Idiopathic pulmonary fibrosis is a life-limiting, incurable condition. Recent guidance from NICE identifies priorities of care for this group of patients.

**Management of idiopathic pulmonary fibrosis**

**In this article...**
- Definition of idiopathic pulmonary fibrosis
- Priorities for managing patients with this condition
- The role of the nurse and multidisciplinary team

**Clinical features**
There are over 150 ILDs, including IPF. Patients usually present with shortness of breath and cough, sometimes with other symptoms. For example, Raynaud’s disease is seen in ILD associated with connective tissue disorders, while lethargy and tiredness are often experienced in sarcoidosis. Some types of ILD, such as IPF, do not respond to treatment and affected patients will ultimately need palliative care (Duck, 2009).

In IPF, the lungs gradually become scarred, causing them to become smaller and stiffer, making it difficult to inhale. On a high-resolution CT scan, honeycomb scarring in the periphery of the lung can be seen. For this reason, IPF is often referred to as “honeycomb lung disease”.

Many patients describe a protracted time to diagnosis, and those with IPF are often misdiagnosed with asthma or COPD, causing frustration and loss of confidence in health professionals (Schoenheit, 2011). NICE advises clinicians to be aware of the clinical features of IPF when assessing a patient and refer for a chest X-ray or to a respiratory specialist if they have concerns. Box 1 outlines the key signs and symptoms.

**Diagnosis of IPF**
As IPF is difficult to diagnose and can be confused with common interstitial pneumonias or ILDs, NICE (2013a) recommends that all patients suspected of having IPF are referred to an ILD specialist multidisciplinary team (MDT) for diagnosis. This team should consist of a:

- Consultant respiratory physician with expertise in ILD;
- Consultant radiologist with expertise in ILD;

**5 key points**

1. **Definition of idiopathic pulmonary fibrosis**
   - Idiopathic pulmonary fibrosis is a rapidly progressive interstitial lung disease

2. **Priorities for managing patients with this condition**
   - There is no cure and patients have a prognosis of 2-4 years

3. **The role of the nurse and multidisciplinary team**
   - Five thousand new cases of IPF are diagnosed annually

4. **Clinical features**
   - Patients with IPF are often misdiagnosed with asthma or COPD

5. **Diagnostic of IPF**
   - All patients suspected of having IPF should be referred to an ILD specialist multidisciplinary team

**Box 1. Signs and symptoms of IPF**
- Persistent breathlessness on exertion
- Persistent cough
- Bilateral inspiratory crackles when listening to the chest with a stethoscope
- Clubbing of the fingers
- Normal or impaired spirometry usually with a restrictive but sometimes with an obstructive pattern

The condition occurs typically in people aged over 45 years.
ILD specialist nurse; MDT coordinator.

A consultant histopathologist with expertise in ILD and a thoracic surgeon should be included in the MDT if patients have had a bronchoalveolar lavage or are likely to need an open lung biopsy to confirm diagnosis.

Information and support
ILD nurses are essential to the MDT as they provide patients with information about investigations, diagnosis and prognosis.

Most patients have never heard of IPF and are unlikely to be prepared for bad news. The five-year survival rate is worse than in all but two cancers – pancreas and lung (Vancheri et al, 2010). Supportive nurses are critical to helping patients adjust and develop coping strategies.

Pulmonary rehabilitation
NICE (2013a) recommends offering pulmonary rehabilitation to patients throughout their disease pathway. This should be tailored as the educational needs of IPF patients are different from those with other lung diseases such as COPD.

Supportive care
Nurses are crucial in providing best supportive care, which should be tailored to disease severity and rate of progression, and patient preferences from diagnosis through to end-of-life care.

Since IPF generally affects older people, patients may have comorbidities, take a range of medication and have social issues that also affect quality of life.

Breathlessness on exertion is a characteristic of this disease. Regular assessment for exercise-induced hypoxia and prompt referral for ambulatory oxygen assessment enable patients to engage in normal activities inside and outside the home for as long as possible.

Treatment for cough varies, but it can be eased by mucolytic drugs (which help loosen and clear the mucus from the airways by breaking up the sputum), codeine linctus or oral morphine. Thalidomide improves cough (Horton et al, 2012) but can have unpleasant side-effects and should only be prescribed by specialist doctors.

Palliative care teams can advise on symptom management; referral should be considered for patients who are distressed by symptoms that are difficult to manage.

Monitoring disease progression and withdrawing ineffective therapies that could cause harm or further impair the patient’s quality of life should be considered.

Pharmaceutical interventions
Patients need information about possible treatments. There is no cure for IPF but pirfenidone (Esbriet) has been shown to slow disease progression (Noble et al, 2011). NICE (2013b) has set out criteria for its use in England. It can only be prescribed by registered prescribers and patients have to demonstrate a forced vital capacity (FVC) of 50-80% predicted for age and sex. FVC is the maximum amount of air a person can expel from the lungs after a maximum inhalation. The drug must be stopped if the FVC drops by more than 10% in a year, as this indicates the drug is not working. Patients need support during treatment to manage common side-effects, which can include nausea, loss of appetite, weight loss, lethargy and photosensitivity skin rashes.

There is limited evidence to support other pharmaceutical interventions for IPF so NICE (2013a) reviewed drugs traditionally used to treat the condition. The guideline group felt that, until more evidence became available, most drugs used to treat IPF should not be offered as some have substantial side-effects. Drugs include ambrisentan, azathioprine, bosentan, co-trimoxazole, myophenolate mofetil, prednisolone, sildenafil and warfarin.

Some patients have already been prescribed a triple therapy (prednisolone, azathioprine and n-acetylcycteine) and, if they are stable, the patient and the clinical team should discuss the pros and cons of continuing the regimen. The Data Management Committee recently stopped the triple therapy arm in a research study (Panther study) due to an increased incidence of mortality in the triple therapy arm.

The N-acetylcycteine and placebo arm continued and the results of this study should be available in May this year.

Lung transplantation
All physicians should discuss lung transplantation with IPF patients 3-6 months after diagnosis if they have no absolute contraindications (Orens et al, 2006); these include:

- Malignancy in the last two years, except squamous and basal cell tumours;
- Significant chest wall/spinal deformity;
- Unstable psychiatric/psychological conditions that might affect adherence to follow-up and treatment regimens;
- Substance addiction, including alcohol, tobacco and narcotics, unless substance free for at least six months;
- Advanced dysfunction of major organs (heart, liver or kidney);
- Coronary heart disease not amenable to bypass grafting or percutaneous intervention, or associated left ventricular dysfunction;
- Infection with HIV, hepatitis C and/or hepatitis B;
- Absence of consistent social support.

Clinicians are advised to contact their local lung transplant centre for advice and should expect a response within four weeks.

Review and follow-up
Patients with rapid disease progression should be reviewed every three months, while those who are stable should be reviewed every six months. The review should include:

- Lung function;
- Oxygen assessment;
- Offer of pulmonary rehabilitation;
- Assessment of comorbidities;
- Smoking cessation advice;
- Identifying and assessing any hospital admissions and exacerbations;
- Assessment for palliative care referral.

Conclusion
NICE has gone some way in identifying the needs of patients with IPF and has evaluated the evidence to date to guide clinicians in their management of this fatal disease. There are many research recommendations including the benefit of oxygen, the value of supportive care, symptom control and end-of-life care.

References
National Institute for Health and Care Excellence (2013a) Diagnosis and Management of Suspected Idiopathic Pulmonary Fibrosis. nice.org.uk/gc163