

Prescription of exercise to children with univentricular heart and Fontan circulation – a way to retard decline?

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Introduction, after Fontan completion of the univentricular heart cardiopulmonary fitness is reduced and the maximal aerobic capacity typically remains at 43-63% of normal. Often the measurements of lung function are abnormal. The chronically elevated CVP in combination with single pumping systemic ventricle gradually contributes to gradual hemodynamic failure with troublesome life-threatening chronic complications. It is not known how to prevent or postpone deterioration. Habitual exercise of young Fontan patients may have positive effects on peripheral muscle mass and lung function which may enhance Fontan hemodynamics, patient contention and well-being.

Methods, 18 patients with Fontan circulation at 14.6±2.4 years of age (12 boys), 19±2.7 kg/m² of BMI, and with FM of 18.4±9.5% were recruited. Twelve of the patients had RV- (HLHS) and the rest had LV-systemic ventricle. None of the patients had residual flow through cavoatrial fenestration. The measurements included daily habitual questionnaire, body composition, lung volumetry at rest for forced vital capacity (FVC), forced expiratory volume/sec (FEV₁), FEV₁/FVC, EUROFIT-testing for comprehensive muscle fitness, and spiroergometry using bicycle. The patients have received an individually tailored exercise prescription for six months after which the measurements will be repeated.

Results, the peak heart rate reached at exercise was 166±15 beats/min, and maxVO₂ 31.6 ± 2.1 ml/kg/min. The patients with RV-systemic ventricle indicated significantly less habitual weekly exercise (2.2±1.6 vs. 5.8±3.h/week, p<0.005), had lower limb muscle mass (9.9±1.0 vs. 11.3±1.6 kg/m², p<0.03), lower maxVO₂ (27.1±6.0 vs. 31.6±2.1 ml/kg/min, p<0.05) without difference in peripheral oxygen saturation (92.4±4.2 vs. 93.1±2.5 %, ns.). All patients had restrictive lung function. However, FVC (p<0.0025), and FEV₁ (p<0.0001) were significantly inferior in patients with RV-systemic ventricle without difference in FEV₁/FVC suggesting more pronounced restriction of lung function.

Conclusions, our pediatric and adolescent patients with completed Fontan circulation had reduced exercise capacity from normal. The patients with RV-systemic ventricle showed less habitual exercise, and had inferior attributes of cardiopulmonary exercise capacity and lung function. The measures aiming at rehabilitation and maintenance of the patient's physical fitness need to target both ventilatory as well as hemodynamic qualities.