

Pulmonary arterial stiffness indices assessed by intravascular ultrasound in children with early pulmonary vascular disease: prediction of disease progression and mortality during 20-year follow-up

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Introduction: Prognosis in children with pulmonary vascular disease (PVD) is closely linked to right ventricular (RV) failure due to increased RV afterload. Pulmonary arterial (PA) stiffening is known to occur early in the disease course of PVD and constitutes a main component of RV afterload. We aimed to evaluate the clinical value of indices of PA stiffness in children with early or advanced PVD, by determining the association of such indices with long-term disease progression and mortality.

Methods: Forty-one children with arterial PVD in early or more advanced stages, defined as mean PA pressure ≥ 20 mmHg and/or pulmonary to systemic flow ratio ≥ 1.2 , and mean pulmonary capillary wedge pressure < 15 mmHg, underwent cardiac catheterization with intravascular ultrasound (IVUS) imaging between 1994 and 1997 with follow-up until 2015. Indices of PA stiffness evaluated were compliance and distensibility. During long-term follow-up, transthoracic echocardiography and cardiac catheterization were performed to determine whether PVD had reversed or progressed.

Results: Following baseline cardiac catheterization, 27 (66%) patients underwent closure of a cardiac shunt defect. During a median (interquartile range) follow-up of 19 (18-20) years, 31 (76%) cases of PVD had reversed and 10 (34%) had progressed. Six patients died due to PVD. In addition to conventional hemodynamics, lower compliance and distensibility were significantly associated with PVD progression ($p=0.007$ and $p=0.011$) and mortality ($p=0.007$ and $p=0.009$), also after adjustment for age, sex and end-diastolic PA luminal area. Survival rates differed significantly between patients with high and low compliance ($p=0.011$) and distensibility ($p=0.009$, Figure 1). Also in a subgroup of patients with favorable hemodynamic profiles at baseline, lower compliance and distensibility were associated with progression of PVD during follow up ($p=0.002$ and $p=0.030$).

Conclusions: PA stiffness indices assessed by IVUS are associated with long-term disease progression and mortality in children with PVD and are a valuable complement to conventional hemodynamic evaluation, especially in the early stages of the disease.

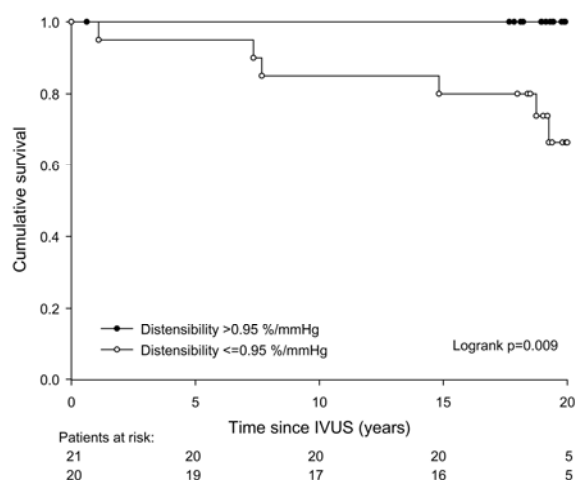


Figure 1. Comparison of survival of patients with high and low distensibility.