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Book Review

Pulmonary Vascular Disorders

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Edited by: M. Humbert, R. Souza and G. Slmonneau; Published by: S. Karger AG, Basel, Switzerland; 2012; Hard Cover; Pages: X+290 [82 figures (21 in colour), 45 tables]; Price: CHF 212.00/EUR 177.00/ USD 249.00; ISBN 978-3-8055-9914-6, E-ISBN 978-3-8055-9915-3

The book provides latest information on the rapidly-growing field of acute and chronic pulmonary vascular disorders. Recent years have witnessed a plethora of information in this field. Topic of pulmonary circulation is ever-challenging for those involved with evaluation, diagnosis and management. Development, optimisation and availability of several tools for diagnosis and management of pulmonary embolism and pulmonary hypertension have provided practicing guidelines to facilitate their uniform application worldwide. Chapters of the book have been written by authorities in their respective fields around the world who are involved in research, education and patient care in the field of pulmonary vascular disorders. This book will be useful to internists, cardiologists, chest physicians and thoracic surgeons, nurses and students.

Chapter 1 details updated clinical classification of pulmonary hypertension. It has 5 tables. Venice clinical classification of pulmonary hypertension (2003) is described in table 1. Table 2 describes updated clinical classification of pulmonary hypertension (fourth World Symposium held in Dana Point, USA, 2008) and the main modifications to the Third World Symposium on Pulmonary Arterial Hypertension (held in Venice, Italy) are set in bold. Tables 3 and 4 detail updated risk factors and associated conditions for pulmonary hypertension and details an anatomic-pathophysiologic classification of congenital systemic-to-pulmonary shunts associated with pulmonary artierial hypertinsion (PAH).

Chapter 2 describes pathology of PAH. This chapter has two useful figures on idiopathic PAH and one figure on pulmonary veno-occulusive disease (PVOD).

Chapter 3 details invasive rest and exercise haemodynamics in the modern management of pulmonary vascular disease. The chapter has 5 figures and 6 tables. Table 1 provides evidence-based clinical indications and utility of invasive haemodynamic measurements. The haemodynamic definition of pulmonary hypertension and pulmonary capillary wedge pressure in this chapter are useful. In addition, this chapter provides useful description of exercise testing in the catherer laboratory.

Chapter 4 provides information on exercise testing in pulmonary arterial hypertension. This chapter highlights the essential role of invasive pulmonary haemodynamics in diagnosis, assessing prognosis and monitoring the effect of therapy in pulmonary vascular disease. Coloured figure 3 of this chapter provides graphic representation of a cardio-pulmonary exercise test in idiopathic pulmonary hypertension and is easy to understand.

Chapter 5 deals with non-invasive exploration of the pulmonary circulation and the right heart. It describes role of three basic non-invasive modalities: echocardiography, magnetic resonance imaging (MRI) and computed tomography (CT). While the role of CT is limited to diagnosis of pulmonary thrombo-embolism and lung disease, echocardiography is an invaluable tool for diagnosis and follow-up of pulmonary hypertension, whereas MRI is a three-dimensional imaging modality that provides an excellent information on measurements of right ventricular volume and mass with greater precision.

Chapter 6 on bio-markers in pulmonary arterial hypertension provides the rationale of integration of existing biomarkers in evaluation and follow-up of PAH patients. This chapter describes several bio-markers including uric acid, troponins, asymmetric dimethylarginine, endothelin, D-dimer, von Willebrand factor and natriuretic peptides.

Chapter 7 details several mutations associated with PAH and provides guidance for screening. Chapter 8 deals with drug- and toxin-induced PAH. This chapter provides an important update on the association of PAH and the use of appetite suppressants and association of PVOD with drugs such as Mitomycin-C etc. Chapter 9 covers description of idiopathic PAH in developed and developing countries. Chapter 10 details PAH complicating connective tissue disorders (CTDs). The chapter specifically describes the pathophysiology, various clinical manifestations and present and promising therapy for CTD-associated PAH with a special emphasis on scleroderma-associated PAH.

Chapter 11 describes role of viral infections in PAH. While the role of human herpes virus-8 (HHV-8) in aetiology of PAH is controversial, a causative role of human immunodeficiency virus (HIV) has been defined. Hepatic disease and/or portal hypertension may have major consequences on pulmonary vasculature. Chapter 12 describes portopulmonary hypertension (PoPH) and

hepatopulmonary syndrome (HPS). Table 1 of this chapter provides important distinctions between HPS and PoPH. The chapter emphasises main role of orthoptic liver transplantation (OLT) in HPS and contraindication in PoPH. Table 2 of chapter 13 on pulmonary hypertension in congenital heart diseases is useful. The chapter quite adequately covers pathophysiology, definition and classification, epidemiology and genetics, assessment and management. Chapter 14 describes pulmonary hypertension in sickle cell disease. Several studies during the last decade have described prevalence of PAH up to 30% based on tricuspid valve regurgitation jet velocity $\geq$ 2.5m/s, however, the chapter also details two studies of low prevalence of PH in sickle cell disease based on right heart catheterisation.

Chapter 15 details schistosomiasis and pulmonary hypertension. The chapter highlights schistosomiasis as the most prevalent cause of worldwide cases of PAH. The chapter reviews schistosomiasis-associated PAH, epidemiology, mechanisms of the disease, clinical and haemodynamic features and management. This chapter is most relevant to endemic areas of schistosomiasis.

Chapter 16 covers a rare disease, Pulmonary Veno-Occlusive disease (PVOD) and emphasises its close similarity with idiopathic PAH and initial and incorrect diagnosis in 5-10% of cases. Table 1 highlights these differences very well and emphasises that high resolution computed tomography (HRCT) may be suggestive but histopathology is required for confirmation. Figures 1 and 2 are reader-friendly for understanding of concepts. PAH-related therapy may deteriorate PVOD with a risk of pulmonary oedema. Chapter 17 reviews pulmonary hypertension and left heart disease. Specifically, it covers epidemiology, pathophysiology, risk factors and treatment controversies concerning related to heat failure with mitral and aortic valvular disease; decreased and preserved left ventricular ejection fraction. Chapter 18 details pulmonary hypertension in chronic obstructive pulmonary disease (COPD). The chapter is important as prevalence of COPD is increasing worldwide. Pulmonary hypertension is a common complication of COPD and its presence is associated with decreased survival. COPD is by far the most common cause of pulmonary hypertension. The chapter describes definitions, prevalence of pulmonary hypertension in COPD, pathology, pathophysiology, diagnosis. In addition, the chapter details main features of PH in COPD, severe and out of proportion PH, evolution and prognosis of PH in COPD and treatment of PH in COPD. Main emphasis of treatment section is on long-term oxygen therapy (LTOT) and further reiterates paucity of drug treatment trials for PH due to COPD.

Chapters 19, 20 describe pulmonary hypertension complicating interstitial and granulomatous lung disease, and high altitude pulmonary hypertension.

Chapters 21 and 22 describe acute pulmonary venous thromboembolic disease and anticoagulation for venous thromboembolism in the modern management era respectively. Both chapters are useful to several disciplines as venous thromboembolism is encountered in several settings. The chapters adequately cover risk factors, diagnosis, risk stratification, and treatment. Long-term outcome is also described. Table 1 of the chapter 22 on anticoagulation reviews the newer oral anticoagulants and is useful to the readers. The chapter highlights the need for head-to-head comparison between new oral anticoagulants. The chapter also discusses several challenges associated with their use. In addition, the chapter provides information on recent improvements and unresolved issues. The chapter does not discuss data from studies in prevention of stroke in atrial fibrillation or in acute coronary syndrome.

Chapter 23 describes chronic thromboembolic pulmonary hypertension. The chapter appropriately defines the disorders as a dual vascular disorder comprising major vessel thrombotic obstruction, and classical small-vessel arteriopathy. The chapter emphasises the underestimation of true incidence of this disorder. Chapter 24 details medical treatment of PAH. Its table 1 is important as it provides useful information on currently licensed treatments for PAH. Description of treatment algorithm for PAH in figure 2 is useful.

Chapter 25 (Lung transplantation and role of novel extracorporeal support in pulmonary hypertension), Chapter 26 (Atrial septostomy), Chapter 27 (Pulmonary vascular disorders in hereditary hemorrhagic telangiectasia) deal with newly emerging and exciting fields. Chapter 28 is important for Future Perspectives in Pulmonary Arterial Hypertension. It describes anticipated approaches to research in pathogenesis, diagnosis, monitoring and novel therapies.

S.K. Sharma
Editor, IJCDAS
and
Chief, Division of Pulmonary, Critical Care and Sleep Medicine
Head, Department of Medicine
All India Institute of Medical Sciences
New Delhi - 110 029
Phone: 91-011-26593303 (O), 91-011-26594415 (O)
Fax: 91-011-26589898
E-mail: sksharma.aiims@gmail.com,
SKSharma.aiims@yahoo.com