Bicuspid Aortic Valve Characteristics in Children

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Abstract

Background- Bicuspid aortic valve (BAV) is the most common congenital heart disease and the most common malformation of aortic valve. In BAV, there are two cusps instead of three cusps in the aortic valve. The objectives of this study were the determination of the aortic root dilatation and other anatomic and hemodynamic characteristics and abnormalities of BAV.

Methods- Thirty patients and 30 control subjects were evaluated. Aortic root dimensions were measured via two-dimensional echocardiography (2-D echo) at 4 levels, including the aortic valve annulus, sinuses of Valsalva, sinotubular junction (STJ) and proximal ascending aorta (AAO). Hemodynamic data and anatomic characteristics were measured using 2D and Doppler-echo. All the findings were matched and indexed for body surface area (BSA) and were compared with the matched data of the control subjects. Clinical and demographic findings of BAV were also determined and collected through a questionnaire.

Results- Among the patients, 70% were male and the mean age and weight of the patients were 7.5 years and 22.13 kg, respectively. 86.66% of the patients had systolic ejection murmur (SEM), 76.66% systolic ejection click (SEC) and 10% had chest pain. Other congenital heart diseases (CHD) were found in 26.96% of the patients, including coarctation of the aorta (CoA) in 23% of the cases. Matched mean anatomic aortic valve area (AAVA) was $2.05cm^2/m^2$, and matched mean effective aortic valve area (EAVA) was $1.41cm^2/m^2$ BSA. Maximum aortic valve pressure gradient (PG max) in systole was 56.56mmHg. Forty percent of the patients had aortic stenosis (AS): mild AS in 16.66%, moderate AS in 13.33% and intermediate AS in 10%. Prevalence of aortic insufficiency (AI) was 36.68%. When the data were compared with the control subjects, all the patients showed a meaningful larger aortic root dimension at all 4 levels (P values are presented in Table IV). Aortic root dilation was at the level of the annulus, sinuses of Valsalva, STJ and proximal AAO in 6.25%, 4.75%, 10.20% and 10.13%, respectively.

Conclusion- These findings support the hypothesis that BAV and aortic root dilation may reflect a common developmental defect. AS and AI are common in BAV. Similar to other obstructive defects of the left heart, BAV is significantly more common in males. Because murmurs and clicks are common in BAV even without AS or AI, all patients with a heart murmur and/or click must be evaluated for BAV ***(Iranian Heart Journal 2008; 9 (1):40 -46).***

Key words: bicuspid aortic valve ■ congenital heart disease ■ aortic root dilation ■ children

The bicuspid aortic valve (BAV) is the most common congenital heart disease (CHD), and is found in 1-2% of the general population. BAV is the most common malformation of the aortic valve.

Patients with BAV are susceptible to infective endocarditis (moderate risk). As the most common CHD and due to its serious complications, BAV must be diagnosed as soon as possible.

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In this study, we describe our findings in patients referred to the pediatric clinic for medical advice.

In BAV, there are 2 cusps instead of 3 cusps in the aortic valve (Fig. 1).

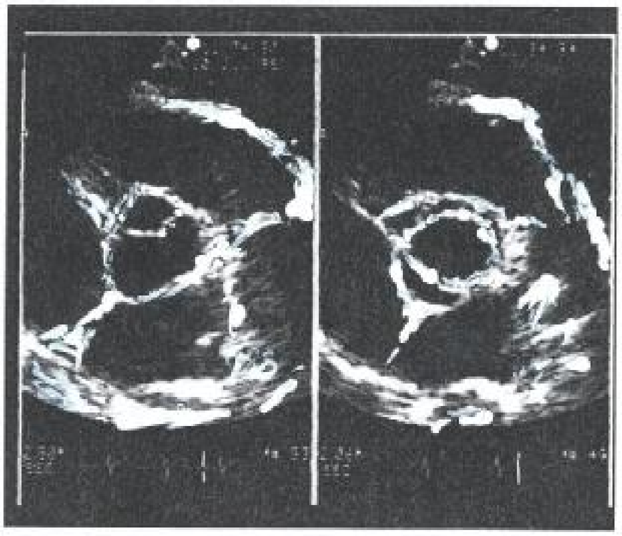

Fig.1. Echocardiographic examinations in parasternal short axis view. Left panel; diastole; the presence of a raphe at 1 o" clock simulates a normal tricuspid aortic valve.

Right panel: midsystole; it is clearly demonstrated that there are only two cusps.

Methods

This was a case-series study with a sample volume of 30 cases, and the duration of the study was 12 months. All the patients were referred to the outpatient pediatric cardiology clinic of Shaheed Rajaie Cardiovascular Medical Center during 2003-2004 (12 months). They were visited by pediatric cardiologists and their assistants. All the demographic data were obtained from the patients and their parents. The patients were examined, and then all the collected data and clinical findings were recorded in special questionnaires after the documentation of BAV via echocardiography.

For the estimation of AAVA and EAVA, we used a continuity equation:

$EAVA = LVOT_{CSA} \times LVOT_{VTI} / AV_{VTI}$ and $\pi d^2/_4$, where EAVA= effective aortic valve area, LVOT= left ventricle outflow tract, AV= aortic valve, CSA= cross-sectional area, VTI= velocity time integral (estimated by Doppler echo), d= diameter of aortic valve annulus and LVOT and π=3.14. Area unit was cm^2, and VTI unit was cm/s (second). For data analysis, we also used T-test and χ^2. This study has been done for the first time in Iran.

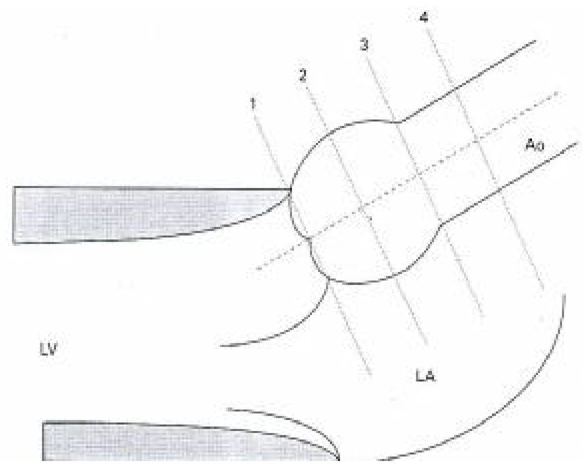

Fig. 2. Diagrammatic representation of the aortic root with the sites of measurement: 1, aortic annulus; 2, sinuses of Valsalva; 3, supra-aortic ridge; 4, proximal ascending aorta. Ao, aorta; LA, left atrium; LV, left ventricle.

Results

70% of the patients were male (M), and 30% female (F). The mean age and weight (wt) of the patients were 7.5 years and 22.13 kg, respectively. About 87% of the patients had systolic ejection murmur (SEM), and 76.66% had systolic ejection click (SEC) and about 10% had chest pain (CP). Other congenital heart diseases (CHD) were found in 26.96% of the patients, including coarctation of the aorta (CoA) 23%, ventricular septal defect (VSD) 6.66%, atrial septal defect (ASD) 3.33% and congenital complete heart block (CHB) 3.33% who underwent permanent pacemaker (PPM) implantation (Table I, Figs. 3, 4).

TableI. Demographic variables arranged according to age and number.

Patient[a]	No.[b]	Age			Sex		Wt (kg)[k]	HT (Cm)[l]	BSA[m] (m^2)	MM[q]		CP[z]	Click[y]	Other CHD	Kind of CHD[w]
		D[c]	M[d]	Y[e]	F[f]	M[g]				SM[p]	DM[s]				
K-B	1	17	-	-	-	+	3	50	0.20	+	-	-	-	+	ASD. CoA
M-Z	2	-	10	-	-	+	8	72	0.45	+	-	-	-	+	VSD. CoA
K-F	3	-	11	-	+	-	8	71	0.44	+	-	-	-	-	-
Z-S	4	-	6	1	+	-	10	88	0.50	+	-	-	+	-	-
A-S	5	-	-	3	-	+	15	110	0.66	+	+	-	+	-	-
M-M	6	-	4	3	-	+	12	92	0.55	+	-	-	+	-	-
B-B	7	-	6	4	+	-	19	124	0.82	-	+	-	+	-	-
H-B	8	-	-	5	-	+	20	122	0.82	+	-	-	+	-	-
F-Gh	9	-	4	5	+	-	18	116	0.76	+	+	-	+	-	-
M-K	10	-	-	5	+	-	15	112	0.66	-	-	-	-	+	CoA
M-M	11	-	9	6	-	+	17	110	0.70	+	-	-	+	-	-
M-T	12	-	-	7	-	+	17	115	0.73	+	+	-	+	-	-
S-M	13	-	3	7	+	-	20	126	0.82	+	+	-	+	-	-
M-K	14	-	5	7	-	+	23	140	0.92	+	-	-	-	-	-
S-S	15	-	2	7	+	-	16	115	0.70	+	+	-	+	-	-
A-A	16	-	10	7	-	+	22	129	0.86	+	-	-	+	-	-
H-A	17	-	10	8	-	+	25	142	0.96	+	-	-	+	-	-
M-N	18	-	10	8	-	+	23	124	0.88	+	-	-	+	+	CoA
H-H	19	-	11	8	-	+	23	130	0.90	+	+	-	+	-	-
H-M	20	-	11	8	-	+	20	120	0.82	+	+	-	+	-	-
A-KH	21	-	-	9	-	+	18	120	0.80	+	-	-	+	-	-
R-S	22	-	-	10	-	+	23	126	0.88	+	-	-	+	-	-
R-A	23	-	2	10	-	+	30	154	1.10	-	-	-	-	-	-
M-V	24	-	-	11	-	+	35	161	1.22	-	-	-	-	+	CHB+PPM
N-B	25	-	2	12	+	-	35	170	1.25	+	+	+	+	-	-
A-S	26	-	-	13	+	-	30	160	1.10	+	-	-	-	+	CoA
A-A	27	-	-	13	-	+	37	150	1.24	+	-	+	+	-	-
H-A	28	-	6	13	-	+	35	165	1.20	+	+	-	+	-	-
S-A	29	-	-	14	-	+	55	170	1.60	+	-	+	+	+	CoA
M-S	30	-	2	14	-	+	32	150	1.12	+	+	-	+	+	CoA-VSD
	30	17	124	218	9	21	22.13 (3-55)	124.46 (50-170)	0.85 (0.20-1.60)	26	11	3	23	8	CoA=7 VSD=2 ASD=1 CHB=1
Consi-deration	Total	7.5 y (17d-14y) Mean			30.00%	70.00%	Mean			86.66%	36.66%	10.00%	76.66%	26.66%	

a: first and last initial; b: number of patient's record; c: day ; d: month; e: year; f: female; g: male; k: weight in kilograms; l: height in centimeters; m: body surface area in square meters; q: murmur; p: systolic murmur; s: diastolic murmur; z: chest pain; ASD: atrial septal defect; CHB: complete heart block; CoA: coarctation of aorta; PPM: permanent pacemaker; VSD: ventricular septal defect

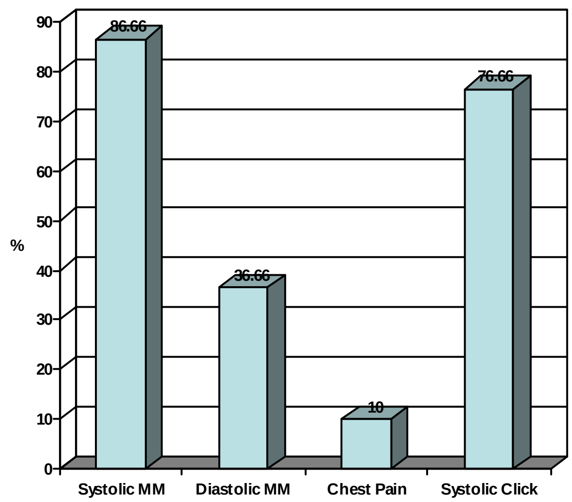

Fig. 3. Prevalence of clinical findings

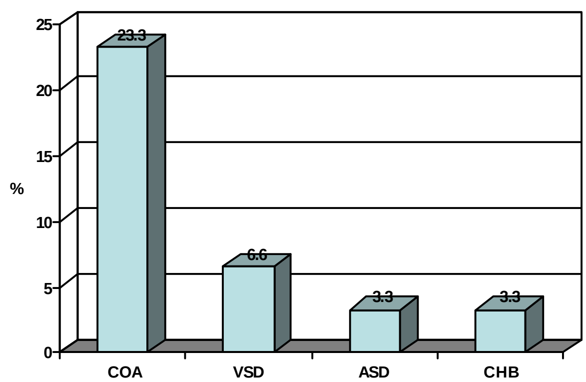

Fig. 4. Prevalence of associated CHD

Matched mean anatomic aortic valve area (AAVA) was 2.05 cm^2/m^2, and matched mean effective aortic valve area (EAVA) was 1.41 cm^2/m^2 BSA (body surface area). EAVA: AAVA ratio was about 0.68, suggesting incomplete aortic valve opening during systole (Fig. 5).

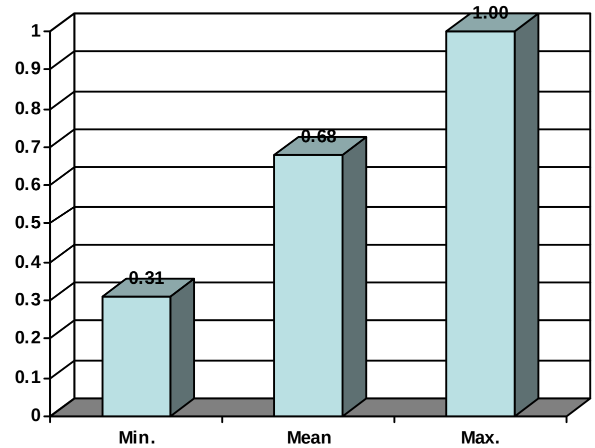

Fig. 5. EAVA/AAVA ratio.

Maximum pressure gradient (PG_{max}) across the aortic valve was 56.56mmHg in systole. Among the patients, 40% had aortic stenosis (AS), of which 16.66% was mild, 13.33% was moderate and 10% was intermediate AS. There was no case with severe AS. Prevalence of aortic insufficiency (AI) was 36.68%, all of which were mild (1^+) to moderate (2^+). When the collected echocardiographic data (Tables II, III) were compared with the control subjects (Table IV), all the patients showed meaningfully larger aortic root dimensions at the aortic valve annulus, sinuses of Valsalva, STJ and proximal AAO (Fig. 2, see Table IV P-values). The percents of the enlargement of the 4 above-mentioned diameters were 6.35% for the annulus, 4.75% for sinuses of Valsalva, 10.20% for sinotubular junction (STJ) and 10.13% for proximal ascending aorta (AAO). All the echocardiographic data were collected and summarized in Tables I-IV.

Table II. Echocardiographic variable arranged according to age and number.

1	2	3	4	5	6	7	8	9	10	File No. / Variables	
3.15	2.04	1.89	2.57	1.05	2.54	2.24	2.27	1.22	1.62	Indexed-AAVA Cm^2/m^2	
3.15	1.43	0.90	0.81	1.15	2.54	2.12	1.51	1.51	1.56	Indexed-EAVA Cm^2/m^2	
1.00	0.70	0.48	0.31	1.00	1.00	0.93	0.66	1.00	0.96	EAVA/AAVA Ratio	
3.15	1.57	1.95	3.28	2.25	3.63	2.35	3.06	1.60	1.93	Indexed-LVOTSA Cm^2/m^2	
1.56	1.50	2.59	3.19	1.99	1.22	1.42	2.11	3.43	1.69	V_{max}- M/s	AOV
1.09	0.93	1.52	2.06	1.37	0.84	1.09	1.27	2.29	1.18	V_{mean}- M/s	AOV
13.43	9.01	26.87	40.73	15.83	5.96	8.16	17.86	47.11	11.50	P_{max}- mmHg	AOV
6.31	4.52	12.50	21.25	9.26	3.34	5.52	8.73	29.39	6.56	P_{mean}- mmHg	AOV
21.10	20.42	38.75	48.38	34.27	20.22	26.67	36.45	55.55	32.84	VTI-Cm	AOV
1.74	1.52	1.05	0.68	0.93	0.83	1.40	1.06	3.23	1.48	V_{max}- M/s	LVOT
1.07	0.91	0.76	0.57	0.79	0.70	1.07	0.71	2.18	1.04	V_{mean}- M/s	LVOT
11.77	9.38	4.45	2.79	3.50	2.78	7.79	4.52	41.72	8.83	P_{max}- mmHg	LVOT
6.20	4.49	2.60	2.26	2.92	2.29	5.55	2.57	24.59	5.26	P_{mean}- mmHg	LVOT
19.92	18.58	18.04	12.01	17.57	14.25	24.11	18.09	52.33	26.47	VTI-Cm	LVOT
0.93	0.93	1.05	1.46	1.38	1.59	1.57	1.79	1.25	1.28	LVOT	Diameters (Cm)
0.66	0.66	1.03	1.28	0.94	1.38	1.57	1.54	1.09	1.17	AV-Ann	Diameters (Cm)
0.74	1.51	1.21	1.71	1.85	1.72	2.48	2.23	1.64	1.55	Vals-Sin	Diameters (Cm)
0.66	1.00	1.02	1.22	0.99	1.22	1.93	1.74	1.19	1.18	STJ	Diameters (Cm)
1.09	1.60	1.33	1.85	1.35	1.38	1.60	1.98	1.92	1.60	AAO	Diameters (Cm)

11	12	13	14	15	16	17	18	19	20	File No. / Variables	
2.90	3.44	1.25	1.78	0.63	1.23	2.31	2.27	2.76	1.44	Indexed-AAVA Cm^2/m^2	
1.30	2.67	1.32	1.78	0.63	1.23	2.31	1.81	1.42	1.92	Indexed-EAVA Cm^2/m^2	
0.44	0.77	1.00	1.00	1.00	1.00	1.00	0.79	0.51	1.00	EAVA/AAVA Ratio	
0.31	2.28	2.82	2.11	1.77	2.62	3.50	2.56	2.42	2.79	Indexed-LVOTSA Cm^2/m^2	
1.65	1.82	1.92	1.44	1.77	3.76	1.97	1.62	2.47	1.75	V_{max}- M/s	AOV
1.08	1.28	1.33	0.97	1.29	2.29	1.28	0.97	1.74	1.16	V_{mean}- M/s	AOV
11.02	12.89	14.57	8.42	12.69	56.56	15.68	10.64	24.46	12.34	P_{max}- mmHg	AOV
5.65	7.91	8.23	4.57	7.75	26.41	8.21	4.91	14.54	0.39	P_{mean}- mmHg	AOV
27.86	25.83	37.93	21.82	38.80	56.40	35.74	22.31	46.00	32.78	VTI-Cm	AOV
0.91	1.81	0.80	1.39	0.77	1.04	1.33	1.21	1.52	1.09	V_{max}- M/s	LVOT
0.68	1.27	0.67	0.91	0.66	0.80	0.86	0.74	1.21	0.80	V_{mean}- M/s	LVOT
3.52	12.08	2.61	7.72	2.36	4.33	7.07	5.94	9.36	4.86	P_{max}- mmHg	LVOT
2.31	7.68	1.98	4.29	2.02	2.90	3.67	2.76	6.92	3.05	P_{mean}- mmHg	LVOT
11.00	30.20	16.98	18.33	12.50	26.44	23.61	15.79	27.03	22.54	VTI-Cm	LVOT
1.72	1.67	1.72	1.58	1.26	1.70	2.07	1.70	1.67	1.71	LVOT	Diameters (Cm)
1.61	1.79	1.31	1.51	1.10	1.20	1.87	1.60	1.78	1.23	AV-Ann	Diameters (Cm)
1.49	2.17	1.98	2.29	1.57	2.02	2.07	2.50	1.86	1.78	Vals-Sin	Diameters (Cm)
1.23	2.00	1.28	1.77	1.17	1.73	1.60	2.02	1.31	1.28	STJ	Diameters (Cm)
1.46	2.17	1.56	1.96	1.20	2.39	1.96	2.46	1.60	1.72	AAO	Diameters (Cm)

21	22	23	24	25	26	27	28	29	30	File no. / variable	
2.54	3.86	2.36	2.14	0.83	0.53	1.65	2.53	1.94	3.17	Indexed-AAVA Cm^2/m^2	
0.86	2.89	1.50	1.41	0.83	0.52	0.83	0.85	1.78	1.38	Indexed-EAVA Cm^2/m^2	
0.33	0.88	0.63	0.66	1.00	0.99	0.44	0.33	0.93	0.43	EAVA/AAVA Ratio	
2.66	3.14	1.66	1.68	2.21	1.60	1.54	2.61	1.76	2.50	Indexed-LVOTSA Cm^2/m^2	
2.37	2.26	1.50	1.80	2.01	3.72	2.92	3.73	1.35	1.47	V_{max}- M/s	AOV
1.62	1.64	0.87	1.17	1.38	2.65	1.92	2.66	1.21	1.20	V_{mean}- M/s	AOV
22.59	20.54	9.02	13.04	16.18	55.41	34.22	55.36	14.89	8.71	P_{max}- mmHg	AOV
13.03	12.62	3.78	6.63	9.31	33.65	18.81	33.80	7.09	6.70	P_{mean}- mmHg	AOV
43.34	46.13	22.12	26.14	39.53	91.87	59.13	91.91	37.91	27.94	VTI-Cm	AOV
0.87	2.03	1.30	1.60	0.84	1.40	1.59	1.41	1.92	3.05	V_{max}- M/s	LVOT
0.67	1.50	0.82	0.98	0.70	1.03	1.29	1.62	1.35	2.91	V_{mean}- M/s	LVOT
3.07	16.54	6.84	10.24	2.82	7.90	10.26	7.90	14.67	45.98	P_{max}- mmHg	LVOT
2.19	10.62	3.32	4.82	2.22	5.19	7.68	5.17	8.75	38.21	P_{mean}- mmHg	LVOT
14.10	42.36	20.00	21.98	14.94	30.40	31.87	30.35	37.90	15.50	VTI-Cm	LVOT
1.65	1.88	1.53	1.62	1.88	1.50	1.56	2.00	1.91	1.89	LVOT	Diameters (Cm)
1.61	1.91	1.82	1.83	1.69	1.73	1.73	1.97	1.99	2.13	AV-Ann	Diameters (Cm)
1.48	2.27	2.00	2.49	1.98	2.28	2.38	2.56	2.74	2.70	Vals-Sin	Diameters (Cm)
1.38	1.92	1.80	1.88	1.60	1.82	1.70	2.83	2.66	1.68	STJ	Diameters (Cm)
1.22	2.45	2.03	2.17	3.52	3.83	2.48	2.83	2.75	2.78	AAO	Diameters (Cm)

Table III. Maximum, Minimum & Mean of variables

Indexed AAVA $^{cm2}/_{m2}$	3.44	3.44	3.44
$^{cm2}/_{m2}$ Indexed EAVA	3.15	3.15	3.15
Ratio $^{EAVA}/_{AAVA}$	1.00	1.00	1.00
$^{cm2}/_{m2}$ Indexed LVOTSA	3.63	3.63	3.63
mmHg $AVPG_{max}$	56.56	56.56	56.56
mmHg $AVPG_{mean}$	33.80	33.80	33.80
m/s AV-VTI	91.91	91.91	91.91
LVOT-VTI m/s	52.33	52.33	52.33
Cm STJ-Dia	2.83	2.83	2.83
AV-An-Diia Cm	2.13	2.13	2.13
Sin-Vals-Dia Cm	2.74	2.74	2.74
A-AO-Dia Cm	3.83	3.83	3.83
Variable / Range	Maximum	Maximum	Maximum

AAO= Ascending aorta= Ascending aorta; AOVSA= AOV surface area = Aortic valve surface area;
AAVA= Anatomic-AV area= Anatomic Aortic valve area;AOV= AV= Aortic valve; EAVA= Effective AV area = Effective aortic valve area; Ra= Ratio; LVOTSA= LVOT surface area = Left ventricular outflow tract surface area; LVOT = LV outflow tract; LV= Left ventricle; Cm^2= Square centimeter; M^2= Square meter;V max= Maximum velocity; V mean= Mean velocity; M/S= Meter per second;mmHg= Millimeter Hg (Mercury); VTI= Velocity time integral; P max= Maximum pressure gradient across AV;
P mean= Mean pressure gradient across AV; AV-Ann= Aortic valve annulus; Sin-Vals= Valsalva sinus;STJ= Sinotubular junction;
AAO= Ascending aorta; Cm= Centimeter; Dia= Diameter;

Table IV. Comparison of matched anatomic variables between patients and control cohort

AAO Diameter	STJ-SAR Diameter	Sinuses Diameter	AV-Annulus Diameter	No	Variables / Cohort
3.15 cm/m^2	2.70 cm/m^2	3.30 cm/m^2	2.55 cm/m^2	30	Patients
2.86 cm/m^2	2.45 cm/m^2	3.15 cm/m^2	2.40 cm/m^2	30	Controls
10.13%	10.20%	4.75%	6.25%	30	Difference
$P<0.05$	$P<0.01$	NS	$P<0.005$	30	P value

Indexed-Matched= All diameters Indexed Per m^2; M^2= Square meter; AV=Aortic Valve; Sinuses=Valsalva Sinuses; STJ-SAR=Sinotubular Junction- Supraaortic Ridge; AAO=Ascending Aorta; BAV=Bicuspid Aortic Valve

Discussion

The findings of our study support the hypothesis that BAV and aortic root dilation may share a common developmental defect. AS and AI are common in BAV. Similar to other obstructive defects of left heart, BAV is significantly more common in males. Even without AS or AI, murmurs and clicks are very common in BAV.

There are few studies about BAV in the literature. In 2003, Nistri et al. reported the results of their study and concluded that in BAV, aortic root dimensions are larger than control healthy subjects. These excess values of diameter for aortic valve annulus, sinuses of Valsalva, STJ and proximal AAO were 7.5%, 11.6%, 15% and 43.9%, respectively [3] (Table V).

Table V. Comparison of our study with others

Hemodynamic variables			Anatomic variables				Variables / Studies
Normal	AI	AS	AAO	STJ	Sinuses	Annulus	
60%	36%	40%	10.13%	10.20%	4.75%	6.25%	Our study
23%	64%	13%	50-60%	57-79%	36-78%	9-59%	Rebecca et al [(2)]
-	-	-	44%	15%	11.60%	7.50%	s. Nistri et al [(3)]

Habn et al. reported that these excess of dimension for the above-mentioned 4 levels were 9-59%, 36-78%, 57-79% and 50-64%. The BAV was more common in males.[2] In another study by Paul, et al. they reported that BAV is the most common CHD and may be a genetic defect. BAV is a disease of the aortic root at all levels.[1]

While our study supports the results of the other studies in other centers, it also shows differences in the dimensions and other values measured by us and others. Because other studies were done in adult subjects in contrast to ours, which was carried out in children and young patients, and because the aortic root dilatation, AS and AI are progressive, the percents of excess in aortic root dimension must be smaller in our study. According to our study and other studies we recommend:

1.Evaluation of all children with heart murmur and / or click to rule out BAV,

2.Follow-up of all patients with BAV for progression of AS, AI and other complications,

3.Screening of the patient’s first-degree relatives for BAV by noninvasive screening and diagnostic tests such as transthoracic echocardiography, and

4.Evaluation of BAV cases for the co-existence of CHD.

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