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Thoracic Aorta Evaluation in Children with a Bicuspid Aortic Valve: Contribution of Magnetic Resonance Angiography

Biküspit Aort Kapağı Olan Çocuklarda Torasik Aort Değerlendirmesi: Manyetik Rezonans Anjiyografinin Katkısı

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ÖZET

Amaç: Biküspit aort kapağı (BAK) erken kapak hasarına ve aort genişlemesine neden olan en yaygın doğumsal kapak anomalisidir. Bu çalışmada BAK'lı pediatrik hastalarda ekokardiyografiye (EK) ek olarak aort görüntülemede kullanılan manyetik rezonans anjiografinin (MRA) katkısı araştırıldı.
Yöntemler: Tek merkezli bir çalışmada, 18 yaşın altında BAK tanısı alan 571 olgunun tıbbi kayıtları geriye dönük olarak incelendi. Bu grupta aort değerlendirmesi için MRA yapılan 32 hasta (21 kız, ortalama yaş 12.66 ± 3.92 yıl) çalışmaya dahil edildi. Faz kontrast yöntemi ile elde edilen MRA'da, anulus, çıkan aort, sinüs valsalva, sinotübüler bileşke, orta çıkan ark, orta ark ve proksimal ark düzeylerinede en büyük çaplar elektronik olarak ölçüldü. EK incelemesi sırasında elde edilen ve kaydedilen vücut yüzey alanı ve arteriyel kan basıncı değerleri elde edildi. Z skorları hesaplandı. EK ile saptanan ejeksiyon fraksiyonu, aort darlığı (AD) ve yetersizlik (AY) kaydedildi. Aortik ark geometrisi ve supraaortik varyasyonlar MRA ile belirlendi. EK ve MRA bulguları, MRA tarafından sağlanan ek bulgular değerlendirildi.
Bulgular: EK'de, katılımcıların% 62,5'inde AY ve% 6,25'inde kombine kapak bozukluğu saptanmıştı. İzole AD saptanmadı, katılımcıların% 31,25'inde kapak patolojisi yoktu. Kapak disfonksiyonunda en önemli çap değişikliği orta ark seviyesinde tespit edildi. En yaygın aortik ark Romanesk özellikteydi (% 53). MRA'da, katılımcıların% 15,5'inde bovin ark varyasyonu ve% 3'ünde izole sol vertebral arter vardı. Kapak bozukluğu, z skorları, arteriyel kan basıncı, vücut yüzey alanı, aort geometrisi, supraaortik varyasyonlar arasında bağlantı bulunmadı.
Sonuç: BAK'lı çocuklarda EK bulgularına ek olarak, MRA, hemodinamik değişiklikleri etkilediği bilinen aortik ark geometrisi ve supraaortik varyasyonları göstermek için yararlıdır.

Anahtar Kelimeler: Bikusbit aorta kapağı, Aorta, Çocuk, Ekokardiografi, MRG

ABSTRACT

Aim: Bicuspid aortic valve (BAV) is the most common congenital valve anomaly causing early valve damage and aortic enlargement. In this study, the contribution of magnetic resonanceangiography (MRA) used for aorta imaging in addition to echocardiography (EC) in pediatric patients with BAV was investigated.
Methods: In a single center study, the medical records of 571 cases with a diagnosis of BAV under 18 years of age were retrospectively reviewed. In this group, 32 patients (21 female, mean age was 12.66 ± 3.92 years) who underwent MRA for aortic evaluation were included in the study. In MRA obtained by phase contrast method, the largest diameters of the annulus, ascending aorta, sinus valsalva, sinotubular junction, middle ascending arch, middle arch and proximal arch levels were measured electronically. Body surface area and arterial blood pressure values obtained and recorded during EC examination were obtained. Z scores were calculated. Ejection fraction, aortic stenosis (AS) and insufficiency (AI) founded with EC were recorded. Aortic arch geometry and supraaortic variations were determined with MRA. The EC and MRA findings, additional findings provided by MRA were evaluated.
Results: In EC, 62.5% of the participants had AI and 6.25% had combined valve disorder. There was no isolated AS, 31.25% of the participants had no valve pathology. The most important diameter change in valve dysfunction was detected at the level of the middle arch. The most common aortic arch was Romanesque (53%). In the MRA, 15.5% of the participants had bovine arch variation and 3% had isolated left vertebral artery. No correlation was found between valvular disorder, z scores measured by MRA, arterial blood pressure, BSA, aortic geometry, supraaortic variations.
Conclusion: In addition to EC findings in children with BAV, MRA is useful for demonstrating aortic arch geometry and supraaortic variations known to affect hemodynamic changes.

Key words: Bicuspid aort valve, Aorta, Children, Echocardiography, MRI

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INTRODUCTION

Bicuspid aortic valve (BAV) is the most common congenital cardiovascular malformation that can be detected in approximately 0.5-2% of the population. Patients with BAV are at risk for valvular and vascular complications. Infective endocarditis, severe aortic stenosis (AS) and aortic insufficiency (AI) can be considered as serious complications (1). The changed elastic properties of the aortic valve may cause valve dysfunction in patients with BAV (2). Aortic pathologies associated with BAV include enlargement of the ascending aorta, aneurysm formation, and dissection (3,4). Therefore, the aortic valve functions of each patient with BAV should be evaluated (5).

Echocardiography (EC) is the standard diagnostic tool to define the morphological features of the BAV and to evaluate complications of the aortic valve and thoracic aorta. Real-time 3D echocardiography can facilitate the anatomical evaluation of the valve. However, it is less useful for evaluation in aortic stenosis (6). ECG-gated Multislice Computed Tomography (MSCT) can demonstrate aortic anatomy by providing high spatial resolution anatomical images of the aortic valve and ascending aorta. It should be kept in mind that ECG-gated MSCT is performed with iodinated contrast media, requiring significant ionizing radiation even when ECG pulse is used to reduce radiation. however, the information obtained does not provide functional information about the severity of the valve disease (5).

Magnetic resonance imaging (MRI) is useful in measuring aortic valve structure, left ventricular volume, the severity of AS and AI, and in the evaluation of the thoracic aorta. The magnetic resonance angiography (MRA) obtained by phase contrast method show highly correlation with Doppler EC findings. Cardiovascular MRI with delayed contrast-enhanced can be used to identify myocardial pathology in patients with severe AS (7).

The purpose of this study is to describe the diameter changes and aortic-supraaortic variations in the thoracic aorta by MRA in children and adolescents who have had BAV by EC.

METHODS

After obtaining permission from the local ethics committee (16.04.2021/3207/130), the medical records of children under age of 18 who were examined in our hospital between January 2013 and January 2021, who had diagnosed with BAV by EC, were retrospectively analyzed. The EC reports of all patients with BAV were obtained from the hospital information system and re-examined by the pediatric cardiologist and pediatric radiologist.

Body weights of all participants were recorded in kilograms and height in cm. With this information, the body surface area (BSA) was calculated using the formula BSA = $0.024265 \times W^{0.5378} \times H^{0.3964}$ (8). This index is required for the interpretation of aorta diameter and left ventricular EF in the growing child. Also, z scores of aortic diameters (in annulus, sinus valsalva, sinotubular junction and middle arch of aorta levels) were calculated with the help of this index (9). The mean systolic and diastolic blood pressure values of the participants obtained during the EC examination taken from the hospital records were recorded.

All individuals underwent comprehensive transthoracic echocardiography using the Philips Affinity 70C ultrasound scanner fitted with S5-1 and S8-3 transducers. Standard echocardiographic evaluations had performed with the participants supine using a conventional M-mode, pulsed-wave Doppler sequence, and the standard imaging techniques recommended by the American Society of Echocardiography guidelines (10). The diameter of the aortic annulus, the maximum diameter of the Valsalva sinus (a representative measure of the anterior and posterior sinus state), the sinotubular ridge and the aortic diameter rising> 1 cm from the sinotubular junction in the parasternal long axis plane had measured during systole, hence with the maximal opening of the aortic valve leaflets. The aortic data had evaluated as recommended by the 2014 European Society of Cardiology guidelines (11).

In patients with aortic z-score greater than 2 in EC and aortic valve dysfunction in EC, MRA was performed to view the aorta. These selected patients examined using 1.5T MRI (Avanto, Siemens AG Healthcare Sector, Erlangen, Germany). In the MRA obtained by phase contrast method examination, the largest diameters of the aorta emerging in the annulus, sinus valsalva, sinotubular junction, middle ascendan arch, middle arch and proximal arch sections were measured by a pediatric radiologist electronically in the Picture Archiving and Communication System (PACS, GE Health Sol. Inc. USA) and recorded in millimeters. Ascending aortic diameter measurements made in EC were recorded by repeating from the same areas on MRA. The Z scores were calculated by MRA measurements (Fig.1). Ascending and aortic arch diameters were measured in the axial plane from unprocessed images of the MRA. The MRA examinations that were not obtained in the appropriate protocol or could not be evaluated due to artefacts were not included in the study. The z scores of these measurements were calculated and recorded. Kappa test was performed for the measurements of ascending and aortic aorta diameters, which were made by EC and MRA, to compare the compatibility of the two imaging methods. The variations of the supraaortic aortic branches detected in the MRA were recorded.

The left ventricular ejection fractions (EF) calculated on the EC reports of these participants were recorded. AS, AI and

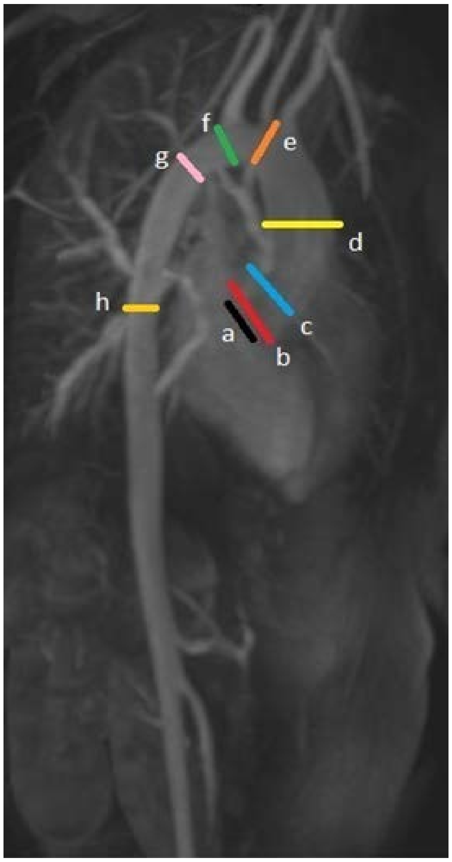

Figure 1. The diameter measurement points in the reconstructed MRA image in the sagittal plane are as follows: a) Annulus (b) Sinus valsalva c) Sinotubular junction d) middle assending aorta: The middle of the distance between the sinotubular junction and the brachiocephalic artery in the ascending aorta e) Proximal arch: Between the brachiocephalic artery and the left common carotid artery origin f) Middle arch: Distal part of the origin of the left subclavian artery. g) Proximal descending aorta: A point 2 cm distal from the left subclavian artery origin. h) Middle descending aorta: In the descending aorta, from the point corresponding to the aortic annulus level.

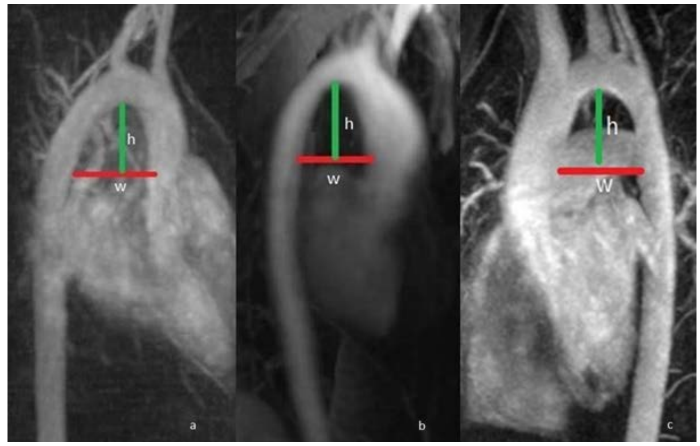

Figure 2. In MRA, the length of the line drawn from the sinus valsalva to the proximal descending aorta was determined as the width of the arch (w). The length of the line drawn perpendicular to the width line from the highest point of the arc was accepted as height (h).
(a) Type 1 arch: Round (if the height of the arch is equal to the width of the arch), (b) Type 2 arch: Gothic-like (if the height of the arch is greater than its width), (c) Type 3 arch: Romanesque-like (if the height of the arch is less in the width of the arch). The isolated left vertebral artery is observed in this patient.

combined valvular dysfunction defined in EC were recorded.

The aortic arches were divided into three types by measuring the height and width of the aortic arch from the MRA sagittal plane view. The height of the arch was measured from the highest point of the arch, the width of the arch was measured from the anteroposterior line drawn from the sinus valsalva to the porximal arch. According to this: Type 1 archus: Round (if the height of the arch is equal to the width of the arch), Type 2 archus: Gothic-like (if the height of the arch is greater than its width), Type 3 archus: Romanesque-like (if the height of the arch is less in the width of the arch) (Fig. 2). In this study, the relationship between BSA, systolic-diastolic values of arterial blood pressure, aortic arch geometry, aortic variations and aortic diameter measurements were investigated in children with BAV.

Statsitics

In calculating the sample size of this study, the Power (Power of Test) for each variable was determined by taking at least 80% and Type-1 error as 5%. Whether the continuous measurements in the study were normally distributed or not was checked with Kolmogorov-Smirnov (n> 50) and Skewness-Kurtosis tests and Parametric tests were applied because the measurements were normally distributed. Descriptive statistics for continuous variables in the study; mean, standard deviation; and for categorical variables, it was expressed as number (n) and percentage (%). "Independent T-test" was used to compare the measurements according to categorical groups. Pearson correlation coefficients were calculated to determine the relationships between continuous measurements. Pearson's Chi-square test was used to determine the relationship between categorical variables. In the calculations, the statistical significance level was taken as (a) 5% and the SPSS (IBM SPSS for Windows, ver.24) statistics package program was used for analysis.

Table 1. The average of the measurements made on the participants.

	MRA		Echocardiography	
	Mean	Std. Dev.	Mean	Std. Dev.
Annulus aorta (mm)	21,09	3,01	21,39	3,02
z-annulus	2,75	0,85	2,65	0,79
Sinus Valsalva (mm)	26,06	4,18	26,01	3,96
z-sinus	2,12	1,18	2,09	1,09
sinotubuler junction (mm)	24,00	4,93	24,01	4,68
z-stj	2,41	1,48	2,39	1,35
Middle asc aorta (mm)	23,72	4,83	23,55	4,06
z-arch	3,02	2,15	2,99	2,01
Prox arch aorta (mm)	19,91	3,90	20,01	3,82
z-prox	2,45	0,68	2,38	0,55
Middle arch aorta (mm)	17,22	2,89	17,05	2,02
z-middle	2,1	1,45	2,01	1,18
Prox desc aorta (mm)	15,63	2,55	15,55	2,42
z-pdesc	2,1	1,25	2,05	1,01
Middle desc aorta (mm)	14,59	2,28	15,01	2,02
z-mdesc	1,72	2,01	1,62	1,92
Systolic TA (mmHg)	107,81	11,77	-	-
Diastolic TA (mmHg)	64,06	13,68	-	-
EF (%)	-	-	70,69	3,37

RESULTS

A total of 571 cases had diagnosed with BAV. Thirty-two of these cases, who had MRA examination, were included in the study. Twenty-one of the participants were girls (66.6%) and 11 were boys (34.4%). The mean age was 12.66±3.92 years. BSA average of the participants 1.5±0.36 m^2. The average value of EF measured in EC was 70.69±3.37%. The mean systolic blood pressure was 107 + 11,77 mmHg, and the diastolic blood pressure was 64,06+ 13,68 mmHg.

Kappa test was performed for the measurements of ascending and aortic aorta diameters, which were made by EC and MRA, to compare the compatibility of the two imaging methods. There was strong agreement in measurements made at the level of annulus, sinus valsalva, sinotubular junction, middle ascending aorta, proximal and middle aortic archs (respectively. κ =0.91, 0.95, 0.87 ,0.86, 0.86 and 0,85).

The mean aortic diameters and z-scores of all participants obtained by MRA are EC shown in Table 1. When these measurements were compared, no statistical relationship was found between gender and aortic diameters and z scores (p=0,327-0,911) (Table. 2). A positive correlation was found between the age of the participants and the diameter measurements obtained from all levels. Aortic diameter increased with increasing age (p=0,005). The z scores of the calculated ascending aorta and aortic arch showed a negative correlation with age. There was no significant relationship

Table 2. Comparison results of measurements by gender.

	Girls		Boys		*p
	Mean	Std. Dev.	Mean	Std. Dev.	
Annulus aorta (mm)	21,24	3,13	20,82	2,89	,714
z-annulus	2,44	,82	2,72	,96	,911
Sinus Valsalva (mm)	25,76	4,19	26,64	4,30	,583
z-sinus	2,46	1,24	2,93	1,05	,293
sinotubuler junction (mm)	23,86	4,95	24,27	5,12	,825
z-stj	2,08	1,49	1,79	2,21	,623
Middle asc aorta (mm)	23,33	5,32	24,45	3,83	,541
z-middle asc aorta	2,92	2,40	2,96	1,51	,327
Prox arch aorta (mm)	19,76	3,75	20,18	4,33	,777
z-prox arch aorta	2,08	1,20	2,1	1,28	,552
Middle arch aorta (mm)	17,38	2,92	16,91	2,95	,669
z-middle arch aorta	1,72	1,05	1,78	1,12	,612

* Significance levels according to Independent T-test results.

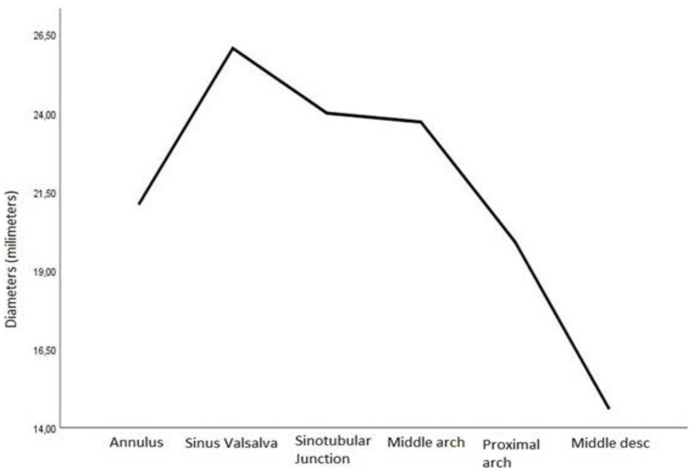

Figure 3. The average of the thoracic aortic diameters in the determined measurement areas is given in the graphic.

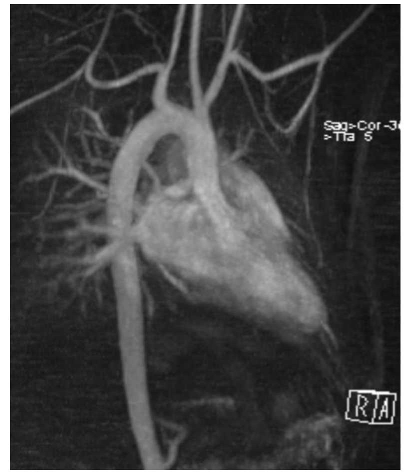

Figure 4. A 7-year-old girl with BAV, the common origin of the left common carotid artery and brachiocephalic artery in the aortic arch is observed. Bovine arch variation is seen.

between aortic valve pathologies and aortic arch geometry in thoracic aortic z scores (respectively p<0,360 and 0,295). The aortic diameters were larger than other aortic sections at sinus valsalva, sinotubular junction and middle arch levels (Fig.3). A statistically significant difference was observed in the presence of combined valve pathology and AI in the diameter measurements performed at the level of the middle aortic arch (p <0.05). This diameters increased in the presence of this valve pathologies. No correlation was found between AS, AI and combined valvular pathology and gender (respectively p=0,245; 0,184 and 0,245). There was no statistically significant difference between age and AS, AI and Mixed valvular pathology (p> 0.05). In EC examinations, 20 of 32 participants (62,5%) had AI, 2 (6,25%) had AS and AI combination (mixed). Isolated AS had not detected. None of the 10 (31,25%) participants had present. Of those with AI, 15 had 1st degree, 3 had 2nd degree, 2 had 3rd degree insufficiency. The most important diameter change in valve dysfunction was detected at the level of the middle arch with both EC and MRA (p=0,028). There was no significant relationship between AI, and combined valvular pathology and other aortic diameters (p> 0.05). Gender distribution, AS and AI rates of all BAV patients in our study are given in Table 3.

When the aortic arch geometry was evaluated, 10 rounded (31%), 5 (16%) Gothic-like and 17 Romanesque-like archs (53%) were observed with MRA. There was no statistically significant association between archs geometry and age, sex, AS, AI and combined valve pathology, EF, systolic and diastolic blood pressure (p=0,05).

In the evaluation made in terms of the variation of supraaortic branches, 5 of 32 patients diagnosed with BAV had bovine arch variation (15.5%) and one had an isolated left vertebral artery (3%). Of these patients, 2 (40%) were Gothic like, 2 (40%) were Romanesque like, 1 (20%) had a round

Table 3. Gender distribution, AS and AI rates of patients with BAV.

		Girls			Boys			**p.	Age		*p.
		N	Line %	Column %	N	Line %	Column %		Mean	Std.Dev.	
AS	(-)	19	63,3	90,5	11	36,7	100,0	,290	12,87	3,85	,245
	(+)	2	100,0	9,5	0	0,0	0,0		9,50	4,95	
AI	(-)	9	60,0	42,9	6	40,0	54,5	,529	11,67	4,50	,184
	(+)	12	70,6	57,1	5	29,4	45,5		13,53	3,20	
Mixed	(-)	19	63,3	90,5	11	36,7	100,0	,290	12,87	3,85	,245
	(+)	2	100,0	9,5	0	0,0	0,0		9,50	4,95	

*Significance levels according to Independent T-test results ** Significance level according to chi-square test results arch. Of those with supraaortic variance, 2 (Type 2 arch) had AI, 1 (Type 2 arch) mixed valvular pathology, 2 (Type 2 arch) none. No statistical relationship was found between this variation and aortic diameters, aortic valve pathologies, systolic-diastolic blood pressures, and arch types ($p > 0.05$).

When the results obtained from the hospital information records were evaluated, no aortic dissection or aneurysm formation had detected in any of the children with BAV included in our study as a result of MRA examination.

DISCUSSION

In our study, in which we measured the diameter of the aorta with EC and MRA in children with BAV, we found high compatibility between the two techniques. In the examination of aortic valve function performed with EC, we found the most common AI, consistent with the medical literature. MRA provided additional information on aortic morphology. Romanesque-like arches were found most frequently. We found no association between gender, mean blood pressure, aortic z-scores, aortic valve dysfunction, and thoracic aortic arch type. Another advantage of MRA over EC was that supraaortic branch variations could be demonstrated with MRA. With this method, we detected the most common bovine arch variation in the aorta.

BAV is a common congenital heart valve anomaly (12). An increase in diameter may occur due to dynamic changes in the ascending aorta of this valve pathology, which causes stenosis, insufficiency or mixed functional abnormalities. In these cases, along with the degree of valvular functional pathology, measurement and follow-up of the aorta diameters are required. It is necessary to evaluate the diameter of the ascending aorta because of aortic dissection and rupture that may develop in patients with BAV (13). It has been reported that changes in diameter due to postvalvular pressure changes in the aorta develop in the ascending aorta. If the diameter of the ascending aorta at the EC is measured greater than normal (z score>2), an MRA may be required to assess the lumen and distal parts of the aorta. Unlike other studies, our study shows the changes detected by MRA in the ascending and aortic arch in children with BAV who were evaluated with this imaging workflow.

According to the medical literature, the most common pathology of aortic valve is reported as a complication of BAV in all age groups (5). If the aortic valve cusps are asymmetrical, the stenosis may develop in childhood (1). In our study, we did not have a patient with BAV under the age of 18 who had isolated AS in the evaluations made with EC. The combined valve lesion had detected in 2 of 32 patients (6.25%). AI can coexist with AS (combined), and the degree of insufficiency is low. AI is a more common pathology in children with BAV than AS (5). Consistent with this result, the most common valvular pathology in children with BAV in our study was AI (62.5%). We found first-degree failure in 75% of these patients. We did not find aortic valve pathology in 33% of the patients with BAV. With phase contrast MRA, the regurgitation fraction in the aortic valve can be calculated (7). We did not evaluate these parameters, as it was not among our study objectives to investigate dynamic changes. These assessments had made with the EC.

Aortic dilatation formation is common in the presence of BAV due to genetic changes in aortic wall morphology and aortic valve dysfunctions (14). Patients with BAV face the risk of aortic diameter increase, aneurysm, and dissection throughout their lives. In patients with BAV, the aortic root and ascending aorta are significantly greater than patients with other valve anomalies. With the latest information, it is assumed that patients with BAV have an aortopathy that causes aortic dilatation (1,4,5). In our study, we found a significant increase in lumen diameter at the mid-arch level in MRA and EC, AI and combined valve pathology. We found no significant change with valvular pathology in other areas where measurements were made. In the study of Warren et al comparing children with pediatric BAV with healthy children, they showed that the z scores of the ascending aorta were significantly higher in children with a diagnosis of BAV than in healthy children (15). They stated that the annulus and sinotubular junction level did not change significantly. Since there was no control group in our study, such a comparison was not made. We concluded that the Z scores obtained for the defined levels of aortic diameters did not differ significantly in terms of aortic valve dysfunctions. On the other hand, in our study, it was observed that the aorta diameter in children with BAV was greater at the sinus valsalva, sinotubular junction and middle arch levels compared to other aorta sections. Fernandes et al. consistent with the results, we showed that the most frequently affected area of dilatation is the assending aorta in children with BAV (16).

Aortic morphology assessment can be performed with MRA examination. Helical flow pattern ensures healthy functioning of cardiovascular dynamics by providing laminar flow of blood and facilitating ventricular ejection (17). In Gothic morphology, while the arch provides more helical flow compared to other geometric types, it has been found to be associated with hypertension (18). It can be concluded that abnormal arch geometry leads to adverse effects on cardiovascular functioning due to an overexpression of the helical flow pattern and to larger pressure variations (17). Based on this hypothesis, the possible connection between BAV and aortic valve dysfunctions caused by aortic geometry was investigated in this study. In MRA examinations, half of the children with BAV had Romanesque-like archs aorta. The most significant result of the pressure analysis is that the

Romanesque arch always shows the smallest pressure gradient when compared to the other geometries. In fact, this geometry is the most comparable to the physiological configuration, in which the heart is optimized to minimize the energy consumption (17). As a result of our study, we found that the geometry of the arch was not associated with the patients' age, gender, blood pressure values, and aortic valve dysfunctions.

In the literature, we could not find any study investigating the changes in the diameter and shape of the aorta with age in the population under the age of 18. Redheuil et al. found that the aortic arch enlarges and the curvature decreases with increasing age in adults(19). They found that this changes were associated with decreased ascending aorta distension, increased aortic flow pressure, and increased systemic arterial blood pressure. In this study, we concluded that there was no relationship between systolic and diastolic blood pressure and aortic geometry in patients under 18 years of age.

The most common variant of the bovine arch is the origin of the left common carotid artery and brachiocephalic artery in the aortic arch. This variation can be found in 15% of the population (in the range of 8-25%). As part of this variation, there is another variation called the truncus bicaroticus, originating from the brachiocephalic artery of the left common carotid artery (20,21). On the MRA examination, we were able to show variations of the supraaortic branches that are difficult or impossible to identify with EC. Chatzistamatiou et al detected bovine archs variation in a female patient with hypertension and BAV in the second decade (22). They reported that early onset atherosclerosis in this patient was due to intraluminal flow dynamics changes caused by BAV. Supports this result in the way, Pham et al, in their histological evaluation of the aortic wall in BAV and in patients with BAV and bovine aortic arch patients, found that both entities affect the vascular wall architecture (23). In our evaluation in terms of supraaortic branch variation, we found bovine arch variation in 5 of 32 patients diagnosed with BAV (Figure 4), and isolated left vertebral artery in one. Early atherosclerosis or vascular wall irregularity accompanied by this variation was not differentiated. In the light of this information in the literature, it is understood that patients with BAV and aortic variations require clinical and radiological follow-up in terms of arterial wall changes caused by flow dynamic changes.

Aortic valve morphology, especially aortic diameters at the level of the annulus, can be evaluated with EC to demonstrate aortic dilatation and complications in children with BAV. The increase in aortic diameters necessitates an advanced examination method. Because EC, which is an ultrasonography method, cannot provide sufficient information in the thorax due to its limitations. In this case, aortic valve morphology, aortic valve dysfunction, aortic diameter measurements, aortic lumen and wall properties can be evaluated by static and dynamic methods with MRI. This method minimizes evaluator dependency in evaluation and allows re-evaluation over images (4,5,7,12). In our study, we were able to show the presence of aortic morphology and supraaortic branch variation with MRA in children with BAV. It is clear that MRA is helpful in the diagnosis of BAV and to show the variation/pathologies associated with it.

One advantage of MRA over EC is that the entire thoracic aorta can be evaluated. Westhoff-Bleck et al. reported that dilatation could be detected in the entire thoracic aorta and pulmonary arteries in patients with moderate/severe AS and AI in their study with BAV in patients of all age groups (24). Therefore, imaging of the entire thoracic aorta is recommended in cases with BAV. This imaging can also be done with MDCT This imaging can also be done with MDCT. However, MRI is a safer option considering the radiation exposure that will be caused by the necessity of repeating the patient's life.

The study has some limitations. The biggest handicap of the study is that the criteria used in cardiac MRI and MRA decision are not clear. In our study, there was no comparison of diameter measurements in distal of the aortic arch, since there was no control group consisting of healthy children who had MRA imaging of the thoracic aorta. No study evaluating the diameters of the distal aorta in patients with BAV could be found in the English language medical literature. Therefore, the measurements obtained can be used for studies with larger patient groups. The study was not designed for direct comparison of EC and MRA. Aorta valve dysfunction and hemodynamic changes were diagnosed with EC. We did not re-evaluate these changes with MRI. Pediatric patients with BAV are followed up with EC. MRI is performed in patients with increased thoracic aorta diameter due to the risk of dissection and aneurysm in a patient with BAV. This indicates that the MRI indication in the BAV is very limited. Therefore, as the most important limitation, the number of our participants was too low to allow statistical evaluation from time to time. In addition, cusps evaluation of the aortic valve was not performed in children with a diagnosis of BAV in EC and MRI. MRA was performed to evaluate aortic diameters and morphology. Only two supraaortic variation was detected in the small study group. Different variations might be detected in a large patient group.

In conclusion; EC can be used as a reliable method for aortic diameter assessment in children with BAV. This method is reliable for evaluating the diameter at the ascending level. Changes in the diameter of the aortic arch in children with BAV can be demonstrated by MRA in the thorax without anatomical limitations. In addition, MRA is useful to show the aortic arch geometry and supraaortic variations known to affect hemodynamic changes.

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