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Prevalence and clinical significancy of bicuspid aortic valve diagnosed in the first year of life: a long term follow up in the paediatric age

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Introduction and objectives: Bicuspid aortic valve (BAV) is the most common congenital malformation, with an estimated prevalence ranging from 0,48 to 2%. Most of the studies describe complications associated with BAV, including aortic stenosis and regurgitation, infective endocarditis and aortic dilation and dissection. Very few studies have been performed in the neonatal and paediatric age, most lacking in long-term follow up. The aim of our study was to explore the prevalence of BAV and the progression of aortic valve disease and aortic dilation in paediatric patients followed from January 1996 to August 2018.

Methods: A prospective collection of Congenital Heart Disease (CHD) diagnosed in the first year of life was performed in our institution from January 1996 to August 2018. Patients affected by BAV were then followed yearly. BAV morphology, aortic valve dysfunction and aortic dimensions were measured. Comparisons were performed between right-left cusp fusion (R-L) and right-non coronary cusp fusion (R-N), according to Shaefer's classification. Rate of change of the ascending aorta size over time was analyzed, and aortic size values were normalized as the number of standard deviations above or below the mean size expected for body surface area (z scores).

Results: A total of 35,310 live births were screened. Incidence rate of total CHD, including small ventricular septal defects and BAV was19,57%. Sixty had BAV (8,73% of CHD), with an incidence of 17/1,000 live births. Male/Female ratio was 1.4. A positive family history was present in 5 (8.3%). In 12 patients (24%) BAV was associated to other CHD (9 Aortic Coarctation, 1 Atrio-Ventricular Septal Defect, and in two cases a critical neonatal aortic stenosis). BAV was isolated in 48 cases (80%), with an incidence of 13.6/1.000 live births. According to Shaefer's classification the most common was type 1 (R-L), with Right to Left cusps fusion (R-L 76%, R-N 21% and L-N 3%). Among patients with isolated BAV, after a median follow up of 11 [1–21] years an aortic stenosis was observed in 5 patients (10.4%, none severe), an aortic insufficiency in 13 (27%, in one case severe). A dilated ascending aorta (z-score$>$2) was found in 11 patients (23%). An indication cardiac surgery due to severe aortic insufficiency was present in one case. No patient had indication for aortic aneurysm surgery.

Conclusions: Paediatric patients with isolated BAV warrant medical follow-up but progression towards severe valve dysfunction and/or dilation of the ascending aorta warranting cardiac surgery is very low.