Progression of aortic valve dysfunction and aortic root dilatation in paediatric patients with isolated bicuspid aortic valve

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Background: Few data exist regarding the rate of progression of aortic valve dysfunction and aortic root dilatation in paediatric patients with isolated bicuspid aortic valve (BAV).

Methods: A total of 179 paediatric patients (median [95% IQ] age at enrolment 7.8 [2.7-12.0] years, 76.5% male) with isolated BAV were prospectively followed. Patients with severe valve stenosis or regurgitation at baseline were excluded. Progression of aortic stenosis (AS) or regurgitation (AR) from baseline to follow-up was defined as an increase by ≥1 grade according to a three-level scale severity. Progression of aortic dimensions at different levels of the aortic root was estimated by calculating corresponding changes in the Z-scores. A Z-score >2 was considered significant.

Results: At baseline, AS was present in 25 (14.0%) patients (mild in 23, moderate in 2), whereas AR was present in 87 (48.6%) patients (mild in 79, moderate in 8). Mean diameters at the level of annulus, Valsalva sinuses (VS), sino-tubular junction (STJ), and proximal ascending aorta (AA) were 17.4±4.5 mm, 22.2±5.6 mm, 19.3±5.1 mm, and 21.7±6.2 mm, respectively. The corresponding proportions of subjects with significant enlargement (Z-score >2) at baseline of 24.6%, 15.1%, 22.4% and 39.1%.

After a median follow-up of 5.4 [2.3-9.2] years, AS was present in 28 (15.6%) patients (mild in 21, moderate in 6, and severe in one), whereas AR was present in 102 (57.0%) patients (mild in 75, moderate in 27). A progression ≥1 grade in AS and AR was observed in 9 (5.0%) and 29 (16.2%) patients, respectively.

Mean diameters at the level of annulus, VS, STJ, and AA were 20.4±5.0 mm, 26.7±6.2 mm, 21.9±5.9 mm, and 26.4±6.8 mm, respectively. The corresponding proportions of subjects with Z-score >2 were 18.4%, 11.2%, 17.3%, and 49.2%. A progression from normal dimensions (Z-score ≤;2) to significant enlargement (Z-score >2) was observed in a minority of patients (10.6%, 5.6%, 9.5%, and 19.0% of patients, respectively).

Conclusions: The rate of progression of aortic valve dysfunction and aortic root dilatation in paediatric patients with isolated BAV is relatively slow. These findings may be taken into account to better guide risk assessment and clinical follow-up in these patients.