Care planning in idiopathic pulmonary fibrosis

Initial assessment
At 59 years of age, Henry Spencer (not his real name) started to experience increasing breathlessness when walking up hills, was unable to walk up several flights of stairs and had gained weight. Initially, he dismissed his symptoms as age related, but then began to struggle to play his saxophone. Having developed a cough and intermittent chest tightness, he went to see his GP in 2011. The GP diagnosed a chest infection and prescribed antibiotics, but the symptoms did not resolve so Mr Spencer returned to his GP two weeks later.

Mr Spencer exhibited some clinical features of IPF (Box 1): he was aged over 45 years and had persistent breathlessness on exertion, although cough and clubbing were not present. Auscultation of the chest revealed fine crackles at the lung bases, so the GP referred Mr Spencer for a chest X-ray. When lung disease is suspected, spirometry can help determine the pattern. Characteristic changes in the lung volumes allow the condition to be classified as obstructive, restrictive or mixed. IPF is a restrictive lung disease, but it occasionally has an obstructive pattern if emphysema is present. Spirometry must always be interpreted within the clinical context. The quality of spirometry in general practice is variable. In this instance, the GP referred Mr Spencer to the lung function laboratory at the secondary care centre where results indicated a very mild restrictive pattern.

Mr Spencer’s chest X-ray suggested IPF so the GP referred him to a chest physician, requesting a high-resolution CT (HRCT) scan of the thorax. IPF can sometimes be identified on a HRCT because of its characteristic honeycomb appearance. However, IPF is the commonest type of ILD, and results in scarring in the lungs.

In this article...
- Description of idiopathic pulmonary fibrosis
- Recommended treatment regimen for IPF
- Case study of a patient with IPF

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Abstract

Patients with idiopathic pulmonary fibrosis require accurate diagnosis and treatment to improve their quality of life and prognosis. This case study follows a patient’s care from presentation to diagnosis and management over four years. It illustrates the recommended care pathway, the complexity of the condition and the importance of shared care between specialist and local services. See page 16 for the latest NICE standards for IPF care.

Keywords: Idiopathic pulmonary fibrosis/supportive care/diagnosis

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5 key points
1. Idiopathic pulmonary fibrosis is a progressive lung disease with a poor prognosis
2. Careful assessment is crucial to accurate diagnosis of IPF
3. A new drug treatment, pirfenidone, was approved by the European Commission in 2011 and gained NICE approval in 2013
4. The progressive nature of IPF means care should be shared between specialist and local services
5. More research is needed on non-pharmacological management of IPF
Mr Spencer did not have this presentation and was advised to have a lung biopsy. Mr Spencer’s biopsy confirmed a diagnosis of IPF and he was prescribed 5mg prednisolone once a day to slow progression of the fibrosis. At this time, the evidence to support the use of corticosteroids in IPF was weak but, in the absence of any alternative therapy, corticosteroids were considered a reasonable option in some patients (Raghu et al, 2011). Box 2 gives further information about investigations.

**Assessment and diagnosis**

Following referral to a specialist ILD centre, Mr Spencer was admitted for assessment. Taking a detailed history and recording symptoms are essential parts of the assessment; symptoms mark progression, and assist in shaping the management plan and in identifying possible triggers to assist in establishing the diagnosis. Mr Spencer had asymptomatic coronary artery disease and well-controlled type 2 diabetes. He lived with his wife, who was fit and well. He had an allergy to cats, but there were no pets or birds in the house, nor was there any damp or mould. This helped to rule out hypersensitivity pneumonitis, an inflammatory lung condition caused by inhaling foreign substance. He had worked in the army in ancillary roles and had been exposed to asbestos. This helped to rule out asbestosis. He had never smoked, never drank alcohol and had no exposure to tuberculosis.

Mr Spencer occasionally experienced gastro-oesophageal reflux disease (GORD). If untreated, this is thought to contribute to IPF progression, possibly due to micro-aspiration of gastric content (Lee et al, 2010). His shortness of breath and exercise tolerance gradually worsened, and he experienced occasional chest tightness, which he described as “difficulty getting breath in”. It is important to capture patients’ experience in their own words; an objective measure of breathlessness such as the Modified Medical Research Council (MRC) scale provides a reliable indication of progression of IPF (Mura et al, 2012).

ILD clinical nurse specialists have a valuable role in helping patients to understand their diagnosis and uncertain prognosis, and to offer information about investigations and management. A CNS should be available throughout the care pathway.

It was agreed Mr Spencer’s disease-specific care would aim to optimise the management of his pulmonary fibrosis. Lung transplantation was discussed but, in view of his diabetes, age and asymptomatic coronary artery disease, it was thought he was unlikely to benefit from a transplant.

**Pharmacological management**

Mr Spencer had been started on omeprazole by his local chest physician to treat his GORD but, after the side-effect of diarrhoea caused him to lose 14lb, he was switched to ranitidine. He was also prescribed prednisolone, along with calcium and alendronic acid to prevent corticosteroid-induced osteoporosis; calcium and vitamin D levels must be monitored in patients taking long-term steroids, and replacement therapy provided.

Because of his high cholesterol levels, Mr Spencer had been prescribed simvastatin by his GP. Statins have been linked with the development of fibrotic lung disease (Fernandez et al, 2008), although others dispute this (Santana et al, 2008) more research on this is needed. Since the symptoms of his lung disease began shortly after he started a statin, this treatment was discontinued. Current guidance (National Institute for Health and Care Excellence, 2014) does not advocate an alternative.

**Triple therapy**

Traditionally, ILD was treated with triple therapy of prednisolone, n-acetylcysteine (NAC) and azathioprine (AZT), but this has been superseded by new therapies. Some patients with complex diagnosis may take a combination of old and new therapies. Mr Spencer was initially prescribed:

- Prednisolone – increased to 10mg daily;
- NAC 600mg three times a day – this is an antioxidant used as adjunct therapy to AZT and is given to stabilise an oxidant-antioxidant imbalance thought to occur in IPF;
- AZT starting at 50mg daily for one month and titrating over a three-month period to a maximum 150mg (American Thoracic Society and European Respiratory Society, 2002), with full blood count and liver function tests monitored weekly for at least three months by the GP. AZT is an immunosuppressant and combining it with corticosteroids was thought to slow down disease progression.

Mr Spencer’s care plan included six-monthly clinical reviews with pulmonary function tests. Forced vital capacity is used as a marker of disease progression, enabling intermittent recalibration of life expectancy and supportive care (NICE, 2015). Mr Spencer’s first-six-month review coincided with a press release on the National Heart, Lung and Blood Institute’s PANTHER trial, designed to determine whether azathioprine and oral corticosteroids +/- NAC could slow IPF progression (NHLBI, 2011). The triple therapy (NAC, prednisolone and azathioprine) arm of the trial was stopped prematurely following...
BOX 2. INVESTIGATIONS

When patients are referred to the specialist interstitial lung disease centre, blood tests and/or a diagnostic bronchoscopy with bronchoalveolar lavage (BAL) may be required (the latter if there is diagnostic uncertainty). BAL involves a bronroscope being passed down the bronchus into the lungs where fluid is instilled then withdrawn for examination. While BAL fluid containing white blood cells can be non-specific, it is helpful in establishing a diagnosis. For example, raised lymphocytes in the lavage is more likely to indicate hypersensitivity pneumonitis than IPF but may indicate other conditions (ATS/ERS, 2002).

BAL can help multidisciplinary teams diagnose without biopsy. Video-assisted thoracoscopic lung biopsy carries risks, for example of pneumothorax, and should be performed only when clinically necessary.

New treatment

Pirfenidone was emerging as a new treatment (Noble et al, 2011) but had not then been approved by NICE. In the interim, the manufacturer provided the drug free of charge for 12 months through a named patient programme, on which Mr Spencer was enrolled in 2012. Patients had to agree that after the 12 months they would stop therapy or pay for it themselves if their local commissioning body would not fund it.

The uncertainty was stressful but, fortunately, Mr Spencer was granted an additional 12 months; the therapy would cease if there was a significant decline in FVC >10% in the year. He received ongoing therapy following approval by NICE (2013).

Pirfenidone can have significant side-effects such as skin rashes, gastro-intestinal problems and abnormal liver function tests; some people cannot tolerate these.

With the introduction of the pirfenidone and in view of Mr Spencer’s diabetes, prednisolone was reduced to 5mg daily. Mr Spencer was reviewed every six months at the specialist centre, with pulmonary function tests; the long-term plan was to move towards shared care involving the local chest physician and monitoring by GP services. The progressive and uncertain nature of IPF mean it is important to develop shared care and to ensure open and effective communication between specialist centres and local services.

Exacerbations

Mr Spencer was tolerating pirfenidone well but, at his first six-month review, he reported increasing breathlessness; he was walking at a slower pace and had a slightly worse cough with persistent white purulent phlegm. A six-metre walk test showed a drop in oxygen saturations; he had been started on ambulatory oxygen by his community respiratory team. In the clinic, his oxygen saturations were 92% and his pulmonary function test had declined. Patients need to know the positive effects of pirfenidone take about six months to appear.

It was difficult to tell whether the slight decline was the start of an exacerbation of IPF or early signs of infection. Best practice suggests that suspected infection should be treated quickly and he was started on oral antibiotics pending HRCT to assess for pulmonary embolism and progression of fibrosis and an echocardiogram to detect signs of pulmonary hypertension.

An HRCT did not indicate pulmonary embolus, and there were no significant changes in Mr Spencer’s IPF. The echocardiogram indicated early pulmonary hypertension, a common complication of IPF. It was decided to continue all medication as prescribed, with a review in four months.

At his next review, Mr Spencer’s lung function had stabilised; he was feeling less breathless and playing the saxophone again. His repeat six-metre walk test indicated desaturations to 86% and he was advised that he could benefit from ambulatory oxygen for exertion or if feeling breathless on exercise. Although his community respiratory team had prescribed oxygen, he had not used it because he had felt well. Overnight oximetry with a home monitor indicated he did not desaturate and did not require overnight oxygen at this stage. He was placed back on six-monthly reviews but could contact his CNS should his symptoms worsen.

Mr Spencer has now been taking pirfenidone for two and a half years and his general quality of life has improved. His oxygen requirements have gradually increased and lung function has gradually decreased. He has been referred to his local palliative care team to optimise supportive care.

Conclusion

The CNS is integral to the multidisciplinary team in the care of patients with IPF. The landscape for the care of these patients has changed rapidly with the emergence of new therapies, and clinical trials continue. However, non-pharmacological management remains challenging due to the lack of evidence. The time for nurse-led research has never been better. NT

● The latest NICE standards for IPF care, page 16.

References


National Institute for Health and Care Excellence (2013) Idiopathic Pulmonary Fibrosis: Diagnosis and Management of Suspected IPF. nice.org.uk/cg163


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