Abstract: **P956**

**Prognostic role of right ventricular hypertrophy in anderson fabry disease**

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Background: Right ventricular hypertrophy (RVH) is a common finding in Anderson Fabry disease (AFD). In other infiltrative and storage cardiomyopathies right ventricular (RV) involvement may influence prognostic stratification. Nevertheless, the prognostic implications of right ventricle involvement in AFD have never been assessed.

Purpose: Evaluation of the prognostic significance of RVH and RV systolic function in AFD cardiomyopathy.

Methods: Forty-five AFD patients (56% male) with extensive baseline evaluation, including assessment of RVH and RV systolic function, were followed-up for an average of 44.9±8.5 months. RV systolic function was assessed by standard and tissue Doppler echocardiography and quantified using RV fractional area change (RVFAC), tricuspid annular plane systolic excursion (TAPSE) and Systolic tissue Doppler velocity of the tricuspid annulus (RV Sa). Cardiovascular events were defined as new-onset atrial fibrillation, heart failure or pacemaker implantation; renal events were defined as progression to dialysis and/or renal transplantation or significant worsening of glomerular filtration rate; cerebrovascular events were defined as stroke or transient ischemic attack. The outcome was defined as the time to death or the first event in any of the above predefined categories.

Results: Fourteen patients (31.1%) presented RVH, while RV systolic function was normal in all cases. During the follow-up period, 13 patients (28.8%, 11 male) experienced major events including two deaths. Pacemaker implantation (6 cases) was the most common type of event. At univariate analysis several variables were associated with the occurrence of events, including RVH and indexes of RV systolic function. However, at multivariate analysis only proteinuria (HR:8.3, 95% CI: 2.88 to 23.87, p ?0.001) and LV mass indexed (HR: 1.01, 95% CI: 1.00 to 1.03, p=0.03) were independent predictors of outcome.

Conclusions: The presence and extension of RVH is not associated with outcome in AFD. Our study confirms that at variance with other infiltrative or storage cardiomyopathies, RV involvement in AFD is an innocent bystander and does not influence prognosis.