

Invasive assessment of pulmonary endothelial function in Fontan patients

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Background: Pulmonary endothelial dysfunction due to a loss of pulsatile pulmonary blood flow is thought to be a major factor in the development of increased pulmonary vascular resistance (PVR) in Fontan patients causing significant morbidity and mortality. We therefore aimed to assess pulmonary endothelial function by vasodilator response to acetylcholine administered in segmental pulmonary arteries in children and adolescents with Fontan hemodynamics.

Methods: Twenty-four Fontan patients (mean age 12.0 ± 7.8 years, 10 females) underwent invasive cardiac catheterization including intra-arterial Doppler- and pressure measurements in a segmental pulmonary artery. Pulmonary flow reserve (PFR) was quantified as the change of average peak velocity (APV) in response to acetylcholine (Ach) infusion and local PVR was assessed by local pressure divided by velocity.

Results: APV significantly increased from 16.8 ± 7.5 to 20.3 ± 6.9 cm/s ($p=0.01$) resulting in a mean PFR of 1.35 ± 0.70 . Local PVR dropped from 0.72 ± 0.32 to 0.57 ± 0.20 mmHg/cm/s ($n=18$; $p=0.004$). A significant relationship was found between baseline local PVR and PFR ($r=0.73$, $p=0.0006$) as well as between local PVR and the percent decrease in local PVR ($r=0.52$, $p=0.02$). No relationship was found between PFR as well as local PVR with global hemodynamics such as cardiac output and global PVR.

Conclusions: This study is the first assessing local pulmonary endothelial function in Fontan patients. Given the difficulties in the assessment of global PVR in these patients, measurement of local pulmonary blood flow velocity and PVR may be helpful in the evaluation of adverse pulmonary vascular remodelling. Future research is required to study whether impaired local pulmonary endothelial function may be predictive for Fontan failure.