Abstract

This volume presents a global overview of the role of ion flux via transmembrane ion channel proteins in the regulation of pulmonary vascular tone and in the vascular remodeling processes associated with pulmonary vascular disease—offering a comprehensive review of the multiple families of ion channels that have been identified and characterized in pulmonary artery smooth muscle, as well as a practical discussion of experimental tools for the study of ion channel physiology and molecular biology.


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Emerging ion channel targets. The identification of heterozygous loss-of-function mutations in the KCNK3 (potassium channel subfamily K member 3) gene that encodes TWIK-related acid-sensitive potassium channel 1 (TASK1) as a cause for PAH has revived interest in the concept of channelopathy [62]. In addition to voltage-gated potassium channels, different types of transient receptor potential channels, calcium sensor proteins and calcium-activated chloride channels have been implicated in PAH pathogenesis [63–67]. This metabolism–epigenetics axis facilitates adaptation to a changing environment in the pulmonary vasculature and right ventricle, providing a potential novel therapeutic target.