

The Impact of Pulmonary Insufficiency on Patients with Surgically Repaired Pulmonary Stenosis vs. Tetralogy of Fallot by Cardiac Magnetic Resonance

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Background: Patients with repaired pulmonary stenosis (PS) and tetralogy of Fallot (TOF) both develop pulmonary insufficiency (PI) leading to right ventricular (RV) dilatation and dysfunction. Cardiac magnetic resonance (CMR) plays a key role in determining timing of pulmonary valve replacement (PVR) in TOF, but it is unclear whether these criteria can be extrapolated to patients with PS. We aimed to compare the differential effect of pulmonary insufficiency on RV volume and systolic and diastolic function in patients with surgically repaired PS and TOF.

Methods: All patients with surgically repaired PS, undergoing CMR from 2007-2013, were matched 1:2 by age and pulmonary regurgitant fraction with TOF patients. Patients with prior PVR were excluded. Right and left ventricular (LV) ejection fraction (EF), end-diastolic volume (EDV), and presence of end-diastolic forward flow in the pulmonary artery (EDFF) were compared. Using feature tracking software (Tomtec, Unterschleissheim, Germany), RV longitudinal systolic strain and diastolic strain rate were measured from a 4-chamber slice, and RV circumferential strain from the most basal short-axis slice with circumferential RV myocardium.

Results: In 24 PS patients (mean $40.7 \pm SD 13.3$ years old, 41.7% male) with pulmonary regurgitant fraction of $41.5 \pm 17.1\%$ and 47 TOF patients (mean $39.1 \pm SD 12.5$ years old, 55.3% male) with pulmonary regurgitant fraction of $40.9 \pm 16.3\%$, there was no difference in RV EDV (153.3 vs. 153.5 mL/m², $p = 0.99$), presence of EDFF (83.3 vs. 61.7% , $p = 0.10$), or RV longitudinal diastolic strain rate (1.27 vs. 1.13 , $p = 0.13$). However, PS patients had preserved RV EF (54.3 vs. 48.0% , $p < 0.0001$) and LV EF (57.7 vs. 54.8% , $p = 0.05$) compared to TOF patients, predominantly due to difference in RV circumferential (-15.8 vs. -11.8 , $p < 0.0001$) rather than longitudinal strain (-18.0 vs. -15.9 , $p = 0.04$).

Conclusions: With the same degree of PI, PS patients have similar RV dilatation and diastolic dysfunction compared to TOF patients. However, RV EF is preserved, predominantly due to differences in outflow tract function, as is LV EF. This may reflect different degrees of scarring and ventricular-ventricular interaction, and suggests CMR criteria for PVR may not be identical for patients with PS vs. TOF.