P-216

The functional right ventricle in patients with Ebstein´s anomaly is not small

Nerad M.(1), Mueller D. (1), Buchner C.(1), Rieger H.(1), Steinlechner E(1)., Meierhofer C.(1), Martinoff S.(2), Hess J(1), Stern H.(1), Fratz S.(1)
(1) Department of Paediatric Cardiology and Congenital Cardiac Defects, TUM, Deutsches Herzzentrum München
(2) Department of Radiology and Nuclear Medicine, TUM, Deutsches Herzzentrum München

INTRODUCTION:
Contemporary opinion regarding the size of the functional right ventricle (RV) in patients with Ebstein´s anomaly is that the functional RV is small. However, only few have attempted to measure the size of the functional RV in post-mortem hearts of patients with Ebstein´s anomaly. Therefore, in vivo size of the functional RV in patients with Ebstein´s anomaly is actually still unknown. Therefore, the aim of this study was to determine the size of the functional RV of patients with Ebstein´s anomaly without previous cardiac surgery by cardiovascular magnetic resonance.

METHODS:
Cardiovascular magnetic resonance of 36 consecutive patients with unoperated Ebstein´s anomaly (mean age 33±20yrs, 23 female, February 2006 till December 2010) determined functional RV volume. Ventricular volumes were measured using standard analysis software.
Additionally, in a subgroup of 11 patients with Ebstein´s anomaly short axis and axial slices were performed to measure ventricular volumes and to determine inter- and intra-observer variances.

RESULTS:
Mean Right Ventricular Enddiastolic Volume (RVEDV) was 256±141ml; RVEDV indexed to body surface area (RVEDVI) was 154±70ml/m². Maximum values were 656ml and 365ml/m², respectively. Only 4 Patients had normal RVEDVI<90ml/m². RVEDV: LVEDV ratio was 2.9±1.7 : 1. Maximum RVEDV:LVEDV was 8.8 : 1.
Inter- and intra-observer variance were not significantly different between axial and short axis slice measurements.

CONCLUSIONS:
Our results show that the volume of the functional RV in patients with Ebstein´s anomaly is not small. Only 4 patiens had normal RVEDVI. None of the patients had a small functional RV. Therefore, contemporary opinion regarding the size of the functional RV in patients with Ebstein´s anomaly should be revised.