Consistent improvement of right ventricular (RV) size and function after late pulmonary valve replacement following complete repair of Tetralogy Fallot (TOF): A 20-year experience

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Introduction: Pulmonary valve insufficiency (to various degrees) is unavoidable for most patients after TOF repair, but can be well tolerated for several years. However, longstanding pulmonary valve insufficiency can lead to progressively severe RV dilatation and eventual dysfunction, which may predispose to heart failure, arrhythmias, and sudden death. Pulmonary valve replacement (PVR) is advocated in an increasing over time proportion of patients with prior TOF repair, aiming to prevent (and hopefully reverse) further excessive RV enlargement and dysfunction. We report our experience with PVR late after complete repair of TOF.

Methods: Between 1/9/1997 and 1/11/2017, 60 patients (45 male - 15 female), median age 12 years (range 5 - 46) underwent PVR late after complete TOF repair. Most patients (n=36) had undergone total TOF repair at other institutions, while twenty four were among the more than 300 TOF repairs performed by our group over the same time period. Median time interval between repair and PVR was 10.5 (range 1-43) years. Over the study period, we have used uniform indications for PVR, including symptoms, increase in RV end-diastolic-volume-index (RVEDVI) to 160ml/m2 or more, and/or reduction of RV ejection fraction (RVEF) below 50%, as measured by MRI. Heterologous valved conduits (ContegraR, diameter 18-22mm), were used in 19 patients, 2 patients received HancockR conduits (16 and 22mm), and all 39 remaining patients had bioprosthetic valves (Edwards-MagnaR, 19-29mm). Concomitant residual lesions (tricuspid or aortic valve insufficiency, residual VSD, RVOTO or pulmonary artery stenosis) were repaired in 23 patients.

Results: There was no early or late mortality. All symptomatic patients improved their NYHA functional class from median II to I (p<0.001). At follow up (median 8 years, range 1 to 20), RVEDVI improved from 165ml/m2±15 to 132ml/m2±16 (p=0.003), and RVEF from 42%±7 to 49%±7 (p=0.007). No patient has required redo PVR.

Conclusions: Late PVR after prior complete TOF repair can be performed safely, and results not only in stabilization but actually in improved patient functional status, reduced RV size and also in improved RV function.