Bicuspid aortic valve morphology: Does the pattern of leaflet fusion determine aortic and valvar pathology?

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OBJECTIVE: Congenital bicuspid aortic valve may appear as an isolated lesion or in association with other congenital malformations, hemodynamic derangements, and aortic dilation. The purpose of this study is to determine the association between types of leaflet fusion and valve dysfunction and aortic pathology in patients with bicuspid aortic valve.

METHODS: We performed a retrospective review of 518 patients who presented between 1994 and 2014. Echocardiography was used to diagnose bicuspid aortic valve leaflet fusion and associated abnormalities.

RESULTS: Median age was 13 years and 81% of the patients were aged <18 years. Right and left coronary leaflet fusion was the most prevalent subtype (right/left, n=330, 63.7%; right/non, n=181, 34.9%; left/non, n=7, 1.4%). Moderate or greater aortic stenosis (right/non, n=81, 45% vs. right/left, n=88, 27%, p<0.001) and mild or greater aortic insufficiency (right/non, n=101, 56% vs. right/left, n=88, 27%, p<0.001) were observed more often in patients with right and non-coronary leaflet fusion. Similarly, right and non-coronary leaflet fusion was associated with mild or greater ascending aortic dilation (right/non, n=96, 53% vs. right/left, n=71, 22%, p<0.001). Conversely, the majority of patients with aortic coarctation had fusion of the right and left coronary leaflets (right/left, n=136, 41% vs. right/non, n=41, 23%, p<0.001).

CONCLUSIONS: Our study suggests that aortic valve morphology may be a determinant of AS and AI in pediatric patients with BAV. We found that fusion of the R-N leaflets was associated with twice the risk of AS and AI compared to R-L fusion. Although patients with R-L leaflet fusion had less aortic valve and ascending aortic pathology, they were more likely to have CoA.
The etiological factors that determine the formation of R-N and R-L BAVs may also be involved in the occurrence and progression of the pathologies associated with each subtype. A patient's BAV morphology may influence their need for future interventions as well as the longevity of certain operations such as the Ross procedure.