BOOK REVIEW

Pulmonary Hypertension
Edited by M. Humbert and J.P. Lynch
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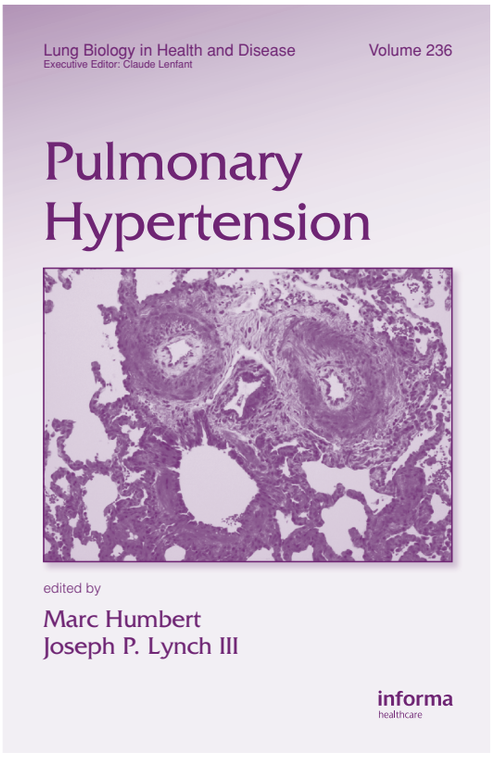

Pulmonary hypertension (PH) is an area of clinical medicine and medical science that is exemplary in many aspects, with tremendous progress having been made in a relatively short period of time. Identification of this condition was made possible in 1950 with the introduction of right heart catheterisation. The interest of the medical community really began in 1973 with the first international meeting dedicated to "primary" pulmonary hypertension, when the incidence of PH had notably increased due to the use of the anorexigen aminorex fumarate. Although idiopathic pulmonary arterial hypertension (PAH) is a rare condition, the global burden of PH as a whole is largely underestimated. In 2009, PH is no longer only a disease common in young females often related to the use of anorexigen, but may be encountered as a result of a growing list of conditions or risk factors. When present, PH has been proven to affect the long-term prognosis of many chronic diseases, including chronic obstructive pulmonary disease and idiopathic pulmonary fibrosis. With progress in pathophysiology, diagnosis, classification, treatment, management and prognosis, pulmonary hypertension has been "adopted" by the medical community, and is no longer an orphan disorder. Given the amount of recently accumulated data and recent changes in classification and treatment recommendations, the release of a comprehensive textbook will undoubtedly be useful for the growing number of physicians with an interest in pulmonary vascular medicine.

This book is part of a series of monographs edited by C. Lenfant, entitled *Lung Biology in Health and Disease*, which fully reaches its double objective of: 1) providing an up-to-date and comprehensive review of clinical management from diagnosis to treatment; and 2) critically reviewing the pathobiology of the disease, with a large section dedicated to the generation of hypotheses and suggestions for future investigations.

All the authors are well recognised leaders in their field and have contributed to the ever growing literature about PH. They review the evidence and share their experience when evidence is poor. As opposed to previous editions on "primary" PH in the series, the many aetiologies of PH are covered in this completely new version.

The first section of the book reviews the recently updated "Dana Point" classification of PH and the epidemiology of disease, and reflects changes regarding the newly recognised causes of PH (*e.g.* use of metamphetamine, left heart diastolic dysfunction), recent improvement in genetics of PH (*e.g.* hereditary PAH), and more detailed aetiological descriptions (*e.g.* congenital heart disease) or new subcategories (*e.g.* chronic haemolytic anaemia). Pathophysiology of PH is reviewed in depth, with contributions from both animal models and human studies, which have led to establishment of the role of site-specific angioproliferation and endothelial cell dysfunction resembling that of cancer growth in many aspects, and the role of the immune system, and that have paved the way for the currently used therapy and a number of potential targets for future therapy. The chapter on pathology of PH is particularly enjoyable and informative, with attractive images. The chapter on genetics exemplifies how identification of mutations in rare familial forms of the disease has prompted significant progress in understanding the molecular basis of PAH as a whole.

The subsequent chapters review, in a very clear and informative manner, how PH may be suspected, investigated and diagnosed, with the role of echocardiography for screening and noninvasive assessment, and the decisive and undisputed importance of right heart catheterisation for establishing the diagnosis and contributing to aetiological and prognostic assessment.

The authors of the 12 subsequent chapters systematically review all the clinical aetiological variants of PH, *i.e.* idiopathic, associated with connective tissue disease, congenital heart disease, HIV infection, portopulmonary hypertension, schistosomiasis, chronic haemolytic anaemia, pulmonary veno-occlusive disease/pulmonary capillary haemangiomatosis and chronic mountain sickness. These chapters will be tremendously useful for clinical practice, as they provide in-depth understanding of how the aetiological context affects prognosis and management decisions, with much progress having been made in recent years. For example, screening for PH is recommended in patients with systemic sclerosis or dyspnoeic HIV-infected patients; also, particular caution is required when considering PAH-specific therapy in patients with pulmonary veno-occlusive disease.

The chapter on chronic thromboembolic PH, the only curable variant of the disease, is of particular interest given the specificities of management with need of referral to a specialised centre to consider thromboendarteriectomy; the comprehensive and well-illustrated chapters on imaging of PH and of surgical management of PH complement this approach. The following chapters allude to areas yet to be explored in

more detail, such as severe PH associated with chronic lung disease (chronic obstructive pulmonary disease, idiopathic pulmonary fibrosis, combined pulmonary fibrosis and emphysema and histiocytosis X, to list a few). PH associated with chronic heart disease, which may occasionally develop with severe pre-capillary PH out of proportion to the left heart disease, may have been added to this list, although the subject is controversial and borderline to the areas covered by this book.

Therapy of PH, often too briefly covered in reviews, is nicely detailed in this book within nine chapters, which clearly describe the rationale and the current evidence for the use of prostacyclins, endothelin receptor antagonists, phosphodiesterase inhibitors and calcium channel blockers. Important chapters relate to the current recommendations, the current evidence and treatment decisions regarding combination therapy, and reflections about the treatment goals, which are now becoming more ambitious with the objective of improving long-term survival and quality of life. Much clinical research needs to be performed to help treatment decisions in clinical practice, as experience and opinion-based medicine still largely contribute to the medical choices.

The chapter about the design of past and future therapeutic trials will undoubtedly be useful for critical appraisal of available evidence and planning of future, and increasingly sophisticated, trials. So-called "conventional therapy" is also reviewed, as well as various difficult and challenging situations, such as PH in children, PH in pregnancy and management of PH in the intensive care unit. Separate chapters deal with the role of atrioseptostomy and lung transplantation.

In conclusion, this state-of-the-art and comprehensive review of PH has set the stage for future research, be it clinical, fundamental or translational. M. Humbert, J.P. Lynch and the authors should be commended for reaching the challenging objective of this book being both a basis for translational research and a useful textbook for clinicians managing all aspects of PH.

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