NATIONAL INSTITUTE FOR HEALTH AND CLINICAL EXCELLENCE

SCOPE

1 Guideline title

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

1.1 Short title

Idiopathic pulmonary fibrosis

2 The remit

The Department of Health has asked NICE: 'To produce a clinical guideline on the diagnosis and management of suspected idiopathic pulmonary fibrosis.'

3 Clinical need for the guideline

3.1 Epidemiology

a) Idiopathic pulmonary fibrosis (IPF) used to be called cryptogenic fibrosing alveolitis. It is a severe progressive lung disease, in which fibrous tissue forms in the lungs. Smoking is believed to be a risk-factor but the exact cause is unknown.

b) Most people with idiopathic pulmonary fibrosis experience worsening breathlessness leading to respiratory failure. Average survival is around 3 years, and mortality rates are comparable to many solid cancers.

c) The median age of presentation is 70 years. It is rare in people younger than 45.

d) Idiopathic pulmonary fibrosis is becoming more common. The incidence is around eight to nine per 100,000 person years, which

means more than 4000 new cases occur in the UK each year. The prevalence is around 15 to 25 per 100,000 and increases with age. The average hospital with a catchment of 500,000 will have 35 new cases a year and the average GP surgery of 10,000 patients will have two new cases every 3 years.

3.2 Current practice

a) Idiopathic pulmonary fibrosis is one of several interstitial lung diseases that tend to present in a similar manner with breathlessness, bibasal chest crepitations and diffuse chest X-ray changes. Idiopathic pulmonary fibrosis has the poorest prognosis of these disorders, so establishing a timely, confident diagnosis is important. A confident diagnosis needs careful integration of clinical, radiological (high-resolution CT scans) and pathological data and there is evidence that this is best achieved in a specialist multidisciplinary setting.

b) To manage IPF, there is evidence to support a role for some types of best supportive care, such as smoking cessation, pulmonary rehabilitation, withdrawal of ineffective therapy, oxygen therapy and palliation of symptoms.

c) Currently, there is no proven effective drug therapy for IPF. Corticosteroids and azathioprine are often used. A recent trial suggests the addition of N-acetylcysteine to prednisilone and azathioprine may slow the rate of disease progression more than prednisolone and azathioprine alone.

d) There are some emerging therapies for the disease. Some of these are costly, and all are as yet unproven but they may change the treatment landscape.

e) Lung transplantation is a valuable resource for selected patients. It is suitable for only a minority of patients with idiopathic pulmonary fibrosis, and the number of patients that die waiting for a lung

transplant is proportionately higher than any other patient group. Efforts should be made to identify which patients would benefit most.

f) Access to pulmonary rehabilitation services and palliative care for idiopathic pulmonary fibrosis is not uniform. The past few years have seen several ad hoc specialist centres emerge, often with limited or no resource support.

g) The British Thoracic Society guidelines were published in 2008. There is an urgent need for guidance on initial diagnosis and the management of idiopathic pulmonary fibrosis because the ongoing burden of disease has significant resource implications, and because of the imminent emergence of new potential therapies.

4 The guideline

The guideline development process is described in detail on the NICE website (see section 6, 'Further information').

This scope defines what the guideline will (and will not) examine, and what the guideline developers will consider. The scope is based on the referral from the Department of Health.

The areas that will be addressed by the guideline are described in the following sections.

4.1 Population

4.1.1 Groups that will be covered

a) Adults (18 and older) with suspected or diagnosed idiopathic pulmonary fibrosis.

b) No patient subgroups have been identified as needing specific consideration.

4.1.2 Groups that will not be covered

a) Children and young people (younger than 18).

b) People with a diagnosis of pulmonary fibrosis as a complication of:

- connective tissue disorders (for example, systemic lupus erythematosus, rheumatoid arthritis, scleroderma, polymyositis and dermatomyositis)
- a known exogenous agent (for example, drug-induced disease or asbestosis).

4.2 Healthcare setting

All healthcare settings.

4.3 Clinical management

4.3.1 Key clinical issues that will be covered

a) Diagnosis:

- high resolution computed tomography (CT) scanning
- biopsy (bronchoalveolar lavage and surgical lung biopsy)
- multidisciplinary teams to achieve a consensus diagnosis
- pulmonary function tests.

b) Prognosis:

- pulmonary function tests (resting spirometric and gas transfer measurement)
- sub-maximal exercise testing
- echocardiography.

c) Treatment of the disease with the following drugs:

- prednisolone
- mycophenolate mofetil
- warfarin

- azathioprine
- N-acetyl cysteine
- proton-pump inhibitors
- co-trimoxazole
- ambrisentan
- bosanten
- sildenafil.

Note that guideline recommendations will normally fall within licensed indications; exceptionally, and only if clearly supported by evidence, use outside a licensed indication may be recommended. The guideline will assume that prescribers will use a drug's summary of product characteristics to inform the decisions they make with patients.

d) Symptom relief:

- lung transplantation timing and referral
- best supportive care (benzodiazepines, oxygen therapy and palliative care)
- non invasive and invasive ventilation
- pulmonary rehabilitation (breathlessness management).

e) Patient review and follow-up.

4.3.2 Clinical issues that will not be covered

a) Therapies for pulmonary hypertension as a complication of idiopathic pulmonary fibrosis.

b) Treatment of lung cancer as a complication of idiopathic pulmonary fibrosis.

c) Lung transplantation, other than timing and referral.

4.4 Main outcomes

a) Lung capacity: measurement of vital capacity (VC) or forced vital capacity (FVC).

b) Gas transfer: measurement of the carbon monoxide diffusing capacity of the lungs (T_LCO).

c) Change in health-related quality of life measured using the Short Form-36 or Saint George's Respiratory Questionnaire and/or a measure of function such as the 6 minute walk test or EQ 5D.

d) Hospitalisations due to exacerbation of the disease.

e) Mortality.

f) Adverse events.

4.5 Economic aspects

Developers will take into account both clinical and cost effectiveness when making recommendations involving a choice between alternative interventions. A review of the economic evidence will be conducted and analyses will be carried out as appropriate. The preferred unit of effectiveness is the quality-adjusted life year (QALY), and the costs considered will usually only be from an NHS and personal social services (PSS) perspective. Further detail on the methods can be found in 'The guidelines manual' (see ‘Further information’).

4.6 Status

4.6.1 Scope

This is the final scope.

4.6.2 Timing

The development of the guideline recommendations will begin in September 2011.

5 Related NICE guidance

5.1 Published guidance

- Lung cancer. NICE clinical guideline 121 (2011). Available from www.nice.org.uk/guidance/CG121
- Tuberculosis. NICE clinical guideline 117 (2011). Available from www.nice.org.uk/guidance/CG117
- Chronic obstructive pulmonary disease. NICE clinical guideline 101 (2010). Available from www.nice.org.uk/guidance/CG101
- Smoking cessation services. NICE public health guidance 10 (2008). Available from www.nice.org.uk/guidance/PH10.

5.2 Guidance under development

NICE is currently developing the following related guidance (details available from the NICE website).

- Opioids in palliative care. NICE clinical guideline. Publication expected May 2012.

5.2.1 NICE guidance to be incorporated

This guideline is intended to incorporate the following NICE guidance, subject to a technology appraisal consultation:

- Pirfenidone for the treatment of idiopathic pulmonary fibrosis. NICE technology appraisal. Publication date to be confirmed.

6 Further information

Information on the guideline development process is provided in:

- 'How NICE clinical guidelines are developed: an overview for stakeholders' the public and the NHS'
- 'The guidelines manual'.

These are available from the NICE website (www.nice.org.uk/GuidelinesManual). Information on the progress of the guideline will also be available from the NICE website (www.nice.org.uk).