ILD highlights from the ATS 2017 congress
May 19th – May 24th / Washington, DC

Developments in diagnosis and management

Background

IPF is an incurable disease with a disease course that can vary greatly between patients.[1] While there is no cure, pharmacological and non-pharmacological therapy options exist and, in an effort to optimise treatment of IPF patients, this area is being actively researched and is constantly developing.[2]

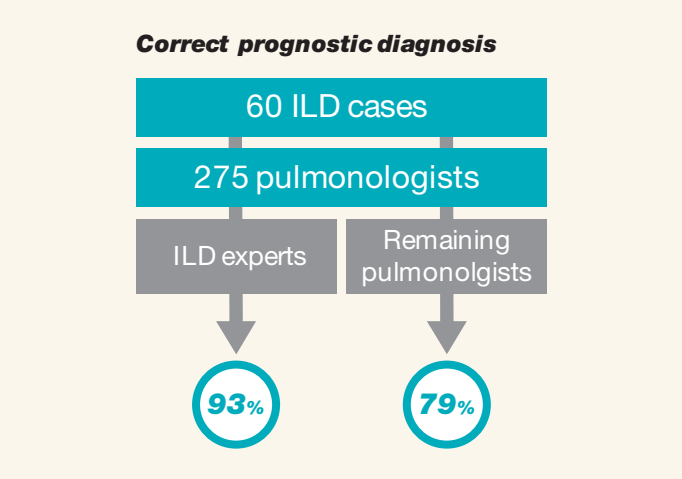
Diagnosis of idiopathic pulmonary fibrosis (IPF) is challenging and requires careful evaluation of clinical, laboratory, radiological and/or pathological data and systematic exclusion of other disease aetiologies. Thus, accuracy of IPF diagnosis improves with multidisciplinary discussion between pulmonologists, radiologists, and pathologists, particularly if the radiological and histopathological data are inconsistent.[3]

The importance of MDD in IPF diagnosis

The usefulness of multidisciplinary discussion (MDD) in IPF diagnosis was addressed in many presentations at this year's ATS congress.

MMD was found to influence diagnostic as well as therapeutic decisions.[4]

A study to evaluate accuracy of clinical IPF diagnosis found that prognostic accuracy was generally high, with ILD experts correctly diagnosing IPF more often than non-expert pulmonologists.[5]



It was also shown that interobserver agreement for IPF diagnosis is moderate among pulmonologists and greater between ILD experts, while interobserver agreement is poorer for conditions such as NSIP (idiopathic non-specific interstitial pneumonia) and HP (hypersensitivity pneumonitis).[6]

The addition of a rheumatologist to a multidisciplinary team (MDT) can lead to increased diagnostic accuracy through more effective differentiation between IPF and CTD (connective tissue disease). Additionally, rheumatological evaluation could help to reduce invasive procedures.[7]

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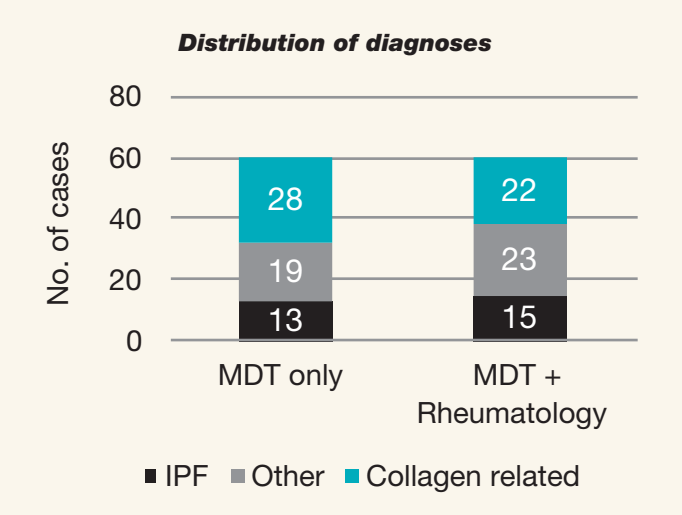

Interstitial lung abnormalities on CT

Interstitial lung abnormalities (ILA) visualised by computed tomography (CT) can indicate precursors of pulmonary fibrosis and should be closely followed over time.

CT scan data from the COPDGene study showed that ILAs on CT are associated with lower lung function, decreased quality of life and higher mortality. More interstitial changes were associated with a genetic component, the MUC5B promoter polymorphism rs35705950, especially in non-Hispanic whites.[8]

Combining clinical characteristics, MUC5B promoter genetic variant, and peripheral blood mononuclear cell (PBMC) gene expression profiles was most reliable in predicting early pulmonary fibrosis.[9]

In a cohort of patients with surgical resections for pulmonary nodules, ILA on chest CT were associated with subpleural fibrosis and fibroplastic foci.[10]

Awareness and treatment compliance

Although IPF and NSCLC (non-small cell lung cancer) have similar survival outcomes, fewer IPF patients initiate and continue treatment.[11] Compliance to antifibrotic treatments is relatively low (60% of commercially insured population, 51% or less in Medicare population).[12] Better awareness and education of physicians and patients on the benefit of early antifibrotic treatment and treatment persistence might improve outcomes for IPF patients.

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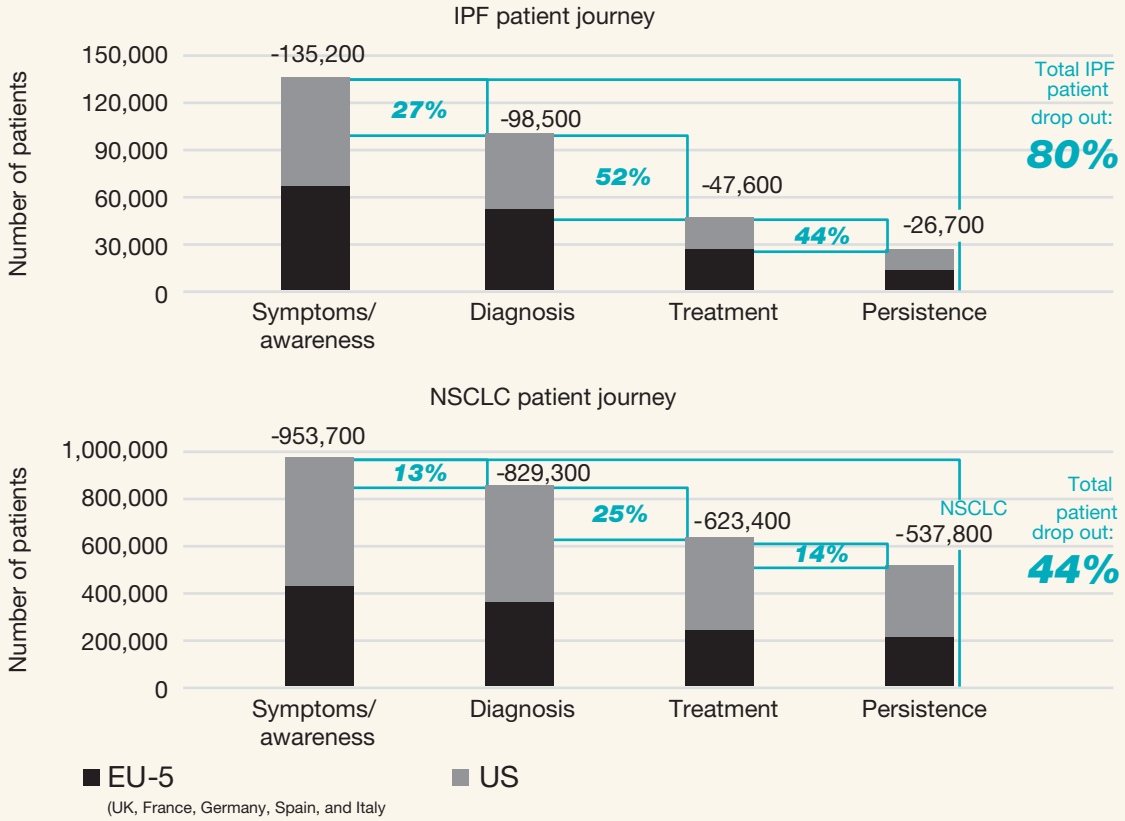
Patient 'drop out' at each stage of the journey



Quality of life in ILD patients

Several presentations at the ATS 2017 addressed the impact of ILD-associated symptoms and co-morbidities on disease related quality of life (QoL) in ILD patients. In a cohort of IPF and SSc-ILD (systemic sclerosis-associated ILD) patients, cough was more frequent, more severe and more often productive in IPF patients. Cough severity was related to dyspnoea and pulmonary function. In both cohort groups, cough was associated with poor quality of life.[13]

A study on the impact of depression on patients with mild-to-moderate IPF revealed that, although 39% of included patients had significant depressive symptoms, none were taking antidepressant medication. A strong association was found between depressive symptoms and disease severity, symptom burden and health-related quality of life (HRQoL).[14]

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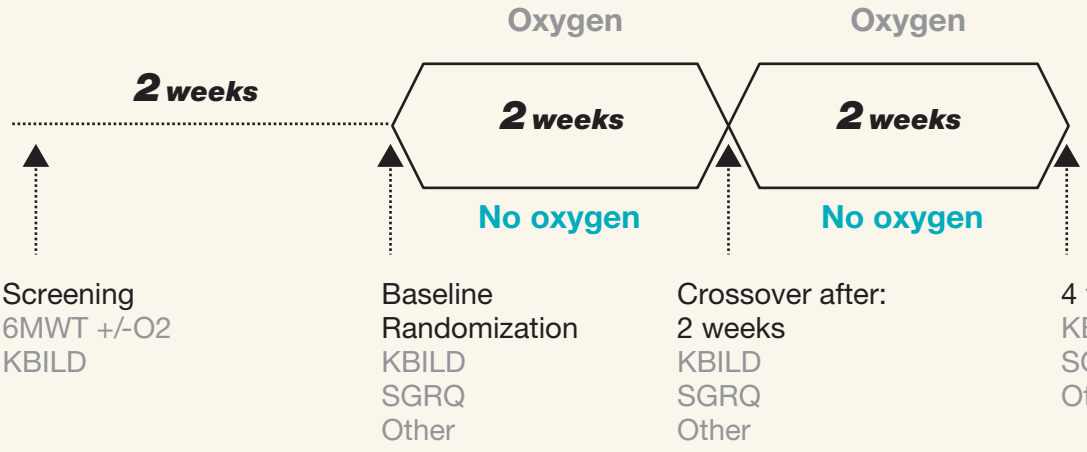
AmbOx – ambulatory oxygen in fibrotic lung disease

The AmbOx trial is a multicentre, randomised, cross-over controlled trial that assessed the effect of ambulatory oxygen on quality of life in patients with fibrotic ILD over two weeks. The beneficial influence of oxygen therapy on symptoms and quality of life in patients with fibrotic ILD shows that ambulatory oxygen use should be considered in future ILD-specific guidelines.[15]

AmbOx trial design



6MWT = 6-Minute Walking Test, K-BILD = King's Brief Interstitial Lung Disease Questionnaire

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