

European Respiratory Society Annual Congress 2013

Abstract Number: 3080

Publication Number: P3404

Abstract Group: 4.3. Pulmonary Circulation and Pulmonary Vascular Disease

Keyword 1: Extrapulmonary impact **Keyword 2:** Physiology **Keyword 3:** Pulmonary hypertension

Title: Load-independent right ventricular function and idiopathic pulmonary arterial hypertension severity

Pia 16832 Trip p.trip@vumc.nl MD ¹, Silvia 16833 Rain s.rain@vumc.nl MD ^{1,2}, Harm-Jan 16834 Bogaard hj.bogaard@vumc.nl MD ¹, Nico 16835 Westerhof n.westerhof@vumc.nl ¹, Jolanda 16836 van der Velden j.vandervelden@vumc.nl ², Frances S. 16838 de Man fs.deman@vumc.nl ¹ and Anton 16849 Vonk-Noordegraaf a.vonk@vumc.nl MD ¹. ¹ Pulmonology, VU University Medical Center, Amsterdam, Netherlands and ² Physiology, VU University Medical Center, Amsterdam, Netherlands .

Body: Rationale Right ventricular (RV) failure is the main mortality determinant in idiopathic pulmonary arterial hypertension (iPAH). However, little is known about adaptive and maladaptive changes in RV performance during disease progression. This study aims to characterize global RV function in mild, intermediate and severe iPAH based on patient survival duration from iPAH diagnosis. Methods Right heart catheterization and MRI were performed in 67 individuals (Con=10, Mild=28, Intermediate=11, Severe=18). iPAH severity was defined as: less than 6 months survival (severe), between 1-3 years survival (intermediate) and more than 5 years survival (mild). RV systolic elastance (Ees), arterial elastance (Ea), RV coupling (Ees/Ea) and diastolic elastance (Ed) were quantified by pressure-volume analysis. Results RV systolic and diastolic elastance were significantly increased in iPAH compared to controls. RV coupling was maintained in mild and intermediate iPAH, however significantly decreased in severe iPAH. RV diastolic elastance was preserved during mild iPAH, but significantly increased in intermediate and severe disease. Conclusions Systolic RV adaptation to the increased afterload is only compromised in severe iPAH. However, RV diastolic impairment occurs already from intermediate iPAH.