

Should Bicuspid Aortic Valve Morphology be Incorporated into the Criteria of Shone's Complex?

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Objectives: Shone's complex consists of multilevel left sided obstructions. Bicuspid aortic valve (BAV) is a frequent finding in patients with Shone's complex. The purpose of this study was to determine the prevalence of BAV in patients with Shone's complex and in those patients with BAV, to characterize the sub-type of BAV leaflet fusion.

Methods: We performed a retrospective review of 32 patients who presented with Shone's complex between 1977 and 2014. Patients with multilevel left sided obstructions confirmed by echocardiogram were included. Qualifying lesions we included were mitral stenosis, parachute mitral valve, supra-mitral ring, subaortic stenosis, aortic stenosis, supra-valvular aortic stenosis, and coarctation. BAV was categorized by leaflet fusion into right and non-coronary (R/N) versus right and left leaflet (R/L) fusion.

Results: 15 (47%) of the 32 patients were male. Median age at time of presentation to our institution was 15 days (mean age, 5±21 days; range, 1 day to 10 years). Of the 32 patients, mitral valve abnormalities were present in 30 (94%), aortic abnormalities excluding BAV were present in 30 (94%), coarctation was present in 26 (87%). BAV was present in 30 (94%) of 32 patients. In patients with BAV, R/L fusion was the dominant subtype, present in 29 (97%) of these patients while R/N fusion was present in only 1 (3%) patient. Only 4 (13%) patients with BAV had aortic insufficiency.

Conclusions: R/L fusion BAV appears to be the predominant subtype of BAV in patients with Shone's complex. The etiological factors that determine the formation of R/L BAVs may also be involved in the occurrence and progression of the pathologies associated with Shone's complex. We suggest that bicuspid aortic valve morphology be considered in the criteria for Shone's complex.