Diffuse Parenchymal Lung Disease

Progress in Respiratory Research

Vol. 36

Series Editor *Chris T. Bolliger*, Cape Town

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Volume Editors *Ulrich Costabel*, Essen
Roland M. du Bois, London
Jim J. Egan, Dublin

144 figures, 15 in color, and 47 tables, 2007

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Prof. Dr. Ulrich Costabel
Department of Pneumology and Allergy
Ruhrlandklinik
Tueschener Weg 40
DE–45239 Essen (Germany)

Prof. Dr. Roland M. du Bois
Interstitial Lung Disease Unit & Clinical Genomics Group
Royal Brompton Hospital
Sydney Street
London SW3 6NP (UK)

Dr. Jim J. Egan
The Mater Misericordiae Hospital
University College Dublin
Eccles Street
Dublin 7 (Irland)

Library of Congress Cataloging-in-Publication Data

Diffuse parenchymal lung disease / volume editors, Ulrich Costabel, Roland M. du Bois, Jim J. Egan.
p. ; cm. – (Progress in respiratory research, ISSN 1422-2140 ; v. 36)
Includes bibliographical references and indexes.
ISBN-13: 978-3-8055-8153-0 (hard cover : alk. paper)
1. Interstitial lung diseases. I. Costabel, Ulrich. II. Du Bois, Roland M. III. Egan, Jim J. IV. Series.
[DNLM: 1. Lung Diseases. 2. Lung Diseases, Interstitial. W1 PR681DM v.36 2007 / WF 600 D5695 2007]
RC776.I56D54 2007
616.2′4–dc22

2007010588

Printed in Switzerland on acid-free paper by
Reinhardt Druck, Basel
ISBN 978–3–8055–8153–0, ISSN 1422–2140

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Foreword

The book series *Progress in Respiratory Research* was started in 1963 and is enjoying increasing success. Since I took over from my predecessor Prof. H. Herzog in 1997, my vision was to cover the whole area of thoracic medicine. In contrast to standard text books, however, this series aims at providing cutting-edge knowledge including the most recent advances in the field discussed. This necessitates three preconditions: top quality experts as volume and chapter authors, a strict enforcement of submission deadlines, and a fast printing process by the publisher. I am happy to say that all of these requirements have been ful-filled in this current 36th volume of *Progress in Respiratory Research* devoted to diffuse parenchymal lung disease (DPLD). The choice of the topic was made easy as none of the previous volumes have ever addressed DPLD, an important area in pulmonary medicine, where a lot of progress has been made during the last couple of years. It was therefore a timely topic to choose. The choice of terminology for the volume title was a deliberate one; the classic designation interstitial lung disease has been replaced by the more modern and more accurate term DPLD.

As usual, the most important task of the Editor-in-Chief after the choice of the topic of a volume was the choice of the volume editor(s). I was fortunate enough to have the immediate support of three well-known experts in DPLD – Ulrich Costabel, Jim Egan and Ron Dubois – who shared the task of compiling the content and choosing the best possible authors to write the individual book chapters. As you the reader can easily see, the result has been a fantastic book covering all the important aspects and including the majority of leaders in DPLD at a global level. True to the vision of the book series, authors were instructed to include the latest references available at the time of writing. The book is aimed at all doctors from general practitioners to pulmonary physicians with a special interest in DPLD; there is something for everyone. For a more specific introduction of the various topics please see the preface by the volume editors.

My thanks go to the three editors as well as all the chapter authors, and, once again, to the editorial staff at the publishing house, S. Karger AG, Basel, Switzerland. The publisher has proved his quality once again by printing a high-quality book in the shortest possible time after receipt of the final manuscript, well done guys!

C.T. Bolliger
Cape Town

Preface

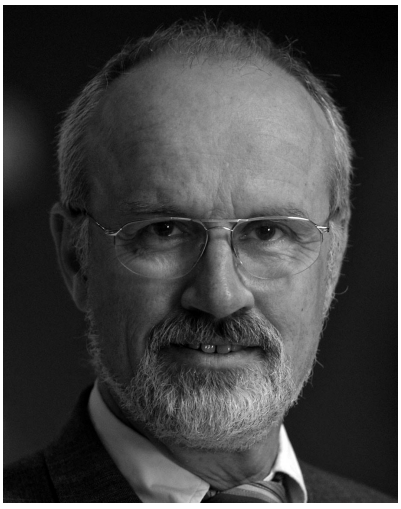
Diffuse parenchymal lung diseases (DPLD) represent a large and heterogeneous group of disorders, many of them belonging to the category of orphan diseases. Our knowledge on DPLD has expanded greatly during the last decade. New insights into the pathogenesis and new techniques such as high-resolution CT scanning have made significant contributions to a better understanding of these less common disorders which remain an intellectual challenge to clinicians, radiologists and pathologists. DPLD comprise over 200 entities of known and unknown causes, with or without associated systemic disease, of acute or chronic onset, of indolent or rapidly progressive course, and wide variations in treatment response. The management of DPLD remains difficult, but correct diagnosis, appropriate treatment and a balanced assessment of prognosis are important. This issue of *Progress in Respiratory Research* aims to provide valuable information on the rapid advances in this field.

The first section covers general topics including the most recent classification system, the general diagnostic approach, and the clinical evaluation of the patient including radiology, histopathological patterns, and bronchoalveolar lavage findings. The concept of classification of DPLD has undergone significant change over recent years. The idiopathic interstitial pneumonias now comprise seven distinct diagnoses which have all been previously regarded as forms of idiopathic pulmonary fibrosis/cryptogenic fibrosing alveolitis. Even the current classification is controversial, since disorders related to cigarette smoking such as DIP and RBILD are included without being truly idiopathic. In the diagnostic evaluation CT scanning can sometimes provide highly disease-specific information so that surgical lung biopsy can be avoided. Diagnosis cannot, however, be based on imaging or histopathology alone, but needs precise clinical information, so that the necessity of a multidisciplinary clinical/radiological/pathological approach in making the final diagnosis has been emphasized.

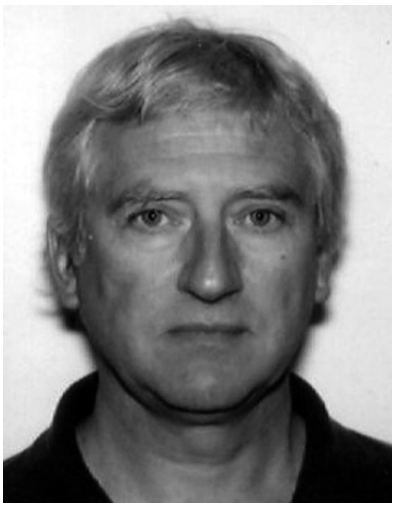
Several chapters deal with the basic aspects of DPLD. Genetic factors that pre-dispose to DPLD and define clinical disease phenotypes have been characterized in sarcoidosis and systemic sclerosis, whereas this is far less advanced in idiopathic fibrosing lung diseases. The basics of granuloma formation, fibrogenesis in idiopathic pulmonary fibrosis, and mechanisms of vasculitis are highlighted. A special chapter addresses novel aspects of treatment for interstitial lung diseases with an emphasis on idiopathic pulmonary fibrosis.

A large section of the book is devoted to specific diseases, covering granulomatous disorders, idiopathic pulmonary fibrosis and other idiopathic interstitial pneumonias, collagen vascular diseases, drug-induced infiltrative lung disease, diffuse alveolar hemorrhage, and even rarer disease entities such as the eosinophilic pneumonias, Langerhans' cell histiocytosis, lymphangioleiomyomatosis and pulmonary alveolar proteinosis. Bronchiolitis and lymphoproliferative lung disorders are also included, since their clinical and radiological manifestations mimic diffuse parenchymal lung disease. The last two chapters are focused on DPLD in children and lung transplantation for end-stage fibrosis, two topics which deserve special consideration.

The editors have recruited international experts from the various disciplines involved in these disorders to



U. Costabel, Essen



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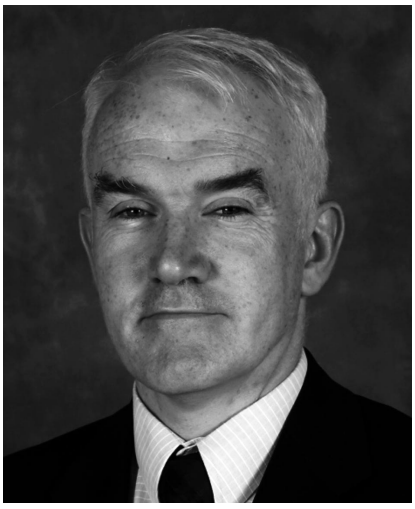

J.J. Egan, Dublin

contribute state-of-the-art reviews to this book. We hope that the readers will find it a helpful tool in the daily management of their patients with DPLD and an invaluable information source.

We thank Chris Bolliger for providing us with the opportunity to update a very exciting area in the field of lung diseases. We are grateful to S. Karger publishers for being extremely supportive in getting the book published. Our special thanks go the authors; we appreciate the high quality of their contributions and also their willingness to add the latest and hottest news to the bibliography of their chapters.

Ulrich Costabel, Roland M. du Bois, Jim J. Egan