Aortic Diameter In Children With Bicuspid Aortic Valves

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Introduction: Prediction of aortic growth in patients with bicuspid aortic valve (BAV) is essential to identify patients at risk for dissection, but data in children are scarce.

Methods: We retrospectively identified all children with a BAV from an echocardiography database between 2005-2013. Medical records were reviewed and echocardiographic images re-measured. Aneurysm ($z >2$) was based on two different z-score equation methods (Gautier et al. vs Campens et al.).

Results: In total 251 BAV patients were identified, mean age 5.8 years (64% males). Valve pathology (stenosis and/or regurgitation) occurred in 100% of Sievers' type 2, 62% of type 1 and 46% of type 0 ($p=0.001$). In 234 patients aortic diameters were measured, mean follow-up was 4.7 years. The two z-score equation methods disagreed significantly for aortic sinus and, in patients below age 1.5 years for ascending aorta (AA). AA aneurysm was present in 24% (Gautier) vs 36% (Campens) at inclusion. Mean z-scores progressed significantly with age: 0.67 to 1.24 vs 0.45 to 1.29 at 5 and 15 years, respectively. Associations for AA growth were an initial z-score $>2$ ($p<0.001$) and Sievers’ type 2 ($p<0.05$). Dissection or preventive aortic surgery did not occur. AA measurements on MRI and echocardiography agreed well (mean difference 1.1 mm, $p=0.09$).

Conclusions: Aortic complications seem no major threat in paediatric BAV patients. Ascending aorta growth, based on existing z-score equations, is faster than expected. BAV morphology according to Sievers et al. seems predictive for valvular and aortic prognosis. MRI appears redundant for paediatric aortic diameter evaluation.