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Clinical Classification of Pulmonary Hypertension

I read with interest the recently published Dana Point clinical classification of pulmonary arterial hypertension (PAH) in the *Journal* supplement (1). Any globally relevant scheme of clinical classification cannot and should not lose sight of the prevalence of the disease state worldwide. Although the classification is not based on prevalence, and causes such as left ventricular systolic and diastolic dysfunction, in which PAH is not of primary importance, are kept in group 2 only, downgrading PAH associated with congenital heart disease (CHD) in their classification scheme to 1.4.4 seems unjustified. Although precise epidemiological data are not available for most causes of PAH, it is easy to understand the importance of PAH due to CHD on a global scale. In 2009, nearly 80% of the world's population lives in the less and least developed regions of the world. These regions also have a younger population profile, higher growth rates despite high infant mortality, and poor facilities for the management of CHD. Because the incidence of CHD is more or less uniform at 6 to 8 per 1,000 live births, these data easily would translate into a very high burden of PAH associated with CHD in the world, more than any cause listed in group 1.4. Most patients with CHD in the Western world undergo surgery early in life, and therefore this facet of CHD is less and less evident there, but the various aspects of the pathogenesis of PAH associated with CHD are also far from resolved. In view of the continued epidemiological, scientific, and pedagogical importance, PAH associated with CHD should be classified as 1.4.1 or even higher.

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Reply

The relevance of congenital heart disease (CHD) in the setting of pulmonary arterial hypertension (PAH) is unquestionable, as stated in the letter by Dr. Kothari. However, it is important to emphasize that the core structure of the updated clinical classification of pulmonary hypertension (1) is not based on the relevance of the topic or on the prevalence of the disease; thus, there is no hierarchical level within each one of the groups. The fact that schistosomiasis-associated PAH is classified as item 1.4.5 does not make it more or less important than human immunodeficiency virus infection–associated PAH, classified as item 1.4.2. The same can be directly extrapolated to the other 4 groups. Consequently, one could not consider that CHD-associated PAH has been downgraded from the previous classification. This classification has even strengthened the role of CHD by describing not only the anatomic-pathophysiologic classification of congenital systemic-to-pulmonary shunts associated with PAH (see Table 4 in the classification [1]) but also the clinical classification of congenital systemic-to-pulmonary shunts associated with PAH (see Table 5 in the classification [1]) to disseminate the current knowledge on the management of such relevant conditions.

The prevalence of the different forms of pulmonary hypertension is extremely important for the appropriate understanding of the whole pulmonary hypertension scenario, mainly considering that regional characteristics, such as local altitude, sanitary conditions, and health care infrastructure and organization, may play a significant role (2). Even considering that this is not related to the order in the updated classification, the commentary from Dr. Kothari should be considered as a reinforcement of the need for robust registries of the different forms of pulmonary hypertension (3) to serve as a basis for international collaborative efforts in the pulmonary hypertension field.

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