INTERSTITIAL LUNG DISEASES 1: DIAGNOSIS AND INVESTIGATION

The interstitial lung diseases (ILDs) are a heterogeneous group of over 150 disorders. They have clinical, radiological and pulmonary function features in common and, if untreated, can lead to lung fibrosis or scarring. They represent about 15% of the workload of respiratory physicians in secondary care, and the end stages are a substantial burden on primary care. The ILDs are not neoplastic but some have a poorer prognosis than some cancers.

An estimated 10,000 cases of ILD are diagnosed in the UK each year. Idiopathic pulmonary fibrosis (IPF) – formerly known as cryptogenic fibrosing alveolitis (CFA), sarcoid and hypersensitivity pneumonitis – also known as extrinsic allergic alveolitis – occur most frequently (Johnston et al, 2006).

For many respiratory nurses and chest physicians, ILDs are a confusing set of conditions. There are many reasons for this.

First, none of them are ‘common’ conditions so clinicians outside specialist centres will rarely see enough of any one condition to be completely comfortable with diagnosis and management. There are so many types that clinicians usually only encounter a few.

Second, classification of ILDs is somewhat complex. Names are long and complicated and it can be difficult to separate one type from another without expert opinion. In addition, the classification of a subgroup of ILDs (the idiopathic interstitial pneumonias) changed in 2002 and not all doctors and nurses are familiar with the new concepts.

Finally, there is no definitive, evidence-based management strategy so, even after securing a confident diagnosis, the most suitable strategy is uncertain. Clearly this is frustrating and unsatisfactory for both clinicians and patients.

For further reading, Ellis (2005) gives an excellent description of the types of ILD.

DEVELOPMENT OF UNDERSTANDING

Until relatively recently a clinical history, physical examination, a few relevant blood tests and a chest X-ray were the only tests available to differentiate the ILDs. In many cases no disease-causing agent could be identified so no specific clinical disease was identified. Clinicians were faced with a breathless patient, who may or may not have finger clubbing, crackles on listening to their lungs, a ‘hazy’ chest X-ray and often restrictive spirometry (although some demonstrate obstructive spirometry).

This led to a range of ILDs being grouped together and often labelled with the ‘older’ term of CFA or as lung or pulmonary fibrosis or fibrosing alveolitis. This was particularly seen in patients with different forms of idiopathic interstitial pneumonias (IIPs).

Treatment was often attempted with immunosuppressant drug combinations using prednisolone, azathioprine and/or cyclophosphamide. Many pioneering clinical teams carried out small-scale studies including Raghu et al (1991), Johnson et al (1989) and Turner-Warwick et al (1980). However, response rates were relatively poor, with only around one-third of patients at best responding to these often toxic drug combinations. Many experienced major side-effects with little objective clinical benefit. Understandably, some clinicians began to take the attitude that the treatment was not worth the associated risk. Although understandable, this was unhelpful to patients experiencing rapidly increasing breathlessness, fear for the future and feelings of hopelessness.

Thankfully, understanding has improved. Factors that have probably contributed significantly to this include:

- High-resolution CT scanning, which allows subtle differences in the pattern and distribution of lung damage to be identified between different diseases;
- Video-assisted thoracoscopic (VATS) lung biopsy techniques, which are safer and a less invasive way of obtaining lung tissue;
- Developments in histopathology allowing pathologists to differentiate small changes between lung biopsies and attribute them to separate disease processes;
- Clinical teams recognising the different clinical patterns seen in different diseases.

These factors led to a panel of experts re-examining and reclassifying the ILDs – particularly the IIPs. Idiopathic pulmonary fibrosis has also been reviewed recently.

Consensus documents were published by the American Thoracic Society and the European Respiratory Society (ATS and ERS, 2002; ATS, 2000), making evidence-based treatment and management recommendations. These should form the basis of all diagnostic, assessment and treatment plans.

DIAGNOSIS

Correct diagnosis is always important. It gives patients a label they can begin to understand and research and can help them come to terms with their disease. The clinical team can decide on treatments and give patients some idea of their prognosis.
It is not uncommon, however, for patients to be misdiagnosed because of the complexities in differentiating between ILDs and this may have consequences.

For example, clinicians may decide not to treat IF with prednisolone due to a perceived poor response rate and high side-effects from steroid use. This might be acceptable if the diagnosis is correct but, if patients have a different ILD, they may be denied drugs that could slow their disease progression.

Most regions in the UK have tertiary centres with teams specialising in ILD, and referral to these can permit review of cases and clarification about specific diagnosis.

A significant number of ILDs are likely to respond to immunosuppressant drugs or disease-specific therapies to some extent. This means it may well be worth a trial of treatment in many cases. However, the response may not always be overt, such as a clear improvement in lung function, quality of life or exercise capacity. In certain ILDs, the best to be expected from treatment is a slowing down of disease progression.

Some ILDs, in particular IF, appear resistant to current therapies so it is important to separate these from other forms of ILD. The current consensus is that patients with IF should be considered for ongoing clinical trials of new medications rather than simply being given usual immunosuppressant drugs such as prednisolone and azathioprine. Local tertiary referral centres will probably know which trials are available.

Johnston et al (2006) recommended that all patients with suspected ILD should, as a matter of course, be referred to a specialist centre for diagnosis and overall/shared management. There is evidence from lung cancer models that patients benefit from being grouped and treated together – even if there are no good treatment options. They provide support for each other and they also value advice from a range of clinicians who have specialist knowledge and feel comfortable diagnosing and managing patients within their disease specialty.

This approach also has advantages for clinical teams. They have the opportunity to acquire knowledge and experience quickly as they see large numbers of patients sharing the same patterns and problems. This aids their accumulation of knowledge of the rarer conditions and can lead to the development of new disease-specific management strategies. These might include production of information leaflets, education programmes, advice networks, patient support groups and links with palliative care services for management of end-stage disease or distressing symptoms. In addition, having larger groups of patients concentrated in one centre opens up the opportunity to be involved in trials of experimental treatments.

INVESTIGATING ILDs

Investigations undertaken on new patients might include:
- Medical history, including presenting problem, time and onset of symptoms;
- Drug, chemical or fumes exposure;
- Occupational history;
- Family history of lung disease;
- Physical examination;
- Blood tests/immunological screen to detect coexisting connective tissue diseases, rheumatoid arthritis or sensitisation to birds or infective agents such as aspergillus fungus (precipitins);
- Full pulmonary function tests including transfer factor;
- Exercise testing, such as shuttle walk test or six-minute walk test;
- Oxygen assessment, in particular ambulatory assessment;
- Chest X-ray;
- High-resolution computerised tomography;
- Bronchoalveolar lavage;
- Open lung biopsies or VATS biopsy (in some cases).

Once assessed, patients should ideally be discussed in an ILD multidisciplinary team meeting. Gathering the relevant information to make a specific ILD diagnosis is more likely to be secured within this environment.

The team should include at least one nominated respiratory physician, an experienced thoracic radiologist and a pathologist – all of whom should have a special interest in ILD – and a specialist respiratory nurse (Johnston et al, 2006).

The team will usually come to a consensus opinion on diagnosis and generate a management plan and appropriate treatment strategy.

More detailed information on ILD diagnosis and management can be found in the following documents: ATS (2000); ATS and ERS (2002); and British Thoracic Society (1999).

Part 2 of this unit, which examines the management of the three most common ILDs, will be published next week.

KEY REFERENCES


The full reference list for this part of the unit is available in Portfolio Pages on nursingtimes.net