

Idiopathic pulmonary fibrosis in adults: diagnosis and management

Clinical guideline

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Your responsibility

The recommendations in this guideline represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, professionals are expected to take this guideline fully into account, alongside the individual needs, preferences and values of their patients or service users. The application of the recommendations in this guideline are not mandatory and the guideline does not override the responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or their carer or guardian.

Local commissioners and/or providers have a responsibility to enable the guideline to be applied when individual health professionals and their patients or service users wish to use it. They should do so in the context of local and national priorities for funding and developing services, and in light of their duties to have due regard to the need to eliminate unlawful discrimination, to advance equality of opportunity and to reduce health inequalities. Nothing in this guideline should be interpreted in a way that would be inconsistent with compliance with those duties.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

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This guideline is the basis of QS79.

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.

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](#).

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)
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After clinical evaluation, baseline lung function and CT	<p>Consultant respiratory physician</p> <p>Consultant radiologist</p> <p>Interstitial lung disease specialist nurse</p> <p>Multidisciplinary team coordinator</p>
<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>
When considering results of bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy	<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>
See chapter 6.5 (Multidisciplinary Team) in full guideline 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.

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, see the NICE technology appraisal on [pirfenidone for the treatment of idiopathic pulmonary fibrosis](#). For guidance on nintedanib, see the NICE technology appraisal on [nintedanib for the treatment of idiopathic pulmonary fibrosis](#).
- 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
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 [full guideline](#) for more information on the expertise of the multidisciplinary team.

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.

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^[1] 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, see the NICE technology appraisal on [pirfenidone for the treatment of idiopathic pulmonary fibrosis](#). For guidance on nintedanib, see the NICE technology appraisal on [nintedanib for the treatment of idiopathic pulmonary fibrosis](#).
- 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.
- 1.5.13 Advise the person that oral N-acetylcysteine^[2] 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.

More information

You can also see this guideline in the NICE pathway on [idiopathic pulmonary fibrosis](#).

To find out what NICE has said on topics related to this guideline, see our web page on [respiratory conditions](#).

See also the guideline committee's discussion and the evidence reviews (in the [full guideline](#)), and information about [how the guideline was developed](#), including details of the committee.

^[1] 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.

^[2] 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.

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 *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.

2.2 *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.

2.3 *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.

Update information

May 2017: Recommendation 1.5.11 was amended to add a link to the NICE technology appraisal on [nintedanib for the treatment of idiopathic pulmonary fibrosis](#). Two outdated research recommendations were removed.

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Accreditation

