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ORIGINAL ARTICLE

Right Ventricular Outflow Tract Enlargement Prior to Pulmonary Valve Replacement is Associated with Poorer Structural and Functional Outcomes, in Adults with Repaired Tetralogy of Fallot

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Background Pulmonary valve replacement (PVR) is commonly performed late after Tetralogy of Fallot (TOF) repair. We examined the effects of PVR on cardiac structure, function and exercise capacity in adults with repaired TOF.

Methods Eighteen adult patients with repaired TOF and severe pulmonary regurgitation (PR) with right ventricular (RV) dilatation requiring PVR for clinical reasons (age; 25 ± 8 years) were recruited to undergo cardiac MRI (1.5 T) and cardiopulmonary exercise testing before and 14 ± 3 months after PVR.

Results Reduced indexed RV end-diastolic volume (RVEDVi; 186 ± 32 mL/m^2 pre-op vs 114 ± 20 mL/m^2 post-op, $p < 0.001$) was observed after PVR. ''Normalisation'' of RVEDVi ($\leq$108 mL/m^2) was achieved in only seven of 18 patients. Pre-PVR RVEDVi correlated with post-operative change in RVEDVi (change = −72.1 ± 20.4 mL/m^2, $r = -0.815$, $p < 0.001$). Exercise capacity remained high-normal post-PVR (% predicted maximal workload: 93 ± 16% vs 91 ± 12%, $p = 0.5$). Regional RV volumes were assessed; RV outflow tract (RVOT) volumes were compared to the RV muscular corpus. Large pre-PVR RVOT volumes correlated negatively with post-surgical RV ejection fraction, peak VO_2 and delta VO_2 at anaerobic threshold ($p < 0.05$ for all).

Conclusions Normalisation of RV volume is unlikely to be achieved above a pre-PVR RVEDVi of 165 mL/m^2 or more. In particular, an enlarged RVOT prior to PVR predicts suboptimal structural and functional outcomes.

Keywords Tetralogy of Fallot • Pulmonary valve replacement • Adult congenital heart disease • Right ventricular dilatation • Cardiopulmonary exercise test • Cardiac magnetic resonance imaging

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Introduction

Trans-annular patch repair of Tetralogy of Fallot (TOF) leaves patients with pulmonary regurgitation (PR) which, in the long-term, leads to right ventricular (RV) dilatation, which in turn has been associated with exercise intolerance, arrhythmia, RV dysfunction and sudden cardiac death [1–6]. As such, pulmonary valve replacement (PVR) is a very common late procedure in young adults with repaired TOF and has been shown to be safe and effective in reducing pulmonary regurgitation, RV volume, QRS duration and improving exercise capacity [7–14]. However, these beneficial results need to be balanced against the risk associated with repeat PVR due to a limited lifespan of the implanted valve and the lack of conclusive evidence for improved survival [15–18]. Thus, the timing of PVR is of great importance and is a point of current conjecture [9,10,18–20].

Previous reports have documented the effects of PVR in adults with impaired exercise capacity prior to PVR [8–13] and have failed to consider regional aspects of RV structure in assessing the responses to PVR surgery. In particular, we were concerned that dilatation of the RV outflow tract (RVOT) would not necessarily diminish after PVR (as opposed to likely diminution in size of the muscular RV corpus [RVMC]) and that residual enlargement of the RVOT would be associated with (i) failure to normalise RV volumes after PVR and (ii) failure of exercise capacity to improve. We therefore studied regional RV volumes and exercise capacity, in adults with PR after TOF repair, before and after clinically indicated PVR.

Methods

Patient Population

Between April 2009 and December 2012, a total of 18 repaired TOF patients referred to our adult congenital heart service for a cardiac MRI and cardiopulmonary exercise test (CPET) were prospectively recruited to undergo these assessments both before and approximately one year after surgical PVR (Table 1). Ethical approval was obtained from the Royal Prince Alfred Hospital Human Research Ethics Committee, Sydney, and all participants gave informed consent.

Table 1 Cohort Characteristics.

Participants (*n*)	18
Age (years)	25 ± 8
Males (*n*)	14
Height (cm)	174 ± 8
Weight (kg)	74 ± 14
Age at TOF repair (years)	2 ± 1
PVR details	
Bioprosthesis (*n*)	15
Pulmonary homograft (*n*)	2
Aortic homograft (*n*)	1
Cross clamp time (min)	47 ± 18
Pulmonary artery augmentation (*n*)	10
Transventricular incision (*n*)	18
Years between TOF repair and PVR	23 ± 7

PVR, pulmonary valve replacement; TOF, Tetralogy of Fallot.

Cardiovascular MRI Protocol

MRI was performed using a 1.5 T MR scanner (GE Medical Systems).

Assessment of ventricular volumes and function using cine MRI

Vertical long-axis, four-chamber and short-axis views of the heart were acquired over the entirety of both ventricles (9–12 slices) using retrospectively gated steady-state free precession (FIESTA) cine MRI images. Image parameters: TR = 3.2 ms; TE = 1.6 ms; flip angle = 78°; slice thickness = 8 mm; matrix = 192 × 256; field of view = 300–380 mm; and temporal resolution = 40 ms, acquired during a single breath-hold. Manual segmentation of short-axis cine images with endocardial outline at end diastole and end systole was used to assess RV and LV volumes (performed in Osirix software, version 4.0 32bit). Regional analysis of the RV volumes in systole and diastole was then performed, where the RV was divided into the fibrous RVOT and RVMC components. The delineation of these segments was based on the following criteria. The fibrous RVOT was contoured from the pulmonary valve leaflets superiorly, to the fibrous-muscular transition zone within the RVOT anteriorly based on delayed enhancement studies. Posteriorly the fibrous RVOT was contoured to include volume until the superior aspect of the interventricular septum. An arbitrary line was contoured between the lower anterior and posterior landmarks to define the lower border zone of the fibrous RVOT. Fibrous RVOT measurements were independently analysed in all subjects by two observers with an intraclass correlation coefficient of 0.977 for end-systolic measurements and 0.923 for end diastolic measurements. Simpson's rule was used to calculate end-diastolic and end-systolic volumes for both ventricles and the RVOT; ejection fractions were calculated from these volumes.

Magnetic resonance angiography protocol

Pulmonary artery (PA) morphology was assessed with a dedicated aortic MRA. Administration of intravenous contrast (0.2 mmol/kg of gadolinium pentatate, Magnevist) was triggered visually and was acquired in a 15 s breath-hold. The two-phased MRA was performed with a T1 weighted spoiled gradient echo sequence. The following imaging parameters were used: TR = 7 ms; TE = 3.5 ms; flip angle = 30°; matrix = 256 × 192; field of view = 480 mm; slice thickness = 3 mm, zip 2.

MR flow calculation

A phase-sensitive (VENC 200) gradient-echo sequence (TR, <5 ms; TE, <3 ms; flip angle, 15°; slice thickness, 7 mm; field of view = 300–380 mm matrix, 256 × 240, temporal resolution = 30 ms) was acquired during a breath-hold in order to obtain PA and aortic flow data. The respective midpoint of the main PA and ascending aorta (sino-tubular junction) was used as image planes. Retrospective cardiac gating was used to