

## Pulmonary Vascular Disorders

# Progress in Respiratory Research

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**Chris T. Bolliger** Cape Town

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# Pulmonary Vascular Disorders

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## Foreword

The aim of *Progress in Respiratory Research*, the so-called blue series, has always been to cover a broad range of topics in respiratory medicine. When looking for a topic for the next volume to follow the most recent one on *Antituberculosis Chemotherapy* (vol. 40), I realized that we never had an entire book dedicated to pulmonary vascular disorders during my tenure as series editor. I actually had to go back all the way to 1990, i.e. 22 years ago, to find the last such volume entitled: *Pulmonary Blood Vessels in Lung Disease*. While paging through it, I was stunned to realize how much progress has been made during the last two decades. On the therapeutic side for primary pulmonary hypertension, the only options that were discussed back then were the use of warfarin and a first hint at calcium channel blockers, which probably would have to have been administered in high doses. It was thus clear to me that we urgently needed a state-of-the-art update in that field.

To ask Marc Humbert to become a volume editor was an easy choice, and to my delight he not only accepted the task but also brought along two other eminent specialists, Rogerio Souza and Gérald Simonneau, as volume co-editors. As usual for *Progress in Respiratory Research*, the

'who's who' were invited from all over the world to contribute chapters to the current book and cover the entire range of pulmonary vascular disorders. Careful attention was paid to have the latest scientific findings reported.

The result is an outstanding stand-alone volume which – as the volume editors mention in the following Preface – should appeal to health care providers interested in the field, ranging from nurses to top specialists in the field.

I hope that you as the reader will enjoy the read, and would like to express my gratitude to the volume editors and chapter authors, as well as the editorial team at Karger Publishers, in particular Linda Haas and Stefan Sessler, who helped us all to have this book out in time.

*C.T. Bolliger, Cape Town*

## Preface

World health is generally improving with fewer people dying from infectious diseases, and therefore chronic diseases are more prevalent. In that context, the burden of chronic respiratory diseases is increasing and conditions such as asthma and chronic obstructive pulmonary diseases are clear public health priorities worldwide. Pulmonary vascular diseases and their consequences on right heart function also contribute markedly to the global burden of chronic pulmonary diseases.

Pulmonary hypertension has previously been called an orphan disease, that is, a condition which affects few individuals and is overlooked by the medical profession and pharmaceutical companies. Although undoubtedly rare, the concept that pulmonary hypertension is overlooked cannot be considered to be the case today. Indeed, there have been a number of important discoveries in recent years that have significantly improved our understanding of the disease, helped guide patient management, and laid foundations for future research. Since the middle of the twentieth century, amazing achievements have been made in the field from the development of right heart catheterization techniques to the first description of 'primary' pulmonary hypertension and the progress achieved as a result of the National Institute of Health Registry and the World Pulmonary Hypertension conferences which have taken place only four times: in 1973 (Geneva, Switzerland), 1998 (Evian, France), 2003 (Venice, Italy), and 2008 (Dana Point, California, USA). The more recent set of European guidelines have been jointly published by the European Society of Cardiology and the European Respiratory Society and endorsed by the International Society for Heart and Lung Transplantation. These guidelines give a robust hemodynamic definition of pulmonary hypertension as an

increase in mean pulmonary arterial pressure  $\geq 25$  mm Hg at rest, as assessed by right heart catheterization. These guidelines also provide a very clear classification of the five major clinical subcategories of pulmonary hypertension. Of these, pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension have seen the most rapid advancements in terms of knowledge and treatment options in the past decades.

By contrast, pulmonary venous thromboembolic disease is one of the most frequent diseases encountered in clinical practice, and diagnostic and treatment options have been revolutionized by large studies allowing robust recommendations in terms of diagnosis, prevention, and treatment. Only a small minority of patients with acute pulmonary embolism (sometimes subclinical) will develop chronic thromboembolic disease, a major cause of pulmonary hypertension which can be cured by pulmonary endarterectomy, an outstanding but difficult surgical procedure developed in a few expert centers.

We have come a long way since 1973 when the World Health Organization sponsored the first international meeting on a mysterious condition named 'primary pulmonary hypertension', spurred by the interest created by the sudden increase in patients who had used the anorexigen aminorex fumarate. It remains widely believed that pulmonary hypertension is a rare disease. Although true for idiopathic pulmonary arterial hypertension, a condition affecting less than 15 individuals per million inhabitants in Europe, the true burden of pulmonary hypertension is currently unknown and largely underestimated. In the developing world, highly prevalent diseases such as schistosomiasis in Brazil are associated with an increased risk of pulmonary hypertension. In addition, patients with



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sickle cell disease, human immunodeficiency virus infection, liver cirrhosis, autoimmune diseases, and congenital heart diseases are at risk for pulmonary hypertension. Moreover, hypoxia is a major worldwide risk factor for pulmonary hypertension. The predominant causes of hypoxia are inadequate oxygenation of arterial blood as a result of either lung disease, impaired control of breathing, or residence at high altitude. Indeed, more than 140 million individuals live above 2,500 m worldwide, including more than 80 million in Asia and 35 million in South America.

Altogether, the burden of pulmonary vascular diseases is certainly underestimated worldwide and requires much attention from the medical community. Better characteri-

zation and management of the subjects displaying pulmonary vascular conditions is of major importance in order to improve patients' outcomes. We hope that this book will be helpful to nurses, medical trainees, and medical doctors from various fields (cardiology, internal medicine, intensive care medicine, pneumology, thoracic surgery, rheumatology, etc.) who witness the burden of pulmonary vascular diseases in their daily practice.

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