

Right Ventricular Outflow Tract Growth in Palliated Cyanotic Tetralogy of Fallot

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Background: Chronic pulmonary insufficiency following transannular patch repair of tetralogy of Fallot with pulmonary stenosis (TOF/PS) is associated with adverse outcomes. Our institutional strategy for symptomatic neonates with TOF/PS and severely hypoplastic pulmonary valves (PV) consists of palliation with a modified Blalock-Taussig shunt (mBTS), with the goal of increasing the likelihood of a pulmonary valve sparing (PVS) repair later in infancy. As little contemporary data are available regarding pulmonary valve and pulmonary artery growth in palliated patients with TOF/PS, we performed a retrospective study examining longitudinal growth of these structures by echocardiography.

Methods: We reviewed surgical data for all patients who underwent repair of TOF/PS at our center from 2000 to 2012. In the subset of patients initially palliated with a mBTS, pulmonary valve annulus and main and branch pulmonary artery dimensions were assessed by echocardiography prior to mBTS and prior to surgical repair. Patients who underwent right ventricular outflow tract intervention prior to repair and patients without echocardiograms available for review were excluded.

Results: Of 172 patients with repaired TOF/PS, 40 (23%) were initially palliated with a mBTS shunt, of which 31 (78%) met inclusion criteria. Palliated patients had significantly hypoplastic PV and mildly hypoplastic main (MPA) and branch pulmonary arteries (RPA, LPA) at baseline (z-scores: PV -3.0 ± 0.9 , MPA -2.3 ± 0.8 , RPA -1.9 ± 1.1 , LPA -1.4 ± 0.8). All structures had significant absolute growth prior to surgical repair ($p < .001$ for all). PV and MPA z-scores were unchanged (PV z-score = -3.3 ± 1.2 , $p = .28$; MPA z-score = -2.4 ± 1.4 , $p = .46$), whereas the branch pulmonary arteries showed significant improvement in z-scores (RPA z-score = -1.2 ± 1.5 , $p = .03$; LPA z-score = -0.8 ± 1.3 , $p = .008$). PVS repair was performed in 10 of 31 palliated patients (30%), a far higher rate of PV annulus preservation than that reported in most published series of primary neonatal complete TOF repairs.

Conclusions: Palliation with a mBTS in cyanotic patients with TOF/PS is associated with absolute growth of right sided structures and allows for normalization of branch pulmonary artery size. Although pulmonary valve z-scores did not improve after mBTS, absolute growth may facilitate valve sparing repair in one third of patients.