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The unnatural course of right ventricular dilatation in patients with severe pulmonary regurgitation after repair of Tetralogy of Fallot

Hoelscher M. (2,3), Bonassin F. (1,2,3), Oxenius A.(1,2,3), Greutmann M. (1,2,3) Leonardi B. (4), Kellenberger C.J. (3,5), Valsangiacomo Buechel ER. (2,3)

1 Clinic for Cardiology, Cardiovascular center, University Hospital Zurich, Switzerland

2 Paediatric Heart Centre, University Children's Hospital, Zurich, Switzerland

3 Children's Research Centre, University Children's Hospital, Zurich, Switzerland

4 Department of Cardiology and Cardiac Surgery Bambino Gesù Children's Hospital, IRCCS Rome, Italy

5 Department of Diagnostic Imaging, University Children's Hospital, Zurich, Switzerland

Background:

Pulmonary valve regurgitation (PR) and related right ventricular (RV) dilatation are frequent sequelae after surgical repair for Tetralogy of Fallot (TOF). Timing of pulmonary valve replacement is still subject of debate and little is known about the rate of progression of RV dilatation. We sought to analyze the natural course of RV dilatation using serial magnetic resonance (CMR) data.

Methods:

This retrospective multicentric case control study included all TOF patients with significant PR and RV dilatation, who underwent serial CMR scans between 2002 and 2016 without any intervention between CMR exams. RV volumes and degree of PR were quantified in both centers using the same CMR technique. Clinical and volumetric CMR data were analyzed regarding progression of ventricular dilatation and related potential risk factors.

Results:

87 consecutive patients (82 TOF, 3 DORV, 2 PA/VSD) underwent at least 2 (range 2-6) CMR examinations. Median age at first CMR was 13.7 yrs (range 1.4 – 49 yrs), time interval to last CMR was 3.3 yrs (6 months-10.5 yrs).

In the overall group no significant change was observed between first and last CMR for RV end-diastolic volume (RVEDV 151±32 vs 155±34 ml/m²), RV end-systolic volumes (RVESV 79±28 vs 80±28 ml/m²), RV function (EF 49±8% vs 49±8%)

Progression of dilatation, defined as an increase >20ml/m², occurred in 20 patients (22%). In this group RVEDV increased from 153±31 ml/m² to 183±30 ml/m² (p<0.001); RVESV changed from 89±33 ml/m² to 105±30 ml/m² (p<0.001); ejection fraction did not deteriorate significantly (46±9% vs 46±7%). Presence of a transannular patch, initial RV volume and age at repair were not predictive for progressive RV dilatation. In contrast longer time interval between repair and first CMR (p 0.01) and to last CMR (p 0.03) were correlated to increasing RV dilatation.

Conclusion:

During a follow-up of 3.3 yrs, only in 22% of all TOF patients with PR experience significant RV dilatation.

Time since surgical repair was the only factor predictive for progression of dilatation.