

# Mast Cells as a Therapeutic Target in Pulmonary Arterial Hypertension (PAH)

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#### Technology description

The inventor has discovered that targeting mast cells, part of the inflammatory response in pulmonary hypertension, can significantly attenuate disease development.

#### **Technology Overview**

Pulmonary hypertension is a progressive disease of various origins associated with poor prognosis and results in right heart dysfunction. In all its variant presentations it is estimated to affect up to 100 million people worldwide. Pulmonary arterial hypertension (PAH) is a life-threatening disease that is characterized by excessive pulmonary vasoconstriction and abnormal vascular remodeling where inflammation plays a major role. Increased pulmonary resistance owing to progressing vascular remodeling, can ultimately lead to right heart failure and death. There is growing evidence that release of local mediators in the lung are responsible for the inflammation and vascular remodeling that characterize PAH. The right ventricular response to increased pressure is recognized as critical to survival in patients with PAH.

The inventor has identified that by targeting mast cells, a hitherto unknown contributor to PAH, and therapeutically stabilizing, blocking the effect of their mediators and/or products, or diminishing their number, then the excessive pulmonary vasoconstriction, abnormal vascular remodeling, and right ventricular hypertrophy that occur in PAH can be attenuated and/or prevented. This can be achieved using a HIF stabilizer.

#### **Disease Prevalence**

The global prevalence of PAH is projected to be in the region of 100,000 – 200,000, that is about 15 - 50 patients per million of the population. Despite the current treatments available mortality remains high and there is a considerable unmet medical need in the management of this disorder. The increasing occurrence of cardiac diseases will drive the growth of the overall pulmonary arterial hypertension (PAH) market, which was valued at US\$5 billion in 2015. Owing to the sedentary lifestyles, growing consumption of fast food, and lack of exercise, the global numbers for the patients with cardiac diseases is expected to rise substantially thereby, increasing the susceptibility of pulmonary arterial hypertension among the global population.

Institution

**Cornell University** 

Inventors

Randi Silver

## 联系我们



### 叶先生

电话: 021-65679356 手机: 13414935137

邮箱: yeyingsheng@zf-ym.com