Influence of non-invasive hemodynamic CMR parameters on maximal exercise capacity in surgically untreated patients with Ebstein’s anomaly

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Introduction: Ebstein’s anomaly is often associated with RV dysfunction. Data on RV function in surgically untreated patients are however rare. Since a good quality of life correlates with a good exercise capacity in daily life, we investigated non-invasive quantitative data derived from cardiovascular magnetic resonance (CMR) and its impact on maximal exercise capacity in patients with surgically untreated Ebstein’s anomaly.

Methods: We investigated 54 unoperated patients with Ebstein’s anomaly, age 5 to 69 years (median 30 years) and examined these patients with CMR and cardiopulmonary exercise testing (CPET). We compared seventeen CMR parameters with CPET parameters. We performed univariate and multivariate analysis with the focus on the maximal exercise capacity in these patients. For the maximal exercise capacity peak oxygen uptake as the percentage of normal (peakVO₂%) was selected. The following CMR volume and flow parameters were correlated to peakVO₂%. Both right and left ventricular ejection fraction (RVEF and LVEF), the indexed enddiastolic and endsystolic volumes (RVEDVi, RVESVi, LVEDVi and LVESVi) as well as the indexed stroke volumes (RVSVi and LVSVi), the total normalized right and left heart volumes (volume of the atrium and the ventricle together) as well as the total right to left heart volume ratio (R/L-ratio). Indexed flow data as the antegrade (PA ante, Aorta ante) and the indexed net flow (PA netto, Aorta netto) in the pulmonary artery and the aorta as well as its normalized values on heart rate (CI-PA, CI-Aorta) were used.

Results: RVEF ($r^2 0.2788$), PA netto ($r^2 0.2330$), and PA ante ($r^2 0.1912$) showed the best correlation with peakVO₂% (all $p<0.001$). Further significant linear correlation could also be demonstrated with CI-PA, LVEF, LVSVi, Aorta netto, RVESVi and Aorta ante. All other parameters did not show a significant correlation with peakVO₂%. Multivariate analysis for RVEF and PA netto revealed a $r^2$ of 0.4350.

Conclusions:
CMR parameters reflecting cardiac function as RVEF and LVEF and flow data of cardiac forward flow best correlate to peakVO₂%. The evaluation of the indexed net flow in the pulmonary artery and the overall function of the right ventricle (RVEF) best predicts the maximal exercise capacity in patients with Ebstein’s anomaly.