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Ventricular function, myocardial delayed enhancement and patient-reported quality of life in adolescents and adults with repaired tetralogy of Fallot

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Introduction

In patients with repaired tetralogy of Fallot (TOF), right ventricular myocardial delayed enhancement (MDE) and diastolic dysfunction are common, and have been associated with decreased exercise capacity and increased arrhythmia. Predictors of quality of life (QOL) have not been reported in this population.

Purpose

We assessed the hypothesis that a greater degree of MDE in adolescents and adults with repaired TOF would correlate with diastolic dysfunction and decreased QOL.

Methods

Patients with repaired TOF ≥ 14 years old undergoing cardiovascular magnetic resonance (CMR) had ventricular volumes measured from SSFP cine images in the short axis plane, and peak early and atrial filling rates, early filling fraction and deceleration time calculated from right ventricular inflow curves, generated by the sum of tricuspid inflow and pulmonary insufficiency measured from phase contrast imaging. MDE was scored 0-3 (none, <2 cm, 2-3 cm, >3 cm) in each of 6 regions of the right ventricle and added for a total score. Patients completed the Short Form 36, version 2, a QOL assessment validated in multiple populations with a normalized scale.

Results

Forty-seven patients (32 \pm 15 years old, 62% male) underwent CMR. MDE was present in 42/46 (91%), with enddiastolic forward flow (EDFF) in the pulmonary artery in 29/45 (64%). Mean right ventricular ejection fraction (RVEF) was $45 \pm 8\%$, with mean left ventricular ejection fraction (LVEF) 54 ± 8%. MDE inversely correlated with RVEF (r = 0.30, p = 0.04). Patients with MDE scores above the median had a lower early filling fraction (35.5 ± 11.9% vs. $44.1 \pm 14.6\%$, p = 0.04), with a trend toward higher atrial filling rates (357 \pm 123 vs. 287 \pm 128 ml/s, p = 0.08). MDE did not differ in patients with or without EDFF, and did not correlate with pulmonary regurgitant fraction or volume. QOL physical and mental summary scores did not differ from population-derived norms. Physical functioning subscale scores trended lower than the general population (47.6 \pm 9.7 vs. 50.0 \pm 10.0, p = 0.10) and correlated positively with higher MDE scores (r = 0.32, p = 0.03). Role-physical subscale scores correlated with higher LVEF (r = 0.46, p = 0.001) and longer deceleration time (r = 0.31, p < 0.05) and trended with higher RVEF (r = 0.27, p = 0.07). QOL scores did not differ with EDFF, pulmonary regurgitation, or end diastolic volume.

Conclusion

Patients with greater degree of MDE demonstrated right ventricular systolic dysfunction and evidence of impaired relaxation, but did not have lower QOL scores. LVEF was the best predictor of patient-reported QOL, underscoring the importance of preserving LVEF in this population. The influence of pulmonary valve replacement on QOL remains unclear.

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