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Exercise capacity correlates with ventricle size in adult operated tetralogy of Fallot
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Purpose: Cardiopulmonary exercise testing (CPET) and cardiac magnetic resonance imaging (CMR) are used to time pulmonary homograft replacement in operated TOF patients with significant pulmonary regurgitation (PR). We correlated CPET and CMR parameters in these patients.

Methods: Single-centre retrospective analysis of 36 operated TOF patients (post-repair 21±7 yr; 30 NYHA I, 6 NYHA II; age 28±11 yr; 29 males) with at least moderate PR on CMR who underwent CPET within 15 median (5, IQR 1-7) months of CMR. CPET parameters were compared with 30 age- and sex-matched healthy controls (age 30±10 years, male=24).

Results: Peak oxygen consumption (VO2max) (29.0±7.1 ml/kg/min, p=0.001) and peak oxygen pulse (POP) (11.6±3.8 vs 13.9±2.9 ml/beat, p=0.008) are significantly lower in TOF group vs control. No significant difference in anerobic threshold (AT) and VE/VCO2 slope seen. Univariate analyses show negative correlation between PR fraction and AT; positive correlation between indexed left (LV) and right (RV) ventricular end-diastolic volumes and VO2max (see table). These remain significant after age and sex adjustments. RV restriction has no correlation with CPET results.

Conclusions: TOF subjects have near normal exercise capacity but significantly lower VO2max and POP vs controls. Increased PR fraction is associated with lower AT; increased RV and LV diastolic volumes, unexpectedly with higher VO2max. The latter may represent adaptive LV and RV remodeling in well-compensated TOF subjects whose exercise habits were not studied here.

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The impact of pulmonary valve replacement on left ventricular mechanical efficiency in adult patients with tetralogy of fallot: a study with the bi-axial energetics
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Purpose: Pulmonary valve replacement (PVR) improves right ventricular (RV) function in patients with tetralogy of Fallot (TOF). However, the impact of PVR on left ventricular (LV) function and the mechanism of improvement are still unclear. The purpose of this study is to clarify the impact of PVR on LV mechanical efficiency in adult patients with TOF.

Methods: Fourteen patients with TOF (age 30±1.1±11.5 years) who underwent PVR for significant pulmonary insufficiency were enrolled in this study. We measured left ventricular end-diastolic volume index (LVEDVI), left ventricular end-systolic volume index (LVESEVI), contractility (end-systolic elastance; Ees), afterload (effective arterial elastance; Ea), and mechanical efficiency (ventricular-aortic coupling; Ea/Ees and the rate of stroke work and pressure-volume area; SW/A) from echocardiographic data before, after, and later after PVR. Ees, Ea, and SW/PVA were approximated as follows: Ees=mean arterial pressure/minimal ventricular volume, Ea = maximal ventricular pressure / (maximal ventricular volume-minimal ventricular volume), and SW/PVA=1 / (1 + 0.5 Ea/Ees).

Results: LVEDVI, stroke volume and ejection fraction increased significantly after PVR. Contractility (Ees) did not change but afterload (Ea) decreased, and LV mechanical efficiency (Ea/Ees and SW/PVA) improved significantly. These changes were observed just after PVR, and further improvement was not noted in the late follow-up.

Conclusions: LV efficiency improves after PVR. Increased pulmonary forward flow and relief of the compression from the RV increase LVEDVI and stroke volume. LV afterload decreases to adapt the increase of stroke volume without the change of contractility. These changes result in the improvement of LV mechanical efficiency.

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Impaired preload reserve during exercise limits exercise capacity in the better Fontan patients
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Purpose: Intuition in patients with univentricular heart disease the total pulmonary connection (TCP) has been the preferred technique for the last 20 years. Since the introduction, mortality and morbidity of the Fontan operation have improved vastly. Limited exercise tolerance remains a problem. It is not clear if exercise tolerance differs between both available techniques (i.e. intra-atrial lateral tunnel (ILT) or extracardiac conduit (ECC)) in the modern, 2-stage approach of the Fontan circulation. Purpose of this study was to compare exercise testing results between these two groups.

Methods: 82 Fontan patients (50 male) with a TCP, age 12±4±2 years, age at TCP completion 3±2±1 years, successfully underwent cardiopulmonary exercise testing (CPET) (peak respiratory exchange rate (RERpeak) <1.00) on a bicycle ergometer. Peak workload (Wpeak), peak heart rate (HRpeak), peak VO2 (VO2peak) and VE/VCO2-slope were determined. Predicted values were derived from a group of healthy controls. A distinction was made between ILT (n=33) and ECC (n=49) modifications of the TCP-technique.

Results: For the entire group mean RERpeak was 1.08±0.05, mean Wpeak was 70±16%, mean VO2peak was 73±10%, median VE/VCO2 slope was 107% (inter-quartile range (IQR) 16%) and mean HRpeak (170±18/min) was 91±10% of the predicted value. There was no difference in age at time of the test between the 2 groups (12±7±3 IQR 9.9 vs. 11.9±2.2 (ECC) years, p=0.77). Outcomes for the ILT and ECC group were comparable for percentage of predicted values of Wpeak (66±17% vs. 71±15%, p=0.134), HRpeak (90±6% vs. 92±11%, p=0.358) and VO2peak-slope (105% (IQR 24%) vs. 109% (IQR 14%), p=0.98). The reached percentage of predicted VO2peak was lower for the ILT group compared to the ECC group (69±14% vs. 76±16%, p=0.046).

Conclusion: CPET parameters Wpeak, VO2peak and HRpeak and VO2peak-slope are impaired in contemporary Fontan patients. The results are comparable for ILT and ECC techniques concerning most values. However the percentage of predicted VO2peak was lower in patients with an intra-atrial lateral tunnel. These results show that reduced exercise capacity remains an important issue in Fontan patients. The ECC modification might have a slightly more favorable outcome for exercise capacity at medium term follow up.