Earlier Conduit Placement in Down Syndrome Patients Following Tetralogy of Fallot Repair

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Background: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect and is relatively common in patients with Down syndrome (DS) either with or without concomitant atrioventricular septal defect. Down syndrome has a strong association with pulmonary arterial hypertension, which could contribute to more severe pulmonary regurgitation after TOF repair and possibly result in earlier conduit placement. We sought to compare cardiac magnetic resonance (CMR) measures of pulmonary regurgitation and right ventricular dilation as well as timing of conduit placement between those with and without DS after initial TOF repair.
Methods: A retrospective review of our surgical database from 2000-2015 was performed to identify patients with TOF with pulmonary stenosis. Those with pulmonary atresia, major aortopulmonary collaterals, discontinuous pulmonary arteries, and absent pulmonary valve syndrome were excluded. We also excluded patients who had initial repair with a conduit. Those with DS were compared to those without using Fisher's exact test for categorical outcomes and Wilcoxon rank sum test for continuous outcomes. Cox proportional hazards modeling was performed to determine risk factors for conduit placement. Kaplan Meier curves were created for freedom from conduit placement.
Results: The cohort included 284 patients with 171 (60%) males. There were 35 (12%) with DS and 31 (11%) with another genetic syndrome. The primary cardiac diagnosis was TOF in 262 (92%) and TOF with atrioventricular septal defect in 22 (8%) with 16 (73%) of those having DS. Pre-repair intervention to augment pulmonary blood flow was performed in 60 (21%) and 210 (74%) had a transannular patch repair, with no difference between the DS and non-DS groups. DS showed a greater degree of pulmonary valve regurgitation on CMR (55 ± 14% vs. 37 ± 16%, p=0.01). In multivariable analysis DS (HR 2.3, 95% CI 1.2-4.5, p=0.02) and transannular patch repair (HR 5.5, 95% CI 1.7-17.6, p=0.004) were significant risk factors for conduit placement after repair. In Kaplan Meier analysis those with DS had significantly lower freedom from conduit placement (p=0.03). Freedom from conduit placement at 15 years was 61% (95% CI 46-73%) in the non-DS group and 37% (95% CI 12-63%) in the DS group.
Conclusion: Down syndrome is associated with an increased degree of pulmonary regurgitation and earlier conduit placement after TOF repair. These patients require earlier assessment by CMR to determine timing of conduit placement, and evaluation for and treatment of preventable causes of pulmonary hypertension may be beneficial.