

O10-1

Ventricular dysfunction rather than increased pulmonary vascular resistance is the predominant cause of Fontan failure - Haemodynamic and echocardiographic findings in adults with failing Fontan

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Objectives: Despite its short- and long-term success, the Fontan circulation is palliative and an increasing number of patients will eventually experience Fontan failure during long-term follow-up. Pathophysiology of failing Fontan is complex and conventional treatment options are usually of limited success. Increased pulmonary vascular resistance (PVR) is a frequently incriminated cause of Fontan failure but also ventricular dysfunction is not uncommon. We sought to characterize longitudinal haemodynamic and echocardiographic findings in adult failing Fontan patients to determine the most prevalent causes of haemodynamic failure.

Methods: From the entire cohort of Fontan patients treated in our institution (N=443), adults >18 years of age at last follow-up (n=164) were retrospectively screened for failure (characterized by NYHA III-IV without improvement and/or >2 unscheduled hospitalizations within 12 months and/or active protein-losing enteropathy). Haemodynamic and echocardiographic findings were collected and analysed.

Results: We identified 40 patients (median age 28.5 years [IQR 23.9-36.3], median follow-up after Fontan 21.4 years [IQR 14.8-24.0]). Of these, 29(72.5%) had moderate to severe systolic ventricular dysfunction (ejection fraction $\leq 45\%$, n=15) and/or evidence of diastolic dysfunction (end-diastolic ventricular pressure, EDP ≥ 12 mmHg, n=18). Borderline / elevated PVR (calculated index $2-2.5/\geq 2.5$ WU*m²) was only seen in 5(12.5%) / 7(17.5%) patients, respectively. Overall, ejection fraction declined significantly from $63\pm 6\%$ at early follow-up (2.1 years [IQR 0.6-10.8]) to $48\pm 14\%$ at last follow-up (p<0.001). Pulmonary artery pressure increased from 11 ± 3 to 15 ± 5 and EDP from 6 ± 3 to 12 ± 5 mmHg (both p<0.001), while transpulmonary gradient (5 ± 2 vs. 4 ± 2 mmHg) and PVR index (2.0 ± 1.1 vs. 1.7 ± 0.8 WU*m²) did not change significantly (p>0.05). Mortality in adult failing Fontan patients was substantial, 20(50%) died during follow-up.

Conclusions: Therapeutic options in a truly failed Fontan circulation are limited and sustained recompensation by medical, interventional or surgical means is rare. Therapies targeting reduction of PVR may be of limited success since systolic and, rather underappreciated, diastolic ventricular dysfunction seem to be the predominant causes of Fontan failure. Heart transplantation as ultima ratio should be considered timely, before progressive multi-organ dysfunction impedes successful transplantation, especially in the light of donor organ shortage and extended waiting time, since mortality in failing Fontan patients is high.