International Corner

Connective Tissue Disease-Associated Pulmonary Hypertension

Christopher P. Denton PhD, FRCP
Professor of Experimental Rheumatology
Royal Free Campus, University College London
Head of Scleroderma Service, Royal Free Hospital
London, UK

I was very excited to see a whole issue of *Advances in Pulmonary Hypertension* (Vol. 7, No. 2, Summer 2008) devoted to connective tissue disease associated pulmonary hypertension (PH). As a practicing rheumatologist in a center that manages a large cohort of more than 1000 scleroderma cases, and a center that benefits from having a pulmonary hypertension centre embedded within it, all of the topics covered in this issue were very relevant.

Scleroderma has the highest frequency of pulmonary hypertension of any of the rheumatic disease and is especially challenging to manage as cases often have co-morbidity that affects assessment and may lead to poor outcome. There are also particular challenges for diagnosis and assessment. All of these points were highlighted and I was especially pleased to see the emphasis that was put on autoantibodies as useful tests is assessing risk of developing pulmonary arterial hypertension (PAH) in scleroderma. Less invasive tests focused on at-risk groups will be essential for timely detection and treatment of PAH in connective tissue disease.

Scleroderma and the antiphospholipid syndromes were very clearly reviewed and the articles offered clear insight into the likely differences in frequency of PAH in these diseases and also the potential contribution of thrombosis to contribute. Being mindful of pathogenic mechanisms is likely to underpin better therapy in connective tissue disease associated pulmonary hypertension. One of the challenges faced daily in managing connective tissue disease associated PAH is the co-existence of interstitial lung fibrosis. The degree to which many patients should have PAH or PH secondary to lung fibrosis remains a topic of considerable debate and I was interested to see how this challenge was tackled in some of the major centres in the USA.

Without doubt, the most engaging and relevant part of this excellent issue was the round table discussion. Almost all of the important points raised are as relevant to practice in the UK and other European centres as they are in USA. It is clear from the answers to many key questions that this whole topic requires more research and better information. However, with exciting projects such as PHAROS for scleroderma and complementary exercises ongoing in Europe, I was left with optimism about clinical practice for patients with connective tissue disease-associated pulmonary hypertension, even if the outcomes for many patients do not seem to be as good as in idiopathic PAH.

In conclusion, there are more similarities than differences in the approach to PAH in Europe and the USA based upon these articles. This is a testament to the strong international collaboration in clinical trials and educational programs that exist in the fields of connective tissue disease and pulmonary hypertension.

Address for reprints and other correspondence: Christopher P. Denton, PhD FRCP, Professor of Experimental Rheumatology, Centre for Rheumatology, Royal Free Hospital, London,NW3 2QG. email: denton@rfhsm.ac.uk