

## High Prevalence of Occult Left Heart Disease in Scleroderma-Pulmonary Hypertension

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### **Introduction:**

The association between scleroderma (SCL) and Pulmonary Hypertension (PH) is well established. Typically, differentiation between pulmonary arterial hypertension (PAH) and pulmonary venous hypertension (PVH) is based on the pulmonary arterial wedge pressure (PAWP)  $\leq 15$ mmHg being the current criterion for diagnosing PAH. The 'gold-standard' measure of increased left heart pressure is the Left Ventricular End-Diastolic Pressure (LVEDP). It has been reported that in patients with PH, the PAWP and LVEDP are frequently discordant. Some patients with LV diastolic dysfunction may have normal resting LVEDP, but will show an abnormal increase in LVEDP in response to intravenous fluid loading. The proportion of scleroderma patients suspected of having PAH but in reality having occult PVH is unknown. We investigated the prevalence of discordance between PAWP and LVEDP in the SCL population, studied the frequency of occult PVH as assessed by the saline challenge, and examined the epidemiological factors and echocardiographic correlates of this phenomenon.

### **Methods:**

Retrospective chart review of 107 SCL: patients. All patients with suspected PH had routine right/left heart catheterization with LVEDP measurement pre/post fluid challenge. We extracted demographic, hemodynamic and echocardiographic data. Patients were classified into one of four groups – hemodynamically normal (mPAP $<25$ mmHg), PVH (mPAP $\geq 25$ mmHg, PAWP $>15$ mmHg), occult PVH (mPAP $\geq 25$ mmHg, PAWP $\leq 15$ mmHg, LVEDP $>15$ mmHg before/after fluid challenge) and PAH (mPAP $\geq 25$ mmHg, PAWP $\leq 15$ mmHg and LVEDP $\leq 15$ mmHg before/after fluid challenge).

### **Results:**

Fifty-three of 107 patients had PH. Based on the PAWP-based definition 29/53 had PAH and 24/53 had PVH. After considering the resting and post-fluid-challenge LVEDP, 11 PAH patients were reclassified as occult PVH. The occult PVH group was hemodynamically, echocardiographically and demographically closer to the PVH group than the PAH group.

### **Conclusions:**

PVH had high prevalence in our SCL-PH population. Distinguishing PAH from PVH with only PAWP results in some PVH patients being misclassified as having PAH