P-124

Progression of ascending aorta dimensions in pediatric patients with bicuspid aortic valve

Trippel A. (1), Ammann RA.(1), Bruhin S. (1), Pfammatter J.P. (1), Pavlovic M. (1)
Pediatric Cardiology
University Hospital
3010 Bern
Switzerland

Introduction
Bicuspid aortic valve (BAV) is the most common congenital cardiac defect. Apart from obstructive lesions, a significant part of patients develop dilatation of the ascending aorta. Natural history and significance of aortic dilatation in pediatric BAV patients is not well defined. Therefore, we studied progression of dimensions in these patients with and without coarctation.
Methods
Patients with BAV or with BAV and coarctation (BAV+coarc) were selected from institutional database. Inclusion criteria were: at least three echocardiograms over last 15 years, each at least one year apart. Aortic diameters were measured at 3 levels (annulus, sinus, ascending), and transformed into age- and gender-specific z-scores. Impact of gender, diagnosis (BAV vs. BAV+coarc) and valve function on changes of z-scores over time was analyzed by linear mixed-effects regression.
Results
116 measurements were recorded in 28 patients (9 patients BAV, 19 patients BAV+coarc). Mean age at time of measurements was 12.0 years (range, 0.1 to 23.1 years). Eighty-five measurements were performed in male patients. Aortic regurgitation was present in 29, and mild valvular stenosis in 22 measurements. No deaths were recorded. Overall, mean z-score
for aortic annulus was 1.14 (range, -1.72 to 3.72), for sinus 0.49 (-2.87 to 3.63), and for ascending aorta 1.86 (-2.02 to 5.54). Ascending aorta z-scores increased significantly over time in the entire sample (0.07/year; 95%CI, 0.04 to 0.10; $p<0.001$); in both BAV (0.12/year; 95%CI, 0.05 to 0.18; $p<0.001$) and BAV+coarc (0.06/year; 95%CI, 0.02 to 0.10; $p=0.002$); in boys (0.08/year; 95%CI, 0.04 to 0.11; $p<0.001$) but not in girls (0.03/year; 95%CI, -0.05 to 0.10; $p=0.49$); in patients with (0.10/year; 95%CI, 0.03 to 0.17, $p=0.006$) and without aortic regurgitation (0.07/year; 95%CI, 0.03 to 0.10; $p<0.001$); and in patients without aortic stenosis (0.07/year; 95%CI, 0.04 to 0.11; $p<0.001$) but not in patients with aortic stenosis (-0.02/year; 95%CI, -0.10 to 0.06; $p=0.65$).
Conclusions
Ascending aorta z-scores progressed significantly, particularly in patients with isolated BAV; indicating intrinsic wall abnormalities. Progression was influenced by gender and aortic function. Patients will be further examined for molecular markers, aiming to identify
patients at risk for severe progression, with potentially highest benefit from early medical treatment.