Interstitial lung diseases: a view for the future

Ron du Bois, MD, PhD
Asmarley Professor of Respiratory Medicine
Royal Brompton Hospital & National Heart & Lung Institute
Imperial College of Science, Technology & Medicine, London, UK
e-mail address: r.dubois@rbh.nthames.nhs.uk

Ron du Bois is a Consultant Physician at the Royal Brompton Hospital and Professor of Respiratory Medicine, Imperial College, London UK. He developed his clinical and research interests in the genetics, pathogenesis and treatment of interstitial lung diseases. He is now head of the Clinical Genomics Group at Royal Brompton & National Heart & Lung Institute, London and visiting Scholar at National Jewish Medical & Research Center, Denver, Colorado. He currently serves on the Editorial Boards of several Journals.

Over the last five to ten years considerable progress has been made in our appreciation of the diffuse lung diseases (interstitial lung disease; ILD), particularly idiopathic pulmonary fibrosis (IPF) and sarcoidosis. For example, a number of studies have used the new ILD diagnostic criteria to construct more up to date disease definitions and these have confirmed that IPF, as now defined, carries a significantly worse prognosis than the hitherto accepted median survival of five years; other studies have shown that longitudinal change in lung function is a powerful predictor of survival, not just in IPF, but also in NSIP; and, most gratifyingly, this more robust approach to diagnosis has attracted the interest of the pharmaceutical industry and there have now been completed at least five prospective randomised placebo controlled double blinded studies of novel therapies in IPF – interferon γ; pirfenidone; bosentan; etanercept; and N-acetyl-cysteine with variable outcomes.

In sarcoidosis, new entities are emerging, including small fibre neuropathy and novel approaches to fatigue; genetics studies have resulted in an iterative approach to subsets of sarcoidosis that suggest that the umbrella term “sarcoidosis” shelters a number of distinct disease; and progress is being made in defining severity and more focussed treatment strategies.

This is a good start but what should our vision for the future be? Up to 1:3-4000 of the European population has a diffuse lung disease; although the situation is improving, these individuals remain disenfranchised, ill informed and with little therapy to even arrest the disease despite the progress of the last 10 years? Much resource has been applied to diseases with a much lower mortality rate but which are more attractive to pharmaceutical industry and government for different, and sometimes not so different, reasons. So, what can be done to improve the lot of patients with this group of diseases? The future vision should include:

- The development of consistent international registries, with governmental support, to define the extent of the problem and to collect cohorts of patients for clinical trials of novel therapies and clinical research
- To have National Framework agreements (UK nomenclature but concept applicable internationally) that provide the necessary skilled infrastructure that will allow quality of care to be uniformly of high quality across and between our countries
- This infrastructure will also help to define the health economics of this group of diseases
- A continuing iterative approach to disease classification
- How to define environmental exposures over time
- A DNA bank to provide power for genetic studies
- To refine our diagnostic precision/nomenclature. This will likely result in more weight being applied to CT scan data and longitudinal behaviour
- To look to the development of imaging that can track molecular mechanism; PET/CT is a good start but resolution needs to be even higher
- To dismiss finally the anecdotal approaches to treatment that have bedevilled any understanding of the relative pros and cons of any given agent; all patients who are eligible should be encouraged to enrol in clinical trials.