Determinants of pulmonary valve replacement indication in repaired Tetralogy of Fallot patients: a multicenter experience

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Introduction: Repaired Tetralogy of Fallot (rToF) patients often need pulmonary valve replacement (PVR) due to progressive right ventricular (RV) dilation and biventricular dysfunction. Cardiac magnetic resonance (CMR) represents the gold standard to evaluate RV size and function aiding definition of PVR timing. Determinants of PVR have not yet been described.

Methods: We enrolled patients who underwent hemodynamic evaluation by CMR late after ToF repair (by either transanular or infundibular RV outflow tract reconstruction) in five paediatric cardiology centers, between March 2003 and March 2013. Surgical data (time and types of repair) and CMR parameters were collected. Indications to PVR were centrally reviewed.

Results: 495 patients (60% males) aged 21±11 (range 2-67) years were included. Mean age at repair was 30±54.2 (range 0.2-574.8) months. In the majority of cases (77%) transanular patch was the type of surgical correction performed. Among the study population 222 patients (47%) had RV end-diastolic volume indexed for bsa (RVEDVi)>150 mL/m2 and/or RVEF<47%, whose 70% were males and 86% with transanular patch (TP) repair. In multivariate logistic regression, in TP group, presence of RVEDVi>150 mL/m2 and/or RVEF <47%, whose 70% were males and 86% with transanular patch (TP) repair. In multivariate logistic regression, in TP group, presence of RVEDVi>150 mL/m2 and/or RVEF <47% was significantly correlated with time elapsed from correction to MRI examination (p=0.017) (figure 1a) and to grade of pulmonary regurgitation (PR) (p=0.008). In addition, mean RV EF and LV EF were mildly lower in TP as compared to infundibular patch (IP) group (both p<0.05). In IP group RV dilation was not related to time from surgical correction (p=NS) (figure 1b), but only to grade of PR and to the presence of significant tricuspid regurgitation (p=0.022 and p=0.032, respectively).

Conclusions: The mechanism underling RV dilation in rToF patients appears to have significant differences in the two groups. Indeed, time from surgical repair seem to have a significant impact only in the TP group while tricuspid regurgitation seem to be crucial in the other. Longitudinal follow-up studies are needed to define the role of each factors involved in RV dilation in predicting the adverse outcomes of rToF patients.