

697 - Elevated Pulmonary Vascular Resistance At Rest Is Associated with Exercise Pulmonary Arterial Hypertension in Systemic Sclerosis Patients without Pulmonary Hypertension Hemodynamics At Rest

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Hall F2 - Poster Hall (McCormick Place West)

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Background/Purpose:

Pulmonary arterial hypertension occurs in 1 of 7 patients with systemic sclerosis (SSc), but is the leading cause of death. Patients who do not have pulmonary hypertension (PH) at rest have been shown to have elevated mean pulmonary arterial pressure (mPAP) with exercise. This population represents a group that may be at increased risk for PH, which could have implications on prognosis and treatment. This study analyzes the resting and exercise right heart catheterization (RHC) data in the PHAROS (Pulmonary Hypertension Assessment and Recognition of Scleroderma) registry.

Method:

PHAROS is a multicenter cohort of SSc patients designed to determine the outcomes of high risk SSc patients for the development of PH. Of 390 patients within the registry, 87 patients had RHC at rest and with exercise following local protocols. Analysis of the data was performed with focus on the parameters of mPAP, pulmonary capillary wedge pressure (PCWP), and pulmonary vascular resistance (PVR). The following definitions of pulmonary hypertension were used: pulmonary arterial hypertension (PAH) = mPAP \geq 25mmHg, PCWP \leq 15mmHg; pulmonary venous hypertension (PVH) = mPAP \geq 25mmHg, PCWP > 15mmHg; exercise pulmonary arterial hypertension (ePAH) = mPAP with exercise > 30mmHg, PCWP \leq 18mmHg; exercise pulmonary venous hypertension (ePVH) = mPAP with exercise > 30mmHg, PCWP > 18mmHg.

Result:

There were 87 patients within the PHAROS registry that had an exercise RHC. Of these, 52 patients (60%) had no change between resting and exercise RHC, including patients that remained normal with exercise or had pulmonary hypertension with rest and exercise. Thirty-five patients (40%) had discrepant hemodynamic changes with exercise. Within this group of 35, 21 patients (60%) had normal resting RHC with ePAH, 8 patients (23%) had normal resting RHC with ePVH, and 6 patients (17%) had PAH with ePVH. In total, PAH was diagnosed in 22 patients (25%) and ePAH in an additional 21 (24%). Among patients with a normal resting RHC, a PVR >140 dyn·s/cm⁵ was associated with ePAH, (positive predictive value 76%, negative predictive value 62%). A PCWP >10mmHg was associated with ePVH, (positive predictive value 62%, negative predictive value 100%).

Conclusion:

In patients with SSc, an exercise RHC can reveal underlying pulmonary hypertension despite normal resting hemodynamics. Patients with normal mPAP at rest, but elevated PVR >140 dyn·s/cm⁵ or PCWP >10 mmHg may be at increased risk of ePAH or ePVH respectively. In these patients, pursuing an exercise RHC may lead to earlier diagnosis and treatment.

Keywords: pulmonary complications, scleroderma and systemic sclerosis

Disclosure: **J. Kushi**, None; **M. E. Csuka**, Actelion Pharmaceuticals US, 2, United Therapeutics, 2 ; **L. K. Hummers**, None; **F. M. Wigley**, None; **V. M. Hsu**, Actelion Pharmaceuticals US, United Therapeutics ; **D. Khanna**, Actelion Pharmaceuticals US, 5, United Therapeutics, 5, Pfizer Inc, 5, Novartis Pharmaceutical Corporation, 5, Actelion Pharmaceuticals US, 8, United Therapeutics, 8, Gilead, 8 ; **J. A. Molitor**, Actelion Pharmaceuticals US, 5, United Therapeutics ; **V. D. Steen**, Gilead, 2, United Therapeutics, 2, Pfizer Inc, 2, Actelion Pharmaceuticals US, 2, Genentech and Biogen IDEC Inc., 5, Gilead, 5, Novartis Pharmaceutical Corporation, 5, Pfizer Inc, 5, Gilead, 8, Actelion Pharmaceuticals US, 8 .