

## PW3-8

### **Long term follow up of adolescent and adult grown up congenital heart disease (GUCH) patients after modified Fontan operation**

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**Introduction:** Since the introduction of single ventricle palliation more patients reach adulthood and need specialized supervision and therapy. We analyzed hemodynamics, physical performance and morbidity in GUCH patients after modified Fontan operation.

**Methods:** Seventy patients, who underwent Fontan operation between 1991 and 2010, reached adolescent (15-18 years) or adult age (range: 15-50, median 27 years) during the median follow up of 10.4 (range: 0.6–19) years; 20 of them were older than 30 years. Seventeen patients were operated on as adults (16-37 years). The intraatrial modification was performed in 38 patients and extracardiac Fontan operation (ECFO) in 32. The hemodynamics was analyzed by heart catheterization and MRI. The cardiopulmonary capacity was tested by spiroergometry with monitoring of the oxygen consumption capacity (VO<sub>2</sub>max). Necessity for cardiac medication and the incidence of arrhythmias were checked.

**Results:** There were three late deaths (mortality 4.3%). Patients who underwent heart catheterization (n = 43) showed low pulmonary artery pressure (median 11 mmHg) and low transpulmonary gradient (median 6 mmHg). The cardiac index, measured by MRI (n=29) was in median in adolescents better than in adults (3.1 vs. 2.4 l/min/m<sup>2</sup>, p=0.027). VO<sub>2</sub>max decreased significantly during the follow up (27.1±7.7 ml/min/kg (59.9±17.0%) early vs. 20.2±7.8 ml/kg/min (54.7±17.9%) late, p=0.005). Thirteen patients (19%) developed tachyarrhythmias; 11 patients, all with intraatrial Fontan operation, required a permanent pacemaker due to bradyarrhythmias. Medical treatment of heart failure with more than two drugs was necessary in 41 patients (59%). All patients were on an anticoagulation regime. No clinically relevant thromboses were noted but one thromboembolic event occurred in one patient after intracardiac Fontan. No transplantations were necessary in adult patients after Fontan operation during current follow up.

**Conclusions:** Grown up patients after lateral tunnel or extracardiac Fontan operation reach adulthood with stable hemodynamics and low morbidity. The incidence of arrhythmias after ECFO is lower. Regular checks of the patients' physical exercise capacity and hemodynamics are necessary to optimize the cardiac medication for progressive heart failure and to identify candidates for later heart transplantation.