Long term course of bicuspid aortic valve in patients with and without associated cardiac malformations.

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Introduction: Evolution of bicuspid aortic valve (BAV) in patients with associated cardiac malformations remains largely unknown. We compared the course of isolated BAV with that of BAV associated with other cardiac malformations in a large cohort of patients.

Methods: Single-centre, retrospective cohort study. All patients in whom BAV was diagnosed in a large regional referral university hospital between 01/1989 and 12/2015, and who underwent at least 2 echocardiographic examinations in our centre were eligible. Two groups were defined, namely "isolated BAV", and "associated BAV" (e.g. BAV associated with coarctation, other simple or complex cardiac malformations).

Results: 280 patients were included, 61.8% with isolated BAV and 38.2% with associated BAV, who were mainly children. Mean duration of follow-up (FU) was 9 years (range 0-39 years). Patients with associated BAV were younger and had less complications at diagnosis (p<0.0001 for both). About half the population developed a new complication of BAV during FU, with no difference between groups (p=0.845), but patients with associated BAV had less worsening of their initial complication (p<0.033). At the end of FU, 51.1% of the whole population had an aortic dilatation. More than half of the patients underwent cardiac surgery related to BAV in 50% of cases. Patients with associated BAV underwent more cardiac surgery (p<0.0001), mainly for associated cardiac malformations and at a younger age (p<0.0001). Younger age at diagnosis (odds ratio, OR=0.63; 95% CI=[0.5-0.9] per quartile) was significantly associated with the risk of complications during FU, and the presence of a complication at diagnosis (OR=18,15; 95% CI=[4.2-78.9]) was associated with the occurrence of cardiac surgery related to BAV during FU. Associated BAV (OR=0.01; 95% CI=[0.003-0.7]) had a lower risk of surgery related to BAV. Cumulative incidence of moderate to severe aortic stenosis or regurgitation was 23.9% (95% CI= [19-29.9]) and 45.1% (95% CI= [37.9-53.1]) respectively at 20 and 40 years of age, with no difference between both groups.

Conclusion: In our study, associated BAV was not predictive of a higher risk of complications during FU.