

## Preface

The major function of the lungs is gas exchange and it does this using a low resistance circulation. The pulmonary circulation (or the pulmonary vasculature) is a unique system that differs dramatically from the systemic circulatory system (e.g., coronary, cerebral, renal arteries) in structure, function, and regulation. A typical example of functional differences between the pulmonary and systemic vasculature is that hypoxia causes pulmonary vasoconstriction but systemic vasodilation. Furthermore, in patients with systemic arterial hypertension (e.g., essential hypertension), pulmonary arterial pressure is normal, while in patients with idiopathic pulmonary arterial hypertension (previously referred to as primary pulmonary hypertension), systemic arterial pressure is usually within the normal range. The divergent vascular responses to hypoxia and the alternative existence of systemic or pulmonary arterial hypertension in patients indicate that the pulmonary vasculature or the pulmonary circulation is unique in terms of its anatomic and histological structure, physiological and pharmacological properties, genetic and epigenetic development as well as cellular and molecular determinants for vasoconstriction, vascular-wall remodeling, and embolus formation. Therefore, the pathogenic mechanisms of pulmonary vascular diseases are rather different from those of systemic circulatory disorders. Development of therapeutic approaches and improvement of clinical management for patients with pulmonary vascular diseases should be directed by understanding the unique physiological and pathological features of the pulmonary vasculature at organ, tissue, cell, and molecular levels.

Although many books have addressed clinical aspects of cardiovascular diseases, systemic arterial hypertension and basic science progress about structural and functional studies on systemic arteries (e.g., coronary, cerebral, and other peripheral arteries and microcirculation), very few books have focused on the pulmonary circulation and pulmonary vascular disease. Given the significant differences between the pulmonary and systemic vasculature and between systemic and pulmonary vascular diseases, it is urgent to have a comprehensive reference book specifically designated to describe *a*) basic structure and function of the pulmonary vasculature or the pulmonary circulation, *b*) pathophysiology of the pulmonary circulatory system, and *c*) clinical aspects (diagnosis, treatment, and prevention) of pulmonary vascular diseases.

*Textbook of Pulmonary Vascular Disease* is therefore designed for and is of special interest to *a*) clinicians (pulmonologists, cardiologists, intensive care physicians, cardiothoracic and vascular surgeons, and emergency physicians), *b*) physician-scientists and basic-science researchers in the fields of cardiopulmonary and critical care medicine, vascular physiology and pathophysiology, translational medical research, and bioengineering, *c*) healthcare workers in cardiopulmonary and critical care medicine, and *d*) clinical and research fellows as well as residents, medical and graduate students. *Textbook of Pulmonary Vascular Disease* combines basic scientific concepts and knowledge on the pulmonary circulation with clinical diagnosis and treatment on pulmonary vascular diseases. *Textbook of Pulmonary Vascular Disease* is unique in that no book currently available *i*) focuses on elucidating the cellular and molecular regulation of normal pulmonary vasculature and the pathogenic mechanisms of pulmonary vascular diseases, *ii*) includes advanced techniques and technology for basic and clinical research, and *iii*) includes conventional and molecular approaches currently available for

diagnosis and treatment of patients with pulmonary vascular diseases. Another feature of the book is the inclusion of surgical approaches for treatment of pulmonary vascular diseases, which have not been well described in previously published books.

*Textbook of Pulmonary Vascular Disease* is divided into five parts. Part 1 (Structure, Function and Regulation), consisting of nine sections and twenty-nine chapters, is designated for basic knowledge and recent findings related to pulmonary vascular structure, function, and regulation at levels of molecule, cell, tissue, organ and system. Part 2 (Methodological Approaches for Research) is composed of six sections and sixteen chapters that are designed to provide a basic knowledge and spectrum on the techniques and technology that are commonly used to study genetic, molecular, cellular, systemic and pathophysiological aspects of the pulmonary vasculature. Part 3 (Pathology and Pathobiology) includes four sections and nineteen chapters that discuss the potential mechanisms or sequence of events involved in the initiation and progression of abnormalities in the pulmonary vasculature in patients with pulmonary vascular disease. Part 4 (Pulmonary Vascular Diseases) consists of nine sections and thirty-two chapters devoted to describe pathogenesis, epidemiology and pathophysiology of almost all of the pulmonary vascular diseases identified so far by the World Health Organization (WHO). One of the sections is specifically designated to describe pulmonary vascular disease in pediatric patients. Part 5 (Diagnosis and Treatment) includes three sections and twenty chapters designed to illustrate, in details, the diagnostic and therapeutic procedures currently used for patients with pulmonary vascular disease.

*Textbook of Pulmonary Vascular Disease* is written by more than 220 experts in the field including physicians, surgeons, epidemiologists, bioinformaticians, nurses, physician scientists and investigators. All of the contributors are actively involved in clinical, physiological, and pathophysiological studies on the pulmonary circulation and pulmonary vascular diseases. The vast majority of authors are recognized experts in the research area of the topic on which the chapter is based with many contributors also Fellows of the Pulmonary Vascular Research Institute, a not-for-profit, international scientific association focused on the pulmonary circulation and pulmonary vascular disease. Founded in 2007, the Pulmonary Vascular Research Institute (PVRI) has assembled clinical, epidemiological, translational and basic scientists from around the world to perform research and advance education regarding pulmonary vascular disease and right heart failure. There is a focus on performing research in and providing education and treatment to underserved populations of the world. The unique strength of the PVRI is that it brings together a multidisciplinary faculty from around the world within a single focused institute. PVRI members have the expertise to conduct basic, translational, and clinical research at a level that no single academic institution can offer. The publication of an authoritative textbook on pulmonary vascular disease was an initial goal of the Institute.

*Textbook of Pulmonary Vascular Disease* will not only serve as a reference book for physicians, surgeons, private practitioners, translational medical researchers, clinical and research fellows, and medical and graduate students, but also can be used as a guidance manual for technical and marketing personnel in pharmaceutical and biotechnological companies, that are interested in clinical and basic science research in cardiopulmonary diseases, pulmonary vascular diseases, vascular biology, and lung/heart transplantation.

We hope that this book will also allow readers to foster new concepts and new collaboration and cooperation among clinicians, physician scientists and investigators so as to further understand the pathogenic mechanisms of pulmonary vascular disease and develop novel therapeutic approaches for the disease.

La Jolla, California  
May 9, 2010

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<http://www.springer.com/978-0-387-87428-9>

Textbook of Pulmonary Vascular Disease

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S.L.; West, J.B. (Eds.)

2011, XXXII, 1658 p. 345 illus., 224 illus. in color.,

Hardcover

ISBN: 978-0-387-87428-9