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CLINICAL AND ECHOCARDIOGRAPHIC PROFILE OF BICUSPID AORTIC VALVE

Cardiology

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KEYWORDS

INTRODUCTION:

Bicuspid aortic valve (BAV) disease is the most common congenital heart defect, with a prevalence estimated between 0.5% and 2% [1–5]. There is a male predominance of approximately 3:1 ratio. In adulthood, complications are common[6, 7] and therefore, the burden of disease from BAV disease is more significant than any other congenital cardiac lesion. Despite its importance, our understanding of BAV disease is incomplete and questions remain unanswered about this common condition. Most studies documenting valve morphology have been either in small samples of patients[8, 9]. Therefore, we evaluated BAV morphology by two-dimensional echocardiography and assessed the association between aortic valve morphology and degree of valve pathology.

METHODS:

This was a cross-sectional observational study where we enrolled 135 adults (males – 85 and females – 50) aged ≥14 years in compliance with the Helsinki Declaration, where patients gave consent to the inclusion into data analysis. All echocardiographic measurements were performed according to ASE (American Society of Echocardiography) and EAE (European Association of Echocardiography) recommendations. We defined the BAV as having partial or complete fusion of two of the aortic valve leaflets, with or without a central raphe, resulting in partial or complete absence of a functional commissure between the fused leaflets. To assess morphology of aortic valve, short axis view at aortic valve level and parasternal long axis was used. Aortic measurement was done in parasternal long axis focussed view at mid systole for annulus and end systole for sinus, sinotubular junction (STJ) and ascending aorta. Aortic stenosis was considered significant if there was a Doppler velocity of ≥2 m/s across the aortic valve. Classification of the severity of aortic regurgitation was based on combined evaluation of proximal jet width, abdominal aortic Doppler, and pressure half time. BAV was divided into three groups namely fusion of right and left coronary cusp(LCC), fusion of non and right coronary cusp(RCC) and fusion of left and non coronary cusp(NCC). We found the prevalence of type of bicuspid aortic valve and its associated aortic valve disease.

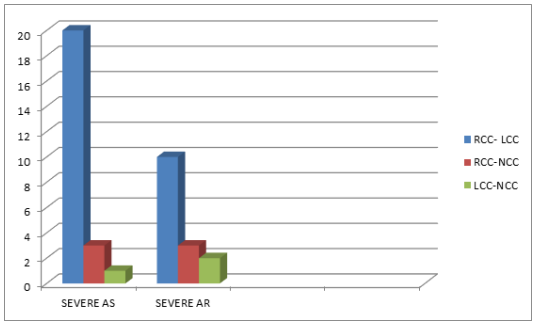
RESULTS:

The mean age of subjects was years and in those with fusion of RCC-LCC was 18.22 years, fusion of RCC-NCC was 22.3 years, fusion of LCC-NCC was 22.3 years. There was male prepondence among all the three groups. Fusion of RCC and LCC was the common type among BAV. There was not much difference of aortic root dimensions among the type of BAV as shown in table 1. Aortic stenosis was the common valvular lesion than regurgitation in all the groups. Majority of the patients with fusion of RCC and LCC had severe aortic stenosis and severe aortic regurgitation. Distribution of severe aortic stenosis and aortic regurgitation is shown graph 1 among various types of BAV disease.

Table: Clinical and echocardiographic parameters among BAV types

	Fusion of RCC-LCC (n-96)	Fusion of RCC-NCC (n-29)	Fusion of LCC-NCC (n-10)
Mean age (years)	18.22	22.3	22.3
Male:Female	64:32	16:13	6:4
Mean annulus size	45mm	46mm	44mm
Mean sinus size	42mm	41mm	42mm
Mean STJ size	39mm	40mm	40mm
Mean AA size	42mm	42mm	41mm
Mild aortic stenosis	21	10	3
Moderate aortic stenosis	20	7	2
Severe aortic stenosis	20	3	1
Mild aortic regurgitation	20	10	3
Moderate aortic regurgitation	18	7	2
Severe aortic regurgitation	10	3	2
Congenital heart disease	1	0	0
Coarctation	2	2	1
Aortic dissection	1	1	0

Graph 1: Distribution of severe aortic stenosis and severe aortic regurgitation among different BAV types



DISCUSSION:

BAV is the most common congenital cardiac malformation present in the general population. Both genetic and environmental causes of the BAV have been suggested, but the pathogenesis remains unclear [8]. The bicuspid valve is typically made of two unequal-sized leaflets. The larger leaflet has a central raphe or ridge that results from fusion of the commissures, and these fused commissures are susceptible to disruption as occurs with balloon valvuloplasty. The morphologic patterns of the bileaflet valve vary according to which commissures have fused, with the most common pattern involving fusion of the right and left cusps. Fusion of the right and left coronary cusps is associated with coarctation of the aorta. Fusion of the right and noncoronary cusps is associated with cuspal pathology. Rarely, the leaflets are symmetrical or there is no raphe ("pure" bicuspid valve). A number of classifications have been used that pertain to the orientation of the

leaflets [9]. BAV is associated with significant complications in one third of patients. Ward [12] suggested that the BAV might be responsible for more deaths and morbidity than the combined effects of all other types of congenital heart defects. Given the significant impact the BAV has on health, it is important to their profile in population. Previously published studies have shown that BAV is a common finding in isolation, identified in 0.5% to 2% of the population and has been observed in conjunction with many other types of congenital heart lesions [4,13]. In addition, there is a well-documented association of BAV with aortic coarctation[14]. Our study found that BAV was occasionally present in many congenital heart disease.The majority of previously published reports have assumed that all types of BAV are similar and have not differentiated between type of leaflet fusion when assessing valve dysfunction and progression [15, 16]. Our study demonstrates that aortic valve morphology is an important determinant of the risk for aortic stenosis and aortic regurgitation, at least inadults. We found that patients with BAV with fusion of the right-coronary and left coronary leaflets had more risk of aortic stenosis and regurgitation compared with other types of BAV.

CONCLUSION:

Our study clearly demonstrates that BAV occurs with associated with aortic valve disease. In addition, BAV morphology is highly correlated with type and severity of valve dysfunction. These findings are helpful in predicting which patients are likely to have a benign cardiac course and which are at greater risk for progressive aortic valve disease.

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