

Idiopathic pulmonary fibrosis: the diagnosis and management of suspected idiopathic pulmonary fibrosis

NICE guideline

Draft for consultation, January 2013

If you wish to comment on this version of the guideline, please be aware that all the supporting information and evidence is contained in the full version.

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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. It also includes the role of multidisciplinary diagnostic and management teams, and specific therapeutic interventions.

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 medicines – guidance for doctors](#) 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.

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](#).

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').

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.

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 the clinical features of idiopathic pulmonary fibrosis for the purpose of performing a chest X-ray and specialist referral. The clinical features may include:
 - 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. **[1.1.1]**

Diagnosis

- Diagnose idiopathic pulmonary fibrosis only with the consensus of the multidisciplinary team, 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]**

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

Stage of diagnostic care pathway	Multidisciplinary team composition
After clinical evaluation, baseline lung function and computed tomography	Consultant respiratory physician Consultant radiologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
After clinical evaluation, baseline lung function, computed tomography, and bronchoalveolar lavage, and/or transbronchial biopsy if performed 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 pathologist Thoracic surgeon as appropriate Interstitial lung disease specialist nurse Multidisciplinary team coordinator
After clinical evaluation, baseline lung function, computed tomography, bronchoalveolar lavage, transbronchial biopsy/no transbronchial biopsy and surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant pathologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator

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 throughout diagnosis and treatment. This should include a clear explanation of the implications of the investigations for both diagnosis and prognosis. **[1.3.1]**
- 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. **[1.3.3]**

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. **[1.5.1]**

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 ineffective therapies
 - end of life care. **[1.6.1]**
- If the person is breathless on exertion consider:
 - assessment for ambulatory oxygen therapy and long-term oxygen therapy **and/or**
 - assessment for pulmonary rehabilitation. **[1.6.2]**

Pharmacological interventions¹

- 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

¹ There is an ongoing technology appraisal for 'Pirfenidone for the treatment of idiopathic pulmonary fibrosis' and subject to timescales, the final version of this guideline will cross refer to the published final technology appraisal guidance.

- prednisolone
- sildenafil
- warfarin. [1.7.2]

Lung transplantation

- Refer for lung transplantation assessment people with idiopathic pulmonary fibrosis who wish to explore lung transplantation and who do not have absolute contraindications. Ask the centre for an initial response within 4 weeks. [1.7.6]

Review and follow-up

- Clinical assessment at follow-up appointments for people with idiopathic pulmonary fibrosis should include:
 - assessment of lung function
 - assessment for oxygen therapy
 - assessment for pulmonary rehabilitation
 - smoking cessation advice in line with [Smoking cessation services](#) (NICE public health guidance 10)
 - identification of exacerbations and previous respiratory hospital admissions
 - assessment for lung transplantation in people who do not have absolute contraindications (see recommendations 1.7.5 and 1.7.6)
 - consideration of referral to palliative care services for people with advancing idiopathic pulmonary fibrosis
 - assessment for comorbidities (which may include dyspepsia, diabetes, lung cancer, ischaemic heart disease and pulmonary hypertension).[1.8.2]

1 Recommendations

The following guidance is based on the best available evidence. The [full guideline](#) [\[hyperlink to be added for final publication\]](#) gives details of the methods and the evidence used to develop the guidance.

1.1 *Awareness of clinical features of idiopathic pulmonary fibrosis*

1.1.1 Be aware of the clinical features of idiopathic pulmonary fibrosis for the purpose of performing a chest X-ray and specialist referral. The clinical features may include:

- 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 to exclude alternative diagnoses, including:
 - exposure to environmental and occupational risk factors
 - symptoms suggestive of connective tissue disease
 - exposure to medication which may cause lung fibrosis **and**
- carrying out a clinical examination (see recommendation 1.1.1 for clinical features) **and**
- performing lung function testing (spirometry and gas transfer) **and**

- reviewing results of chest X-ray **and**
- performing computed tomography of the thorax (including high-resolution images).

1.2.2 At each stage of the diagnostic care pathway the multidisciplinary team should consist of a minimum of the healthcare professionals listed in table 1 with expertise in interstitial lung disease.

1.2.3 Diagnose idiopathic pulmonary fibrosis only with the consensus of the multidisciplinary team, 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
After clinical evaluation, baseline lung function and computed tomography	Consultant respiratory physician Consultant radiologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
After clinical evaluation, baseline lung function, computed tomography, and bronchoalveolar lavage, and/or transbronchial biopsy if performed 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 pathologist Thoracic surgeon as appropriate Interstitial lung disease specialist nurse Multidisciplinary team coordinator
After clinical evaluation, baseline lung function, computed tomography, bronchoalveolar lavage, transbronchial biopsy/no transbronchial biopsy and surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant pathologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator

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.

1.3 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 throughout diagnosis and treatment. This should include a clear explanation of the implications of the investigations for both diagnosis and prognosis.

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.

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. (For circumstances where the 6-minute walk test may be useful, see recommendation 1.5.1.)

1.5 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 Reassess people with idiopathic pulmonary fibrosis for pulmonary rehabilitation (in line with recommendation 1.5.1) 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 where it is easy for people with idiopathic pulmonary fibrosis to get to and have good access for people with disabilities.

1.6 Best supportive care

1.6.1 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 ineffective therapies
- end of life care.

1.6.2 If the person is breathless on exertion consider:

- assessment for ambulatory oxygen therapy and long-term oxygen therapy **and/or**
- assessment for pulmonary rehabilitation.

1.6.3 If the person is breathless at rest consider:

- assessment for ambulatory oxygen therapy and long-term oxygen therapy **and/or**
- benzodiazepines **and/or**
- opioids.

1.6.4 Assess the oxygen needs of people who have been hospitalised with idiopathic pulmonary fibrosis before they are discharged.

1.6.5 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 thalidomide² with a consultant respiratory physician with expertise in interstitial lung disease if the cough is intractable.

1.6.6 Ensure people with idiopathic pulmonary fibrosis, and their families and carers, have access to the full range of services offered by multidisciplinary palliative care teams. Ensure there is collaboration between the multidisciplinary team, community services and the palliative care team.

1.7 Disease-modifying treatments

Pharmacological interventions

There is no conclusive evidence to support the use of any drugs to increase the survival of people with idiopathic pulmonary fibrosis³.

1.7.1 Advise the person with idiopathic pulmonary fibrosis that oral *N*-acetylcysteine⁴ is used for managing idiopathic pulmonary fibrosis, but its benefits are uncertain.

1.7.2 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

² At the time of consultation (January 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 medicines – guidance for doctors](#) for further information.

³ There is an ongoing technology appraisal for 'Pirfenidone for the treatment of idiopathic pulmonary fibrosis' and subject to timescales, the final version of this guideline will cross refer to the published final technology appraisal guidance.

⁴ At the time of consultation (January 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 medicines – guidance for doctors](#) for further information.

- mycophenolate mofetil
- prednisolone
- sildenafil
- warfarin.

1.7.3 If people with idiopathic pulmonary fibrosis are already using combination prednisolone and azathioprine, with or without *N*-acetylcysteine:

- discuss the risks of this treatment **and**
- consider gradual withdrawal of both prednisolone and azathioprine.

1.7.4 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

1.7.5 Discuss lung transplantation as a treatment option for people with idiopathic pulmonary fibrosis who do not have absolute contraindications as advised by regional transplant units. Discussions should:

- take place between 3 to 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 if appropriate.

(See section 1.6 for recommendations about best supportive care.)

1.7.6 Refer for lung transplantation assessment people with idiopathic pulmonary fibrosis who wish to explore lung transplantation and who do not have absolute contraindications. Ask the centre for an initial response within 4 weeks.

Ventilation

- 1.7.7 Do not routinely offer mechanical ventilation (including non-invasive mechanical ventilation) to people with idiopathic pulmonary fibrosis who develop life-threatening respiratory failure.
- 1.7.8 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 section 1.6 for recommendations about best supportive care.)

1.8 *Review and follow-up*

- 1.8.1 Consider follow-up of people with idiopathic pulmonary fibrosis (see recommendation 1.3.3):
- 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.
- 1.8.2 Clinical assessment at follow-up appointments for people with idiopathic pulmonary fibrosis should include:
- assessment of lung function
 - assessment for oxygen therapy
 - assessment for pulmonary rehabilitation
 - smoking cessation advice, in line with [Smoking cessation services](#) (NICE public health guidance 10)
 - identification of exacerbations and previous respiratory hospital admissions

- assessment for lung transplantation in people who do not have absolute contraindications (see recommendations 1.7.5 and 1.7.6)
- consideration of referral to palliative care services for people with advancing idiopathic pulmonary fibrosis
- assessment for comorbidities (which may include dyspepsia, diabetes, lung cancer, ischaemic heart disease and pulmonary hypertension).

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 computed tomography findings are insufficient to support a confident diagnosis?

Why this is important: A confident diagnosis of idiopathic pulmonary fibrosis needs integration of clinical and computed tomography 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.

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 computed tomography findings are insufficient to support a confident diagnosis?

Why this is important: A confident diagnosis of idiopathic pulmonary fibrosis needs integration of clinical and computed tomography 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 patients with idiopathic pulmonary fibrosis*

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

Why this is important: There is evidence that patients with idiopathic pulmonary fibrosis may benefit from pulmonary rehabilitation. However this evidence is mostly derived from programmes designed principally for patients 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 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.

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 consultation on the guideline (January 2013). Further information is available on the [NICE website](#).

Published

- [Opioids in palliative care](#). NICE clinical guideline 140 (2012).

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- [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).
- [Dyspepsia](#). NICE clinical guideline 17 (2004).

Under development

NICE is developing the following guidance (details available from the [NICE website](#)):

- Pirfenidone for the treatment of idiopathic pulmonary fibrosis. NICE technology appraisal guidance. Publication expected April 2013.

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

4.1 *Guideline Development Group*

Nik Hirani (Chair)

Senior Clinical Lecturer and Honorary Consultant in Respiratory Medicine,
University of Edinburgh

Geraldine Burge

Interstitial Lung Disease Specialist Nurse, Heartlands Hospital, Birmingham

Sue Copley

Consultant Radiologist and Reader in Thoracic Imaging, Hammersmith
Hospital, Imperial College Health Care NHS Trust, London

Annette Duck

Interstitial Lung Disease Specialist Nurse, University Hospital of South
Manchester Foundation Trust, Manchester

Nicholas Kim Harrison

Senior Clinical Lecturer and Honorary Consultant Respiratory Physician,
College of Medicine, Swansea University

Melissa Hippard

Patient member

Richard Hubbard

British Lung Foundation Professor of Respiratory Epidemiology, University of
Nottingham, Nottingham

Angela Key

Chief Respiratory Physiologist, Aintree University Hospitals NHS Foundation
Trust, Liverpool

Tessa Lewis

GP, Aneurin Bevan Health Board

Ann Millar

Professor of Respiratory Medicine, University of Bristol, Bristol

Nick Screaton

Consultant Cardiothoracic Radiologist, Papworth Hospital NHS Trust,
Cambridgeshire

Malcolm Weallans

Patient member, The Pulmonary Fibrosis Trust

Patrick Wilson

Senior Respiratory Pharmacist, Glenfield Hospital, Leicester

Co-opted external advisers

Andrew Nicholson

Consultant Histopathologist, Royal Brompton and Harefield NHS Trust and
Professor of Respiratory Pathology, National Heart and Lung Institute Division
of Imperial College School of Medicine, London

Stephen Clark

Consultant Cardiothoracic and Cardiopulmonary Transplant Surgeon, Director
of Cardiopulmonary Transplantation, Clinical Lecturer (Institute of Cellular
Medicine, University of Newcastle), Cardiothoracic Centre, Freeman Hospital,
Newcastle upon Tyne

Sally Singh

Head of Pulmonary and Cardiac Rehabilitation, Department of Pulmonary
Rehabilitation, Glenfield Hospital, University Hospitals of Leicester NHS Trust,
Leicester

4.2 *National Clinical Guideline Centre*

Vanessa Delgado Nunes

Guideline Lead

Nina Balachander

Senior Research Fellow and Project Manager

Vicki Pollitt

Senior Health Economist

Zahra Naqvi

Research Fellow

Izaba Younis

Research Fellow

Joanna Ashe

Information Scientist

4.3 NICE project team

Sarah Willett

Associate Director

Clifford Middleton

Guideline Commissioning Manager

Andrew Gyton

Guideline Coordinator

Judith Thornton

Technical Lead

Jasdeep Hayre

Health Economist

Jaimella Espley

Editor