Factsheet 26

Updated on 11 December 2020

Interstitial Lung Disease (ILD)

This factsheet has been designed for use by healthcare professionals only.

Introduction

Interstitial Lung Disease (ILD) refers to a lung condition that affects the alveoli and associated blood supply and lung tissue, which is different to the airways we associate with COPD or Asthma.
There are over 200 subtypes of ILD. Due to the complexity of the diseases, there is an international guidance classification document and recommendation that all patients with a suspected ILD are referred to a specialist ILD Multi - Disciplinary Team (MDT). This is to ensure the patient gets an accurate diagnosis, appropriate management stream and support.[1]

Broadly speaking the diseases can be subdivided into those with an identifiable cause and those without, known as Idiopathic.

The most common type of ILD is Idiopathic Pulmonary Fibrosis (IPF). This accounts for approximately 40% of all the conditions and is increasing. 6,000 patients a year are diagnosed with IPF and the stark reality is that average survival rate is 3-5 years from diagnosis.[2]

Pathology:

What is happening in the lungs in IPF is complex and poorly understood. We understand that it involves abnormal wound healing, following repeated injury to the alveolar region. The fibroblasts in the lung tissue, which are there to assist in healing, overproduce. This, plus other mechanisms, lead to a change in the normal architecture in the lung and therefore, have an irreversible fibrotic impact. It is thought that inflammation plays a less dominant role in this type of ILD. This is important as it helps us to understand why some of the treatments work and some do not. For example, IPF does not respond to the immunosuppression action of steroids, and has in fact been shown to be detrimental to the patient.

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Presentation:

It is more common in men over the age of 60, exposure to smoking increases their risk. The patient typically describes an insidious onset of cough - this accounts for 80% of patients and/or breathlessness on exertion. There may be weight loss, loss of appetite and fatigue too. Unfortunately, there is evidence that patients can present with these symptoms for up to 3-4 years before diagnosis or referral made to a specialist team.[1]

Patient History:

NICE 2017 advocate that everyone with suspected IPF need a detailed history, a thorough clinical examination and blood tests to help exclude the ILD with a known cause. For example, diseases associated with environmental and occupational exposure, other underlying connective tissue diseases and possible drug therapies.

Exposure history:

Establishing the patient's possible exposure is paramount in any respiratory history taking. We need to consider medications that patients may take that can cause an interstitial type lung disease. **This** is an excellent resource to check. Most common drugs are Nitrofuranotin, Amiodorone and some of the rheumatology drugs.

It is essential to explore what working environment the patient has been in throughout their working career and any hobbies they enjoy, past or present. Smoking history, passive or active, can impact this condition and family history is also important as there is evidence to support a family link in approximately 20% of cases.

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On examination:

Velcro crackles - The most common finding when examining the patient is crackles on inspiration, on both sides that can be described as "Velcro crackles" and is thought to be the earliest sign that can be picked up before the patient becomes much worse. Evidence also shows that 50% of patients may have clubbing of their fingers due to the chronic respiratory disease.[3]

Exertional desaturation:

Due to the damage being in the alveoli and surrounding blood supply, gas exchange can be affected, this is more common on exertion and is very debilitating for the patient. It is therefore useful to measure oxygen levels at rest and after exertion.

Appropriate tests:

Which tests performed in primary care will vary according to local policy.
Bloods are needed to rule out any known cause. These may include; a fibrotic screen, ANA, ANCA Rheumatoid Factor and serum Ace to rule out sarcoid. Allergy precipitins and inflammatory markers from the CRP are also required.
CXR – A chest Xray may or may not show any changes, but in progressed disease it may show fibrosis, particularly in the lower lobes. Occasionally a diagnosis can be picked up incidentally by the patient having a CXR.
HRCT – This is essential to confirm or refute any changes in the interstitial areas and can assist in the classification of the type of disease. The HRCT in ILD can show a very distinctive picture which is described as "honeycomb" at the periphery of the lung fields, again, on both sides.
Lung function - Findings can be different to that of COPD and Asthma. The patient can have normal spirometry, or obstructed spirometry but it typically shows a restrictive picture. This is due to the inability of the fibrosed and stiffened lung tissue being able to expand normally. Therefore, reducing the patient's ability to meet their predicted value of Forced Vital Capacity. (FVC). Once the patient is referred to the specialist team further lung function will be performed to establish

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the patient's ability to take on oxygen across the alveolar membrane. This is the transfer factor and is a good prognostic indicator.

Referral to Specialist team and confirmation of diagnosis:

Diagnosis can be very a difficult and protracted process for the patient, and it is essential they are involved in the decision making and fully aware of what is happening. The MDT have the responsibility of gathering all the information and making the decision regarding the confirmed or likely diagnosis. This would include the respiratory consultant, ILD nurse specialist, radiologist, pathologist, and thoracic surgeon.

If the multidisciplinary team cannot make a confident diagnosis from the patient's clinical features, lung function and radiological findings, some patients may require further investigations such as bronchoscopy. This enables samples of lung tissue and cells to be collected to examine type of changes within the samples. However, some patients will require a surgical biopsy with the thoracic surgeon.

Treatment options:

Drug Therapy - Pirfenidone & Nintedanib

There has been a significant amount of clinical studies and drug trials in ILD and NICE have approved both antifibrotic drugs Pirfenidone & Nintedanib.[4, 5] They have been shown to slow down the pace of the disease progression and therefore reduce the rate of the patients' functional decline. However, there are specific criteria the patient must meet before they can be considered. The patients FVC must be between 50% and 80% predicted.
This type of antifibrotic treatment is initiated in the tertiary centres under a shared care policy with local specialist teams. The patient requires close monitoring and support. There are common side effects that can be managed with specialist support and the patient requires frequent lung function. The guidance suggests that the treatment is stopped if the disease progresses.

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Pulmonary Rehabilitation (PR)

Patients need to be given every opportunity to understand their lung condition and provided with as many of the coping mechanisms possible to live well with ILD. This can be achieved by referring to the PR programme. The benefits of PR are well documented and has the same positive impact for this patient group.[6] The programme does need to reflect the needs of the IPF patient, in particular the education component and severity of exertional desaturation.

Best supportive care

This needs to be implemented and discussed at the point of diagnosis. It is essential that this is patient focused and tailored to the individual's disease severity, rate of progression, and the person's preference. Patients need appropriate and accurate information and support. We as health care professionals are responsible for guiding patients to the appropriate information. ILD generally is a misunderstood condition and there is limited knowledge both in the clinical arena and the public. But there are numerous patient self-help groups that are very proactive in supporting IPF and raising the profile.

- Action for Pulmonary Fibriosis
- British Lung Foundation
- Pulmonary Fibriosis Foundation

Managing difficult symptoms:

Shortness of breath, cough, fatigue, and weight loss can be extremely debilitating for the patient with IPF and they need support to manage the symptoms. Who delivers this support will depend on local knowledge and expertise, but typically, the patient will work closely with the ILD CNS/physio and the palliative care team.

ILD is a condition that requires specialist input; however, patients can present to any health care setting with symptoms of ILD. We need to raise the profile of early diagnosis and understanding of the disease. Therefore, it is everyone's responsibility to get this diagnosis right and quickly for the benefit of our patients.

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