Classification of Pulmonary Hypertension

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Abstract

Pulmonary hypertension refers to a variety of conditions characterized by elevations in pulmonary arterial pressure. Major advances in the understanding of PH have led to the current classification in which PH diseases are grouped into five categories according to cause and therapeutic strategy, with each category subdivided to reflect diverse underlying etiologies and sites of injury. The five major categories of the Venice classification include PAH, pulmonary venous hypertension associated with left heart disease, PH associated
with hypoxemia, PH due to chronic thrombotic and/or embolic disease, and PH due to miscellaneous causes. One notable change in the current nomenclature is that the term “idiopathic pulmonary arterial hypertension” (IPAH) has replaced “primary pulmonary hypertension.” The WHO functional classification standardizes the comparison of clinical severity among patients with PH. Experts have embraced both the clinical and functional classification systems. Future attempts at refining the classification of this constellation of diseases are likely to embrace new insights into the molecular mechanisms and genetics of PH.

**Key Words:** pulmonary hypertension classification; idiopathic pulmonary hypertension; connective tissue disease-associated pulmonary hypertension; chronic thromboembolic pulmonary hypertension; pulmonary circulation; pulmonary vascular pathology.

### 1. INTRODUCTION

The term “pulmonary hypertension” (PH) denotes various conditions in which pulmonary arterial pressure is elevated above normal. The original classification of PH, established at a World Health Organization (WHO) Symposium in 1973, categorized the disorder as “secondary” when an identifiable factor was deemed causal and “primary” when no underlying etiology or risk factor could be identified (1). However, these terms were problematic because the actual “cause” of secondary pulmonary hypertension was often no clearer than with the “primary” group, and the two types were often very similar in presentation, histopathology, and response to therapy. These concerns and the extraordinary advances in the understanding of PH led to a revision of the classification at the Second World Symposium on Pulmonary Hypertension held in Evian, France, in 1998. The Evian group defined PH as a pulmonary artery pressure $\geq 25$ mm Hg at rest or $\geq 30$ mm Hg with exercise and categorized different types of PH based on clinical similarities, focusing on the biological expression of the disease as well as etiological factors. The Evian classification has subsequently been widely accepted, proving particularly useful in clinical practice and for drug evaluation and registration (2).

More recently, PH was again reclassified by a group of experts at the Third World Symposium on Pulmonary Arterial Hypertension held in Venice, Italy, in 2003 (Table 1). The Venice classification preserved the philosophy and architecture of the Evian classification but included important revisions, most notably the abandonment of the term “primary” pulmonary hypertension. In the current classification,