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Interstitial Lung Diseases

Edited by

R.M. du Bois and L. Richeldi

EUROPEAN RESPIRATORY SOCIETY

Interstitial Lung Diseases

European Respiratory Monograph 46
December 2009

Editor in Chief
K. Larsson

This book is one in a series of European Respiratory Monographs. Each individual issue provides a comprehensive overview of one specific clinical area of respiratory health, communicating information about the most advanced techniques and systems needed to investigate it. It provides factual and useful scientific detail, drawing on specific case studies and looking into the diagnosis and management of individual patients. Previously published titles in this series are listed at the back of this book with details of how they can be purchased.

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The Guest Editors

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R.M. du Bois is Professor of Medicine in the Dept of Medicine at National Jewish Health, Denver, CO, USA, and Senior Research Investigator in the Dept of Population Genetics and Gene Therapy, National Heart and Lung Institute, Imperial College, London, UK. He was previously Consultant Physician at the Royal Brompton Hospital and Professor of Respiratory Medicine at Imperial College, London, UK. He qualified from Cambridge University and trained in general and respiratory medicine in London. After a 2-yr period as a visiting scientist in the Pulmonary Branch at the National Institutes of Health, Bethesda, MD, USA, he returned to London to develop his clinical and research interests in the genetics, pathogenesis and treatment of interstitial lung diseases at the Royal Brompton Hospital, where he was head of the Interstitial Lung Disease Unit and the Clinical Genomics Group. More recently, he has been appointed to National Jewish Health, Denver, to work on translational programmes of research.

R.M. du Bois has been Associate Editor for *Thorax* and the *American Journal of Respiratory and Critical Care Medicine*. He is a member of the American Thoracic Society (ATS), the European Respiratory Society (ERS) and the British Thoracic Society (BTS). He has served as a member of the BTS Orphan Lung Diseases Committee, the ATS Planning and Nominating Committees, and the ERS Council.

L. Richeldi is Associate Professor of Respiratory Medicine in the Dept of Oncology, Haematology and Respiratory Diseases, and Director of the Research Centre for Rare Lung Diseases, at the University of Modena and Reggio Emilia, Modena, Italy. Since qualifying with an MD from the University of Modena and gaining his PhD in Cardiopulmonary Pathophysiology from the University of Rome "La Sapienza", he has worked in the Dept of Tuberculosis and Respiratory Diseases at the University Hospital of Modena and been Director of the Postgraduate School in Respiratory Medicine at the University of Modena and Reggio Emilia.

L. Richeldi is a member of the editorial board for the *American Journal of Respiratory and Critical Care Medicine*. He is a member of the ERS study group "Diffuse Parenchymal Lung Disease", and has several roles within the ATS, being a member of the study group "Infections and tuberculosis", a member of the Microbiology, Tuberculosis and Pulmonary Infections Program Committee and a member of the steering committees for ATS guidelines on "Diagnosis of tuberculosis infection" and "Idiopathic pulmonary fibrosis". He is also actively involved within the Italian Society of Respiratory Medicine, the Cochrane Collaboration and the Italian Registry of Diffuse Lung Diseases.

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Preface

It is pleasure to present the fourth 2009 issue of the *European Respiratory Monograph* (*ERM*). For the second time, an issue is dedicated to interstitial lung diseases; the first *ERM* on this topic was published in 2000. As this is a rapidly growing field within respiratory medicine, with continuous novel research achievements, there is high motivation to publish a new *ERM* on interstitial lung diseases after nine years.

The current issue is somewhat differently structured compared with the former version. The editors have chosen an interesting approach in which they, after an introductory chapter written by two prominent scientists within the field, T.E. King Jr and U. Costabel, in the first section have focused on current challenges and questions, in the second section on diseases and in the third part have addressed some international aspects of this heterogeneous group of diseases. In the first part, aspects of morphology, cell biology and biomarkers have been extensively covered, as well as clinical topics such as exercise testing and clinical trials. The disease section has been expanded to include specific chapters on occupational interstitial lung diseases, drug-induced infiltrative lung diseases and interstitial lung diseases in children. In the third section, the situation regarding interstitial lung diseases in India and the Far East is addressed. In these areas, interstitial lung diseases are most likely under diagnosed, due to low awareness and low access to modern technical equipment such as computed tomography.

As the knowledge within the area of interstitial lung diseases is growing and the interest from pulmonary clinicians is increasing, it is felt timely now to publish this *ERM*. I very much appreciate that two of the most outstanding experts in the field accepted to serve as guest editors for this issue and that they were able to engage so many of their expert colleagues in this project. We know, from a recently performed market research survey, that the *ERM* is highly appreciated and read by the members of the European Respiratory Society, and it is therefore a pleasure to present this new *ERM* covering a topic that will be of immense interest to almost everyone who has an interest in pulmonary medicine.

Editor in Chief,
K. Larsson

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INTRODUCTION

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Interstitial lung disease (ILD) represents a heterogeneous group of distinct disorders, which are increasingly attracting the attention of the community of lung specialists. One of the characteristics of this specialist field of respiratory medicine is the rapid emergence and accumulation of new data on both basic mechanisms of disease and clinical interventions. Importantly, in the last decade, ILD entered the arena of randomised controlled trials, as a consequence of increased diagnostic precision combined with the burgeoning interest of pharmaceutical companies. In 2005, an excellent *European Respiratory Monograph* (*ERM*) was published on sarcoidosis, one of the most common and most challenging ILDs in clinical practice; now, in the present *ERM*, the whole spectrum of ILD is addressed in three broad areas: current challenges, international concerns and individual diseases.

The diagnosis and the management of ILD has become an increasingly popular topic for pulmonologists in general, as reflected in the growing body of literature and in the increasing attendances at the large number of scientific sessions that are dedicated to the different aspects of these diseases at major international respiratory conferences. An inevitable effect of this spread of interest has been the emergence of new challenges, ranging from the differential roles of the main diagnostic tools, to the problems raised by the completion of the first randomised trials and the maintenance of the momentum of clinical trials of new therapy in the future.

When we started to think about the structure of this *ERM*, we had a number of clear aims: to address those challenges that continue to be controversial or unresolved, such as the role of different investigational tools and the design of clinical trials; to cover specific diseases in an innovative, less formulaic fashion, with a strong emphasis on translational aspects; and to acknowledge the internationalisation of this increasingly popular subject by having a section that dealt with the concerns felt in different geographic regions. We also wanted to attract an authorship that combined expertise, the younger generation who are specialising in ILD and of course a broad international spread. We certainly feel that we have succeeded in our authorship goal. From the section on challenges and open questions, through the major clinical entities that are more likely to be encountered by respiratory physicians during their daily activity, to the concerns about ILD felt in the international community, we have received contributions that are innovative, comprehensive and thoroughly readable. Specific diseases covered include those of unknown cause, those associated with collagen vascular disorders and those occurring due to occupational or drug exposures; additionally, a chapter is devoted to the important field of paediatric ILD. In our third section, the globalisation of the network of specialists devoted to these diseases is covered. Expert colleagues, living in areas of the globe where the rapidly increasing availability of diagnostic and therapeutic tools is coupled with a solid scientific and clinical background, comment on the challenges represented by the identification and the treatment of patients with ILD in the most populated areas of the planet. This global approach to ILD also raises the

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problem of generalisation of recommendations in guideline documents and from clinical trials, and this topic is addressed objectively in the closing chapter. In this context, it is encouraging that the forthcoming new guidelines on the diagnosis and treatment of idiopathic pulmonary fibrosis will be the first authentically global document of this sort that involved the input of experts and scientific societies from all over the world.

We are honoured to have had the opportunity to be guest editors of this compilation of such excellent contributions. We are also happy and proud that two internationally recognised experts in the ILD arena accepted our invitation to write the introductory chapter to this *ERM*: their joint authorship spans two continents and is a reflection not only of their high expertise, but also of the positive globalisation process in place in this field of respiratory medicine.

Finally, we thank our wives and families for putting up with our hours in front of the computer, and our junior colleagues for continuing to educate us.