

The Frequency of Bicuspid Aortic Valve in Healthy Young Adults Who Applied to the Health Board: A Tertiary Centre Experience: Retrospective Study

Sağlık Kuruluna Başvuran Sağlıklı Genç Erişkinlerde Biküspit Aort Kapak Sıklığı: Üçüncül Merkez Deneyimi: Retrospektif Çalışma

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ABSTRACT Objective: Bicuspid aortic valve (BAV), which is the one of the most common congenital heart malformation and seen approximately 1-2% of the general population. It has been shown that BAV is associated aortic valve dysfunction (aortic stenosis, aortic regurgitation or both) and increased risk for bacterial endocarditis from adult to the advanced ages. In this study, we aimed to determine the frequency of BAV disease in the asymptomatic young population who is applied to the health board. **Material and Methods:** 5,490 subjects who were consulted to the cardiology outpatient by health board were systematically analyzed from our hospital database system and 15 patients with BAV were identified. The frequency of BAV and aortic valve measurements were recorded. Patients with previous aortic valve intervention were excluded from the study. **Results:** Fifteen patients with BAV were identified (0.3%), all patients were newly diagnosed during screening at the health board. Mean age of the BAV subjects was 23.4±2.45 years. Two of them were female (13.3%) and thirteen of them (86.7%) were male. Aortic insufficiency was the most common pathology, mild in 11 (73.3%), and moderate in 2 (13.3%) cases. Only 2 (13.3%) patients had aortic dilatation. Aortic stenosis was not detected. **Conclusion:** Prevalence of BAV in asymptomatic young adults was lower than the literature.

Keywords: Bicuspid aortic valve disease; aortic valve stenosis; aortic valve insufficiency

ÖZET Amaç: Biküspit aort kapak (BAK) en sık görülen konjenital kalp hastalıklarından biridir ve genel popülasyonda %1-2 oranında görülür. BAK'ın yetişkinlikten ilerleyen yaşlara doğru aort kapak disfonksiyonu (aort stenozu, aort yetersizliği veya her ikisi) ve artmış infektif endokardit riski ile birlikte olduğu gösterilmiştir. Bu çalışmada, sağlık kuruluna başvuran asemptomatik genç erişkinlerde BAK sıklığını araştırmayı amaçladık. **Gereç ve Yöntemler:** Kardiyoloji polikliniğine sağlık kurulu muayenesi amacı ile başvuran 5.490 hastanın verisi tarandı ve 15 BAK hastası tespit edildi. BAK sıklığı ve aort kapak ölçümleri kaydedildi. Daha önceden aort kapak müdahalesi geçiren hastalar çalışmadan dışlandı. **Bulgular:** Sağlık kuruluna başvuran hastaların 15'inde BAK (%0,3) saptandı. Bu hastaların hepsi yeni teşhis edilen hastalardı. Hastaların ortalama yaşı 23,4±2,45 yıl idi. Hastaların 2'si (%13,3) kadın, 13'ü (%86,7) erkekti. En sık patoloji aort yetersizliği idi ve 11 (%73,3) hastada hafif, 2 (%13,3) hastada orta derecede saptandı. İki (%13,3) hastada aort genişlemesi saptandı. Aort darlığı hiçbir hastada saptanmadı. **Sonuç:** BAK prevalansı asemptomatik genç kişilerde literatürde belirtilen orandan daha düşük bulunmuştur.

Anahtar Kelimeler: Biküspit aort kapak hastalığı; aort kapak stenozu; aort kapağı yetersizliği

Bicuspid aortic valve (BAV) has a prevalence of 1-2% in general population and one of the commonest congenital malformation of the heart.^{1,2} Aortic valve dysfunction usually accompanies BAV and it has an increased risk of infective endocarditis especially in older patients.^{2,3} The tissue abnormality is

not limited to the valve leaflets, increased wall stress on the ascending aortic wall due to the cusp fusion abnormality of the valve, causing the development of aortopathy: including dilatation of the aorta and aneurysm formation, dissection, and even sudden death.^{4,5} Despite high incidence of aortopathy, aortic

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dissection is a rare complication for all patients with BAV even though it occurs 5-9 times higher than in the general population.⁶

The diagnosis of BAV can be made in any period of life, from newborns to the elderly, and may present with different clinical findings.^{7,8} BAV might be an incidental finding at physical or echocardiographic examination or it can be presented as severe aortic regurgitation (AR) or aortic stenosis (AS) leading to heart failure and death.^{8,9} Bacterial endocarditis and acute aortic dissection are among the life-threatening complications of BAV.^{6,8-10}

These complications may present at any time in life and may be the first clinical presentation of a known healthy person with unidentified bicuspid status.^{6,10} Therefore, early diagnosis and, if appropriate, early intervention are very important in preventing these complications.

In our country young male and female adults who apply military, police and guard duty undergo comprehensive physical assessment in order to assess their suitability for military, police and guard duty. Therefore, we aimed to determine the frequency of BAV disease in the asymptomatic young population who is applied to the health board.

MATERIAL AND METHODS

Bakırköy Dr. Sadi Konuk Training and Research Hospital is one of the centers for examination of the military and candidate for police department recruits. All candidates were evaluated by echocardiography in the outpatient clinic if they were eligible to recruit because even mild valvular disease is accepted reason for rejection by the relevant institution according to the Turkish Armed Forces Health Regulations and General Directorate of Security Health Regulation. We conducted a cross-sectional study in between April 2013 to January 2018 among asymptomatic population who passed the physical competency test by the relevant institution. 5,490 healthy individuals were included in this study. Bakırköy Dr. Sadi Konuk Training and Research Hospital Clinical Research Ethics Committee was approved the study (date: July 8, 2019, no: 13/2019) and the study was conducted in accordance with the declaration of Helsinki.

Two-dimensional transthoracic echocardiography (TTE) was performed in all patients according to current guidelines.¹¹ Anatomy of the aortic valve, hemodynamic evaluation including peak and mean velocities, mean and maximum pressure gradients were assessed. Left ventricular volume and functional evaluation were also by measuring and left ventricular end-diastolic and end-systolic diameters and left ventricular ejection fraction. Presence and degree of AR or AS were also evaluated. Patients with previous aortic valve interventions were excluded.

Evaluation of aortic valve anatomy was made in the parasternal long- and short-axis views: a BAV defined as the presence of two cusps which were identified in systole and diastole, or both in short-axis view. In addition, eccentric closure, systolic doming or diastolic prolapse of the aortic cusps in parasternal long axis imaging were evaluated as supportive findings. If the diagnosis was not conclusive, transesophageal echocardiography was performed in order to confirm the definitive diagnosis. Based on number of raphe, BAV was categorized as Type 0 (no raphe), Type 1 (only one raphe) and Type 2 (two raphe). On the basis of raphe position with coronary sinuses types 1 and 2 were classified as left (L), right (R) and (N) type.¹² Measurements of aortic annulus, sinus valsalva, sinotubular junction and proximal ascending aorta was made in the parasternal long-axis view perpendicular to aortic long axis.^{13,14} All the measurements were taken three times and the mean of the measurements was used.

Severity of AR was classified into mild, moderate, or severe according to color and continuous flow Doppler criteria. For severity assessment, width of the vena contracta, ratio of regurgitant jet height to left ventricular outflow tract (LVOT) height, deceleration rate of regurgitation jet (pressure half-time), and the presence of holodiastolic retrograde flow in the descending aorta were evaluated. If the ratio of regurgitant jet to LVOT height >65%, pressure half time below 250 msec, and holodiastolic retrograde flow in the descending aorta were present than this regurgitation was considered as severe.¹⁵ Ascendant aortic diameter of 20 mm to 37 mm was considered as normal and the values of greater than thirty-seven

millimeters were considered as ascending aortic dilatation.

STATISTICAL ANALYSIS

The demographic characteristics of the subjects and the collected data were entered in SPSS version 23 (IBM SPSS Statistics for Windows, Version 23.0. Armonk, NY: IBM. Corp). Qualitative variables were characterized using mean and percentage values. Categorical variables were expressed using frequency and percentage, and numerical variables were expressed using mean±standard deviation.

RESULTS

From April 2013 to January 2018, 5,490 young adults were selected for referral to the cardiology outpatient clinic for 2-dimensional echocardiography and medical evaluation from health board. Fifteen patients with BAV were identified (0.3%), all of were newly diagnosed during screening and had Type 1 BAV (RL type BAV). Mean age of the BAV subjects was 23.4±2.45 years. No patient had diabetes mellitus, hypertension and coronary artery disease. Two of them were female (13.3%) and thirteen (86.7%) were male. AR insufficiency was the most commonly hemodynamic impairment in 13 cases (86.7%), mild in 11 (73.3%), and moderate in 2 (13.3%). Only 2 (13.3%) patients had aortic dilatation. AS was not detected. BAV was functioning normally in 2 patients (13.3%). However, the aortic dimensions of BAV patients with mild to moderate aortic regurgitation were not greater than those of patients without regurgitation. Mean aortic diameters in BAV patients were 26.79±3.40 mm at the root of aorta, 33.67±5.91 mm at the mid-ascending aorta. Table 1 shows clinical characteristics of the patients. The patients did not have any co-existing valve pathology such as mitral regurgitation, mitral stenosis or tricuspid regurgitation. Medical treatment was given to the patients who had moderate aortic regurgitation or dilatation of the aorta.

DISCUSSION

In this study, we found the frequency of BAV in 5,490 young adults to be 0.3%. Among them, mild AR was detected in 11 patients and moderate AR

TABLE 1: Clinical characteristics of the patients with bicuspid aortic valve (n=15).

Parameter	
Age (years)	23.4±2.45
Gender (n, %)	
Female	2 (13.3)
Male	13 (86.7)
Aortic insufficiency (n, %)	13 (86.7)
Mild	11 (73.3)
Moderate	2 (13.3)
Aortic stenosis (n, %)	0 (0)
Aortic root diameter (mm)	26.79±3.40
Ascending aorta diameter (nm)	33.67±5.91

was in 2 patients, ascending aortic dilatation was detected in only 2 patients. The prevalence of BAVs is 0.5%-2% in general population, and men are affected 3 times more than women. This disease may be associated with other cardiac and aortic abnormalities as a result of molecular and connective tissue irregularities rather than merely anatomical variation.^{16,17}

The frequency of BAV found in this study may not reflect the accurate prevalence of BAV.¹⁸ This underestimation might be the result of exclusion of patients who had undergone previous surgery or percutaneous intervention. Furthermore the subjects included in the study were composed of healthy young adults without any cardiac complaints. We thought the sample represents the population that totally asymptomatic and well-conditioned population in the same age range.

TTE has a far more better sensitivity in diagnosing BAV compared to physical examination (92% vs 50%, respectively).^{19,20} It is usually the first-choice imaging method in the assessment of aortic valvular function and aortic dimensions. BAV has diversity of clinical presentation from asymptomatic patients with relatively normal valve function to symptomatic patients with severe AS and/or AR. It has been shown that almost 30% of the asymptomatic patients will eventually undergo valve intervention within 20 years.⁸ BAV is frequently accompanied by ascending aortic aneurysm with an estimated prevalence of 15%-45%.^{9,21,22}

However, aortic dissection risk in these patients is found to be relatively low (1%-6%).^{9,20} In the present study all of BAV subjects were asymptomatic.

Fusion of right and left coronary cusps and right and non-coronary cusps are the first and second most common causes of BAV with an estimated prevalence of 59% and 37%, respectively.^{21,23} Fusion of left and non-coronary cusps is the least common form of the disease.²³ In this study, R-L type fusion was found in all young adults with BAV in accordance with the literature.

All of BAVs were newly diagnosed during military, policing evaluation. Since BAV is associated with an increased risk of infective endocarditis, diagnosis of BAV has utmost importance especially in young adults.² Thus, detection of BAV may prevent this ominous complication. Although antibiotic prophylaxis of intermediate risk patients is not recommended in the current guidelines, the importance of dental and cutaneous hygiene is strongly emphasized in patients native valvular disease and BAV.^{24,25} Also, because BAV is an inherited valvular disease, it is recommended that first-degree relatives of newly diagnosed patients should be evaluated for the presence of BAV.²⁶

Most of our patients had mild AR. This finding was consistent with the demonstration that BAV may be an important cause of pure aortic regurgitation, indicating a higher prevalence of regurgitation in younger subjects, in conjunction with the previously reported age dependence of the type of valve complications of BAV.^{27,28}

There were some notable limitations in our study. Firstly, our study is a single-center retrospective cross-sectional study. Apart from the aortic root and ascending aorta diameter, no data on the anatomical features of the aorta were included in the study. Also, subjects do not have long-term follow-up.

CONCLUSION

We found that the prevalence of BAV was lower than the literature and all of subjects with BAV were asymptomatic and healthy. Early BAV detection is important because BAV is related important cardiovascular outcomes. Thus, cautions approach to the patient with a normally functioning bicuspid valve is to educate the patient about the long-term prognosis, emphasize dental hygiene and endocarditis prophylaxis, and follow valve function with periodic echocardiography.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Ersan Oflar, Dilay Karabulut, Abdulcelil Sait Ertuğrul, Alparslan Şahin; **Design:** Ersan Oflar, Dilay Karabulut, Fatma Nihan Turhan Çağlar, Alparslan Şahin; **Control/Supervision:** Ersan Oflar, Dilay Karabulut, Abdulcelil Sait Ertuğrul, Cennet Yıldız, İbrahim Faruk Aktürk; **Data Collection and/or Processing:** Ersan Oflar, Dilay Karabulut, Abdulcelil Sait Ertuğrul; **Analysis and/or Interpretation:** Dilay Karabulut, Cennet Yıldız; **Literature Review:** Ersan Oflar, Dilay Karabulut, Abdulcelil Sait Ertuğrul, Cennet Yıldız, İbrahim Faruk Aktürk, Fatma Nihan Turhan Çağlar, Alparslan Şahin; **Writing the Article:** Dilay Karabulut, Abdulcelil Sait Ertuğrul, Cennet Yıldız; **Critical Review:** İbrahim Faruk Aktürk, Fatma Nihan Turhan Çağlar, Alparslan Şahin; **References and Fundings:** İbrahim Faruk Aktürk, Fatma Nihan Turhan Çağlar.

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