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Study suggests way to prevent rare pulmonary disease

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Research by Vanderbilt scientists suggests that it may be possible to prevent or even reverse pulmonary arterial hypertension (PAH), a rare, progressive disease characterized by narrowing of and high blood pressure in the small arteries of the lungs.

A key player in PAH is the proangiogenic cell (PAC), a cell produced by the bone marrow that normally promotes the growth of new blood vessels.

Last month in the journal *Circulation Research* the researchers reported finding PACs in the stiffened small blood vessels in the lungs of patients with PAH. In cell culture studies, they showed that PACs aggravate damage to the blood vessel lining, leading to scarring and narrowing.

They showed they could both prevent and reverse experimentally induced pulmonary hypertension in a mouse model by killing off the PACs, and also by giving a drug that blocked signaling through a serotonin receptor called 5-HT2B.

"We were quite surprised to find out that the driver of pulmonary hypertension appears to be a very small population of cells from the bone marrow," said the paper's corresponding author, W. David Merryman, Ph.D., professor of Biomedical Engineering, Pharmacology, Medicine and Pediatrics. "It definitely changes the way we think about possible therapies for the disease."

Approximately 500 to 1,000 cases of PAH are diagnosed each year in the United States. Untreated, the disease will eventually damage the heart. Various medications can reduce blood pressure in the lungs and slow the course of the disease but doctors are not yet able to stop or reverse it.

The findings by the Vanderbilt researchers suggest that serotonin signaling triggers the proliferation and homing of PACs. Thus targeting 5-HT2B could potentially lead to the first treatment capable of preventing and reversing PAH.

More information: Bone Marrow-Derived Proangiogenic Cells Mediate Pulmonary Arteriole Stiffening via Serotonin 2B Receptor Dependent Mechanism. *Circulation Research*. DOI: 10.1161/CIRCRESAHA.118.313397

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