

European Respiratory Society Annual Congress 2012

Abstract Number: 3390

Publication Number: P917

Abstract Group: 4.3. Pulmonary Circulation and Pulmonary Vascular Disease

Keyword 1: Pulmonary hypertension **Keyword 2:** Imaging **Keyword 3:** No keyword

Title: Evaluation of acute right ventricular failure in scleroderma and idiopathic pulmonary arterial hypertension

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Body: Background: The ability to maintain adequate right ventricle (RV) function is paramount to survival in pulmonary arterial hypertension (PAH). However, little is known about RV function in patients with established PAH who are hospitalized for acute RV failure. Since RV adaptation is often worse in scleroderma-associated PAH (SSc-PAH) than in patients with idiopathic PAH (IPAH) despite similar afterload, we sought to compare RV function in hospitalized PAH patients using tricuspid annular plane systolic excursion (TAPSE), a non-invasive measurement of RV function that strongly associates with invasive hemodynamics and survival in both SSc-PAH and IPAH. Methods: 47 patients with SSc-PAH or IPAH were evaluated during their first admission to a hospital for RV failure. All patients received transthoracic echocardiography. Vital signs and survival from time of admission were recorded. Results: This cohort was composed of 91% females, with mean age 56 (range 23-81). There were 68% SSc-PAH and 32% IPAH patients. Mean TAPSE was 0.3 cm larger in IPAH compared to SSc-PAH ($1.4\text{cm} \pm 0.424$, vs. $1.1\text{cm} \pm 0.33$, $p=0.02$). This relationship persisted despite controlling for age, admission vitals, and time since diagnosis. 1-year mortality was 47%. Relative risk of mortality was 53% less in the IPAH group than in those with SSc ($10\% \pm 0.32$ vs. $63\% \pm 0.49$, $p=0.004$). Conclusion: SSc-PAH patients admitted with acute RV failure had both lower TAPSE and higher 1 year mortality than those with IPAH. This association suggests that TAPSE may offer prognostic information in patients with acute RV failure. Larger studies are needed to confirm the significance of these findings.