
Foreword

Most of the volumes of the book series *Progress in Respiratory Research* deal with subjects relating to adult pulmonology. To address the needs of the pediatric community the most recent volume, No. 33 in the series, covered a long overdue topic, namely *Paediatric Pulmonary Function Testing*, and was launched just a couple of months ago. When planning the next volume I thought it would be an appropriate and logical sequel to have an update on cystic fibrosis, a topic which interests both pediatricians as well as adult pulmonologists.

True to the vision of our series, the volume should not be yet another textbook, but rather a state-of-the-art overview of the most recent advances in the field. As usual I asked one of the leaders in the area to be the volume editor. When I proposed this to Andi Bush, he enthusiastically accepted but then came back saying he would like to do this book together with four other volume editors. Although I agreed I was quite worried that with so many ‘cooks’, the book would never materialize. How wrong I was! Not only did the five editors manage to share the work without any problems, they also

made sure that the authors delivered their papers on time, and that their contributions represented truly cutting-edge research. This is – among other things – illustrated in the larger number of most recent references of 2004 and even 2005, which has always been my aim for the series.

The usual speed and quality of the publisher, S. Karger AG, Basel, guaranteed that the book was printed within the shortest possible production time.

Looking at the many different aspects of cystic fibrosis covered, the final result is a magnificent book, which will appeal to many more specialists than just pediatric and adult pulmonologists.

All that remains for me to say is well done and a big thank you to the authors, editors, and all people involved at the publisher! The many readers of this volume, No. 34 in the *Progress in Respiratory Research* series, will appreciate its quality.

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Preface

Less than 70 years ago, cystic fibrosis (CF) was a disease that was uniformly fatal in the first year of life, and that could only be differentiated from other gastro-intestinal diseases at autopsy. Over the years, advances such as the development of the sweat test enabled greater diagnostic accuracy and the beginnings of understanding of at least some aspects of pathophysiology.

The real explosion in the knowledge of fundamental airway biology and CFTR function came with the identification of the CFTR gene in 1989. Since then, the tools of molecular and cellular biology, transgenic animals and modern physiology, combined with big strides in modern, multidisciplinary care, have challenged virtually every previously held concept of the disease. The diagnosis is no longer a matter of a positive sweat test as the gold standard: atypical forms of the disease are being recognized. Far from dying in babyhood, patients are increasingly surviving into old age. Treatment goals have moved from the sole (laudable) focus of dealing with symptoms, towards the development of genotype-specific, molecular therapy. Even the pathophysiology is being challenged; the sweat test, which has stood us in such good stead diagnostically, may have deceived us into thinking that all manifestations of CF are related to chloride transport.

There are numerous large and excellent standard textbooks on CF; what is the need for yet another tome? We believe that the rapid advances in CF have reached the point where keeping abreast of research in scientific and clinical

areas has become a major challenge for the individual. There is a need for a concise and up-to-date summary of the current knowledge in all the various areas in which the study of CF is being pushed forward.

In this volume, we feel fortunate to have been able to bring together the finest scientists and clinicians to present a state of the art in their respective fields. They have assumed a basic knowledge of the subject; this is not intended to be a comprehensive text book of CF, and the reader will not find extensive reviews of valuable, but older work. The authors have been tasked to write brief chapters, citing mainly only recent literature, and to make their subject accessible to workers in the field of CF from other disciplines. The aim is that the reader will by the end of the volume be up to date in all of the key areas in this rapidly expanding field. We are very grateful to our authors for the enthusiasm and skill with which they have tackled their tasks. We have certainly learned a huge amount from editing this book, and with due modesty, believe that as a result of their efforts, this volume will be of interest to anyone working, or intending to work, in any area of CF.

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