

# Idiopathic pulmonary fibrosis

Information for the public

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## About this information

NICE clinical guidelines advise the NHS on caring for people with specific conditions or diseases and the treatments they should receive. The information applies to people using the NHS in England and Wales.

This information explains the advice about a lung condition called idiopathic pulmonary fibrosis that is set out in NICE clinical guideline 163.

All of the treatment and care that NICE recommends is in line with the NHS Constitution (<https://www.gov.uk/government/publications/the-nhs-constitution-for-england>). NICE has also produced advice on improving the experience of care for adults using the NHS. For more information, see 'About care in the NHS' on our website ([www.nice.org.uk/nhscare](http://www.nice.org.uk/nhscare)).

## Does this information apply to me?

Yes, if you are 18 years or older and have suspected or diagnosed idiopathic pulmonary fibrosis.

It does not cover other types of pulmonary fibrosis caused by illnesses such as rheumatoid arthritis, or if you have been exposed to a substance that is known to cause pulmonary fibrosis, such as asbestos.

## Idiopathic pulmonary fibrosis

Pulmonary fibrosis is a gradual scarring of the lungs that gets worse over time. 'Idiopathic' means that the cause of the scarring is not known. Idiopathic pulmonary fibrosis used to be known as 'cryptogenic fibrosing alveolitis'.

The scarring makes it more difficult for the lungs to take in oxygen, which can make you more breathless after normal everyday activities, such as walking up the stairs. You may have a cough that does not go away. Over time, as the scarring worsens, the lungs are unable to work properly, making everyday activities more difficult.

At the moment, there is no cure for idiopathic pulmonary fibrosis, but there are treatments that can help lessen the symptoms of the disease.

## Your healthcare team

The various types of tests and treatment described may be provided by a range of healthcare professionals. These could include radiologists (who specialise in interpreting scans and X-rays), lung technicians and surgeons who will help find out what is wrong and specialist doctors, specialist lung nurses and physiotherapists who will work with you to manage your condition.

A member of your healthcare team should discuss idiopathic pulmonary fibrosis with you and explain the tests and treatments for it in detail. You should have the opportunity to ask any questions you have – there are lists of the questions you might like to ask in each section.

**Some treatments described may not be suitable for you, depending on your exact circumstances. If you think that your treatment or care does not match this advice, talk to your healthcare team.**

## Knowing the signs of idiopathic

# pulmonary fibrosis

Idiopathic pulmonary fibrosis is not very common and can be confused with other lung problems. Because of this, NICE has said that it is important for healthcare professionals to know the signs of the condition when they are assessing you. Idiopathic pulmonary fibrosis is more common in people over 45. Not everyone has symptoms but if you notice the following it might suggest you have the disease:

- feeling unusually breathless after normal everyday activities, such as walking up the stairs
- a cough that does not go away or prolonged bouts of coughing
- changes in the finger tips and nails.

Your doctor may also notice the following:

- a crackling sound when they listen to your chest
- test results showing that the lungs are not working as well as they could be.

## Finding out what's wrong

Idiopathic pulmonary fibrosis is difficult to diagnose. You will need to see a specialist doctor who will work closely with radiologists (doctors who specialise in interpreting scans and X-rays) and specialist lung nurses to diagnose the cause of your symptoms. This may give you a 'confident diagnosis' of idiopathic pulmonary fibrosis or identify a different condition.

A 'confident diagnosis' means that your doctor thinks that it is highly likely that you have idiopathic pulmonary fibrosis. Your doctor should discuss the possible benefits of having a 'confident diagnosis' with you and how it could affect your future care. Your specialist doctor should first ask you questions to see if something else could be causing your symptoms. Your doctor will also want to know if you have any other symptoms and about the medicines you have taken. The doctor should look at the results of any recent chest X-rays you have had.

Your specialist doctor will examine you and do tests called spirometry and gas transfer to see how well your lungs are working.

Your specialist doctor may also offer you a type of X-ray known as a CT scan.

## If you need more tests

For some people a 'confident diagnosis' can be made after their doctor has asked them questions and they have had a CT scan. If your specialist doctor can't say for certain that you have idiopathic pulmonary fibrosis based on the information they have, they may suggest further tests. Before you have any of these tests your doctor should discuss the risks and benefits of each of them with you (you may find the list of questions below helpful). They should bear in mind your general state of health and think about what other conditions could be causing your symptoms.

Further tests could include:

- Bronchoscopy – where a narrow, flexible tube containing a small video camera is put down the windpipe and used to look into the lung. During a bronchoscopy tiny pincers or a fine needle at the end of the tube might be used to remove a sample of tissue or the airways might be 'washed' to collect lung cells for examination.
- Lung biopsy – a short surgical procedure that involves removing a small piece of lung tissue, through a small cut in the side of the chest. It is more likely to give you a 'confident diagnosis' than the other tests but there are increased risks because a general anaesthetic is needed. Your healthcare team can explain more about the benefits and risks of lung biopsy.

### Questions you might want to ask

- Can you tell me more about the tests you've offered me?
- What do these tests involve?
- What are the risks and benefits of these tests?
- Will I need an anaesthetic? If so, what type?
- Where will these be carried out? Will I need to have them in hospital?
- How long will I have to wait until I have these tests?

- How long will it take to get the results?
- How important is it to have a confident diagnosis?
- How will having a confident diagnosis affect me and my future care?

## Information and support

Your specialist doctor or the specialist lung nurse should give you (and your family and carers with your permission) accurate and clear verbal and written information about idiopathic pulmonary fibrosis. This should include information about the different tests, your diagnosis and how your condition will be managed. You will have the opportunity to talk about how your idiopathic pulmonary fibrosis is progressing and how severe your disease is. Within 6 months of you being diagnosed with idiopathic pulmonary fibrosis, your doctor should also offer you information about [ventilation](#) and [lung transplantation](#).

A specialist lung nurse should be available to provide information and support to you and your family and carers throughout your illness.

If you smoke, you should also be given advice, treatment and support to help you stop smoking. See [Other NICE guidance](#) for details of our guidance on smoking.

### Questions you might like to ask

- Can you tell me more about idiopathic pulmonary fibrosis?
- Can you tell me more about how my disease is progressing?
- Would it help my condition if I made some changes to my lifestyle?
- Are there any support organisations in my local area?
- Can you recommend any good sources of information about idiopathic pulmonary fibrosis?
- Can you provide any information for my family/carers?

- Can you give me the contact details of a specialist lung nurse who I can contact if I have any questions?
- Is there any additional support that I might benefit from or be entitled to?

## Questions your family or carers might like to ask

- What can I/we do to help and support the person with idiopathic pulmonary fibrosis?
- Is there any additional support that I/we as carer(s) might benefit from or be entitled to?

# How quickly does idiopathic pulmonary fibrosis progress?

The progress of idiopathic pulmonary fibrosis can vary greatly from person to person. This makes it difficult for doctors to predict how fast symptoms may progress and change. You should have repeat spirometry and gas transfer tests 6 and 12 months after diagnosis. You may be invited for more regular tests if there is concern that your idiopathic pulmonary fibrosis is getting worse more quickly than expected.

## Questions you might like to ask

- How severe is my idiopathic pulmonary fibrosis? How quickly is it progressing?
- Are there any options for treatment of my disease?
- How often will I need to come back for tests and appointments?
- How can my illness be managed?

# Easing your symptoms, including breathlessness and cough

As soon as you are diagnosed you should be offered care to lessen the symptoms of idiopathic pulmonary fibrosis and support you through your illness. The care should be tailored to how severe your illness is, how quickly it is progressing and your personal preferences. It may also include treating any other illnesses you may have and stopping treatments that are not working or causing side effects.

## If you are breathless

If you get out of breath your specialist doctor may recommend that you are assessed to see if oxygen could help you. You should be assessed to see if you would benefit most from oxygen to use while resting at home, or a portable supply for everyday activities.

If you become unusually breathless after normal everyday activities, such as walking up the stairs, you may be assessed for pulmonary rehabilitation (see [pulmonary rehabilitation](#)).

If you feel out of breath even when you are resting, your specialist doctor may ask if you need any extra support and discuss how drug treatment could help.

If you have to stay in hospital because of your idiopathic pulmonary fibrosis, your specialist team should check if you will need extra oxygen when you get home.

## If you have a cough

If you have a cough, your specialist doctor may first see if something else could be causing the cough and treat this. If your cough is making you weak and making everyday life difficult, your doctor may suggest treating it with opioids because they can help reduce coughing. If your cough is not responding to any other treatment, your specialist doctor may see if treatment with other drugs could help.

# Pulmonary rehabilitation

Pulmonary rehabilitation sessions teach you about how your lungs work and how to cope

with symptoms such as breathlessness so you can get the most out of life.

When you are diagnosed with idiopathic pulmonary fibrosis, your specialist doctor should offer you assessment to see if pulmonary rehabilitation could help you. Your doctor should offer to repeat the assessments every 6 or 12 months.

If pulmonary rehabilitation could help, you should be able to attend a session designed for people with idiopathic pulmonary fibrosis and tailored to your needs. The sessions should be a mixture of advice and exercise classes. The sessions should be easy for you to get to, even if you have a disability.

## Questions you might like to ask

- Can you tell me why you have decided to offer me this particular type of treatment?
- What will it involve?
- How will it help me? What effect will it have on my symptoms and everyday life? What sort of improvements might I expect?
- How long will it take to have an effect?
- Are there any risks associated with this treatment?
- Is there some other information (like a leaflet, DVD or a website I can go to) about the treatment that I can have?
- What will happen if I choose not to have the treatment you have offered?

## Drugs for treating idiopathic pulmonary fibrosis

There are no drugs that can cure idiopathic pulmonary fibrosis, but there are drugs used to help with the symptoms, although these are not suitable for everyone. Some people with idiopathic pulmonary fibrosis use N-acetylcysteine to help their condition, although it hasn't yet been scientifically proven to work. Pirfenidone is a possible treatment for some

people with idiopathic pulmonary fibrosis. See [Other NICE guidance](#) for details of our guidance on pirfenidone.

Your doctor should not offer any of the following drugs to treat your idiopathic pulmonary fibrosis: ambrisentan, azathioprine, bosentan, co-trimoxazole, mycophenolate mofetil, prednisolone, sildenafil and warfarin. However, your doctor might use some of these drugs to treat other illnesses you may have. If you are already taking prednisolone or azathioprine your doctor should talk to you about the potential risks and benefits of stopping, staying on or changing your treatment.

## Questions you might like to ask

- Why are there no drug treatments for idiopathic pulmonary fibrosis?
- Are there any drugs in development that could treat idiopathic pulmonary fibrosis effectively?

## Lung transplantation

Your specialist doctor and a specialist lung nurse should discuss lung transplantation with you within 6 months of you being diagnosed, unless it is not suitable for you. If you wish, you should be able to invite your family and carers to join these discussions.

If lung transplantation may be suitable for you and you would like to explore the possibility further after talking to the doctor and nurse, your specialist doctor should contact the transplant centre.

## Questions you might like to ask

- Can you explain why a lung transplant is not a suitable treatment for me?
- What are the risks and benefits of lung transplantation?
- When will I hear back about my transplant centre referral?

# Your regular hospital appointments

How often you need to have follow-up appointments will depend on your disease. If your disease is progressing quickly or your symptoms are rapidly getting worse you may be seen at least every 3 months. If your disease is progressing more slowly you may be seen at least every 6 months. If your disease is stable you should be seen every 6 months. If your illness remains stable after a year, you will then be invited back for yearly appointments.

Your follow-up appointments should include:

- having tests to see how well your lungs are working
- being assessed for oxygen therapy
- seeing if you need pulmonary rehabilitation
- advice on stopping smoking, if you need it
- identifying occasions when your idiopathic pulmonary fibrosis got worse and when you needed treatment for it in hospital
- checking if you would like to be assessed for lung transplantation if it is suitable for you (see [lung transplantation](#))
- asking about whether you need any extra support
- assessing you for related illnesses (which may include anxiety, depression, diabetes, heart problems, high blood pressure, indigestion, lung cancer and widening of the airways)
- discussing referral to palliative care services if you have worsening idiopathic pulmonary fibrosis.

## Questions you might like to ask

- How often will I need to come back for appointments?
- Who should I contact if my condition worsens?

- What services are offered by the palliative care team?

## Care at the end of life

If your idiopathic pulmonary fibrosis is advanced you should also be offered end of life care. End of life care helps people live as well as possible at the end of their life.

You and your family and carers should have access to the full range of services offered by the palliative care team.

## Ventilation

Mechanical ventilation is a type of life support where a machine helps a person breathe when their body is unable to. Mechanical ventilation is unsuitable for treating idiopathic pulmonary fibrosis. Because of this, mechanical ventilation should not usually be offered to people with idiopathic pulmonary fibrosis who develop life-threatening breathing problems.

### Questions you might like to ask

- How can I and my family and carers access palliative care services?
- Why is mechanical ventilation unsuitable for people with idiopathic pulmonary fibrosis?
- What other options are there for treating life-threatening breathing problems?

## Sources of advice and support

- British Lung Foundation, 03000 030 555  
[www.blf.org.uk](http://www.blf.org.uk)

- Pulmonary Fibrosis Trust, 0333 2020991  
[www.pulmonaryfibrosistrust.org](http://www.pulmonaryfibrosistrust.org)
- Pulmonary Fibrosis Wales, 01633 411380  
[www.pulmonaryfibrosiswales.co.uk](http://www.pulmonaryfibrosiswales.co.uk)

You can also go to NHS Choices ([www.nhs.uk](http://www.nhs.uk)) for more information.

NICE is not responsible for the quality or accuracy of any information or advice provided by these organisations.

## Other NICE guidance

- Pirfenidone for treating idiopathic pulmonary fibrosis. NICE technology appraisal guidance 282 (2013). See <http://guidance.nice.org.uk/TA282>
- Smoking cessation services. NICE public health guidance 10 (2007). See <http://guidance.nice.org.uk/PH10>

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## Accreditation

