Novel advances in modifying BMPR2 signalling in PAH

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Pulmonary Arterial Hypertension (PAH) is a disease of the pulmonary arteries, that is characterized by progressive narrowing of the pulmonary arterial lumen and increased pulmonary vascular resistance, ultimately leading to right ventricular dysfunction, heart failure and premature death. Current treatments mainly target pulmonary vasodilation and leave the progressive vascular remodelling unchecked resulting in persistent high morbidity and mortality in PAH even with treatment. Therefore, novel therapeutic strategies are urgently needed. Loss of function mutations of the Bone Morphogenetic Protein Receptor 2 (BMPR2) are the most common genetic factor in hereditary forms of PAH, suggesting that the BMPR2 pathway is fundamentally important in the pathogenesis. Dysfunctional BMPR2 signalling recapitulates the cellular abnormalities in PAH as well as the pathobiology in experimental pulmonary hypertension (PH). Approaches to restore BMPR2 signalling by increasing the expression of BMPR2 or its downstream signalling targets are currently actively explored as novel ways to prevent and improve experimental PH as well as PAH in patients. Here, we summarize existing as well as novel potential treatment strategies for PAH that activate the BMPR2 receptor pharmaceutically or genetically, increase the receptor availability at the cell surface, or reconstitute downstream BMPR2 signalling.

Biography

Svenja Dannewitz Prosseda did her MSc Ph.D., at Uniklinikum Freiburg, Baden-Württemberg, Germany, Interested in pulmonary and cardiovascular remodeling in response to hypoxia and inflammation. Our research platform is the world through worldwide collaboration, we can begin to answer the question of a global disease. She is currently working in Cactus communication.

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