Preface

Interstitial lung disease (ILD) is a broad category of lung diseases that includes more than 150 disorders characterized by scarring or fibrosis of the lungs. Even among the many types of the disease, ILD's progression can vary from person to person, and people respond differently to therapy. In the past, emphasis in treating ILDs has focused on the effect on gas exchange and loss of lung volume. This is a direct effect of the damage to the interstitium. However, an important indirect effect is on the pulmonary vasculature with resulting pulmonary hypertension. The association between interstitial lung disease and pulmonary hypertension has long been recognized, it was often associated with hypoxia and fibrosis alone. Recent studies that demonstrate response to pulmonary vasodilators stresses the vascular component of this process. In this book, we examine the various interstitial lung diseases. We also examine the incidence and outcome of pulmonary hypertension in the various interstitial diseases.

The book is divided into two main sections. The first discusses general issues. Drs. Carbone and Bottino introduce both ILD and associated pulmonary hypertension in the first two chapters of the book. The next chapter is by Drs. Meyer and Raghu, who discuss the evaluation of idiopathic interstitial lung diseases. They point out that this includes not only idiopathic pulmonary fibrosis, but other conditions such as nonspecific interstitial pneumonitis and cryptogenic organizing pneumonia. Drs. Moreira and Travis provide a detailed analysis of the pathology of the various ILDs. The pathologist often has the final say about what disease, although a comprehensive approach the clinician, radiologist, and pathologist gives a better definition of many cases. Finally, Drs. Carbone and Bottino summarize the evaluation of pulmonary hypertension. Although most of the information available is from patients with primary pulmonary hypertension, the observations can often be extended to patients with ILD.

The other section of the book deals with specific categories of disease. Dr. Lynch and colleagues discuss bronchiolitis, an increasingly recognized problem leading to airway obstruction and restriction. The use of inspiratory and expiratory high-resolution computed tomography scan has markedly enhanced the recognition of this process. Dr. Selman and his group then discuss hypersensitivity pneumonitis, a diffuse group of diseases bound together by common clinical and pathological features.

Drs. Brown and Strange discuss the collagen vascular diseases. Scleroderma has been one of the most widely studied lung diseases that can cause both interstitial lung process as well as pulmonary hypertension. In the past few years, large clinical trials have been published showing the benefits of some forms of therapy in these diseases. Dr. Martinez discusses the specific problem of pulmonary hypertension with idiopathic pulmonary fibrosis. Because idiopathic pulmonary fibrosis is associated with a high mortality, treatment for this complication may have major impact on the disease. Dr. Lee Newman examines the interstitial lung diseases associated with various occupational exposures. This divergent group can have a quite variable outcome. However, as a group it represents a major part of the differential diagnosis of all patients with interstitial lung diseases.

Dr. Baughman and colleagues discuss sarcoidosis. This multi organ disease affects the lungs in more than 90% of cases. Although most patients do well, there is a group with persistent pulmonary disease. Up to half of these patients will have pulmonary hypertension. Drs. Baughman, Lower, and Engel provide an evaluation for the disease and treatment strategies for the disease and associated pulmonary hypertension.

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