

## MP2-5

### **Clinical significance of left and right ventricle echocardiographic parameters in children with idiopathic pulmonary arterial hypertension.**

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Background: Echocardiography is commonly used for assessment and serial follow-up of right (RV) and left ventricle (LV) function in children with idiopathic pulmonary arterial hypertension (iPAH).

Clinical significance and predictive value of echo parameters have been scarcely characterized.

Objectives: to characterize RV function in children with iPAH in stable and worsening clinical status, and to assess value of echocardiographic indices to predict clinical worsening.

Methods and results: Clinical, biological and echocardiographic variables were prospectively collected in 38 children with iPAH. Patient's median age at inclusion was 6.3 years old, 95%IC [3.2-11.1].

Median follow-up was 15.4 months. Forty seven echo scans were performed in children at time of clinical worsening (TCW) defined as NYHA  $\geq$ III and/ or recent syncope and/or overt RV failure, and 222 echo scans were performed in children in stable clinical status (SC) defined by NYHA  $\leq$ II, without syncope and without RV failure. Median delay between each visit was 54 days, 95%CI [45-71].

Patients at TCW were significantly younger (median age 5.5 years old 95% CI [2.8-8.6],  $p=0.001$ ), and had lower body mass index ( $p=0.002$ ).

TAPSE, RV peak systolic myocardial velocity, pulmonary acceleration time, aortic and pulmonary velocity-time integral (VTI) were significantly reduced at TCW ( $p \leq 0.001$ ). LV filling was also impaired at TCW: mitral early (E)/late(A) ratio and LV myocardial early diastolic velocities (mitral E') were significantly reduced (respectively  $p=0.03$  and  $p=0.01$ ).

In univariate analysis, RV fraction area change and mitral E' were predictive of SC. In multivariate analysis, aortic VTI was predictive of TCW even after adding age and B-type natriuretic peptide into the model (HR=0,7349, 95%CI [0,5616-0,9617],  $p=0,02$ ). ROC curve analysis confirmed the relationship between aortic VTI and TCW (area under curve 0,810, 95%CI [0,722-0,880]). An aortic VTI  $\leq$ 16cm (sensitivity 71%, specificity 83%) predicted TCW.

#### Conclusion:

Echocardiographic parameters of RV function and LV filling were impaired in children with iPAH at TCW. Aortic VTI predicted TCW. Inter-ventricular interactions in this setting might provide further insights into the mechanisms leading to clinical worsening.