NICE National Institute for Health and Care Excellence

NICE Pathways

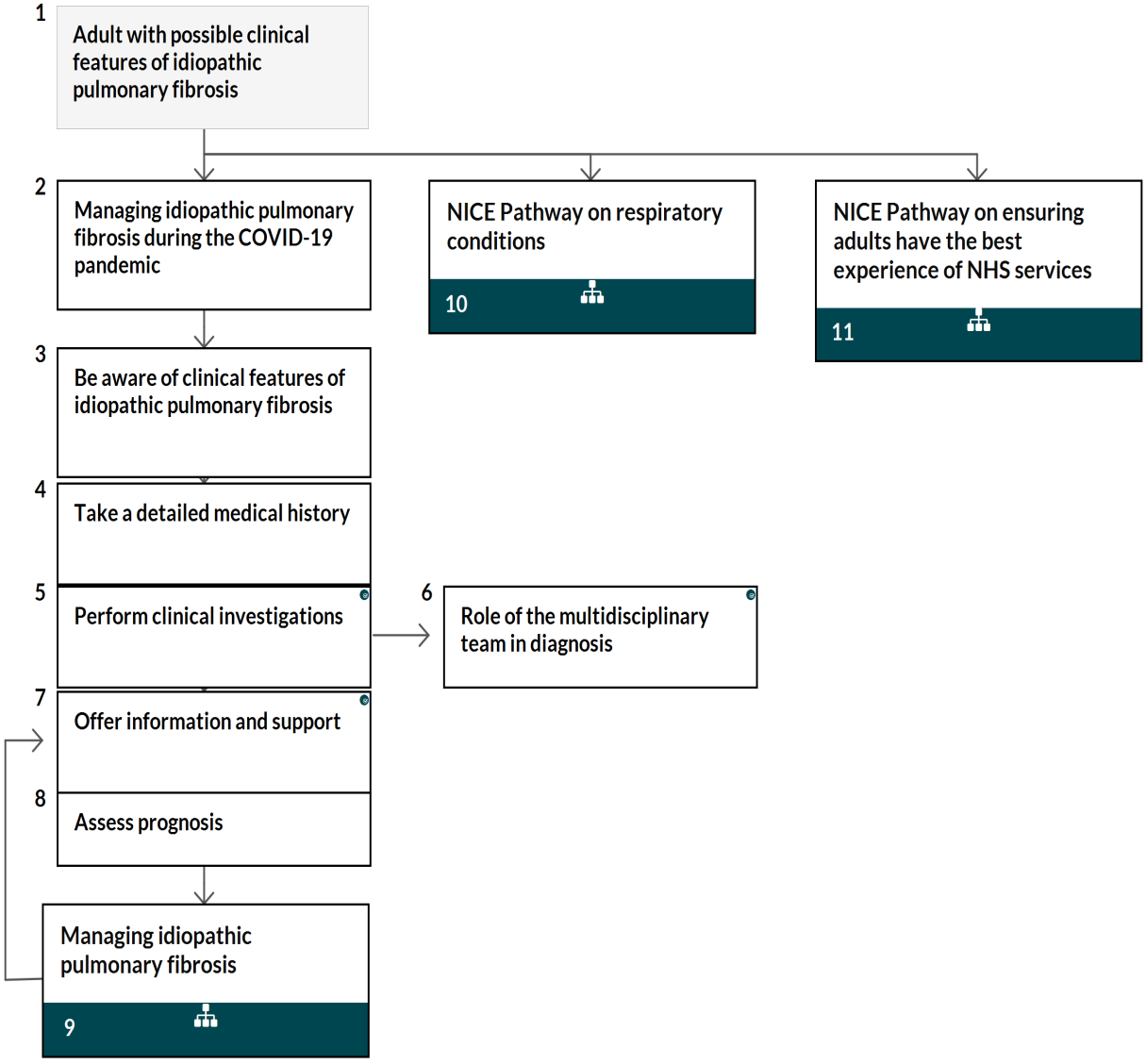
Idiopathic pulmonary fibrosis overview

NICE Pathways bring together everything NICE says on a topic in an interactive flowchart. NICE Pathways are interactive and designed to be used online.

They are updated regularly as new NICE guidance is published. To view the latest version of this NICE Pathway see:

http://pathways.nice.org.uk/pathways/idiopathic-pulmonary-fibrosis
NICE Pathway last updated: 03 November 2020

This document contains a single flowchart and uses numbering to link the boxes to the associated recommendations.



1 Adult with possible clinical features of idiopathic pulmonary fibrosis

No additional information

2 Managing idiopathic pulmonary fibrosis during the COVID-19 pandemic

NICE has produced a COVID-19 rapid guideline on interstitial lung disease. It recommends changes to usual practice to maximise the safety of patients and protect staff from infection during the COVID-19 pandemic.

3 Be aware 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.

4 Take a detailed medical history

Assess everyone with suspected idiopathic pulmonary fibrosis by:

- taking a detailed history, carrying out a clinical examination (see be aware of clinical features of idiopathic pulmonary fibrosis [See page 3]) 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.

5 Perform clinical investigations

Assess everyone with suspected idiopathic pulmonary fibrosis by:

- 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).

Diagnose idiopathic pulmonary fibrosis only with the consensus of the multidisciplinary team, based on:

- the clinical features, lung function and radiological findings
- pathology when indicated.

If a confident diagnosis cannot be made

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.

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.

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.

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

Quality standards

The following quality statement is relevant to this part of the interactive flowchart.

1. Diagnosis of idiopathic pulmonary fibrosis

6 Role of the multidisciplinary team in diagnosis

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. See chapter 6.5 (multidisciplinary team) in the full guideline for more information on the expertise of the multidisciplinary team.

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

Quality standards

The following quality statement is relevant to this part of the interactive flowchart.

1. Diagnosis of idiopathic pulmonary fibrosis

7 Offer 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.

NICE has produced guidance on the components of good patient experience in adult NHS services. Follow the NICE Pathway on patient experience in adult NHS services.

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.

Offer advice, support and treatment to aid smoking cessation to all people with idiopathic pulmonary fibrosis who also smoke, in line with the NICE Pathway on stop smoking interventions and services.

Discussing lung transplantation

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 offer best supportive care and symptom relief.)

Discussing ventilation

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 offer best supportive care and symptom relief.)

Quality standards

The following quality statement is relevant to this part of the interactive flowchart.

2. Access to a specialist nurse

8 Assess prognosis

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.

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.

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 assess for pulmonary rehabilitation programme tailored for idiopathic pulmonary fibrosis.)

9 Managing idiopathic pulmonary fibrosis

See Idiopathic pulmonary fibrosis / Managing idiopathic pulmonary fibrosis

10 NICE Pathway on respiratory conditions

See Respiratory conditions

11 NICE Pathway on ensuring adults have the best experience of NHS services

See Patient experience in adult NHS services

Sources

Idiopathic pulmonary fibrosis in adults: diagnosis and management (2013 updated 2017) NICE guideline CG163

Your responsibility

Guidelines

The recommendations in this guideline represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, professionals and practitioners are expected to take this guideline fully into account, alongside the individual needs, preferences and values of their patients or the people using their service. It is not mandatory to apply the recommendations, and the guideline does not override the responsibility to make decisions appropriate to the circumstances of the individual, in consultation with them and their families and carers or guardian.

Local commissioners and providers of healthcare have a responsibility to enable the guideline to be applied when individual professionals and people using services 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 complying 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.

Technology appraisals

The recommendations in this interactive flowchart represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, health professionals are expected to take these recommendations fully into account, alongside the individual needs, preferences and values of their patients. The application of the

recommendations in this interactive flowchart is at the discretion of health professionals and their individual patients and do 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.

Commissioners and/or providers have a responsibility to provide the funding required to enable the recommendations to be applied when individual health professionals and their patients wish to use it, in accordance with the NHS Constitution. They should do so 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.

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.

Medical technologies guidance, diagnostics guidance and interventional procedures guidance

The recommendations in this interactive flowchart represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, healthcare professionals are expected to take these recommendations fully into account. However, the interactive flowchart 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.

Commissioners and/or providers have a responsibility to implement the recommendations, 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 interactive flowchart 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.