

Aortic valve morphology with special consideration to bicuspid aortic valve –prevalence, subtypes, & associated complications in a tertiary care center of western Rajasthan - An echocardiography based study

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Abstract

The bicuspid aortic valve is composed of two leaflets. Diagnosis of bicuspid aortic valve (BAV) is important because it identifies patients with high risk of valvular and aortic complications. The study was aimed to aortic valve morphology with special consideration to bicuspid aortic valve prevalence, subtypes, & associated complications in a tertiary care center of western Rajasthan by echocardiography based study.

Method: The present study was a Cross-sectional study conducted in the Department of Anatomy S. P. Medical College Bikaner in collaboration with Haldiram Moolchand Cardiovascular Center of P.B.M. Hospital, Bikaner, Rajasthan. A total of 15 subjects of Bicuspid Aortic Valve were identified from 527 echocardiographic reports of patients The patient was clinically examined for the prevalence, subtypes and complications of BAV.

Conclusion: In our study we observed that there is the prevalence of BAV 2.85%, Most common subtype

type1 (A) left-right coronary subtype and complication was aortic stenosis N=3(20%) and Aortic Stenosis with Aortic Regurgitation N=4(26.67%) reported.

Keywords: Bicuspid aortic valve, Prevalence, Subtypes, Complications.

Introduction

The aortic valve normally has three cusps or leaflets /tricuspid, although in 1–2% of the population it is found to have two leaflets/bicuspid. Bicuspid Aortic Valve (BAV) is the commonest congenital cardiac abnormality with an estimated prevalence of 1-2%. It is responsible for more deaths and complications than the combined effects of all the other congenital heart defects. An abnormal aortic valve develops during the early weeks of pregnancy, when the baby’s heart is developing. BAV often runs in families. It is almost 3 times more common in males than females. It is also the most observed cardiac defect in Turner syndrome. 1-2. The Sievers classified bicuspid aortic valve in 3

types: type 0 (no raphe); type 1 (1 raphe); and, type 2 (2 raphes) 3

The critical complications of this condition are well described and include aortic stenosis, aortic regurgitation, infective endocarditis, and aortic dissection.⁴

Transthoracic echocardiogram (TTE) is the routine method used in the study of structure and function of the aortic valve⁵

Echocardiographic screening of FDR (First-degree relatives) is a useful approach for identifying asymptomatic cases, & should therefore be offered to all FDR of BAV individuals.⁶

As there is very less data available on the aortic valve in Rajasthan region so considering the current situation of BAV in Rajasthan, present study aims to investigate the prevalence, subtype and complications of BAV in western Rajasthan people based on a large echocardiographic database.

Materials and Method

The present study was conducted in the Department of Anatomy S. P. Medical College Bikaner in collaboration with Haldiram Moolchand Cardiovascular Center of P.B.M. Hospital, Bikaner, Rajasthan. It was a Cross-sectional study. The study proceeded on 527 patients from outdoor and indoor Department of cardiology in 6 months duration. Prior approval of hospital's ethical committee was taken. Prior consent of individuals under study was also taken in the form of informed consent form.

Inclusion Criteria

Suspected cardiac etiology based on symptoms, signs, or other testing; evaluation and follow-up of subjects with cardiovascular disease (valvular heart disease, congenital heart disease (CHD), coronary artery disease, heart failure, cardiomyopathy, arrhythmia,

hypertension) routine evaluation of cardiac structure and function before noncardiac surgery.

Exclusion Criteria

Operated cases of aortic valve {post Aortic valve replacement}.

Patients with infective endocarditis involving aortic valve.

Materials

Echocardiography machine, Transducer, Gel, Bed, ECG, Digital weighing machine, Height chart and a data sheet, Proforma.

Method

527 individuals were included in the study after obtaining the informed consent and completion of baseline investigation, each individual was interviewed and echocardiography was performed to see the morphology of aortic valve. During the interview, the pre tested pre structured questionnaire was administrated to eligible participants, which contained patient's personal details, details of modifiable risk factor e.g. hypertension, Cholesterol level .

All the patients were examined by their physician before the procedure, full clinical assessment done, reviewing of ECG report before done echocardiography.

The patient was clinically examined for the prevalence, subtypes and complications of BAV. Finding was recorded in preformed proforma.

The Bicuspid Aortic Valve classified into:

Type 0 BAV has by definition no raphe and 2 valve cusps. It further subdivides type 0 into the following 2 subtypes: the more common lateral subtype with right and left cusps and the less common anteroposterior subtype with anterior and posterior cusps.

Type1 BAV has by definition a single raphe and 2 valve cusps making this BAV morph type the most

common. It further subdivided into 3 subtypes: the most common left-right subtype with fusion of the right and left coronary cusps. The less common right-nonsubtype with fusion of the right and noncoronary cusps and the rare nonleft subtype with fusion of the noncusps and left coronary cusps.

Type 2 BAV has by definition 2 raphe and 2 valve cusps making this BAV morphotype the rarest of the 3 types in this classification. Type 2 morphotype had a raphe between the right and left coronary cusps(L-R) and a second raphe between the right and noncoronary cusps(R-N), Classified as a type 2 L-R/R-N BAV.

Procedure

Echocardiography

Echocardiography is a non invasive painless test that uses sound waves to create moving pictures of heart. The pictures show the size and shape of heart. They also show how well heart's chambers and valves are working.

- After explaining about the study, Patient was lie on his back and left side.
- Electrodes (small, adhesive patches) are placed on chest to record his heart's electrical activity (ECG) during the test.
- Gel was applied to his chest.

The transducer was placed on his skin and moved around the area to view the heart from different angles.

Data Analysis

Data thus collected was entered into Microsoft Excel spreadsheet and was presented in the form of tables, figures, graphs & diagrams.

Statistically analyzed with the help of percentage, mean, SD, t-test, and p value of significance to study association of various factors with aortic valve.

Observation

In present study total 527patients were incorporated. They were evaluated and examined for:

1. Bicuspid Aortic Valve prevalence in people of Western Rajasthan on 2-D Echocardiography.
2. To determine subtype of Bicuspid Aortic Valve in people of Western Rajasthan.
3. To study complication of Bicuspid Aortic Valve in people of Western Rajasthan.

Following observations were recorded:-

Table 1: Distribution according to morphology of aortic valve

Aortic valve	Patients	Percentage %
Tricuspid AV	512	97.15%
Bicuspid AV	15	2.85%
Total	527	100%

Table 2: Distribution of Bicuspid Aortic Valve classification according to subtypes and gender

Type N=15	Subtype	Male N=10	Percent %	Female N=5	Percent %
Type 0	Lat (A)	2	13.33%	-	-
	Ant.- post.(B)	1	6.67%	-	-
	Total	3	20%	-	-
Type 1	L-R (A)	5	33.33%	4	26.67%
	R-N (B)	2	13.33%	1	6.67%
	N-L (C)	-	-	-	-
Total		7	46.67%	5	33.33%
Type 2		-	-	-	-
Total		10	66.67%	5	33.33%

Table 3: Distribution of complication associated with Bicuspid Aortic Valve according to reporting Age group.

Age (N=15)	0-20 Years	21-40 Years	41-60 Years	61-81 Years	Total
No complication	2 (13.33%)	3 (20%)	2 (13.33%)	0	7 (46.67%)
Aortic Stenosis with Aortic Regurgitation	-	-	4 (26.67%)	-	4 (26.67%)
Pure Aortic stenosis	-	1 (6.67%)	1 (6.67%)	1 (6.67%)	3 (20%)
Pure Aortic Regurgitation	-	-	1 (6.7%)	-	1 (6.67%)

Table 4: Distribution of Bicuspid Aortic Valve cases according to classification and complication associated with bicuspid aortic valve (N=15).

Subtype of Bicuspid Aortic Valve		Associated complication of Bicuspid Aortic Valve of reported (N=8)		
		Aortic Regurgitation	Aortic Stenosis	Aortic Stenosis with Aortic Regurgitation
Type 0 N=3	Lateral(A)	-	-	1(6.67%)
	Ant.post.(B)	-	1(6.67%)	-
Type 1 N=5	L-R(A)	-	2(13.33%)	1(6.67%)
	R-N(B)	1(6.67%)	-	2(13.33%)
	N-L(C)	-	-	-
Type 2		-	-	-

Discussion

The present Cross-sectional study was conducted after obtaining informed consent of individuals under study, they were questioned and examined for aortic valve morphology. The study proceeded with outdoor & indoor patients of cardiovascular center, in 6 months duration. Prior approval of hospital’s ethical committee was taken.

In our study 15 subjects from 527 echocardiographic reports from the OPD and IPD cardiology were included.

Results obtained were discussed as follows:

A. Prevalence of bicuspid aortic valve

The Prevalence of bicuspid aortic valve N= 15 (2.85%) from 527 echocardiographic reports of patients. Gender Prevalence were found in N=10 (66.7%) Males and N=5 (33.3%) Females. The ratio of Bicuspid aortic valve among male and female is 2:1 respectively which means males with Bicuspid aortic valve were twice in number as compared to females. Mean of aortic root diameter of BAV was 2.27 ± 0.43 cm.

Li yijian (2017)4 et al include the patients between ages 0-83 years in a study conducted in china. It showed the 0.43% (N=668) prevalence of BAV by echocardiographic database.

It found ratio between male and female was 1.9:1. (438:230).

In a study conducted by Movahed M R et al (2006)22 the echocardiographic prevalence of BAV was 0.6% in the larger database and 0.5% in the smaller athletic database.

Basso C et al (2004)18 showed an echocardiography study of 817 asymptomatic children,a BAV was found in 4 of the 817children(0.5%), 3of 4 being found in males.

In an echocardiography study conducted by tutar E et al (2005)²¹ on 1075 neonates, the prevalence of BAV was 4.6 per 1000 live birth. (7.1 per 1000 males and 1.9 per 1000 females).

The prevalence of BAV is approximately 1% of the population with a male/female ratio of between 2:1 and 3:1.^{9, 10,12,14,16}

Author(s)	Year	Study Population	Bicuspid Valve Prevalence(%)
Grant et al ⁹	1928	1,350	0.89%
Roberts ¹⁰	1970	1,440	0.9%
Larson and Edwards ¹²	1984	21,417	1.37%
Data et al ¹⁴	1988	8,800	0.59%
Pauperio et al ¹⁶	1999	2,000	0.65 %
Current study	2019	527	2.85%

B. Bicuspid Aortic Valve Subtype

Total N=15 patients of bicuspid aortic valve were found in study out of which 3 patients had type 0, in which 2 (13.33%) had lateral subtype(A), 1 (6.67%) had Anterior -posterior subtype. Out of N=15,12 patients had type 1,it has 3 subtype in which 9(60 %) had left-right coronary subtype (A), 3(20%) had Noncoronary-left subtype (B)and Noncoronary-left subtype of type1 was not found in study. Type 2 was not found in our study.

1. Bicuspid aortic valve Classified according to reported age group:

Out of N=15 patients of bicuspid aortic valve were found in study out of which 3 patients had type 0 in which N=2 (13.33%) lateral subtype (A), N=1 (6.67%) Anterior-posterior subtype patients had 41-60 years age group. Out of N=15,12 patients had type 1in which N=2 (13.33%) left-right coronary subtype (A) had 0-20

years age group, N=4 (26.67%) left-right coronary subtype (A) had 21-40 years age group, N=2(13.33%) left-right coronary subtype (A) had 41-60 years age group, N=1(6.67%) left-right coronary subtype (A) had 41-60 years age group and N=3 (20%) Noncoronary-left subtype (B) had 41-60 years age group and Noncoronary-left subtype was not found study. Type 2 was not found in study.

2. Classification of bicuspid aortic valve according to gender:

Total N=15 patients of bicuspid aortic valve were found in study. out of N=15, 10 (66.7%) males had Bicuspid Aortic Valve whereas out of N=15, 5 (33.3%) females had Bicuspid Aortic Valve.Out of N=10males, 3 had type 0 in which N=2(13.33%) patients had lateral (A) subtype and N=1 (6.67%) patients had ant.-post. subtype and out of N=10(46.67%)males,7(46.67%) males had type 1in which 5 (33.33%) patients had left right subtype and 2(13.33%) patients had Right-noncoronary subtype.Type 2 was not found in male. out of N=5 (33.33%)female had type 1in which N=4 (26.67%) patients had left right subtype and N=1 (6.67%) patients had Right-noncoronary subtype.Type 0 and Type 2 was not found in female .

Sievers HH et al (2007)²⁴ shows the bicuspid aortic valve were 21 (7%) patients with BAV Type 0 (valve with no raphe).Most patients (n =269, 88%) had BAV type 1 (valve with one raphe). Within this group the predominant finding was type 1, L/R, S (n =119, 39%), which means a BAV with one raphe between the left and right coronary sinuses and with valvular stenosis. There were 14 patients with BAV type 2 (valve with two raphes).

Various classification schemes have been used to characterize the BAV.^{11,24}

Fernandes SM et al (2004)¹⁹ showed a review of echocardiograms of 1135 children with BAV revealed that in 70% of cases ,right coronary and left coronary leaflet fusion was the most common morphologic variant(pattern A).

In a surgical pathology study conducted by Sabet HY, Edwards WD et al (1999)¹⁷ 542 cases of BAV, 86% were noted to have pattern A, 12% pattern B and 3% pattern C.

Fernandes SM et al (2007)²³ showed the valve orientation of BAV may be predictive of clinical outcomes. Fusion of the right and noncoronary valve leaflets has been associated with more rapid progression of aortic stenosis and regurgitation than fusion of right and left coronary leaflets.^{19,23}

C. Complication associated with of Bicuspid Aortic Valve: Total N=15 patients of bicuspid aortic valve were found in study. out of N=15, 4 (26.67%) patients were found aortic stenosis with aortic regurgitation. Out of N=15 ,3 patients had aortic stenosis, the aortic stenosis has 3 type mild ,moderate and severe , 1 (6.67%) patient had mild aortic stenosis , 2 (13.33%) patients had moderate aortic stenosis. Out of N=15, 1 (6.67%) patient had Aortic Regurgitation, it has also 3 type mild, moderate and severe aortic regurgitation .only 1 (6.67%)patient had moderate type aortic regurgitation. no patient was recorded with aortic endocarditis. Out of N=15, 7(46.67%) patients not found any complication.

1. Complication associated with Bicuspid Aortic Valve according to reporting age group:

Out of N=15 patients of bicuspid aortic valve were found in study in which 0-20 years age group was not found complication. In 21-40 years age group had 1 (6.67%) aortic stenosis patient. In 41-60 years age group had 4 (26.67%) aortic stenosis with aortic

regurgitation and 1 (6.67%) Aortic stenosis. In 61-81 year age group had 1 (6.67%) aortic stenosis.

2. Complication associated with Bicuspid Aortic Valve according to gender:

Out of N=15 patients of bicuspid aortic valve were found in study in which N=10 (66.7%) males had Bicuspid Aortic Valve whereas N=5 (33.3%) females had Bicuspid Aortic Valve. Out of 10 males, 1(6.67%) patients had aortic regurgitation, 2(13.33%) patients had aortic stenosis, 3(20%) patients had aortic stenosis with aortic regurgitation and 4(26.67%) patients were not found any complication. Out of N=15,females, 1(6.67%) patients had aortic stenosis, 1(6.67%) patients had aortic stenosis with aortic regurgitation and 3(20%) patients was not found any complication.

3. Bicuspid Aortic Valve cases according classification of bicuspid aortic valve associated with complication:

Out of N=15 patients of bicuspid aortic valve were found in study in which Lateral(A) subtype of type 0, 1(6.67%)patients was found to have aortic stenosis with aortic regurgitation and Ant. post.(B) subtype of type 0 ,1(6.67%) patients was found to have aortic stenosis. In left right subtype (A) of type 1, 1(6.67%) patients was found to have aortic stenosis with aortic regurgitation , 2(13.33%) patients was found to have aortic stenosis and right-noncoronary subtype of type 1, 2(13.33%) patients was found to have aortic stenosis with aortic regurgitation, 1(6.67%) patients was found to have aortic regurgitation.

Li yijian et al (2017)⁴ showed 89 (13.32%) patients were free from aortic valve disease, while 579 (86.68%) patients have various degree of AS or AR. Almost half of the patients who were diagnosed BAV have different degrees of aortic valve disease after 40 years. There were 42.07% patients who had AS accompanied with AR. The number of patients who

have moderate or severe AS or AR increased with age. The percentage of patients with pure AS and pure AR was 27.10% and 17.51%, respectively. They analyzed the constituent ratio of calcific AS in different age groups among 462 patients who have AS. With increasing age, the constituent ratio of calcific AS increased from 0% in group 1 (0 to 20 years) to 50% in group 4 (over 60 years). It shows 59.58% of BAV patients had any degree of AR, and 65.8% (77/117) of patients had mild AR in the pure AR cohort. It also found the incidence of infective endocarditis was 0.68% episodes per patient-years in our study, 4.11% (18/438) and 2.17% (5/230) in men and women, respectively.

Surgical pathology series report the incidence of aortic stenosis for BAV as between 5% and 50%. The relation between age and BAV stenosis, which has the same tendency, is shown in the surgical series: it occurred in 33% of patients aged 21 to 30 years, 60% aged 31 to 40 years, 64% aged 41 to 50 years, 69% aged 51 to 60 years, 60% aged 61 to 70 years, 42% aged 71 to 80 years, and 28% aged 81 to 90 years. 8, 13, 20 Isolated AR in patients with BAV is less common than aortic stenosis, affecting 2% to 10% of patient.⁷

In a surgical series, 49% of patients had BAV among 932 patients with AS who underwent surgical aortic valve replacement. Echocardiographic study has shown that the progression of sclerosis starts in the second decade and calcification often increases prominently by 40 years of age.^{15, 20}

In patients undergoing aortic valve replacement (AVR) for AS, over 50% have a BAV. A BAV is the cause of AS requiring AVR in 60% of patients under age 70 years but also accounts for 40% of cases in those over age of 70.²⁵

Conclusion

We found that in the Haldiram Moolchand Cardiovascular Center of P.B.M. Hospital Bikaner, Rajasthan.

Prevalence of Bicuspid Aortic Valve: The prevalence of Bicuspid Aortic Valve in our study population was n=15(2.85%)

Gender: Bicuspid Aortic Valve was more prevalent in males than female. The ratio between male and female is 2:1 which mean males with Bicuspid aortic valve were twice in number as compared to females.

Subtypes: Most common subtype type I(A) left-right coronary subtype was reported.

Complication – Most common complication was aortic stenosis N=3(20%) and Aortic Stenosis with Aortic Regurgitation N=4(26.67%).

It is suggested that a large population based longitudinal study can reveal future detailed information about bicuspid aortic valve morphology subtypes and associated complication in western Rajasthan.

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