New NICE quality standards for interstitial pulmonary fibrosis aim to improve care standards for all patients diagnosed with this progressive and life-limiting condition

Interstitial pulmonary fibrosis: care standards

In this article...
- Care inequalities for patients with interstitial pulmonary fibrosis
- Implications of new NICE quality standards for IPF
- Role of the clinical nurse specialist in supporting patients

Although idiopathic pulmonary fibrosis (IPF) is considered a rare lung condition, 5,000 new cases are diagnosed each year in the UK. The incidence is higher in men than women, and increases with age (Navaratnam et al, 2011).

The condition is characterised by long periods of stability punctuated by episodes of dramatic acute deterioration, which represent a significant cause of morbidity and mortality (Collard et al, 2008). The exacerbations occur most frequently in the first 18 months after diagnosis (Mura et al, 2012). Death is often triggered by infection and/or coronary artery disease (Coss et al, 2001).

Treatment options for IPF are limited; lung transplantation is the only intervention offering any possibility of added life. However, two new therapies have recently emerged. Pirfenidone is an immunosuppressant drug with both anti-inflammatory and anti-fibrotic effects; it has been shown to slow disease progression in some patients with mild to moderate IPF, and is approved for use in the NHS (National Institute for Health and Care Excellence, 2013a). Ninetedanib is undergoing a scoping exercise and is likely to be available on prescription for IPF early in 2016 (NICE, 2014).

After referral for consultant-led treatment for non-urgent conditions, patients should commence treatment within a maximum of 18 weeks from the date of referral (Department of Health, 2013a). However, a recent survey of patients with IPF and audit of 16 NHS trusts in England, indicated that more than 50% of patients waited longer than three months to see a specialist, with more than 30% waiting longer than six months (British Lung Foundation, 2015).

Care for these patients is subject to special service specification and commissioning arrangements determined by NHS England (2013). This gives them access to a multidisciplinary team for diagnosis, a guarantee of equal access to treatment and a disease-specific management plan.

Since a technology appraisal approved pirfenidone to treat mild to moderate IPF (NICE, 2013a), the number of patients referred to specialist centres has grown exponentially. This has increased demand at a time when the NICE Clinical Guideline (2013b) recognised that there were significant variations in the clinical care of IPF across England and Wales. To improve and standardise care of patients diagnosed with IPF, NICE (2015) has published five quality standards. These are outlined below.

Statement 1: multidisciplinary team
People are diagnosed with IPF only with the consensus of a multidisciplinary team (MDT) expert in interstitial lung disease.
Minimum requirements of the MDT are:
- Consultant respiratory physician;
- Consultant radiologist;
- Consultant histopathologist;
- Clinical nurse specialist (CNS);
- MDT co-ordinator.

The complementary role of the wider interdisciplinary team is also essential. A possible diagnosis of IPF must be considered in any patient aged over 45 years presenting with new persistent breathlessness on exertion and/or unexplained persistent cough. On auscultation of the chest, patients may also have bilateral inspiratory Velcro-type crackles at the lung bases, along with signs of fingernail clubbing and fatigue. The MDT will arrange detailed blood tests, and bronchoalveolar lavage; where diagnostic uncertainty exists a lung biopsy may be required.

Spirometry may be normal in patients with IPF. If it is impaired it usually has a restrictive pattern but sometimes an obstructive pattern is evident.

Chronic obstructive pulmonary disease co-exists in around 8-15% of patients diagnosed with IPF (NICE, 2014), which can make the interpretation of the spirometry challenging. It is therefore important not to rely on spirometry alone when considering a diagnosis of IPF. Chest X-rays may be inconclusive so a high-resolution CT scan of the thorax with specialist radiologist interpretation is needed.

A detailed history about environmental and occupational exposures should be taken by a specialist ILD physician to rule out other conditions, particularly hypersensitivity pneumonitis and asbestosis.

Statement 2: specialist nurse

People with IPF will have an interstitial lung disease specialist nurse available to them. The CNS should provide accurate and clear information to patients and their families about investigations, diagnosis and management. This should be available at all stages of the care pathway to facilitate transitions in care referral for psychological support, and to advise on managing symptoms and the side-effects of and withdrawal from medication. Patients who smoke should also be given smoking cessation advice.

Discussions about lung transplantation should take place three to six months after diagnosis or sooner if clinically indicated; these discussions should be supported by an ILD CNS. Poor outcomes associated with use of mechanical ventilation (including non-invasive mechanical ventilation) for respiratory failure should also be discussed three to six months after diagnosis or sooner if clinically indicated. The CNS is well-placed to liaise with the community respiratory nurse and general practice team about patients’ ongoing needs.

Statement 3: oxygen

People with IPF have an assessment for home and ambulatory oxygen therapy at each follow-up appointment and before they leave hospital following an exacerbation of the disease.

Oxygen therapy is usually effective in relieving symptoms of breathlessness, particularly exertional breathlessness. However, the transition to oxygen therapy has a psychological component – some patients report feeling socially inhibited using oxygen in public, and many see it as a turning point signalling progression of their condition; sensitivity and understanding is needed to ease this transition.

Patients with IPF benefit from ambulatory oxygen therapy (AOT), which improves oxygen levels during activity and can help them to participate in pulmonary rehabilitation (PR). They do not usually require long-term oxygen therapy (LTOT), which is used to stabilise oxygen levels and is used for at least 15 hours per day.

Assessment for AOT is often determined by the six-minute walk test, where levels of oxygen desaturation and time to recover are measured. Patients should be reassessed every three, six or 12 months. Overnight oximetry should be performed to determine the need for nocturnal oxygen.

Statement 4: rehabilitation

Pulmonary rehabilitation programmes should provide services designed specifically for IPF. They should be tailored to patients’ specific needs. Oxygen saturation levels should be monitored during exercise and oxygen therapy adjusted accordingly. Patients should also be regularly reassessed for repeat PR programmes at six- and 12-monthly intervals.

Statement 5: palliative care

People with IPF and their families and carers will have access to services that meet their palliative care needs.

The varying courses of IPF and average life expectancy (generally three to five years from diagnosis) should be discussed in a sensitive manner (NICE, 2014). The symptoms of IPF are often debilitating and difficult to manage and medication may be ineffective, which is why symptom management and control, psychological care and the provision of information should be revisited at every contact. Opioids and benzodiazepines may help when symptoms are debilitating, while effective assessment will ensure that patients can access services to meet their palliative care needs.

Oxygen needs should be reassessed regularly and other causes of breathlessness should be considered as the condition progresses. In some patients, side-effects make it necessary to withdraw some medications to improve quality of life.

Conclusion

NICE guidance addresses the value of drugs and interventions, including oxygen therapy, pulmonary rehabilitation, palliation of breathlessness and cough. However, there remains a paucity of evidence to inform practice. Interdisciplinary care networks are needed to optimise the care pathway for patients with IPF. Management of the condition involves respiratory and palliative care teams in primary, secondary and tertiary care settings, and across other specialties when patients develop secondary complications or have co-existing co-morbidities. The pivotal role of the CNS and the complementary role of the interdisciplinary team must not be under-valued.

References


National Institute for Health and Care Excellence (2013b) Idiopathic Pulmonary Fibrosis: Diagnosis and Management of Suspected IPF. www.nice.org.uk/gs143


For more on this topic go online...
- Principles to effectively manage patients with interstitial lung disease in the community
- Bit.ly/NTILDCommunity