Principles to effectively manage people with interstitial lung disease in the community

Many people with interstitial lung disease have advanced illness. Simple strategies can aid quality of life and help people to stay at home as their condition worsens.

**INTRODUCTION**

Interstitial lung disease is a spectrum of conditions covering more than 150 lung disorders. People with ILD usually present with shortness of breath and/or cough, sometimes with other symptoms, such as Raynaud’s disease in an ILD associated with connective tissue disorders, or lethargy and tiredness often experienced in sarcoidosis.

Some types of ILD do not respond to treatment, such as idiopathic pulmonary fibrosis (IPF). However, others respond well if the disease is diagnosed and treated early. Those with IPF and some advancing ILDs will ultimately need palliative care.

**EDUCATION AND SUPPORT**

Many nurses lack confidence in managing people with ILD because they do not completely understand its complexities.

People may be referred to community teams with labels such as IPF, cryptogenic fibrosing alveolitis (CFA), sarcoidosis or extrinsic allergic alveolitis (EAA). As these are not common respiratory conditions, they can lead to anxiety among health professionals about treatment and management.

The reality is that people with ILD have a range of symptoms, anxieties and needs that most nurses can manage effectively. Their problems reflect those experienced by many with a life limiting illness involving a decline in function and issues around end of life care.

**POWERLESSNESS AND CONTROL**

At the time of diagnosis, many people have advanced disease. Those with a diagnosis of IPF (the most common form of ILD) have a mean life expectancy of 2-4 years from diagnosis (Wells and DuBois, 1994).

As the disease often progresses rapidly, people do not have time to adapt physically or psychologically and many with IPF experience underlying feelings of powerlessness and loss of control.

Nurses can help and support people so they do not feel so overwhelmed by developing a rapport with them; understanding who and they are and were; and asking about their concerns. This support can help people with ILD cope and adapt.

Nurses can empower people and improve their coping strategies through empathetic care and enhance self-esteem by encouraging people to solve problems for themselves. They can also teach cognitive coping strategies by encouraging people to carry on as usual and to modify their lifestyle to adapt to increasing breathlessness.

Encouraging people to engage in social interaction and to use distraction techniques - such as listening to music, reading and going to the cinema - are useful ways of making life worthwhile. As many people with ILD have an unpredictable condition with periods of exacerbations and remission, it is important to plan for the worst but hope for the best.

**MANAGING BREATHELESSNESS**

Breathlessness is usually the major problem reported by people with ILD, and is particularly prevalent on exertion.

Breathlessness resulting from ILD is usually associated with hypoxia, unlike in chronic obstructive pulmonary disease (COPD), when it is usually caused by a combination of de-ranged airflow dynamics, air trapping and, to a lesser extent, hypoxia.

Bronchodilators are the mainstay of treatment in COPD, but giving bronchodilator inhalers to people with ILD has little effect on breathlessness, unless they have coexisting COPD, which may be present in those with a long smoking history.

Lung function tests identify the degree of restrictive versus obstructive lung disease present and determine whether bronchodilator therapy will be helpful.

**Hypoxia and oxygen therapy**

It is important to assess the degree of hypoxia contributing to breathlessness. Pulse oximetry recordings at rest may be normal (93-98%). However, it is essential to take recordings when people are active too.

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**Keywords** Breathlessness | Oxygen Therapy | End of Life | Interstitial Lung Disease

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This article examines the problems of caring for people with interstitial lung disease in primary care. It describes simple strategies that can support people to remain at home during the end stage of their illness.

The diagnosis and investigations of ILD have been covered in previous Nursing Times articles (Duck, 2007a; 2007b).

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**PRACTICE POINTS**

- Accurate diagnosis is vital for the effective management of interstitial lung disease.
- People with the condition may experience feelings of loss of control and powerlessness.
- They should be helped to manage breathlessness.
- Ambulatory oxygen may be required at home to help people maintain normal activities.
- End of life care may need to be considered at the time of diagnosis for those with progressive ILD or with advanced disease at the time of diagnosis.

**Check understanding**

People with ILD should have been given a full explanation of their disease, including the prognosis, in acute care when the diagnosis was made.

It is important for nurses to ask people what they understand about their diagnosis so they will understand what their patients know.

By considering people holistically and separating symptoms from problems and anxieties, nurses can offer effective support.
For example, recording pulse oximetry while people are walking up the stairs helps in assessing hypoxia on exertion and how this affects the ability to maintain normal activities of daily living.

Many people with ILD desaturate (their oxygen levels fall) significantly while performing everyday activities and this impacts significantly on quality of life (Duck, 2006). It is not uncommon for those with ILD to desaturate to an SpO2 of 70% while performing normal activities of daily living.

It may be necessary to prescribe ambulatory oxygen therapy in the home. The British Thoracic Society (2006) guidelines make recommendations for prescribing ambulatory oxygen, including to those with ILD.

There are a variety of new oxygen delivery devices, including high flow nasal cannulas (available from home oxygen suppliers), the OxyArm device and masks that can be used to make it easier to tolerate the higher flow rates that is needed by many people who have ILD (Duck, 2006) (Box 1).

**END OF LIFE CARE**
End of life care starts at diagnosis for some people with ILD who have rapidly progressing illness or advanced disease at the time of diagnosis.

Those with IPF have a life expectancy of 2-4 years from diagnosis and it is important to place them on a general practice supportive care register. This means they have a nominated key worker, preferably a community respiratory nurse, who coordinates all their care and liaises with social and acute care.

Controlling increasing breathlessness is the major challenge for those who have advanced ILD and optimising oxygen therapy is fundamental.

It is possible to provide people with up to 15L of oxygen via concentrators, cylinders and liquid oxygen in the community. As these limits are reached and people start experiencing panic attacks with breathlessness, pharmacological symptom control must be offered. At this point, it is important to check that people have been placed on a supportive care register as it is likely they will die within 6-12 months.

Benzodiazepines are useful for moderating anxiety and panic. Sublingual lorazepam, used in acute breathlessness, is readily absorbed and has a short half life. Diazepam taken orally at night will help to reduce both anxiety and breathlessness. Small doses of morphine used four hourly can help relieve symptoms of breathlessness; however, regular use can cause constipation (Twycross and Wilcock, 1993).

People often need encouragement to use these drugs and nurses should not be frightened of using ones known to have a respiratory depressant effect. Generally speaking, people with ILD have a high respiratory drive that prevents them from retaining carbon dioxide until the final days of life. When people are dying, the priority becomes symptom control and, inevitably, the distressing symptom of breathlessness must be relieved at all costs.

**Liverpool Care Pathway**
The Liverpool Care Pathway (Ellershaw and Wilkinson, 2003) is designed to organise the last days and hours of life. This pathway challenges healthcare teams to notice when death is within days or hours.

In ILD, this requires health professionals to develop their clinical judgement about when this is happening. As oxygen requirements increase, multiprofessional discussions between colleagues in primary and acute care and with people and their families will all help in making decisions around end of life care.

**CONCLUSION**
Managing people who have ILD can be a rewarding experience for nurses. Practitioners should remember that some people do respond to treatment.

Accurate diagnosis can only be made with the input of a specialist radiologist and ILD team.

People with suspected ILD should be given the opportunity to receive care from specialist teams to make sure they receive appropriate treatment.

Nurses in primary care need to be aware of ILD as well as COPD when people present describing breathlessness on exertion. Early ILD is not detected on spirometry, so these people will need a chest X-ray to detect any abnormality.

There are ongoing clinical trials for people with IPF throughout the UK in the hope of developing a treatment for this aggressive disease. In common with people with lung cancer, those with IPF should be given the opportunity to enrol in these trials.

Early and accurate diagnosis will improve the management of those with ILD as, hopefully, will the findings from clinical trials.

**REFERENCES**