Idiopathic Pulmonary fibrosis: Provision of Care in Iraq

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Idiopathic pulmonary fibrosis (IPF) has a prognosis that is worse than many cancers and its behaviour is unpredictable. Patients present with insidious onset of shortness of breath on exertion and dry cough. The FVC tends to decline by 200 ml/year and patients deteriorate over a two to three year period. The natural progression of the disease is hastened with development of acute exacerbations which carry a fifty percent one month mortality.[1] The majority of patients die from respiratory failure without treatment. Early diagnosis and treatment is essential to make an impact on survival and prognosis.

Approved treatments for IPF include, the anti-fibrotics, pirfenidone[2] and nintedanib.[3] Both drugs reduce the rate of decline of FVC by half compared to placebo. Real world data show that both anti-fibrotic medications are associated with a lower risk of all-cause mortality and hospitalizations compared to no treatment.[4] With the availability of these two treatments there may be an opportunity to influence the natural progression of this disease.

There is evidence to suggest that delayed diagnosis impacts patient survival.[5] Early referral to a specialist centre needs to be undertaken for those with known or suspected interstitial lung disease (ILD) since patients have a better prognosis than those who are referred late to the same centre regardless of the severity of the disease.

Specialist centres have access to multidisciplinary team (MDT) evaluation which is considered to be a reference standard for ILD;[6] it provides a forum that enables the integration of all available clinical, radiological and pathological data for diagnosis and subsequent management. It improves diagnostic confidence and agreement compared to individual participants of the MDT, thoracic radiologist and pathologist and ILD clinician working in isolation.[7] It allows confident diagnosis of IPF to be made and also rule out other diagnoses including connective tissue disease related ILD which have different treatment pathways.

Recently, there have been changes in the guidelines for the diagnosis of IPF. The high-resolution computed tomogram (HRCT) is important for the diagnosis of IPF; the radiological patterns include usual interstitial pneumonia (UIP) pattern, probable UIP pattern, indeterminate pattern and alternative diagnosis. The same terminology in these guidelines is applied for microscopic analysis of biopsied tissue. For patients with probable UIP, indeterminate UIP or alternative diagnosis, the guidelines provide conditional recommendation for the use of broncho-alveolar analysis or surgical biopsy.

In addition to improved access to anti-fibrotic medications and ILD MDT discussion, specialised centres may also provide increased access to cardiothoracic surgeons providing biopsies, access to clinical trials of novel therapies and robust links with lung transplantation units.

IPF patients have a heavy symptom burden,[8] often with significant fatigue, anxiety and depression. Patients should therefore be encouraged to be actively engaged and be supported to make informed decisions about their own care. Symptom control and management of breathlessness using oxygen therapy, psychological input, cognitive therapy, pulmonary rehabilitation program and morphine and anxiolytics are equally important.

It is also important to identify other causes of breathlessness in these patients. IPF is an illness of the elderly, occurring more predominantly in men and over the age of 60 years. These patients frequently have comorbidities including coronary artery disease, pulmonary hypertension and lung cancer,[9] that may be

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contributing to their symptomatology.

Recently the Bristol Interstitial Lung Disease had the ability to raise the profile of this disease and other ILDs in Baghdad with the support of Saifee-Burhani Medical Association UK and Royal College of Physicians London UK. It was evident that there was an enormous appetite to raise the standards of ILD patients in Iraq. Its imperative the infrastructure of this ILD Service is made available throughout the country; resourcing of equipment, post-graduate training of thoracic radiologists, pathologists, rheumatologist with specialist interest in connective tissue disease and ILD physicians and medications will result in provision of better equitable care of Iraqis with this condition.

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