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%CI [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 ≥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≤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≤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≤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.