# DOWNLOAD PDF PULMONARY ARTERIAL HYPERTENSION, AN ISSUE OF CLINICS IN CHEST MEDICINE

## Chapter 1 : High Treprostinil Doses Delay PAH-Related and All-Cause Hospitalizations

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Genetics[ edit ] Mutations in several genes have been associated with this condition [21] [22] these include bone morphogenetic protein receptor type 2 BMPR2 and eukaryotic translation initiation factor 2 alpha kinase 4 gene EIF2AK4. Right ventricle on left side Micrograph showing arteries in pulmonary hypertensive with marked thickening of the walls. The pathogenesis of pulmonary arterial hypertension WHO Group I involves the narrowing of blood vessels connected to and within the lungs. This makes it harder for the heart to pump blood through the lungs , much as it is harder to make water flow through a narrow pipe as opposed to a wide one. Over time, the affected blood vessels become stiffer and thicker, in a process known as fibrosis. This further increases the blood pressure within the lungs and impairs their blood flow. In common with other types of pulmonary hypertension, these changes result in an increased workload for the right side of the heart. As such, the right ventricle cannot cope as well with higher pressures, and although right ventricular adaptations hypertrophy and increased contractility of the heart muscle initially help to preserve stroke volume , ultimately these compensatory mechanisms are insufficient; the right ventricular muscle cannot get enough oxygen to meet its needs and right heart failure follows. This blood may also carry less oxygen than normal. Therefore, it becomes harder and harder for the left side of the heart to pump to supply sufficient oxygen to the rest of the body, especially during physical activity. Instead, the left heart fails to pump blood efficiently, leading to pooling of blood in the lungs and back pressure within the pulmonary system. This causes pulmonary edema and pleural effusions. However, in some patients, the raised pressure in the pulmonary vessels triggers a superimposed component of vessel narrowing, which further increases the workload of the right side of the heart. This phenomenon is called hypoxic pulmonary vasoconstriction and it is initially a protective response designed to stop too much blood flowing to areas of the lung that are damaged and do not contain oxygen. When the alveolar hypoxia is widespread and prolonged, this hypoxia-mediated vasoconstriction occurs across a large portion of the pulmonary vascular bed and leads to an increase in pulmonary arterial pressure, with thickening of the pulmonary vessel walls contributing to the development of sustained pulmonary hypertension. This combination of vessel occlusion and vascular remodeling once again increases the resistance to blood flow and so the pressure within the system rises. These results in a severe vasoconstriction and vascular smooth muscle and adventitial hypertrophy characteristic of patients with PAH. Activated PKG promotes vasorelaxation via a reduction of intracellular calcium levels , alters the expression of genes involved in smooth muscle cell contraction, migration and differentiation , and inhibits platelet activation. It acts on the endothelin receptors ETA and ETB in various cell types including vascular smooth muscle cells and fibroblasts, leading to vasoconstriction, hypertrophy, proliferation, inflammation, and fibrosis. It also acts on ETB receptors in endothelial cells; this leads to the release of both vasoconstrictors and vasodilators from those cells, and clears endothelin-1 from the system. In vascular smooth muscle cells, prostacyclin binds mainly to the prostaglandin I receptor. This sends a signal to increase adenylate cyclase activity, which leads to increased synthesis of cyclic adenosine monophosphate cAMP. This in turn leads to increased cAMP-dependent protein kinase or PKA protein kinase A activity, ultimately promoting vasodilation and inhibiting cell proliferation. Prostacyclin signaling also leads to anti-thrombotic, anti-fibrotic, and anti-inflammatory effects. Levels of cAMP which mediates most of the biological effects of prostacyclin are reduced by phosphodiesterases 3 and 4. In PAH, the balance is shifted away from synthesis of prostacyclin towards synthesis of thromboxane. However, several other pathways have been identified that are also altered in PAH and are being investigated as potential targets for future therapies. For example, the mitochondrial enzyme pyruvate dehydrogenase kinase PDK is pathologically activated in PAH, causing a metabolic shift from oxidative phosphorylation to glycolysis and leading to increased cell proliferation and impaired

apoptosis. Focusing only on the pulmonary vasculature provides an incomplete picture of PAH; the ability of the right ventricle to adapt to the increased workload varies between patients and is an important determinant of survival. The molecular pathology of PAH in the right ventricle is therefore also being investigated, and recent research has shifted to consider the cardiopulmonary unit as a single system rather than two separate systems. Importantly, right ventricular remodeling is associated with increased apoptosis; this is in contrast to pulmonary vascular remodeling which involves inhibition of apoptosis. The jugular venous pulse tracing demonstrates a prominent a wave without a c or v wave being observed. The phonocardiograms fourth left interspace and cardiac apex show a murmur of tricuspid insufficiency and ventricular and atrial gallops. Pulmonary artery catheter Severe tricuspid regurgitation In terms of the diagnosis of pulmonary hypertension, it has five major types, and a series of tests must be performed to distinguish pulmonary arterial hypertension from venous, hypoxic, thromboembolic, or unclear multifactorial varieties. PAH is diagnosed after exclusion of other possible causes of pulmonary hypertension.

## Chapter 2 : Pulmonary hypertension - Wikipedia

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