

**The role of regular physical activity in Fontan Circulation**

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Introduction: Functional capacity in Fontan circulation is commonly impaired. Cardiopulmonary exercise test (CPET) variables are prognostic in these patients. We sought to undertake a regular CPET evaluation of Fontan patients followed in outpatient clinic at a tertiary hospital and find predictors of functional capacity.

Methods: Forty-four Fontan patients followed in outpatient clinic were prospectively evaluated with CPET and transthoracic echocardiogram (TTE). Regular physical activity was defined as extracurricular organized sports participation or at least two 45-minute periods of physical activity each week. Regarding atrioventricular (AV) dominant valve regurgitation on TTE, patients were classified by two experienced operators in 2 groups: those without significant regurgitation (none or mild) and those with significant regurgitation (moderate or severe). Statistical inference was performed using R CRAN version 3.5.0. Linear regression and ANOVA were used for continuous variable correlation, Chi-squared test and logistic regression were used for binomial variable correlation.

Results: Median age at Fontan completion was 6 years (SD 3) and median age at current evaluation was 19 years (SD 7). Sixteen patients (36%) had regular physical activity. Mean peak oxygen uptake (peak VO<sub>2</sub>) was 27.6 ml/kg/min (SD 6), peak VO<sub>2</sub> as percentage of predicted value was 67% (SD 15), VO<sub>2</sub> at ventilatory threshold was 16 ml/kg/min (SD 3), VE/VCO<sub>2</sub> slope was 36.7 (SD 7), respiratory exchange ratio (RER) was 1.06 (SD 0.08), maximal heart rate (HR<sub>max</sub>) was 164 bpm (SD 26) and peak oxygen saturation was 87% (SD 8). Peak VO<sub>2</sub> was inversely correlated with age (p=0.006) and significant AV regurgitation (p=0.003) and had a strong positive relation with the practice of regular physical activity (p=0.001). Multivariate analysis showed that higher peak VO<sub>2</sub> was independently associated with regular physical activity and absence of significant AV regurgitation (R<sup>2</sup> = 0.327, F test with p = 0.001).

Conclusion: Peak VO<sub>2</sub> is a marker of functional capacity and prognosis in Fontan patients, usually impaired. Lower peak VO<sub>2</sub> was related with significant AV regurgitation. We identified a strong positive relation between peak VO<sub>2</sub> and regular physical activity, which is relevant for patient guidance and recommendations.