

海外招請講演

海外招請講演3 (IL-03)

AEPC-YIA 記念講演

座長:安河内 聡 (長野県立こども病院 循環器センター)

Wed. Jul 6, 2016 6:00 PM - 6:40 PM 第D会場 (オーロラ イースト)

I-IL-03

6:00 PM - 6:40 PM

[I-IL-03-2]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 occurs early in the course of PVD and constitutes a main component of RV afterload. This study evaluates the clinical value of PA stiffness in children with early or advanced PVD, by determining the association of PA-stiffness 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

Results: During follow-up, 31 (76%) cases of PVD had reversed and 10 (34%) had progressed. Six patients died due to PVD. Patients with progressed PVD showed significantly lower compliance ($p=0.001$) and distensibility ($p=0.002$). Also in a subgroup of patients with favorable hemodynamics at baseline, lower compliance and distensibility were associated with progression of PVD during follow-up ($p=0.002$ and $p=0.030$). Survival rates differed significantly between patients with high and low compliance ($p=0.011$) and distensibility ($p=0.009$).

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