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Research Article

Bicuspid Aortic Valve Leaflet Morphology in Patients with and without Coarctation of the Aorta

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• Bicuspid aortic valve; Coarctation of aorta; Aortic stenosis; Aortic insufficiency

Abstract

Objective: Bicuspid aortic valve (BAV) is the most common congenital heart defect. BAV is often seen with other left-sided structural abnormalities, such as coarctation of the aorta. Furthermore, diagnosing coarctation of the aorta can sometimes be challenging with transthoracic echocardiography. This study aimed to assess if the presence of coarctation of the aorta is associated with a specific BAV morphology.

Methods and Materials: All patients referred to Ghaem Mashhad Hospital for echocardiography were included in the study if they had a BAV. The morphology of the aortic valve was classified based on the fusion of the cusps (anteroposterior opening and mediolateral opening), and the existence of coarctation of aorta was determined based on the findings in echocardiography or the CT angiography. Then, the data were compared in two groups.

Results: Out of 134 patients (104 men and 30 women), 81 had anteroposterior morphology, and 53 had mediolateral morphology. Among the patients with anteroposterior morphology, 43 patients (69.4%) had coarctation of aorta, and 38 patients (52.8%) had no coarctation of aorta. Among patients with mediolateral morphology, 19 (30.6%) were diagnosed with coarctation of the aorta, while 34 (47.2%) showed no evidence of coarctation of the aorta ($p = 0.049$).

Conclusion: The present study showed that the morphology of the aortic valve contribute to predicting the presence of coarctation of the aorta.

INTRODUCTION

Bicuspid aortic valve (BAV) is a congenital heart defect affecting around 1-2% of the population. This condition is linked to a higher likelihood of developing issues related to the aortic valve in the future [1,2]. Approximately 95% of BAVs include a third rudimentary commissure, commonly known as a raphe, which can vary in height. Functionally, considerable aortic stenosis often occurs between the ages of 50 and 60 years, whereas regurgitation tends to develop around the age of 30 years [3,4]. Crucially, BAV is not limited to a singular disease entity. Instead, it entails several disorders that differ in genetic and molecular aspects, resulting in a diverse range of alterations in the development of the heart and aorta [5,6].

Coarctation of the aorta (CoA) is well-acknowledged to occur concurrently in many of these patients. Before the development of repair techniques, aortic dissection accounted for 19% of deaths among individuals with coarctation of the aorta. This risk was notably higher in those with a bicuspid aortic valve (BAV) [5]. The prevalence of aortic coarctation of the aorta in patients with BAV varies between 22% and 36%, depending on the age groups investigated. Nevertheless, the incidence of aortopathy in patients with BAV who also have a coarctation of aorta (cBAV) is currently unknown [6-8].

The present study aimed to compare the bicuspid aortic morphology in patients with and without the coarctation of the aorta. Because some patients have a poor view and diagnosis of the coarctation in inexperience people are so hard so we want to study if the morphology of the aorta was helpful to possible prediction of the presence of coarctation or not.

METHODS AND MATERIALS

Study design

Patients over 17 years old who had been referred to Ghaem Hospital for echocardiography during two years and who had bicuspid aortic valve were included in the study. According to the latest guidelines, the echocardiographic studies were conducted with patients in the left lateral position using Siemens and Philips devices and a 2.5-4 MHz probe. Aortic morphology was observed during systole, and in the case of a fusion between left coronary and right coronary cusps, the patient was labeled as anteroposterior

fusion, and in the case of fusion between right coronary and non-coronary cusps or left coronary and non-coronary cusps, the patient was labeled as mediolateral fusion.

Before do the echocardiography whole physical exam was done for all patients including; synchronous pulses of radial and femoral artery and rediofemoral delay was assessed also systolic blood pressure of upper limb and lower limb was measured. For the purpose of assessing the coarctation of the aorta, the patients were examined from the suprasternal view. The presence of the coarctation of the aorta was defined as a peak aortic gradient greater than 20 mmHg; in case necessary, other modalities such as CT angiography were used. In cases that have typical finding of coarctation in echocardiography (such as peak gradient more than 20 mmHg and other typical findings) we confirm our data with CT angiography. Patients with a previous history of surgery due to coarctation of the aorta or stent implantation who had no aortic valve replacement were not excluded from the study since a surgery or stent implantation would not change the artic morphology.

Statistical analyses

The continuous variables exhibited a non-parametric distribution and were therefore presented as median and interquartile range (IQR), whereas categorical data were provided as frequency and percentage. The Mann-Whitney U test was used to compare groups based on continuous data, whereas Fischer's exact tests were used for categorical variables. Post hoc analyses were conducted using multiple Fischer's exact tests with Bonferroni correction. A significance level of $p<0.05$ was deemed statistically significant. The statistical analyses were conducted using SPSS V.25.0. The data registry of the echocardiography department at Mashhad University of Medical Sciences was utilized for data collection

Ethics

The Ethics Committee of Mashhad University of Medical Sciences approved the present study (IR.MUMS.fm.REC.1395.262).

RESULTS

In the examined samples, 104 patients (77.6%), were men, and 30 patients (22.4%), were women. Fifteen (11.2%) were smokers, and 7 (5.2%) were diabetic; fourteen (10.4%), had hyperlipidemia, and seven patients (5.2%) had ischemic heart disease.

Comparing the two morphological groups regarding gender, 58.7% of men had an anteroposterior valve, and 41.3% had a mediolateral valve (p-value: 0.42). 66.7% of women had an anteroposterior valve, and 33.3% had a mediolateral valve. Also, the comparison of two groups with and without coarctation of the aorta in terms of sex showed that 44.2% of men had coarctation of the aorta Furthermore, 53.3% of women had coarctation of aorta (p-value: 0.378) (Table 1).

Out of 81 patients with anteroposterior morphology (60.4% of the total), 43 patients (53.0%) had coarctation of aorta. In the mediolateral morphology group with 53 patients (39.6% of the total), 19 (35.8%) had coarctation of aorta. There was a significant difference between the two morphology groups regarding the coarctation of the aorta (p-value: 0.049) (Table 2).

In the other word, of 62 patients with coarctation of the aorta, 69.4% had an anteroposterior valve, and 30.6% had a mediolateral valve. In the group of patients without coarctation of the aorta, 52.8% had an anteroposterior valve, and 47.2% had a mediolateral valve (Table 2).

Among patients with anteroposterior morphology, 12.5% had no aortic regurgitation, 37.5% had mild regurgitation, 32.5% had moderate regurgitation, and 17.5% had severe regurgitation. The corresponding proportions in the mediolateral morphology group were 7.5%, 35.8%, 35.8%, and 20.8%, respectively. There were no significant differences between the groups in the frequency of aortic regurgitation (p-value: 0.789) (Table 2).

In the anteroposterior group, 38.6% had no aortic stenosis, 6.8% had mild aortic stenosis, 6.8% had moderate

Table 1: The association between sex and aortic morphology and presence of coarctation

		Male	Female	p-value
Morphology	Anteroposterior	61 (58.7)	20 (66.7)	0.429
	Mediolateral	43 (41.3)	10 (33.3)	
Coarctation of aorta	Yes	46 (44.2)	16 (53.3)	0.378
	No	58 (55.8)	14 (46.7)	

Table 2: The association between aortic morphology and aortic regurgitation, stenosis, and dilation

		Anteroposterior	Mediolateral	p-value
Coarctation	Yes	43 (53%)	19 (35.8%)	0.049
	No	38 (46.91%)	34 (64.1%)	
Aortic regurgitation	None	10 (12.3%)	4 (7.5)	0.789
	Mild	31(38.2%)	19 (35.8)	
	Moderate	26 (32%)	19 (35.8)	
	Severe	14 (17.2%)	11 (20.7)	
Aortic stenosis	None	51 (62.9%)	29 (54.7%)	0.231
	Mild	11(13.5%)	11 (20.7%)	
	Moderate	9 (11.1%)	3 (5.6%)	
	Severe	10 (12.3%)	10 (18.8%)	
Aortic dilation	Yes	39(48.1%)	25 (52.8)	0.886
	No	42 (51.8%)	28 (52.8)	

aortic stenosis, and 7.6% had severe aortic stenosis. In the group with mediolateral morphology, 22% had no aortic stenosis, 8.3% had mild aortic stenosis, 2.3% had moderate aortic stenosis, and 7.6% had severe aortic stenosis. No significant differences were observed in the frequency of aortic stenosis between the groups (p-value: 0.231). About 47.4% of patients with anteroposterior valve had dilated aorta, and 52.6% of them did not have dilated aorta. 46.2% of patients with mediolateral aortic valves had dilated aorta (p-value: 0.886) (Table 2).

DISCUSSION

Although the connection between BAV and CoA is well-known, the previous repair of CoA has typically had little influence on treating the bicuspid valve. The significance of the simultaneous existence of the two criteria is not fully understood. The results of the present study showed that the frequency of coarctation of the aorta was significantly different between the morphology groups. However, no significant differences were observed between the groups regarding aortic regurgitation, stenosis, and aortic dilation. On the other hand, the results of the present study showed a significant difference between the presence of increased blood pressure among those with and without coarctation of the aorta.

Data shows that non-valvular cardiovascular anomalies linked to BAV, including coarctation of aorta, coronary anomalies, sinus of Valsalva aneurysm, aortic aneurysm, aortic dissection, supra-valvular aortic stenosis, patent ductus arteriosus, ventricular septal defect, Shone complex, familial aneurysm syndrome, thoracic aortic dissection, and Turner syndrome [9]. The present study found coarctation of aorta, aortic regurgitation, stenosis, and dilation as the complications observed in patients with BAV. Previous studies have shown aortic stenosis to be more common among the AP BAV subtype [10,11]. Although the higher number of aortic stenosis cases had an AP BAV phenotype in this study, the difference was not significant.

The present study showed that the anteroposterior and mediolateral phenotypes of BAV constituted 60% and 40% of the subtypes, respectively. Most previous studies have shown the dominance of the AP subtype, except for a few studies [12]. Furthermore, it was shown that these subtypes are not significantly associated with gender, a finding supported by a previous study [13]. Furthermore, this study showed a significant association between the AP and ML morphology types and the presence of coarctation of the aorta among BAV patients. A study in 2018 showed similar results; Tabrizi et al., studied 300 adults with BAV to evaluate the association between valvular dysfunction and aortopathies and BAV subtypes [10]. Their findings showed that the number of those with an AP morphology was significantly higher compared to ML among those with coarctation of the aorta, consistent with the present study's findings.

Nevertheless, the mechanism by which aortic aneurysms develop in these individuals remains uncertain. The correlation between aortopathies and various manifestations of BAV can be ascribed to either genetic factors [14,15], or biomechanical variables that result in an increase in asymmetrical shear stress on the aortic wall, which is caused by eccentric turbulent flow through the BAV. This pathological flow can elucidate the varying segmental dilatation patterns of the aorta resulting from different BAV phenotypes [16-21]. In conditions such as AP BAV and ML BAV, the atypical flow of blood is caused by the differing positions of the valve cusps, resulting in the flow being directed towards the right-anterior and right-posterior walls of the aorta, respectively [10].

The present study had some limitations. Given the rarity of coarctation of aorta, the present study found a limited number of events, necessitating the results to be interpreted cautiously. Furthermore, the size of the ascending aorta is dependent of age, which could impact the results in a small sample size. Also, the individuals included in our study were patients who were receiving medical attention at a specialized facility. As a result, the findings of our study may not apply to a broader community.

In conclusion, the results of the present study showed that coarctation of the aorta was more frequently observed among those with AP BAV phenotype. Furthermore, increased blood pressure was more commonly observed among those with coarctation of aorta.

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