

FOURTH EDITION

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PULMONARY CIRCULATION

DISEASES AND THEIR TREATMENT

Edited by
Andrew J Peacock
Robert Naeije
Lewis J Rubin

 **CRC Press**
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Dedication

To my wife Jila and my children Leila, Johnnie, and Vita, who have put up with so much over the years and never questioned my love for science and medicine even though their own interests lay elsewhere.

Andrew J. Peacock

To my wife Marie-Thérèse, my preferred coworker, and to mentor Henri Denolin (1915–2005), who was one of the first presidents of the European Society of Cardiology and constantly reminded his colleagues of the importance of the pulmonary circulation and the right ventricle.

Robert Naeije

To my family for their support, and to our patients, who serve as a constant reminder of the importance of our commitment.

Lewis J. Rubin

We would like to thank our colleagues from around the world who have been an inspiration to this book and in many cases contributors to it. Whether we work in Respiratory Medicine or Cardiology, we all share the view that the connection between the heart and lungs is important.

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Preface

The consequences of disturbed function of the pulmonary circulation remain an enigma to most clinicians. There is disturbance of pulmonary circulatory function in nearly all cardiac and pulmonary disease, yet this is rarely recognized or treated. The reasons for this relative obscurity when compared, for example, with the systemic circulation, are clear. The pulmonary circulation is difficult to examine clinically, and the tools we have for pulmonary circulatory measurement are either crude or invasive, or both. Even when we make invasive measurements with a cardiac catheter, we learn only about the circulation in artificial surroundings (the catheter laboratory), in an unrepresentative position (supine on the table), and in a state of artificially restricted activity. Clearly, we need clinical measurement techniques that will allow us to pursue pulmonary circulatory function in all states of human activity, both in normal people and in those with cardiorespiratory disease.

These are coming, but even now we know much about the pulmonary circulation that can help us in looking after patients in the ward and in the intensive care unit. Fortunately, there has been a great deal of research into the structure and function of the pulmonary circulation. We already understand much of its physiology and pathophysiology, and recently a number of effective therapies have been developed, tested, and put into clinical use with great effect. In this book, a distinguished group of authors, most of whom have a clinical background, have presented what is known about the pulmonary circulation in a readable and, more important, clinically relevant fashion, so that the information can be understood by practicing physicians. This is the fourth edition, with many completely new sections but a similar though more clinically orientated format. The book has been written especially with busy clinicians in mind, particularly those in respiratory medicine, cardiology, pediatrics, and intensive care. It has been deliberately structured so that a subject can be appreciated at any level from the purely clinical right down to the biochemical. This allows the reader to start with a chapter about a particular clinical issue and then, if interested, to pursue that subject from the clinical to the physiological to the biochemical level as he or she desires. Each chapter stands alone, so some repetition is inevitable, for which we make no apology, but this hierarchy of structure will, we hope, make the book accessible without diminishing the quality of the information that is presented.

For the fourth edition of *Pulmonary Circulation*, we have included the latest clinical, pathophysiological, and pathological research on pulmonary circulatory disorders. The editors believe that the brisk pace of discovery and development warranted updating the prior edition of *Pulmonary Circulation* in order to preserve its value as a resource for those interested in this field. In particular, we have provided a greater emphasis on the role of the right ventricle in pulmonary vascular disease, updated knowledge on pathobiology and genetics, and added new material regarding imaging and other diagnostic modalities. Perhaps the most significant advances in this field over the past few years have been achieved in medical therapy, and we have provided new sections on guanylate cyclase activators and combination therapy, in addition to updates on other therapeutics classes and treatment algorithms. Other therapeutics advances, such as balloon dilatation for inoperable chronic thromboembolic disease, are discussed in detail. We have also included the new classification and all the recommendations from the World Conference on Pulmonary Circulation in Nice, France, in 2013 and the latest guidelines from the European Society of Cardiology and the European Respiratory Society.

We are deeply appreciative to our colleagues who have agreed to serve as contributing authors to this book, most of whom are internationally recognized for the work about which they have written. We recognize the commitment of time and effort required to produce a scholarly chapter, and it is their willingness to contribute their expertise that makes this text authoritative.

We hope that *Pulmonary Circulation* will remain, in this fourth edition, a useful reference for the pulmonary hypertension specialist as well as clinicians and trainees alike in the fields of pulmonology, cardiology, pediatrics, and intensive care.

Finally, we wish to express our deep appreciation and gratitude to Kay Conerly and Jennifer Blaise at Taylor and Francis, our publishers, for their support and guidance through the editing process. Their expertise and professionalism helped make an arduous task easier.

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