

Right Ventricular End-Systolic Remodeling Index in the Assessment of Pediatric Pulmonary Arterial Hypertension - Data from the European Pediatric Pulmonary Vascular Disease Network

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Objectives: In adults with pulmonary hypertension (PH) determination of the RV end-systolic remodeling index (RVES RI) with echocardiography was found to be of clinical value for the assessment of disease severity. We aimed to determine if increased RVES RI values will also occur in pediatric PH. We further aimed to correlate these RVES RI values with conventionally used echocardiographic variables such as the tricuspid annular plane systolic excursion (TAPSE), the (S'), the left ventricular eccentricity index (LVEI), the pulmonary artery acceleration time (PAAT), and hemodynamic values such as the mean pulmonary artery pressure (mPAP) and the pulmonary vascular resistance index (PVRi).

Methods: We conducted an echocardiographic study in 49 children with PH, and in 123 healthy children.

Results: The RVES RI was increased in children with PH compared to 49 matched healthy children (1.45 ± 0.16 vs. 1.16 ± 0.06 ; $p < 0.01$). The RVES RI negatively correlated with RV systolic function parameters (TAPSE, $r = -0.487$, $p < 0.001$; S', $r = -0.471$, $p = 0.001$). RVES RI positively correlated with the magnitude of RV-LV interaction (LVEI, $r = 0.633$, $p < 0.001$) and the PAAT ($r = -0.343$, $p = 0.016$). The RVES RI was positively associated with the invasive hemodynamic variables mPAP ($r = 0.464$, $p = 0.004$) and PVRi ($r = 0.579$, $p < 0.001$), demonstrating the RVES RI to reflect disease severity in our pediatric PH patient cohort.

Conclusions: The RVES RI is increased in children with PH compared to age-matched healthy subjects and correlates with conventional echocardiographic and invasive hemodynamic variables of RV function, PAP and PVR. The RVES RI is a simple, reproducible, and clinically relevant echocardiographic measure for the assessment of PH in children.