

**Long-term outcome and late morbidity of adolescent and adult patients with Fontan circulation**

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**Objectives:** Outcomes of the Fontan palliation for univentricular heart defects have substantially improved and most patients will now reach adulthood. However, given the significant intrinsic limitations of the Fontan circulation, we face a continuously growing population of adolescent and adult Fontan patients at risk of experiencing severe complications. We assessed long-term outcome of these patients in our institutional follow-up program aiming at early detection of complications and prompt interventions to optimize Fontan hemodynamics.

**Methods:** From 1990-2013, 205 patients underwent a modified Fontan operation at our institution; patients having reached adolescence or adulthood (n=78) were included. Follow-up, including annual clinical exams, echocardiography and 24h-ECG-monitoring continued for a median of 14.6 years. Moreover follow-up was complemented by exercise capacity testing and patients were encouraged to complete standardized questionnaires exploring their quality of life (SF-36). MRI and cardiac catheterizations were performed according to clinical findings.

**Results:** There were 5 late deaths, one patient received heart transplantation due to ventricular failure. 15-year survival estimate was 94%. Severe complications included late Fontan failure (n=8), dysrhythmias (n=29) and thromboembolic events (n=8). 34 patients (47.2%) received catheter interventions and 15 (20.8%) re-operations including 5 Fontan conversions. Overall late hemodynamic results were good (median pulmonary artery pressure 11mmHg and transpulmonary gradient 6mmHg). Median maximal oxygen consumption was 21ml/min/kg (51% of reference). Severely decreased oxygen consumption (<40% of reference) and chronotropic competence (peak heart rate <120/min) were present in 19.6% and 30.4%, respectively. Ventricular function and Fontan flow examined by MRI were satisfactory (median ejection fraction 53%, cardiac index 2,5l/min/m<sup>2</sup>). Overall quality of life assessed by the SF-36 was good.

**Conclusions:** Stable long-term results demonstrate adequate patient selection and follow-up monitoring correlating with a good quality of life. Nevertheless, long-term morbidity remains a significant issue in adolescent and adult Fontan patients. A considerable fraction of patients displays severely decreased oxygen consumption and chronotropic competence which have recently been shown to be significant risk factors for late mortality. While inadequate Fontan hemodynamics due to increased pulmonary vascular resistance may be improved by medical, surgical or catheter intervention, in ventricular failure therapeutic options are limited and transplantation may be ultima ratio.