

Influence of truncal root dilation on truncal valve insufficiency

Willes, RJ. Hill, Garick

Introduction: Truncus arteriosus, while a relatively uncommon congenital heart defect, is associated with significant morbidity. Re-intervention for truncal valve repair or replacement is common. While aortic root dilation has been described in this population, the relationship between root dilation and truncal valve insufficiency has not been studied.

Methods: We performed a retrospective review of all patients with a diagnosis of Truncus arteriosus with at least one echocardiogram available at the Herma Heart Center at the Children's Hospital of Wisconsin between January 2000 and December 2015. Data collected included demographic data, comorbid conditions, the number and timing of surgeries, type of truncus arteriosus, valve morphology, number and timing of catheter procedures, and major adverse events. Serial echocardiograms were measured to determine truncal root and valve dimensions and the degree of truncal insufficiency.

Results: In the 79 patient cohort the median age was 10.5 (range 1.5-48 years). There was a male predominance with 50 (63%) male. There were a total of 204 surgical and catheter interventions including 6 patients who required truncal valve replacement due to severe truncal insufficiency. There were a total of 7 deaths, with 1 prior to surgical intervention. Six of the total cohort underwent truncal valve replacement. At the time of their most recent echocardiogram 30 (38%) patients had trivial or no truncal insufficiency, 17 (37%) had moderate insufficiency and 8 (10%) had severe truncal valve insufficiency. The median valve annulus measured 2.8 cm (range 0.77-4.79) Truncal root measurements consistently demonstrated Z scores > 5 but Z-scores did not increase over time. Valve annulus, valve morphology, and 22q11 deletion syndrome were not associated with moderate or severe truncal insufficiency in univariate analysis.

Conclusions: Significant truncal valve insufficiency is common in long term follow up. While truncal roots are universally dilated, the dilation is not progressive and is not associated with truncal valve insufficiency. Further study into those who develop truncal valve insufficiency is needed to determine a cause.