses.

How can I help myself?

There are a number of things you can do yourself to stay healthy.

**Diet**

Your body is working harder and you use more calories completing basic activities than people who do not have lung disease. Eating a balanced healthy diet that contains adequate calories is essential and will provide the nutrition you need to stay healthy.

**Underweight**

Being breathless can make it difficult to eat and reduce your appetite even though you need a high calorie intake. This combination can result in loss of weight. If you need help your doctor will be able to refer you to a dietitian for support and advice. There are however some simple steps you can take to help yourself.

- Try and sit at the table to eat your meals where possible
- Small more frequent meals are easier to manage. Avoid overloading your plate it can be very off putting.
- Chose foods that are easy to chew and digest.
- Avoid drinking prior to your meal as this will fill you up
- Supplement your diet with small nutritious snacks
- Fortify your diet with extra calories by for example adding cream or cheese to dishes or using full cream milk.
- If you are still unable to maintain your weight supplement drinks can be prescribed by your doctor but this should be the last resort.

**Overweight**

Some people have the opposite problem and can gain weight. This can be made worse by some drug treatments such as steroids which increase the appetite. Carrying extra weight can impact on your general fitness and if it is distributed mainly around your middle it can put extra pressure on your lungs and make it more difficult to breathe. Unfortunately there are times when treatment with steroid tablets is necessary but talk to your doctor if this is causing you to gain weight. Your doctor will also be able to refer you to a dietitian for help with loosing weight.
Exercise
People with lung disease often limit exercise to avoid breathlessness. As a consequence inactivity weakens the muscles and they become less efficient which can make basic daily activity, such as washing and dressing, more difficult. Staying in shape can help minimise this. Most people with lung disease can exercise safely and should endeavour to do so but if you have any concerns discuss them with your doctor before you start. In addition, if your oxygen level drops during exercise and you require supplementary ambulatory oxygen your doctor can arrange this if necessary. Your GP can refer you to a community based exercise scheme there are a number designed specifically for people with health problems at local leisure centres. However if you have not exercised for some time you may prefer to exercise under medical supervision. Ask your doctor to consider referring you to a pulmonary rehabilitation programme.

In addition rethinking the way you do things may make the task easier, more efficient, use less energy and reduce the resultant feeling of breathlessness. Indeed some tasks may not be necessary at all! These changes are commonly grouped under headings known as the 5 P’s: Prioritising; Planning; Pacing; Positioning and Permission to do things differently or not at all.

Stop Smoking
If you are still smoking the most important thing you can do is stop. This can help prevent further damage to your lungs. Having pulmonary fibrosis does not mean you are not at risk of developing other diseases such as COPD or lung cancer. In addition you will not be eligible for lung transplantation if this is a treatment option for you if you continue to smoke. Tobacco is however very addictive which can make stopping difficult. Your doctor will be able to refer you to a local smoking cessation service for support.

Breathing Techniques and Relaxation
Breathing techniques and relaxation can help you cope with the panic of breathlessness when it occurs. There are a number of different techniques that can be utilised. The best one is the one that works for you and you can remember when you need it most. It is important to practice your chosen technique when you are relaxed and calm so it becomes second nature when needed. Your Respiratory Nurse or Specialist Respiratory Team will be able to help find the best one for you. Ask your doctor for further information.

Home Adaptations and Equipment
Small changes to your home such as additional hand rails on the stairs and in bathrooms, shower and bath aids, toilet aids, perching stools, stair lifts and ramps can make it easier to remain independent and cope with everyday activity. Many of these are available free of charge following assessment by an occupational therapist. Although dependent on personal circumstances there may be a cost contribution for bigger items such as stair lifts. Your doctor or nurse will be able to refer you for an assessment.

Welfare Benefits
Living with lung disease can impact on your ability to work limiting your income or cause additional costs which may bring about financial hardship. There are a range of benefits such as disability living allowance, attendance allowance, carers allowance that you may be entitled to if you cannot work, have care and mobility needs or are caring for someone with a lung condition.

Understanding what you are eligible for and completing the application can be bewildering. www.gov.uk/browse/benefits/disability is a good source of information as are The British Lung Foundation, Age UK and Citizens Advice. The latter in addition all provide individual advice, help and support with applications. Some palliative care teams have a welfare officer. Your GP or specialist may also be able to help.

w w w.ageuk.org.uk H elpline 0800 169 6565
w w w.blf.org.uk H elpline 03000 030 555
w w w.citizensadvice.org.uk H elpline 08444 772020
What about getting out and about?

There is no need to put your life on hold because you have a chronic lung disease. The impact the disease has on you and your quality of life can to a degree be influenced by you and your determination to continue to do the things you love that make life worth living. Whether that is a round of golf, gardening, participating in groups, family events or holidays most activities can continue with some adjustment and planning.

If you are receiving long term oxygen therapy or ambulatory oxygen therapy you should still be able to participate in your chosen activity. There are a range of different modes of delivery available. Your local oxygen service will be able to assess which is the best for you and your particular activity needs.

Mobility Aids

Having a wheel chair may allow you to continue to go out with assistance if your mobility is severely restricted by breathlessness. Wheel chairs and other equipment can be loaned from the British Red Cross for short term use. They have a number of offices. Contact www.redcross.org.uk/Where-we-work/In-the-UK/Wales-and-Western-England/Wales/LocalServices/Medical-equipment for your nearest equipment store. For longer term needs your doctor or nurse can arrange an assessment and provide one for you.

Mobility scooters are a more independent form of assisted mobility but are not available on the NHS. You may however be able to use your disability living allowance to fund one if you are receiving it.

Motoring Benefits

There are a number of schemes and concessions you may be eligible for if you have a disability that makes it difficult to walk and get around. This includes severe breathlessness. You will only be eligible for some if you are in receipt of disability living allowance others are more widely available and may be run by local councils. Some examples are the blue badge scheme, motability, tax disc concession, toll concession installation of dropped curbs. Further information and links to apply are available at www.gov.uk/driving-medical-conditions/blue-badge-and-motability

Going on Holiday

Holidays are important. Most people can travel somewhere but you may need to consider a location with few hills, good transport links and a hotel close to amenities. Think about the facilities at your chosen destination. Are there lots of steps? Is there a lift? Are facilities accessible? Is disabled access good if you need it? What’s the weather like too hot or too cold? How will I get there? Can I fly? Can I travel with my oxygen? What if I’m unwell on holiday? Can I get Travel insurance?

There is lots of advice available the key is planning. The British Lung Foundation web site provides advice for people travelling with lung disease. Download their holiday check list. In addition you can discuss any specific concerns with your doctor or nurse. A good travel agent and personal recommendations can be invaluable. www.lunguk.org/you-and-your-lungs/living-with-a-lung-condition/going-on-holiday-with-a-lung-condition

Tel: 03000030555

Travel Insurance

Medical costs within the EU are covered for UK citizens with a European Health Insurance Card. This is available from www.dh.gov.uk/travellers or telephone 08456062030. However you will still need to purchase travel insurance in case you are taken ill and need to be flown back and for travel to other parts of the world.

Finding reasonably priced travel insurance can cause difficulty. As this is constantly changing it is not possible to be specific in this booklet. The advice is to shop around and start early. The British Lung Foundation and Age UK are often a good starting point. It is important to be honest in your declaration of current health problems as a failure to do so could affect your cover if you become unwell and require medical attention on holiday.
Travel with Oxygen

One of people’s greatest concerns is being able to continue to travel, stay with friends or family at home or abroad with oxygen. Both are possible but will require thought and planning. It may be advisable to plan your first holiday in the UK so you are able to become more aware of your travel needs and where it is easier to deal with problems if they occur. This will help develop your confidence before planning further travel. If you have any concerns contact your oxygen service for advice.

Your current oxygen provider will be able to liaise with the local supplier to deliver oxygen to meet your current prescription to your chosen holiday accommodation. Remember to check that your destination will accept oxygen on the premises. If you currently have liquid oxygen this may not be available at your holiday destination or for short breaks. In such cases it will be substituted with conventional cylinders. When you have finalised your travel plans contact your supplier with your holiday dates, the address, a contact name and number and your booking reference if applicable. The company will require sufficient notice of your travel plans usually two-four weeks. This will vary between companies.

The oxygen supply companies do not advise taking supplied equipment out of the country as they are unable to provide support services or replenish supplies. Oxygen can be ordered for your holiday destination abroad but there will be a charge. Within the European Union this can be claimed back on your return to the UK provided oxygen is arranged through an approved route and you keep receipts. Oxygen can also to arranged for cruise holidays a good alternative for those who prefer not to fly.

- **Cruise: contact Cruise:** Omega Advanced Aeromedical
  Email: info@omegaoxygen.com
  Tel: 01273 308176
- **Destination Country:** Oxygen can be ordered for the country of destination - contact www.nhs.uk/healthcareboard then click on country of destination to see how to arrange oxygen.
- **EU Cost Reimbursement:** contact The Department of Work and Pensions, Tel: 0191 218 1999
- **Outside Europe:** For oxygen outside Europe you will need to contact local oxygen contractor.

Travel by Car

Give yourself plenty of time and break your journey into short manageable sections. If sat for long periods of time it is advisable to complete simple leg exercises to maintain a good circulation get out and walk around when comfort breaks allow. If you require oxygen ensure you have more than you need for the journey in case of unplanned traffic delays or problems that extend the length of your journey. You are able to take medical oxygen in your car your oxygen supplier will provide advice on safe transportation. If travelling to Europe you can take medical oxygen in your car if using the channel tunnel.

Travel by Train

This provides a good option for those who are unable to fly or want to avoid a long car journey. There are excellent train services in Europe and a number of good web sites to help you plan. You are able to take medical oxygen if you travel to Europe via the channel tunnel.

Travel by Air

If you are planning to fly discuss this with your doctor as soon as possible. If your oxygen saturation is below 95% your doctor may refer you for a flight test. This is because there is less oxygen available in an aircraft cabin than on the ground which could result in breathlessness and distress during a flight. The test involves breathing oxygen via a mask that simulates the conditions you will experience in the cabin and testing the level of oxygen in your blood. If this is lower than recommended your doctor will advise you how much oxygen you need during the flight. The airline will require a fitness to fly letter which your doctor can provide.

You are then able to order in flight oxygen from the airline however this service varies between airlines, it has to be booked in advance and may carry an additional charge. There may also be restrictions on the number of people with oxygen carried on a single flight and the prescription available. Always plan ahead, contact the airlines directly and explain your needs before making your booking. Further information is available on the British Lung Foundation and European Lung Foundation websites but is constantly changing so check with the airline you plan to travel with. There is no oxygen available in the airport building you will need to take your own if needed pre-flight. Airports will provide assistance to and from the plane if you contact them in advance and advise them of your needs. Ensure you take your medication and inhalers in your hand luggage you may need them during the flight.
It is also advisable to carry a letter from your doctor detailing your illness and confirming your medication. Prevent in-flight dehydration by drinking plenty of fluids and avoiding alcohol. Move around the cabin and complete frequent in seat exercises as sitting in one place can lead to blood clots in the leg.

**In Flight Oxygen:** contact [http://www.european-lung-foundation.org/7029-airline-index-page.htm](http://www.european-lung-foundation.org/7029-airline-index-page.htm) or [www.blf.org.uk/Page/Airline-oxygen-policies](http://www.blf.org.uk/Page/Airline-oxygen-policies) then contact the airline directly.

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**What Support is available?**

**Specialist Nurse**
Access to a Respiratory Nurse Specialist varies. They provide a service which may include patient assessment, specific disease monitoring, oxygen assessment, advice, support, welfare advice, home assessment, support groups and referral to other services and specialities. Referral can be discussed with your hospital specialist.

**Support Groups**
Support groups provide an opportunity to meet and talk to other people who understand the daily challenges of living with pulmonary fibrosis and access useful information and advice. Your specialist will be able to advise you of groups in your area.

**Current support groups in Wales**

**Pulmonary Fibrosis Wales**
A group set up and run by patients and their families to raise public awareness of the interstitial lung diseases that cause pulmonary fibrosis and provide support to people and their families. The group meets in the evening on the first Tuesday of every month in the Hilton Hotel in Newport which is just off the M4.
Tel: 01633 411380  Email: info@pulmonaryfibrosiswales.co.uk

**Llandough Hospital Pulmonary Fibrosis Patient Support Group**
A support group for patients under the care of Cardiff & Vale University Health Board. The group meets bi-monthly on the last Thursday of the month 1.30pm - 3pm at Llandough Hospital.
Julie Hocking Respiratory Nurse Specialist (ILD)
Tel: 02920716419  Email: Julie.hocking@wales.nhs.uk

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**Breathe Easy**
These are British Lung Foundation patient run support groups. They welcome people with all chronic lung diseases but are not specific to pulmonary fibrosis. For your nearest group contact the British Lung Foundation website.
[www.blf.org.uk](http://www.blf.org.uk)