

# Idiopathic pulmonary fibrosis

## The diagnosis and management of suspected idiopathic pulmonary fibrosis

Issued: June 2013

**NICE clinical guideline 163**

[guidance.nice.org.uk/cg163](http://guidance.nice.org.uk/cg163)

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## Introduction

Idiopathic pulmonary fibrosis is a chronic, progressive fibrotic interstitial lung disease of unknown origin. It is a difficult disease to diagnose and often requires the collaborative expertise of a consultant respiratory physician, radiologist and histopathologist to reach a consensus diagnosis. Most people with idiopathic pulmonary fibrosis experience symptoms of breathlessness, which may initially be only on exertion. Cough, with or without sputum, is a common symptom. Over time, these symptoms are associated with a decline in lung function, reduced quality of life and ultimately death.

The median survival for people with idiopathic pulmonary fibrosis in the UK is approximately 3 years from the time of diagnosis. However, about 20% of people with the disease survive for more than 5 years. The rate of disease progression can vary greatly. A person's prognosis is difficult to estimate at the time of diagnosis and may only become apparent after a period of careful follow-up.

This guideline contains recommendations on the diagnosis of idiopathic pulmonary fibrosis and delivery of care to people with idiopathic pulmonary fibrosis, from initial suspicion of the disease and referral to a consultant respiratory physician, to best supportive care and disease-modifying treatments.

The guideline will assume that prescribers will use a drug's summary of product characteristics to inform decisions made with individual patients.

This guideline recommends some drugs for indications for which they do not have a UK marketing authorisation at the date of publication, if there is good evidence to support that use. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. The patient (or those with authority to give consent on their behalf) should provide informed consent, which should be documented. See the General Medical Council's [Good practice in prescribing and managing medicines and devices](#) for further information. Where recommendations have been made for the use of drugs outside their licensed indications ('off-label use'), these drugs are marked with a footnote in the recommendations.

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## Patient-centred care

This guideline offers best practice advice on the care of people with idiopathic pulmonary fibrosis.

Patients and healthcare professionals have rights and responsibilities as set out in the [NHS Constitution for England](#) – all NICE guidance is written to reflect these. Treatment and care should take into account individual needs and preferences. Patients should have the opportunity to make informed decisions about their care and treatment, in partnership with their healthcare professionals. If someone does not have the capacity to make decisions, healthcare professionals should follow the [Department of Health's advice on consent](#) and the [code of practice that accompanies the Mental Capacity Act](#) and the supplementary [code of practice on deprivation of liberty safeguards](#). In Wales, healthcare professionals should follow [advice on consent from the Welsh Government](#).

NICE has produced guidance on the components of good patient experience in adult NHS services. All healthcare professionals should follow the recommendations in [Patient experience in adult NHS services](#).

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## Key priorities for implementation

The following recommendations have been identified as priorities for implementation.

### ***Awareness of clinical features of idiopathic pulmonary fibrosis***

- Be aware of idiopathic pulmonary fibrosis when assessing a patient with the clinical features listed below and when considering requesting a chest X-ray or referring to a specialist:
  - age over 45 years
  - persistent breathlessness on exertion
  - persistent cough
  - bilateral inspiratory crackles when listening to the chest
  - clubbing of the fingers
  - normal spirometry or impaired spirometry usually with a restrictive pattern but sometimes with an obstructive pattern.

### ***Diagnosis***

- Diagnose idiopathic pulmonary fibrosis only with the consensus of the multidisciplinary team (listed in table 1), based on:
  - the clinical features, lung function and radiological findings (see [recommendation 1.2.1](#))
  - pathology when indicated (see [recommendation 1.2.4](#)).

#### **Table 1 Minimum composition of multidisciplinary team involved in diagnosing idiopathic pulmonary fibrosis**

Stage of diagnostic care pathway	Multidisciplinary team composition (all healthcare professionals should have expertise in interstitial lung disease)
After clinical evaluation, baseline lung function and CT	Consultant respiratory physician Consultant radiologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering performing bronchoalveolar lavage, and/or transbronchial biopsy or surgical lung biopsy  Only some patients will have bronchoalveolar lavage or transbronchial biopsy but they may be being considered for surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant histopathologist Thoracic surgeon as appropriate Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering results of bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant histopathologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
See chapter 6.5 (Multidisciplinary Team) in <a href="#">full guideline</a> for more information on the expertise of the multidisciplinary team.	

## Information and support

- The consultant respiratory physician or interstitial lung disease specialist nurse should provide accurate and clear information (verbal and written) to people with idiopathic pulmonary fibrosis, and their families and carers with the person's consent. This should include information about investigations, diagnosis and management.
- An interstitial lung disease specialist nurse should be available at all stages of the care pathway to provide information and support to people with idiopathic pulmonary fibrosis and their families and carers with the person's consent.

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## ***Pulmonary rehabilitation***

- Assess people with idiopathic pulmonary fibrosis for pulmonary rehabilitation at the time of diagnosis. Assessment may include a 6-minute walk test (distance walked and oxygen saturation measured by pulse oximetry) and a quality-of-life assessment.

## ***Best supportive care***

- Offer best supportive care to people with idiopathic pulmonary fibrosis from the point of diagnosis. Best supportive care should be tailored to disease severity, rate of progression, and the person's preference, and should include if appropriate:
  - information and support (see [recommendation 1.3.1](#))
  - symptom relief
  - management of comorbidities
  - withdrawal of therapies suspected to be ineffective or causing harm
  - end of life care.
- If the person is breathless on exertion consider assessment for:
  - the causes of breathlessness and degree of hypoxia **and**
  - ambulatory oxygen therapy and long-term oxygen therapy **and/or**
  - pulmonary rehabilitation.

## ***Disease-modifying pharmacological interventions***

- For guidance on pirfenidone for the management of idiopathic pulmonary fibrosis, refer to [Pirfenidone for the treatment of idiopathic pulmonary fibrosis](#) (NICE technology appraisal guidance 282).
- Do not use any of the drugs below, either alone or in combination, to modify disease progression in idiopathic pulmonary fibrosis:
  - ambrisentan



- 
- azathioprine
  - bosentan
  - co-trimoxazole
  - mycophenolate mofetil
  - prednisolone
  - sildenafil
  - warfarin.

## ***Lung transplantation***

- Refer people with idiopathic pulmonary fibrosis for lung transplantation assessment if they wish to explore lung transplantation and if there are no absolute contraindications. Ask the transplant centre for an initial response within 4 weeks.

## ***Review and follow-up***

- In follow-up appointments for people with idiopathic pulmonary fibrosis:
  - assess lung function
  - assess for oxygen therapy
  - assess for pulmonary rehabilitation
  - offer smoking cessation advice, in line with [Smoking cessation services](#) (NICE public health guidance 10)
  - identify exacerbations and previous respiratory hospital admissions
  - consider referral for assessment for lung transplantation in people who do not have absolute contraindications (see [recommendations 1.5.16 and 1.5.17](#) )
  - consider psychosocial needs and referral to relevant services as appropriate
  - consider referral to palliative care services

- assess for comorbidities (which may include anxiety, bronchiectasis, depression, diabetes, dyspepsia, ischaemic heart disease, lung cancer and pulmonary hypertension).

# 1 Recommendations

The following guidance is based on the best available evidence. The [full guideline](#) gives details of the methods and the evidence used to develop the guidance.

The wording used in the recommendations in this guideline (for example, words such as 'offer' and 'consider') denotes the certainty with which the recommendation is made (the strength of the recommendation). See [About this guideline](#) for details.

## **1.1 Awareness of clinical features of idiopathic pulmonary fibrosis**

1.1.1 Be aware of idiopathic pulmonary fibrosis when assessing a patient with the clinical features listed below and when considering requesting a chest X-ray or referring to a specialist:

- age over 45 years
- persistent breathlessness on exertion
- persistent cough
- bilateral inspiratory crackles when listening to the chest
- clubbing of the fingers
- normal spirometry or impaired spirometry usually with a restrictive pattern but sometimes with an obstructive pattern.

## **1.2 Diagnosis**

1.2.1 Assess everyone with suspected idiopathic pulmonary fibrosis by:

- taking a detailed history, carrying out a clinical examination (see [recommendation 1.1.1](#) for clinical features) and performing blood tests to help exclude alternative diagnoses, including lung diseases associated with

environmental and occupational exposure, with connective tissue diseases and with drugs **and**

- performing lung function testing (spirometry and gas transfer) **and**
- reviewing results of chest X-ray **and**
- performing CT of the thorax (including high-resolution images).

1.2.2 Diagnose idiopathic pulmonary fibrosis only with the consensus of the multidisciplinary team (listed in table 1), based on:

- the clinical features, lung function and radiological findings (see [recommendation 1.2.1](#))
- pathology when indicated (see [recommendation 1.2.4](#)).

1.2.3 At each stage of the diagnostic care pathway the multidisciplinary team should consist of a minimum of the healthcare professionals listed in table 1, all of whom should have expertise in interstitial lung disease.

**Table 1 Minimum composition of multidisciplinary team involved in diagnosing idiopathic pulmonary fibrosis**

Stage of diagnostic care pathway	Multidisciplinary team composition (all healthcare professionals should have expertise in interstitial lung disease)
After clinical evaluation, baseline lung function and CT	Consultant respiratory physician Consultant radiologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator

<p>When considering performing bronchoalveolar lavage, and/or transbronchial biopsy or surgical lung biopsy</p> <p>Only some patients will have bronchoalveolar lavage or transbronchial biopsy but they may be being considered for surgical lung biopsy</p>	<p>Consultant respiratory physician</p> <p>Consultant radiologist</p> <p>Consultant histopathologist</p> <p>Thoracic surgeon as appropriate</p> <p>Interstitial lung disease specialist nurse</p> <p>Multidisciplinary team coordinator</p>
<p>When considering results of bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy</p>	<p>Consultant respiratory physician</p> <p>Consultant radiologist</p> <p>Consultant histopathologist</p> <p>Interstitial lung disease specialist nurse</p> <p>Multidisciplinary team coordinator</p>
<p>See chapter 6.5 (Multidisciplinary Team) in <a href="#">full guideline</a> for more information on the expertise of the multidisciplinary team.</p>	

## If a confident diagnosis cannot be made

1.2.4 If the multidisciplinary team cannot make a confident diagnosis from clinical features, lung function and radiological findings, consider:

- bronchoalveolar lavage or transbronchial biopsy **and/or**
- surgical lung biopsy, with the agreement of the thoracic surgeon.

1.2.5 Discuss with the person who may have idiopathic pulmonary fibrosis:

- the potential benefits of having a confident diagnosis compared with the uncertainty of not having a confident diagnosis **and**
- the increased likelihood of obtaining a confident diagnosis with surgical biopsy compared with bronchoalveolar lavage or transbronchial biopsy **and**
- the increased risks of surgical biopsy compared with bronchoalveolar lavage or transbronchial biopsy.

1.2.6 When considering bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy take into account:

- the likely differential diagnoses **and**
- the person's clinical condition, including any comorbidities.

1.2.7 If a confident diagnosis cannot be made continue to review the person under specialist care.

## **1.3 Information and support**

1.3.1 The consultant respiratory physician or interstitial lung disease specialist nurse should provide accurate and clear information (verbal and written) to people with idiopathic pulmonary fibrosis, and their families and carers with the person's consent. This should include information about investigations, diagnosis and management.

1.3.2 NICE has produced guidance on the components of good patient experience in adult NHS services. Follow the recommendations in [Patient experience in adult NHS services](#) (NICE clinical guideline 138).

1.3.3 An interstitial lung disease specialist nurse should be available at all stages of the care pathway to provide information and support to people with idiopathic pulmonary fibrosis and their families and carers with the person's consent.

1.3.4 Offer advice, support and treatment to aid smoking cessation to all people with idiopathic pulmonary fibrosis who also smoke, in line with [Smoking cessation services](#) (NICE public health guidance 10).

## **1.4 Prognosis**

1.4.1 Measure the initial rate of decline in the person's condition, which may predict subsequent prognosis, by using lung function test results (spirometry and gas transfer) at:

- diagnosis **and**

- 
- 6 months and 12 months after diagnosis. Repeat the lung function tests at shorter intervals if there is concern that the person's condition is deteriorating rapidly.
- 1.4.2 Discuss prognosis with people with idiopathic pulmonary fibrosis in a sensitive manner and include information on:
- the severity of the person's disease and average life expectancy
  - the varying courses of disease and range of survival
  - management options available.
- 1.4.3 Do not use the 6-minute walk distance at diagnosis to estimate prognosis. (The 6-minute walk test may be useful for other purposes, see [recommendation 1.5.1.](#))

## **1.5 Management**

### **Pulmonary rehabilitation**

- 1.5.1 Assess people with idiopathic pulmonary fibrosis for pulmonary rehabilitation at the time of diagnosis. Assessment may include a 6-minute walk test (distance walked and oxygen saturation measured by pulse oximetry) and a quality-of-life assessment.
- 1.5.2 Repeat the assessment for pulmonary rehabilitation for people with idiopathic pulmonary fibrosis at 6-month or 12-month intervals.
- 1.5.3 If appropriate after each assessment, offer pulmonary rehabilitation including exercise and educational components tailored to the needs of people with idiopathic pulmonary fibrosis in general.
- 1.5.4 Pulmonary rehabilitation should be tailored to the individual needs of each person with idiopathic pulmonary fibrosis. Sessions should be held somewhere that is easy for people with idiopathic pulmonary fibrosis to get to and has good access for people with disabilities.

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## Best supportive care

- 1.5.5 Offer best supportive care to people with idiopathic pulmonary fibrosis from the point of diagnosis. Best supportive care should be tailored to disease severity, rate of progression, and the person's preference, and should include if appropriate:
- information and support (see [recommendation 1.3.1](#))
  - symptom relief
  - management of comorbidities
  - withdrawal of therapies suspected to be ineffective or causing harm
  - end of life care.
- 1.5.6 If the person is breathless on exertion consider assessment for:
- the causes of breathlessness and degree of hypoxia **and**
  - ambulatory oxygen therapy and long-term oxygen therapy **and/or**
  - pulmonary rehabilitation.
- 1.5.7 If the person is breathless at rest consider:
- assessment for the causes of breathlessness and degree of hypoxia **and**
  - assessment for additional ambulatory oxygen therapy and long-term oxygen therapy **and**
  - the person's psychosocial needs and offering referral to relevant services such as palliative care services **and**
  - pharmacological symptom relief with benzodiazepines and/or opioids.
- 1.5.8 Assess the oxygen needs of people who have been hospitalised with idiopathic pulmonary fibrosis before they are discharged.
- 1.5.9 If the person has a cough consider:



- treatment for causes other than idiopathic pulmonary fibrosis (such as gastro-oesophageal reflux disease, post-nasal drip)
- treating with opioids if the cough is debilitating
- discussing treatment with thalidomide<sup>[1]</sup> with a consultant respiratory physician with expertise in interstitial lung disease if the cough is intractable.

1.5.10 Ensure people with idiopathic pulmonary fibrosis, and their families and carers, have access to the full range of services offered by palliative care teams. Ensure there is collaboration between the healthcare professionals involved in the person's care, community services and the palliative care team.

### **Disease-modifying pharmacological interventions**

There is no conclusive evidence to support the use of any drugs to increase the survival of people with idiopathic pulmonary fibrosis.

1.5.11 For guidance on pirfenidone for the management of idiopathic pulmonary fibrosis, refer to [Pirfenidone for the treatment of idiopathic pulmonary fibrosis](#) (NICE technology appraisal guidance 282).

1.5.12 Do not use any of the drugs below, either alone or in combination, to modify disease progression in idiopathic pulmonary fibrosis:

- ambrisentan
- azathioprine
- bosentan
- co-trimoxazole
- mycophenolate mofetil
- prednisolone
- sildenafil
- warfarin.

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- 1.5.13 Advise the person that oral N-acetylcysteine<sup>[2]</sup> is used for managing idiopathic pulmonary fibrosis, but its benefits are uncertain.
- 1.5.14 If people with idiopathic pulmonary fibrosis are already using prednisolone or azathioprine, discuss the potential risks and benefits of discontinuing, continuing or altering therapy.
- 1.5.15 Manage any comorbidities according to best practice. For gastro-oesophageal reflux disease, see [Managing dyspepsia in adults in primary care](#) (NICE clinical guideline 17).

## Lung transplantation

- 1.5.16 Discuss lung transplantation as a treatment option for people with idiopathic pulmonary fibrosis who do not have absolute contraindications. Discussions should:
- take place between 3 and 6 months after diagnosis or sooner if clinically indicated
  - be supported by an interstitial lung disease specialist nurse
  - include the risks and benefits of lung transplantation
  - involve the person's family and carers with the person's consent.

(See [recommendations 1.5.5 – 1.5.10](#) about best supportive care.)

- 1.5.17 Refer people with idiopathic pulmonary fibrosis for lung transplantation assessment if they wish to explore lung transplantation and if there are no absolute contraindications. Ask the transplant centre for an initial response within 4 weeks.

## Ventilation

- 1.5.18 A respiratory physician or specialist nurse with an interest in interstitial lung disease should discuss the poor outcomes associated with mechanical ventilation (including non-invasive mechanical ventilation) for respiratory failure with people with idiopathic pulmonary fibrosis. These discussions should

ideally take place between 3 to 6 months after diagnosis or sooner if clinically indicated. (See [recommendations 1.5.5 – 1.5.10](#) about best supportive care.)

- 1.5.19 Do not routinely offer mechanical ventilation (including non-invasive mechanical ventilation) to people with idiopathic pulmonary fibrosis who develop life-threatening respiratory failure.

## 1.6 Review and follow-up

- 1.6.1 In follow-up appointments for people with idiopathic pulmonary fibrosis:

- assess lung function
- assess for oxygen therapy
- assess for pulmonary rehabilitation
- offer smoking cessation advice, in line with [Smoking cessation services](#) (NICE public health guidance 10)
- identify exacerbations and previous respiratory hospital admissions
- consider referral for assessment for lung transplantation in people who do not have absolute contraindications (see [recommendations 1.5.16 and 1.5.17](#) )
- consider psychosocial needs and referral to relevant services as appropriate
- consider referral to palliative care services
- assess for comorbidities (which may include anxiety, bronchiectasis, depression, diabetes, dyspepsia, ischaemic heart disease, lung cancer and pulmonary hypertension).

- 1.6.2 Consider follow-up of people with idiopathic pulmonary fibrosis:

- every 3 months or sooner if they are showing rapid disease progression or rapid deterioration of symptoms **or**
- every 6 months or sooner if they have steadily progressing disease **or**

- 
- initially every 6 months if they have stable disease and then annually if they have stable disease after 1 year.

<sup>[1]</sup> At the time of publication (June 2013), thalidomide did not have a UK marketing authorisation for this indication. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's [Good practice in prescribing and managing medicines and devices](#) for further information.

<sup>[2]</sup> At the time of publication (June 2013), N-acetylcysteine did not have a UK marketing authorisation. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's [Good practice in prescribing and managing medicines and devices](#) for further information.

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## 2 Research recommendations

The Guideline Development Group has made the following recommendations for research, based on its review of evidence, to improve NICE guidance and patient care in the future. The Guideline Development Group's full set of research recommendations is detailed in the full guideline.

### ***2.1 Bronchoalveolar lavage in the diagnosis of idiopathic pulmonary fibrosis***

What is the value of bronchoalveolar lavage in people in whom idiopathic pulmonary fibrosis is considered the most likely diagnosis when clinical and CT findings are insufficient to support a confident diagnosis?

#### **Why this is important**

A confident diagnosis of idiopathic pulmonary fibrosis needs integration of clinical and CT findings in a multidisciplinary setting. However, a consensus diagnosis cannot always be made with confidence. In some people with 'probable idiopathic pulmonary fibrosis', bronchoalveolar lavage alone may help attain a more confident diagnosis while in others, a subsequent surgical lung biopsy may be needed. It is not known whether the benefits of attaining a more confident diagnosis by bronchoalveolar lavage outweigh the risks of the procedure. A randomised controlled trial should be conducted to determine the potential benefits and risks of bronchoalveolar lavage with regard to increasing diagnostic certainty and avoiding the need for surgical lung biopsy. The study should incorporate outcomes that include diagnostic certainty (sensitivity, specificity), mortality (all-cause and idiopathic pulmonary fibrosis-related), health-related quality of life and change in lung function. Adjustments should be made for differences in baseline clinical and radiological features. Clinical studies should be of sufficient power and duration and include a health economic evaluation.

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## ***2.2 Surgical lung biopsy in the diagnosis of idiopathic pulmonary fibrosis***

What is the value of surgical lung biopsy in people in whom idiopathic pulmonary fibrosis is considered the most likely diagnosis when clinical and CT findings are insufficient to support a confident diagnosis?

### **Why this is important**

A confident diagnosis of idiopathic pulmonary fibrosis needs integration of clinical and CT findings in a multidisciplinary setting. However, a consensus diagnosis cannot always be made with confidence. In such cases of 'probable idiopathic pulmonary fibrosis', surgical lung biopsy may be indicated to allow a diagnosis to be made with greater confidence. It is not known whether the benefits of attaining a more confident diagnosis outweigh the risks of surgical lung biopsy. A randomised controlled trial should be conducted to determine the potential benefits and risks of biopsy with regard to diagnostic certainty (sensitivity, specificity), mortality (all-cause and idiopathic pulmonary fibrosis-related), health-related quality of life and change in lung function. Adjustments should be made for differences in baseline clinical and radiological features. Clinical studies should be of sufficient power and duration and include a health economic evaluation.

## ***2.3 Pulmonary rehabilitation to improve outcomes in people with idiopathic pulmonary fibrosis***

Does pulmonary rehabilitation improve outcomes for people with idiopathic pulmonary fibrosis?

### **Why this is important**

There is evidence that people with idiopathic pulmonary fibrosis may benefit from pulmonary rehabilitation. However this evidence is mostly derived from programmes designed principally for people with chronic obstructive pulmonary disease. It is likely that the needs of people with idiopathic pulmonary fibrosis and chronic obstructive pulmonary disease differ. Randomised controlled trials should be carried out to determine the effects of pulmonary rehabilitation programmes tailored to idiopathic pulmonary fibrosis, compared with currently offered pulmonary rehabilitation programmes, on quality of life, walking distance and lung function with analysis adjusting for confounding factors appropriately. Trials should analyse benefits of the different aspects of pulmonary rehabilitation including the components, setting and location of the

programme, and healthcare resources involved. End points may include: 6-minute walk distance; breathlessness score; a measure of health-related quality of life (ideally employing a tool validated in people with idiopathic pulmonary fibrosis); mortality (all-cause and idiopathic pulmonary fibrosis-related); hospitalisation (all-cause, non-elective and idiopathic pulmonary fibrosis-related); lung function (vital capacity and diffusion capacity for carbon monoxide). Studies should be of sufficient power and duration and include a health economic evaluation.

## ***2.4 Ambulatory oxygen to improve outcomes in idiopathic pulmonary fibrosis***

Does ambulatory oxygen improve outcomes in idiopathic pulmonary fibrosis?

### **Why this is important**

People with idiopathic pulmonary fibrosis frequently demonstrate a fall in oxygen saturation during exercise even though they are not hypoxic at rest. In such people, ambulatory oxygen is often provided to improve exercise capacity, enhance mobility and enable activities of daily living in order to improve quality of life. However, there are no randomised controlled trials to demonstrate that ambulatory oxygen therapy is effective in achieving these aims in patients with idiopathic pulmonary fibrosis. A randomised controlled trial should be conducted to determine the effects of ambulatory oxygen on quality of life in people with idiopathic pulmonary fibrosis and consideration given to the use of a placebo arm. This should include a standardised protocol for assessing exercise such as the 6-minute walk test. The end points may include 6-minute walk distance; breathlessness score; a measure of health-related quality of life (ideally employing a tool validated in idiopathic pulmonary fibrosis patients). Phase III trials should have a duration of greater than 12 months and include a health economic evaluation.

## ***2.5 Anti-reflux therapy as a treatment for idiopathic pulmonary fibrosis***

Is anti-reflux therapy an effective treatment for idiopathic pulmonary fibrosis?

### **Why this is important**

There is evidence from observational studies, and uncontrolled interventional trials, that microaspiration of gastric/oesophageal contents contribute to disease progression, and perhaps

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even cause idiopathic pulmonary fibrosis. There have been no randomised controlled trials of anti-reflux therapy in idiopathic pulmonary fibrosis but proton-pump inhibitors are often prescribed for symptoms of acid-reflux. A randomised, placebo-controlled trial of adequate power and duration of greater than 12 months should be undertaken to determine the benefits and side effects of anti-reflux therapy, including proton pump inhibition in people with a confirmed diagnosis of idiopathic pulmonary fibrosis. Appropriate end points may include mortality (all-cause and idiopathic pulmonary fibrosis-related); hospitalisation (all-cause, non-elective and idiopathic pulmonary fibrosis-related); lung function (vital capacity and diffusion capacity for carbon monoxide); 6-minute walk distance; breathlessness score; a measure of health-related quality of life (ideally employing a tool validated in idiopathic pulmonary fibrosis patients). Phase III trials should include a health economic evaluation.



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## 3 Other information

### 3.1 Scope and how this guideline was developed

NICE guidelines are developed in accordance with a [scope](#) that defines what the guideline will and will not cover.

#### How this guideline was developed

NICE commissioned the National Clinical Guideline Centre to develop this guideline. The Centre established a Guideline Development Group (see [section 4](#)), which reviewed the evidence and developed the recommendations.

The methods and processes for developing NICE clinical guidelines are described in [The guidelines manual](#).

### 3.2 Related NICE guidance

Details are correct at the time of publication of the guideline (June 2013). Further information is available on the [NICE website](#).

#### Published

- [Pirfenidone for treating idiopathic pulmonary fibrosis](#). NICE technology appraisal guidance 282 (2013).
- [Opioids in palliative care](#). NICE clinical guideline 140 (2012).
- [Patient experience in adult NHS services](#). NICE clinical guideline 138 (2012).
- [Lung cancer](#). NICE clinical guideline 121 (2011).
- [Tuberculosis](#). NICE clinical guideline 117 (2011).
- [Chronic obstructive pulmonary disease](#). NICE clinical guideline 101 (2010).
- [Medicines adherence](#). NICE clinical guideline 76 (2009).
- [Smoking cessation services](#). NICE public health guidance 10 (2008).

- [Varenicline for smoking cessation](#). NICE technology appraisal guidance 123 (2007).
- [Brief interventions and referral for smoking cessation](#). NICE public health guidance 1 (2006).
- [Living-donor lung transplantation for end-stage lung disease](#). NICE interventional procedure guidance 170 (2006).
- [Dyspepsia](#).

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## 4 The Guideline Development Group, National Collaborating Centre and NICE project team

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## About this guideline

NICE clinical guidelines are recommendations about the treatment and care of people with specific diseases and conditions in the NHS in England and Wales.

NICE guidelines are developed in accordance with a [scope](#) that defines what the guideline will and will not cover.

This guideline was developed by the National Clinical Guideline Centre / NICE Internal Clinical Guidelines Programme, which is based at the Royal College of Physicians. The Collaborating Centre worked with a Guideline Development Group, comprising healthcare professionals (including consultants, GPs and nurses), patients and carers, and technical staff, which reviewed the evidence and drafted the recommendations. The recommendations were finalised after public consultation.

The methods and processes for developing NICE clinical guidelines are described in [The guidelines manual](#).

## ***Strength of recommendations***

Some recommendations can be made with more certainty than others. The Guideline Development Group makes a recommendation based on the trade-off between the benefits and harms of an intervention, taking into account the quality of the underpinning evidence. For some interventions, the Guideline Development Group is confident that, given the information it has looked at, most patients would choose the intervention. The wording used in the recommendations in this guideline denotes the certainty with which the recommendation is made (the strength of the recommendation).

For all recommendations, NICE expects that there is discussion with the patient about the risks and benefits of the interventions, and their values and preferences. This discussion aims to help them to reach a fully informed decision (see also [Patient-centred care](#)).

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## **Interventions that must (or must not) be used**

We usually use 'must' or 'must not' only if there is a legal duty to apply the recommendation. Occasionally we use 'must' (or 'must not') if the consequences of not following the recommendation could be extremely serious or potentially life threatening.

## **Interventions that should (or should not) be used – a 'strong' recommendation**

We use 'offer' (and similar words such as 'refer' or 'advise') when we are confident that, for the vast majority of patients, an intervention will do more good than harm, and be cost effective. We use similar forms of words (for example, 'Do not offer...') when we are confident that an intervention will not be of benefit for most patients.

## **Interventions that could be used**

We use 'consider' when we are confident that an intervention will do more good than harm for most patients, and be cost effective, but other options may be similarly cost effective. The choice of intervention, and whether or not to have the intervention at all, is more likely to depend on the patient's values and preferences than for a strong recommendation, and so the healthcare professional should spend more time considering and discussing the options with the patient.

## ***Other versions of this guideline***

The full guideline, [Idiopathic pulmonary fibrosis: the diagnosis and management of suspected idiopathic pulmonary fibrosis](#), contains details of the methods and evidence used to develop the guideline. It is published by the National Clinical Guideline Centre / NICE Internal Clinical Guidelines Programme.

The recommendations from this guideline have been incorporated into a [NICE Pathway](#).

We have produced [information for the public](#) about this guideline.

## ***Implementation***

[Implementation tools and resources](#) to help you put the guideline into practice are also available.

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## ***Changes after publication***

**December 2013:** Minor maintenance

## ***Your responsibility***

This guidance represents the view of NICE, which was arrived at after careful consideration of the evidence available. Healthcare professionals are expected to take it fully into account when exercising their clinical judgement. However, the guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer, and informed by the summaries of product characteristics of any drugs.

Implementation of this guidance is the responsibility of local commissioners and/or providers. Commissioners and providers are reminded that it is their responsibility to implement the guidance, in their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity and foster good relations. Nothing in this guidance should be interpreted in a way that would be inconsistent with compliance with those duties.

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0845 003 7780

ISBN: 978-1-4731-0179-1