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sclerodermanews.com: Borderline Pulmonary Pressure in Scleroderma Patients Linked to a Pre-Pulmonary Arterial Hypertension Condition

A review study recently published in the journal Arthritis Research & Therapy suggests that borderline pulmonary pressure in patients with scleroderma can represent a condition of pre-pulmonary arterial hypertension (PAH). The study was conducted by researchers at the Medical University of Graz and the Ludwig Boltzmann Institute for Lung Vascular Research in Austria and is entitled " Borderline pulmonary pressures in scleroderma — a 'pre-pulmonary arterial hypertension' condition? "

Scleroderma is a rare, chronic autoimmune disease in which the body's own immune system attacks healthy tissues resulting in a hardening and tightening of the skin and connective tissues. The disease usually affects the skin, but it can also affect internal organs such as the lungs, blood vessels and the digestive tract.

Patients with scleroderma may develop borderline pulmonary arterial pressure, a condition that has been suggested to represent a transitory state between normal pulmonary arterial pressure (equal or less than 20 mmHg) and pulmonary arterial hypertension (PAH; mean value equal or higher than 25 mmHg). PAH is a life-threatening condition characterized by the increase of blood pressure in the pulmonary arteries that supply blood to the lungs, and it can lead to difficulties in breathing, right-sided heart failure and eventually death. There is an urgent need for an early diagnosis and treatment of PAH.

The DETECT study was a large, international study with more than 400 scleroderma patients across 18 different countries, with a goal of developing a screening algorithm for PAH in scleroderma patients. Among the DETECT patient cohort, 15% were found to have borderline pulmonary pressure hemodynamics, representing a substantial subgroup of this population.

In the review, the team calls the attention to other hemodynamic condition in scleroderma patients, which is an increase in exercise-induced pulmonary arterial pressure. Previous studies have suggested that this might be a frequent condition among scleroderma patients, and that clinical deterioration and PAH development are frequent in this population. In addition, a strong correlation was shown between the two hemodynamic conditions, where patients with resting borderline pulmonary pressure and patients who exhibit a significant pulmonary arterial pressure increase during exercise, strongly overlap.

The authors concluded that borderline pulmonary arterial pressure increase has a relevant clinical significance in scleroderma patients, and that it can be considered a pre-PAH condition and therefore a potential prognostic marker of the disease, allowing an early diagnosis.

According to the authors, one question remains: whether PAH therapy should be given to scleroderma patients who present borderline pulmonary pressure or an exercise-induced increase in pulmonary arterial pressure. Clinical trials should be conducted to address this relevant question.

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