Recognising and managing patients with interstitial lung disease

Annette Duck

Interstitial lung disease is the collective name given to over 200 different types of parenchymal lung disorders (Demedts et al, 2001) (these are disorders that affect the functioning parts of an organ — in this case the lungs — as opposed to the structural elements, such as connective tissue etc). They are different from the more common respiratory diseases such as chronic obstructive pulmonary disease (COPD) and asthma in that they affect the alveoli, small airways, interstitium and surrounding capillary circulation. Interstitial lung disease causes scarring, which makes it difficult for oxygen to diffuse into the circulating blood, resulting in breathlessness.

RECOGNISING INTERSTITIAL LUNG DISEASE

As opposed to COPD and asthma, which are obstructive lung conditions, interstitial lung diseases are known as restrictive lung diseases, so will not often be detected by primary care spirometry until they are quite advanced. Obstructive lung diseases include asthma and COPD and are characterised by air becoming trapped in the lungs, which then grow in size — in asthma this is caused by bronchial irritation and inflammation; whereas in COPD, many of the alveoli will have broken down, trapping air inside the lungs and causing hyperinflation (difficulty in breathing more air in until some has been released).

By contrast, the restricted lungs seen in interstitial lung disease are smaller and shrunken due to scarring and loss of elasticity, making it difficult for them to expand and take air in. Spirometry is not sensitive enough to pick up interstitial lung diseases until the lungs are already very restricted and many patients with mild-to-moderate disease will present with normal spirometry results. This is especially true if they have been smokers and have both COPD and interstitial lung diseases, as each disease is ‘working against’ the other.

Some of the interstitial lung diseases, such as sarcoidosis (this is where small granulomas develop in the body’s organs) and hypersensitivity pneumonitis (where the lungs become inflamed after breathing in foreign substances), are treatable if diagnosed and addressed early before significant scarring has occurred.

Unfortunately, some forms of interstitial lung disease have a very poor prognosis. Idiopathic

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THE SCIENCE — HOW DO THE ALVEOLI WORK?

The alveoli are tiny sacs located in the lungs that allow oxygen and carbon dioxide to move between the lungs and a person’s bloodstream. The alveoli are especially developed to make gaseous exchange in the lungs take place easily and efficiently.

The alveoli provide the lungs with a large surface area and have moist, thin walls, which are only one cell thick. They are filled with a mass of capillaries and the gases move by diffusion, from a high concentration to a low concentration. Oxygen diffuses from the air in the alveoli and thence into the blood, whereas carbon dioxide diffuses from the blood into the air.

Interstitial lung disease is a wide-ranging category of respiratory illness that includes many different lung conditions, such as interstitial pneumonia, idiopathic pulmonary fibrosis, non-specific interstitial pneumonitis and hypersensitivity pneumonitis. All interstitial lung diseases affect the interstitium, a delicate network of tissue that extends throughout the lungs and which supports the microscopic air sacs within the lungs (alveoli). Unfortunately, some forms of interstitial lung disease have a very poor prognosis, therefore it is important that community nurses are aware of the signs and symptoms of interstitial lung disease so that prompt referral, assessment and treatment can be started.

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pulmonary fibrosis (where the alveoli and surrounding tissue become damaged) is one of the more common types of interstitial lung disease and is fatal, with a mean life expectancy of 2–4 years from diagnosis (Wells and Dubois, 1994). More recently, drugs have been developed that have been shown to slow down the rate of disease progression, offering more promise to patients with this disease.

**SIGNS AND SYMPTOMS**

**Breathlessness**

Like COPD and asthma, the main presenting symptom of interstitial lung disease is breathlessness. Patients usually report breathlessness on exertion, which recovers quickly once they stop. At first they may suspect that this is part of the ageing process, or due simply to being unfit or overweight, and usually do nothing about it until it gets worse or they notice a sudden decline. Patients with interstitial lung disease rarely describe breathlessness at rest until the disease is well advanced.

**Cough**

Some, but not all patients describe having a cough — this can be dry or involve sputum production. Some patients will describe this cough in terms of being a nuisance rather than a real symptom, i.e. as being ‘a silly little cough’. However, for many, this is the first and only sign that something is wrong (Duck et al, 2015). Many patients report having been left with a cough after a chest infection, and which does not seem to go away.

**Infections**

Some patients with interstitial lung disease may report having had repeated chest infections over many years, with successive courses of antibiotics prescribed in the absence of a respiratory diagnosis. Some of these patients who are smokers or ex-smokers will get labelled with a diagnosis of COPD; some may even have co-existing COPD and interstitial lung disease.

However, COPD has a much more gradual onset and patients can have it for many years before presenting. Interstitial lung disease, on the other hand, has a more rapid onset, particularly in the case of idiopathic pulmonary fibrosis. There may also be some other subtle systemic symptoms such as rash or fatigue in sarcoidosis, or Raynaud’s disease (a common phenomena that affects the blood supply to certain parts of the body) in connective tissue-related interstitial lung disease.

Asking about work and leisure activities might provide the community nurse with clues as to whether a patient has interstitial lung disease or COPD. For example, people whose work or hobbies involve dusty environments, such as carpenters or pigeon-keepers, are more likely to have interstitial lung disease. Many non-smokers may also report being labelled with COPD and, therefore, often do not receive a thorough assessment and investigation of their breathlessness.

**Clubbing and other finger anomalies**

Some patients with interstitial lung disease will exhibit a widening and curving of their finger and toenails known as ‘clubbing’ (Figure 1), although the reasons for this are unknown. Others might report having cold hands with fingers that become white in cooler weather (Raynaud’s disease).

Community nurses should also look out for a ‘shiny’ or taut quality to the skin on the patient’s fingers and/or toes, which might indicate a connective tissue-related interstitial lung disease. Sometimes, interstitial lung disease can be related to rheumatoid arthritis, therefore caution should be taken in patients with rheumatoid arthritis who start reporting breathlessness.

‘Crackles’ on auscultation

When listening to the chest with a stethoscope, the community nurse might notice the sound of bilateral inspiratory ‘crackles’ that do not improve over time or after an infection. Crackles are more common in the lower bases of the lung but can be located throughout. These are sometimes called ‘Velcro® crackles as they sound like the sound of Velcro being separated. An example of inspiratory crackles in idiopathic pulmonary fibrosis can be heard at: www.soundsofipf.inipf.com

**DIAGNOSIS**

In primary care, the main objective is to be alert to the signs and symptoms of interstitial lung disease; to perform a chest X-ray; and refer the patient to a respiratory physician. On reviewing a chest X-ray, the radiologist may notice increased interstitial markings or subtle shadowing, which could suggest an interstitial lung disease. If the radiologist suspects an abnormality, he or she can recommend refer on to the respiratory physician and/or recommend a high-resolution computed tomography (HRCT) scan for a more detailed assessment.

In 2013, the National Institute for Health and Care Excellence (NICE, 2013a) published guidelines on idiopathic pulmonary fibrosis, which suggested that all patients suspected of having the condition be referred to a specialist interstitial lung disease centre for diagnosis and management. NICE recognised the difficulty in distinguishing idiopathic pulmonary fibrosis from the other interstitial lung diseases, which means that most people now suspected of interstitial lung disease or idiopathic pulmonary fibrosis are referred to one of the specialist interstitial lung disease centres throughout the UK.

Diagnosis and management is made by the multidisciplinary team, including an interstitial lung disease physician; a radiologist; a pathologist; a rheumatologist; a respiratory physician; a community nurse with clues as to whether a patient has interstitial lung disease or COPD. For example, people whose work or hobbies involve dusty environments, such as carpenters or pigeon-keepers, are more likely to have interstitial lung disease. Many non-smokers may also report being labelled with COPD and, therefore, often do not receive a thorough assessment and investigation of their breathlessness.

**Figure 1.**

*Patient exhibiting phenomenon known as ‘clubbed fingers’.*
Coughing can be a crucial early sign of interstitial lung disease and should not be ignored, especially if it has been present for any length of time. Smaller irritating coughs are easy for patients to ignore, but should still be investigated by the community nurse, e.g. questions asked about the duration of the cough; when it started; whether it is a ‘dry’ cough.

with specialist interstitial lung disease expertise; and an interstitial lung disease nurse.

Once a correct diagnosis is made, the most suitable pharmacological treatment and management plan can be started.

TREATMENT

Treatment for interstitial lung disease depends on the particular diagnosis. The British Thoracic Society’s (BTS, 2008) interstitial lung disease guidelines direct clinicians on how to treat and manage the different interstitial lung diseases. Most are treated with steroids or steroid-sparing immunosuppressant drugs, which need careful monitoring and management.

Treatments for idiopathic pulmonary fibrosis

Pirfenidone

Pirfenidone has recently been licensed for the treatment of idiopathic pulmonary fibrosis (NICE, 2013b), however, it is only available in specialist centres throughout the UK.

Pirfenidone has been shown to slow down disease progression. A recent 52-week study using lung function, exercise tolerance and progression-free survival as outcome measures demonstrated that when compared to those patients on placebo, pirfenidone slowed disease progression (King et al, 2014).

The most common side-effects of pirfenidone are gastrointestinal problems and skin sensitivity, although most are manageable and the drug is reasonably well-tolerated by most patients (King et al, 2014).

Nintedanib

Nintedanib is another drug that has been shown to slow disease progression in idiopathic pulmonary fibrosis. One study compared nintedanib with a placebo to examine the decline of forced vital capacity (FVC) in lung function (forced capacity is the maximum amount of air someone can expel from the lungs after a maximum inhalation) (Richeldi et al, 2014). The adjusted annual rate of change in FVC was 114.7ml in patients using nintedanib versus 239.9ml in the placebo, figures that are consistent with a slowing of disease progression.

This means that nintedanib offers another potential treatment for people with idiopathic pulmonary fibrosis, but like pirfenidone it does have some side-effects — the most common of which is diarrhoea (Richeldi et al, 2014). Nintedanib is not currently available on the NHS.

NURSING MANAGEMENT

It is not necessary for community nurses to have a full understanding of all the different interstitial lung diseases in order to care for those affected (Duck, 2008). However, patients do expect nurses to have some understanding of their condition.

Community nursing support — from diagnosis to end-of-life care — is vital to the well-being and adjustment of patients with idiopathic pulmonary fibrosis, in particular (Duck, 2014). The main reason why nursing support is so vital for patients with idiopathic pulmonary fibrosis is because of the rapidity of the disease progression. Patients have little time to adjust to the limitations imposed by idiopathic pulmonary fibrosis and they need psychological support in particular, as there are few treatment options and most people will die very quickly. Idiopathic pulmonary fibrosis has been likened to cancer, however, there are few interstitial lung disease specialist nurses available compared to the resources available for cancer patients.

Unfortunately, there is often only one interstitial lung disease nurse specialist working out of each regional centre. Therefore, to improve the help on offer to patients with advancing disease in their own homes, it is important that there is some outreach from the interstitial lung disease specialist nurse to the local community nurses.

Breathlessness

Many community nurses are used to managing patients who have breathlessness associated with COPD and heart failure, and some of the pharmacological and non-pharmacological strategies that work for these patients will also work for patients with interstitial lung disease. These include:

- Pacing and prioritising activities: it is important that patients do not over-exert themselves, or place too high an expectation on themselves to achieve. It is also important to work out which activities, such as shopping or cleaning, can be delegated or left to others
- Relaxation: patients should be advised to try to reduce the ‘workload’ of breathing by relaxing their shoulders and arms, and if possible breathing in a slower and more focused way
- Visualisation: relaxation can be achieved by encouraging patients to focus on a time when they felt ‘well’ and were happy with life.
- Often the physiological response to these memories will help to reduce breathlessness
- Cognitive behavioural therapy (CBT) (Heslop, 2009): this can help patients to feel more ‘in control’ of what is happening to them. Patients often know the solutions to their own problems; they just need to be empowered to act upon this knowledge
- Pharmacological preparations that reduce the sensation of breathlessness (Jennings et al, 2002) (see below).

Oral preparations of opioids such as morphine will help with breathlessness and occasionally coughing — these should begin with small doses (Jennings et al, 2002). Sub-lingual lorazepam can also help

Red Flag Cough

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reduce the anxiety associated with breathlessness.

**Pulmonary rehabilitation**

Patients with interstitial lung disease can also benefit from pulmonary rehabilitation as recommended by NICE (2013a). Patients are often paralysed by the fear of breathlessness, and feel frightened to do things. Like patients with COPD, it is important that they continue to exercise to maintain muscle strength and prevent de-conditioning (deconditioning in this context means to lose muscle and the ability to perform exercise and activities of daily living). They can learn to feel more confident in exercising and moving around despite being breathless. It is important that patients feel in control of breathlessness.

**Ambulatory oxygen therapy**

Ambulatory oxygen can make a big difference to patients with idiopathic pulmonary fibrosis and interstitial lung disease (Duck, 2008). Idiopathic pulmonary fibrosis, in particular, can progress quickly as patients do not de-condition as quickly as patients with COPD. Therefore, it is important to offer ambulatory oxygen before they de-condition.

However, breathlessness associated with exercise-induced hypoxia can mean that patients are unable to move without supplementary ambulatory oxygen. In these cases, oxygen diffusion across the interstitial space will have been reduced by scarring in the lungs, causing extreme breathlessness on movement. It is not uncommon for oxygen saturation to drop down to 60–70% on movement, and then recover to 94% once the patient is stationary again (Frank et al, 2012).

For this reason, patients must be assessed for ambulatory oxygen on exertion and not at rest. In the community it would be reasonable to measure oxygen saturations while a patient walks upstairs to identify how far the levels fall. The BTS (2006) home oxygen guidelines recommend that ambulatory oxygen is offered to all those people whose oxygen saturations drop below 90% on walking.

Oxygen assessment centres throughout the country (which were set up in the main to assess people with COPD), must assess patients with interstitial lung disease for ambulatory oxygen if patients are describing exercise-induced breathlessness. This is most likely to be caused by hypoxia and is easily corrected by supplemental ambulatory oxygen. Patients not only need ambulatory oxygen while they are walking outside etc; it is also needed indoors for activities of daily living, such as showering and housework, and generally moving around to go to bed or to the toilet, for example.

Patients with interstitial lung disease rarely meet the criteria for long-term oxygen therapy until they have very advanced disease. However, they will meet the criteria for ambulatory oxygen with only moderate disease. Therefore, they often benefit from installation of an oxygen concentrator purely for ambulatory purposes (Duck, 2008).

It is important not to assess patients’ ambulatory oxygen needs based on a reading of their resting oxygen saturation, which may be within normal ranges. Instead, patients should be tested as normal but after a six-minute walk or shuttle walk — this will demonstrate any oxygen de-saturation as a result of exercise (the only difference between six-minute and shuttle walk tests is that the latter is not self-paced but uses ‘bleeps’ to dictate the pace of the patient’s walking).

Patients with interstitial lung disease may need two oxygen prescriptions — one for resting, sitting and sleeping (if they meet the long-term oxygen therapy criteria); and another for moving around or ambulation. They usually need ambulatory oxygen before they meet the criteria for long-term oxygen therapy or resting oxygen (Duck et al, 2015).

**Palliative care and symptom control**

Treatment for interstitial lung disease and palliative care can run concurrently if patients are struggling with symptoms, such as breathlessness, cough and psychological distress.

Edmonds (2001) found that end-of-life care patients with COPD had similar needs to those patients dying from lung cancer and in the author’s opinion, many patients living with and dying from interstitial lung disease and idiopathic pulmonary fibrosis also need symptom management and psychosocial support.

Schoenheit et al (2011) described how patients with idiopathic pulmonary fibrosis have a protracted time to diagnosis, are often misdiagnosed and have many unmet

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**Expert commentary**

_Sandra Olive, respiratory nurse specialist, Norfolk and Norwich University Hospital_

*Interstitial lung diseases are poorly understood and recognised and because of this are often diagnosed at a late stage. Early referral to specialist services is essential to give patients the best chance of appropriate management.*

This article clearly highlights the valuable role of community nurses in identifying the early signs and symptoms of interstitial lung diseases, which might require further investigation. Additionally, it describes issues which affect this group of patients in advanced disease — community nurses are key players in the provision of end-of-life care and an understanding of the problems specific to those living and dying with interstitial lung diseases can help to reduce their distress.
support needs. For these reasons, they would benefit from more support, including earlier diagnosis and more specialist follow-up with less bureaucracy.

MANAGING ADVANCING DISEASE

In the author’s clinical experience, when interstitial lung disease becomes advanced, it is characterised by increasing breathlessness and the need for oxygen. It is not unusual for at-rest patients to be taking in oxygen at eight litres per minute via a high flow nasal cannulae. In addition to hypoxia-related breathlessness, patients may start to experience a particular type of breathing problem, often described as a difficulty ‘getting air in’. This is likely to be as a result of extremely scarred lungs and a concurrent loss of elasticity, causing the lungs to ‘stiffen’ when inhaling and exhaling.

This phenomena can be very frightening for patients. However, community nurses can help by changing the oxygen supply from a nasal cannula to a venturi oxygen mask (sometimes known as an air-entrainment mask) — this will deliver an increased total gas flow by drawing surrounding air into the oxygen system.

Patients at this stage often need a 60% mask with two high-flow concentrators delivering a total of 16 litres/min. Entrained air into the venturi system will further increase the total gas flow into the nose and mouth, which will help reduce the sensation of breathlessness. In addition, the use of a fan can relieve breathlessness by working on the sensory receptors around the mouth and nose (Galbraith et al, 2010).

The use of oramorph (starting 2.5mgs/5mls twice-daily rising to as required), and sublingual lorazepam (0.5mgs as required) can also help with breathlessness and panic (Jennings et al, 2002).

The use of a syringe driver to deliver midazolam and diamorphine may be needed to relieve extreme breathlessness in the final days of life, as well as avoiding unpleasant respiratory arrest when no more domiciliary oxygen can be delivered (Jennings et al, 2002).

Communication

Discussions about preferences for end-of-life care need to be had with the patient well in advance, bearing in mind that idiopathic pulmonary fibrosis is ‘predictably unpredictable’ and a patient may have an accelerated decline, often prompted by an acute exacerbation or chest infection.

In order to coordinate their health and social care, patients should be placed on the palliative care Gold Standard Framework (GSF) once they start having problems with completing activities of daily living and/or leaving the home (www.goldstandardsframework.org.uk).

CONCLUSION

It is important that primary and community nurses are aware of the signs and symptoms of interstitial lung disease so that prompt referral, assessment and treatment can be started. Currently, many patients are being misdiagnosed, and referred too late when scarring is well established and only supportive therapy can be offered. Even for patients with idiopathic pulmonary fibrosis, the future is looking better than ever with the prospect of two new pharmacological agents, pirfenidone and nintedanib, that can increase life expectancy.

REFERENCES


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