Bicuspid aortic valve syndrome as a centre of congenital heart defects of left ventricle outflow tract

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Introduction: Although bicuspid aortic valve (BAV) is not included in epidemiological studies of congenital heart defects (CHD) in children and is stated that the predictive factor for development of serious complications in BAV is the age over 30, our observations indicate that BAV is also a significant factor in CHD pathology in children. Here we tried to determine that significance.

Methods: all children examined in our institution during 11 years period (2000-2011) with diagnose of bicuspid aortic valve, isolated and/or associated with other congenital heart defects were included.

Results: we have found 229 BAV patients. The most common associated LVOT disorder was coarctation of aorta (32.75%). In our study 42.7 % patients with an isolated coarctation had BAV. 29.7% of all BAV patients had aortic stenosis (AS), aortic insufficiency (AI), and/or ascending aortic dilatation (DAA). Percentage of BAV patients with isolated aortic stenosis or insufficiency was the same (7%), BAV with stenosis and insufficiency in 12.7%, AS and DAA in 9.17% patients. Of all the children with BAV, 62.44% had hemodynamic alterations on the aortic valve which manifested themselves as AS and/or AI. In our study progression of aortic stenosis was common finding during childhood while insufficiency mostly stayed mild. Ascending aortic dilatation showed progression with age in significant number of patients. Significant number of patient needed interventional and/or cardiosurgical treatment with increasing number of interventions with age, in accordance with the expected progression of pathological changes (AS, AI, DAA).

Conclusion: Bicuspid aortic valve is a congenital heart defect with a progressive development of hemodynamic changes in the left ventricular outflow tract which may become haemodynamically relevant in childhood age already. Therefore, BAV should be regularly controlled, and dilatation of the ascending aorta should be prevented with beta blockers or ACE-inhibitors. For all the mentioned reasons, we believe that bicuspid aortic valve should be included in the epidemiological studies of congenital heart defects.