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Title: Characteristics of a stable and satisfactory condition in long term survivors of pulmonary arterial hypertension

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Body: Introduction The aim of the study is to perform a longitudinal data comparison between long term stable and unstable patients with pulmonary arterial hypertension (PAH). Methods Included patients were 9 long term stable (stable survivors during 10 year follow-up), 12 long term unstable (died after 8 years) and 9 short term non-survivors (died ≤ 1.5 years) with idiopathic PAH diagnosed between 1999-2003 and followed to 2013. All patients underwent baseline right heart catheterization, cardiac magnetic resonance and 6 minute walk testing (6MWT). Unstable patients had additional measurements after 1, 3 and 7 years and stable patients underwent further evaluation at 10 years. Results Although baseline parameters identified short term non-survivors, these characteristics did not discriminate between long term stable and unstable patients (Fig 1). During follow-up, hemodynamics and 6MWT were similar in both long term follow-up groups (Fig 1A-C). Despite similar baseline values, long term stable patients had unchanged right ventricular (RV) volumes over time whereas unstable patients showed progressive dilatation (p int: 0.03; Fig 1D). Stable patients showed a higher RV ejection fraction (RVEF) than non-stable patients at baseline and over time (p<0.05; Fig 1E).

Conclusions The absence of progressive RV dilatation and preserved RVEF predict a long term stable, satisfactory condition in PAH patients.