British Thoracic Society
London, 4–6 December

Biomarkers: key to new therapies for IPF

The main challenge facing respiratory specialists is that by the time they get to see patients with idiopathic pulmonary fibrosis (IPF), the lung tissue bears little or no resemblance to the normal lung. So said Dr Toby Maher of the National Heart and Lung Institute, London, as he made the case for building better clinical trials and validating biomarkers of disease progression and treatment response.

IPF causes approximately 4000 deaths in the UK every year. Median survival is approximately 3 years from diagnosis.

'There are several problems that we have encountered in trying to move IPF clinical trials forward,' he said. 'Defining the diagnosis is difficult. As it stands, we have a diagnostic classification system that doesn't work properly, and disenfranchises about 25% of patients from a definitive diagnosis of IPF or an idiopathic fibrosis of any other cause.'

However, he conceded, understanding of the pathogenesis of IPF has come on leaps and bounds.

'The problem is that there are very many mediators and pathways involved in this process, and we are not very good at discriminating between those that are upstream or downstream,' said Dr Maher. 'We don't have the pre-clinical tools to identify which are the key targets we should be pursuing.'

Dr Toby Maher, Honorary Senior Lecturer, National Heart and Lung Institute, Imperial College, London

He outlined details of a collaborative project involving respiratory disease centres at Nottingham and the Royal Brompton Hospital, London, the main aim of which is to identify new biomarkers that might be used to develop targeted drug therapies.

'Ultimately,' Dr Maher said, 'the integration of 'omics data, together with careful longitudinal phenotyping, is going to change our understanding of IPF, thereby aiding drug discovery and improving trial design.'

Stephen Pinn

Two-fifths of idiopathic pulmonary fibrosis patients may be denied optimal treatment

By following the 2013 National Institute for Health and Care Excellence (NICE) guideline and using only forced vital capacity as a basis for treating idiopathic pulmonary fibrosis, as many as two out of every five patients who might benefit from such treatment are missing out.

This was the conclusion of a presentation by Dr Nazia Chaudhuri, of Wythenshawe Hospital, Manchester, based on retrospective data from 50 patients on the hospital's named patient programme.

Dr Chaudhuri reminded her audience that following NICE approval, pirfenidone was not recommended for treating idiopathic pulmonary fibrosis patients with a forced vital capacity of 50–80%.

However, she and her colleagues estimated that 20 of these patients (40%) would not have been prescribed pirfenidone as per the NICE guideline – because their average forced vital capacity was significantly higher than to those who did meet the NICE criteria (97.6% *vs* 65.48%, $P<0.001$). In contrast, those 20 patients had a moderate, but not significant reduction in transfer factor (42.35 *vs* 44.74).

Although the presence of emphysema was lower in those who met the NICE criteria than in those who did not (three and eight patients respectively, $P=0.0409$), the average transfer factor was not significantly different between those with and without emphysema (35.75% *vs* 46.75%).

Dr Chaudhuri concluded that as measuring forced vital capacity alone appears to disadvantage a substantial number of patients with regard to eligibility for pirfenidone, both forced vital capacity >50% and diffusing capacity of the lungs for carbon monoxide >35% should be used in assessing these patients.

Stephen Pinn

Pirfenidone could reduce hospital costs for patients with severe lung disease

High levels of access to health services by the most severely ill cohort of idiopathic pulmonary fibrosis patients are stretching NHS resources – but introducing pirfenidone, an antifibrotic agent, into therapeutic regimens may help reduce this financial burden.

Dr Helen Parfrey from Papworth Hospital NHS Foundation Trust in Cambridge, highlighted this possibility after outlining data from a multicentre, retrospective cohort review undertaken across four NHS trusts.

Her conclusions were based on findings from 100 patients (76% male, mean age 69.3 years, 62 patients on oxygen therapy, mean predicted forced vital capacity 70%) at baseline, for which data were available for 67 at 9 months.

In the first 6 months, 11 patients had 15 idiopathic pulmonary fibrosis-related hospitalizations, of which six were for an acute exacerbation. One patient was hospitalized in the 6–9-month period.

Dr Parfrey emphasized that the possible beneficial effects of pirfenidone on hospital resources were still speculative, but warranted further assessment.

Stephen Pinn

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