



Endothelial cells in the pathogenesis of pulmonary arterial hypertension

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Dysfunctional endothelial cells play an important role in initiating and progressing PAH by impacting on multi-aspects of vascular remodelling and vasoconstriction. EC-targeted therapy may be effective in inhibiting vascular remodelling to treat PAH. <https://bit.ly/3nXPvMB>

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Abstract

Pulmonary arterial hypertension (PAH) is a devastating disease that involves pulmonary vasoconstriction, small vessel obliteration, large vessel thickening and obstruction, and development of plexiform lesions. PAH vasculopathy leads to progressive increases in pulmonary vascular resistance, right heart failure and, ultimately, premature death. Besides other cell types that are known to be involved in PAH pathogenesis (*e.g.* smooth muscle cells, fibroblasts and leukocytes), recent studies have demonstrated that endothelial cells (ECs) have a crucial role in the initiation and progression of PAH. The EC-specific role in PAH is multi-faceted and affects numerous pathophysiological processes, including vasoconstriction, inflammation, coagulation, metabolism and oxidative/nitrative stress, as well as cell viability, growth and differentiation. In this review, we describe how EC dysfunction and cell signalling regulate the pathogenesis of PAH. We also highlight areas of research that warrant attention in future studies, and discuss potential molecular signalling pathways in ECs that could be targeted therapeutically in the prevention and treatment of PAH.

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