Variability in surveillance practice for patients with diagnosis of bicuspid aortic valve syndrome

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Abstract

Background/Aim: In patients with bicuspid aortic valves, guidelines call for regular follow-up to monitor disease progression and guide timely intervention. We aimed to evaluate how closely these recommendations are followed at a tertiary care center. Methods: This was retrospective cohort study at a tertiary care center. Among 48,504 patients who received echocardiograms between 2013-2018, 245 patients were identified to have bicuspid aortic valve. Bivariate analyses compared patient and echocardiographic characteristics between patients who did and did not receive follow-up by a cardiovascular specialist. Results: The mean age of the cohort was 55.2 ? 15.6 years and 30.2% were female. During a median follow-up of 3.5 ? 2.2 years, 72.7% of patients had at least one visit with a cardiovascular specialist after diagnosis of bicuspid aortic valve by echocardiogram. Patients followed by specialists had a higher proportion of follow-up surveillance by echocardiogram (78.7% vs. 34.3%, p< .0001), or by CT or MRI (41.0% vs. 3.0%, p < .0001), and were more likely to undergo valve or aortic surgery compared with patients not followed by specialists. Patients with moderate to severe valvular or aortic pathology (aortic stenosis/regurgitation, dilated ascending aorta) were not more likely to be followed by a cardiovascular specialist or receive follow-up echocardiograms. Conclusions: Follow-up care for patients with bicuspid aortic valve was highly variable, and surveillance imaging was performed sparsely despite guidelines. There is an urgent need for surveillance and clinical follow-up mechanisms to monitor this patient population with increased risk of progressive valvulopathy and aortopathy.

Variability in surveillance practice for patients with diagnosis of bicuspid aortic valve syndrome

Running title: Bicuspid aortic valve surveillance

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Abstract

Background/Aim: In patients with bicuspid aortic valves, guidelines call for regular follow-up to monitor disease progression and guide timely intervention. We aimed to evaluate how closely these recommendations are followed at a tertiary care center.

Methods: This was retrospective cohort study at a tertiary care center. Among 48,504 patients who received echocardiograms between 2013-2018, 245 patients were identified to have bicuspid aortic valve. Bivariate analyses compared patient and echocardiographic characteristics between patients who did and did not receive follow-up by a cardiovascular specialist.

Results: The mean age of the cohort was 55.2 ± 15.6 years and 30.2% were female. During a median follow-up of 3.5 ± 2.2 years, 72.7% of patients had at least one visit with a cardiovascular specialist after diagnosis of bicuspid aortic valve by echocardiogram. Patients followed by specialists had a higher proportion of follow-up surveillance by echocardiogram (78.7% vs. 34.3%, p< .0001), or by CT or MRI (41.0% vs. 3.0%, p < .0001), and were more likely to undergo valve or aortic surgery compared with patients not followed by specialists. Patients with moderate to severe valvular or aortic pathology (aortic stenosis/regurgitation, dilated ascending aorta) were not more likely to be followed by a cardiovascular specialist or receive follow-up echocardiograms.

Conclusions: Follow-up care for patients with bicuspid aortic valve was highly variable, and surveillance imaging was performed sparsely despite guidelines. There is an urgent need for surveillance and clinical follow-up mechanisms to monitor this patient population with increased risk of progressive valvulopathy and aortopathy.

Introduction:

Bicuspid aortic valve (BAV) is the most common congenital heart disease, with a prevalence of 0.5-1-2% and a slight male predominance[1-5]. Many patients with BAV are asymptomatic and often present in adulthood as an incidental finding on echocardiogram. While survival in adult patients with BAV may not differ significantly from that of the general population (potentially due to the efficacy of AVR and similar interventions)[3,6-8], patients with BAV are at an increased risk for various aortic pathologies including aortic stenosis (AS), aortic regurgitation (AR), aortic root dilation, aortic aneurysm, and aortic dissection[1,3,9]. A systematic review of 11,000 patients during 2-12 years of follow-up reported that aortic aneurisms were presents in 20-40% of patients with BAV, though less than 0.5% suffered a dissection[5]. Other studies report that up to 84% of patients with BAV may eventually develop an aortic aneurysm over the course of their lifetime, though less than 5% will have an aortic dissection[10,11]. The risk of various aortic pathologies has prompted guidelines to recommend surveillance of patients with BAV to guide timely intervention, but how closely these recommendations are being followed remains unknown.

The 2018 American Association for Thoracic Surgery (AATS) guidelines for the management of BAV recommend serial evaluations of the aorta by transthoracic echocardiogram (TTE) with intervals tailored to the presence and severity of aortic dilation[9]. The 2020 American College of Cardiology/American Heart Association (ACC/AHA) guidelines suggest lifelong surveillance in patients with aortic dilation [?]4.0cm, and

MRI or CT evaluation of difficult to assess structures[12]. Additionally, surgery is often based on the severity of valvular pathologies[7], for which the American Society of Echocardiography gives specific guidelines to assess and categorize[13]. Overall, all major cardiology/ cardiac surgery societies recommend careful clinical and imaging surveillance in BAV patients.

Although there is consensus on the necessity to carefully follow these patients, it is unknown how well current guidelines are adopted into clinical practice for incidentally detected BAV. Therefore, we aimed to understand the extent of clinical gap in implementation of guidelines-based surveillance for BAV patients at a tertiary care health system.

Materials and Methods:

Patient population

This was a single center retrospective study of adult patients with bicuspid aortic valve diagnosed by inpatient or outpatient transthoracic echocardiogram (TTE) or transesophageal echocardiogram (TEE) during 2013-2018 in a tertiary care center. Yale Institutional Review Board approved this study and individual consent was waived (IRB ID: 2000020356). Among 48,504 unique patients who underwent echocardiogram during that period, 245 adult patients were identified to have BAV by screening for the words "bicuspid aortic valve", "the valve has two cusps", or "bicuspid valve" in the echo report. For patients who were included in this study, the first diagnosis of BAV captured by our system was defined as the first echo report within the time period that mentions BAV. The echocardiogram report was manually reviewed for each patient to confirm the case definition. We recorded cases where the echo report was equivocal for the diagnosis of bicuspid valve and used the term 'possible bicuspid aortic valve' (Figure 1). The final date of follow-up for chart review was January 23, 2020.

Collected data and outcomes

The following patient data were collected: demographics, comorbidity, cardiology follow-up (defined as at least one visit to an outpatient cardiovascular specialist), follow-up imaging studies (echocardiogram, CT, and MRI as recommended by current guidelines), aortic diameter at imaging (dilated aorta was defined as >40mm at the aortic root or ascending aorta), the presence of other valvular pathologies, and whether the patient underwent aortic or aortic valve surgery during the study time period. CT and MRI studies were recorded based on if the indication for the study was to evaluate the valve or aorta. Cardiovascular specialist follow-up was defined as at least one outpatient cardiology or cardiothoracic surgery visit following the initial ECHO. Data collected only reflects what was captured in our health system.

To characterize follow-up patterns, we compared patients who received cardiovascular specialist follow-up to those who did not. In order to characterize how patients were followed based on aortic and valvular pathology, we also compared follow-up patterns between patients with normal versus dilated aortic diameters, as well as between patients with varying levels of aortic stenosis or regurgitation.

Statistical analysis

Chi-squared analysis for categorical variables, and two-tailed t-tests for continuous variables were used to evaluate whether patient and echocardiographic characteristics differed between patients who were diagnosed with bicuspid aortic valve by echocardiogram. P value of <0.05 defined statistical significance. We used GraphPad Prism for analysis (version 8, GraphPad Software, San Diego, CA).

Results:

The mean age of the cohort was 55.2 +- 15.6 years and 30.2% were female. During a median follow-up of 3.5 +- 2.2 years, 72.7% of patients had at least one visit in an outpatient cardiovascular clinic after the initial diagnosis of bicuspid aortic valve by echocardiogram. Patients followed by a cardiovascular specialist had a higher proportion of patients receiving at least one follow-up echocardiogram (78.7% vs. 34.3%, $p< .0001$), or at least one CT or MRI (41.0% vs. 3.0%, $p < .0001$), and were more likely to undergo corrective surgery (39.3% vs 4.5%, $p < .0001$), compared with patients not followed by a cardiovascular specialist (Table I).

Thirty-five percent (N=86) of patients in our study had a previously known BAV (as per their medical records), while the rest were given their diagnoses during the study period. Of patients with previous diagnosis of BAV, 84% (N=72) were followed by a cardiovascular specialist, while 67% (N=106) of new diagnoses were followed. In addition, many of the patients who had echocardiograms that showed a potential, but unclear bicuspid valve never received a follow-up ECHO in order to confirm or deny the diagnosis (17.9%, N=44), or never received a firm diagnosis even after multiple echocardiograms (8.6%, N=21) (Figure 1). Furthermore, follow-up echocardiograms did not always provide both aortic dimensions (aortic root or ascending aorta diameter) and mean valvular gradients (N=109, 67% of the final echocardiograms in the study period) and only 16% (N=40) of patients ever received a report on the orientation of their bicuspid valve (Type 0=2, Type 1=36, Type 2=1).

Among patients who were followed by a cardiovascular specialist, the average duration between echocardiograms was 1.11 +- 0.98 years. Patients with more severe aortic disease were not more likely to receive cardiovascular specialist follow-up than patients without severe disease. Patients with dilated aorta were not more likely to receive specialist follow-up but were more likely to receive a follow-up echocardiogram (74.2% vs. 61.8%, p=0.047), a CT or MRI (44.1% vs. 22.4%, p=0.0003), and surgery (41.9% vs. 21.1%, p=0.0005) than patients with normal aortic diameters. (Table II).

Patients were then stratified by aortic diameter and valve function to assess how these patients were followed up. Patients with moderate to severe aortic valve dysfunction (stenosis and/or regurgitation) were not more likely to have more frequent follow-up than patients with none-mild aortic valve dysfunction. Patients with moderate to severe aortic valve dysfunction were not more likely to be followed by a specialist (76.9% vs. 69.5%, p=0.198) or receive a follow-up echocardiogram (71.2% vs. 63.1%, p=0.188). However, they were more likely to receive a CT or MRI to evaluate the aorta or aortic valve (37.5% vs. 25.5%, p=0.045) and/or undergo surgery (51.9% vs. 12.1%, $p < .0001$). (Table II)

We assessed the impact of clinical follow-up by cardiovascular specialist on timely intervention on the valve and/ or aorta. Overall, 28.9% (N=71) of patients underwent aortic and/ or aortic valve surgery, 47.9% (N=34) of whom had already known about diagnosis prior to their inclusion in our study. Among surgical patients, 95.8% (N=68) of surgical patients were followed by cardiovascular specialist, 88.7% (N=63) received at least one follow-up echocardiogram, and 60.6% (N=43) received a CT or MRI to evaluate the aorta or aortic valve. The most common indications for surgery were ascending aortic aneurysm with or without stenosis or regurgitation (N=25, 34%), aortic stenosis (N=24, 34%), aortic regurgitation (N=10, 14%), and endocarditis with or without stenosis or regurgitation (N=5, 7%). After surgery for BAV syndrome, 85.9% (N=61) received at least one repeat echocardiogram, and 35.2% (N=25) received at least one CT or MRI for the purpose of aortic or valve surveillance. (Table III)

Conclusions:

In this study, follow-up care for patients with diagnosis of bicuspid aortic valve by echocardiogram was highly variable. Current guidelines from the The American Association for Thoracic Surgery state that the interval of follow-up imaging should be based on severity of disease (especially based on aortic dilation)[9], and research has shown that bicuspid aortic valves will often progress in severity as patients age[1,3,8]. Therefore, once patients are diagnosed with BAV (or have a possible BAV found by echocardiogram), guidelines recommend that they should be followed by a cardiovascular specialist (cardiology/cardiac surgery) in order to determine the best schedule for imaging and/or surgical intervention. Our study shows that specialist follow-up and imaging surveillance may vary widely, and we must ensure care for patients with a possible bicuspid aortic valve diagnosis.

In this study, we found that over the mean follow-up of 3.5 years, more than a quarter of patients were never seen by a cardiovascular specialist after bicuspid aortic valve diagnosis by echocardiogram. In addition, a third of patients did not receive a follow-up echocardiogram, and only less than half of the patients who may have benefitted from CT or MRI surveillance according to some guidelines (2018 AATS) received it. Furthermore, many unclear bicuspid diagnoses, such as those labeled as "possible BAV" or "unable to

rule out BAV", did not receive a follow-up echocardiogram and/or a firm diagnosis following the initial echocardiogram. These data beg the question of how the bicuspid valves of the other patients progressed, and whether those patients would have benefitted from earlier intervention or earlier acknowledgement of the potential complications that bicuspid aortic valves can present.

Current guidelines suggest that the frequency and type of surveillance should be based on severity of aortic dilatation. Specifically, the 2018 AATS guidelines push for comprehensive serial evaluation. After the initial evaluation of the valve morphology, these guidelines state that normal aortic diameters should receive echo surveillance every 3-5years, stable aortic dilation (40-49mm) should be evaluated every 2-3 years (after an initial check at 12 months), and more advanced aortic dilation (>50mm) should be imaged yearly. It is also further recommended that aortic dilation >40mm should be investigated by echocardiogram-gated MRI or CT angiography[9]. ACC/AHA guidelines suggest a slightly more flexible pattern of surveillance[14-15], with 2020 ACC/AHA guidelines suggesting MRI/CT for difficult to assess structures, then lifelong surveillance of patients whose aortic diameter [?]4.0cm, with intervals determined by family history and progression rate. Additionally, these guidelines suggest lifelong surveillance after aortic valve replacement if the aortic diameter is [?]4.0cm. These guidelines also suggest considering a screening TTE in the first-degree relatives of patients with BAV[12]. These guidelines state that TTE is usually adequate for hemodynamics and evaluation of anatomy, while TEE can provide improved 2D and 3D images. Cardiac MRI or CT provides better images of the aorta (including the sinotubular junction, sinuses, or ascending aorta) when both of those imaging modalities are not adequate to evaluate valve and aorta morphology.

In our study, patients with aortic dilation >40mm were more likely to receive a follow-up echocardiogram, CT, or MRI, but they were not more likely to have outpatient specialist follow-up. Furthermore, the severity of valvular disease at presentation (aortic stenosis or regurgitation) did not significantly affect clinical follow-up or imaging surveillance patterns. (Table II).

Patients with BAV are at risk for aortic dilation independent of valvular dysfunction, even beginning in childhood[16], and aortic dilation can progress even with normally functioning valves[17,18]. At the same time, valvular dysfunction (especially aortic stenosis) is an independent risk factor for dissection[6]. It would follow that severity of disease should impact the level of outpatient surveillance by cardiovascular specialists so that both patients and providers can be aware of risks and potential complications over time and manage imaging appropriately. Unfortunately, we found that increased severity in general did not seem to lead to increased follow-up by a specialist.

Our study speaks to the stark gap in adoption of guidelines and ensuring optimal implementation in the clinical setting. They also provide a window of opportunity to improve system wide screening and institution of diagnosis triggered alerts to the right clinical practices so BAV patients are provided optimal care. This gap in quality of care attests to the importance of interdisciplinary communication between cardiology, radiology, and cardiac surgery to provide optimal care for patients with bicuspid aortic valve syndrome.

Limitations

This study has the following limitations. Due to the retrospective nature of this study, the availability of information such as previously known diagnoses were limited by explicit documentation in available notes. In addition, this was a single-center study, which limits generalizability and raises the possibility of not capturing outside imaging or follow-up in our analysis, although extensive search was conducted using our electronic medical record system.

Overall, follow-up and use of surveillance imaging of the aorta or the aortic valve may be variable despite awareness of guideline recommendations. There is an urgent need for systematic surveillance and implementation of clinical follow-up mechanisms to monitor this patient population with increased risk of progressive valvulopathy and aortopathy.

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Table I: Demographics of patients who received cardiology follow-up

	All Patients (N=245)	**All Patients (N=2**
Patient characteristics	Not followed by cardiac specialist (n=67)	Not followed by card
	N	*%, [SD]*
Age at presentation, mean	56	[18.4]
Male	48	[19.6]
Race		
White	51	76.1%
African American	9	13.4%
Other	7	10.4%
Follow-up at institution (yrs)	2.24	[2.3]
Studies		
BAV previously known	14	20.9%
Follow-up echocardiogram	23	34.3%
Follow-up CT or cMRI	2	3.0%
Comorbidities		
Hypertension	40	59.7%
Coronary artery disease	10	14.9%
BMI, mean (SD)	28.2	[7.3]
Family history of heart disease	21	31.3%
Conditions at initial presentation		
Aortic stenosis or regurgitation (mod-severe)	24	35.8%
Ascending aorta (diameter, cm) mean	3.71	[0.7]
Aortic root (diameter, cm) mean	3.41	[0.7]
Aortic root [?] 3.5cm	23	34.3%
Ejection Fraction at initial echo, mean	58.2	[10.6]
Underwent surgery (valve, root, and/or ascending aorta)	3	4.5%
Overall mortality	16	23.9%

BAV=bicuspid aortic valve, BMI=body mass index, CT=computerized tomography, echo=echocardiogram, cMRI=cardiac magnetic resonance imaging, SD=standard deviation, yrs=years.

Table II: Follow-up patterns of patients based on initial valve or aortic dysfunction

	All patients (N=245)	**All patients (N=245)**
Patient characteristics	None to mild aortic stenosis or regurgitation	None to mild aortic stenosis or regurgit
	N	*%, [SD]*
	141	57.6%

	All patients (N=245)	**All patients (N=245)**
Age at presentation, mean (SD)	55.2	[15.7]
Male	98	69.5%
Follow-up		
Followed by cardiac specialist	98	69.5%
Follow-up echocardiogram	89	63.1%
Follow-up CT or cMRI	36	25.5%
Underwent surgery	17	12.1%
	Normal aortic diameters at initial echo	Normal aortic diameters at initial echo
	N	*%, [SD]*
	152	62.0%
Follow-up		
Followed by cardiac specialist	107	70.4%
Follow-up echocardiogram	94	61.8%
Follow-up CT or cMRI to evaluate	34	22.4%
Underwent surgery	32	21.1%

BAV=bicuspid aortic valve, CT=computerized tomography, echo=echocardiogram, MRI=cardiac magnetic resonance imaging, Stdev or SD=standard deviation, yrs=years.

Table III: Demographics of patients who underwent surgery

	All patients (N=245)	**All patients (N=245)**
Variables	Received surgery	Received surgery
	N	*%, [SD]*
	71	29.0%
Age at time of surgery, mean (SD)	54.5	[13.1]
Male	51	71.8%
Race		
White	61	85.9%
African American	3	4.2%
Other	7	9.9%
Previously known BAV	34	47.9%
Post-surgery studies		
Follow-up echocardiogram	61	85.9%
Follow-up CT or cMRI	25	35.2%
Type of surgery		
AVR	38	53.5%
Valve and ascending aorta	12	16.9%
Valve and root	6	8.5%
Valve and root and ascending aorta	8	11.3%
Ascending aorta repair	5	7.0%
TAVR	2	2.8%
Indication for surgery		
Aortic stenosis	24	33.8%
Aortic regurgitation	10	14.1%
AS and AR	1	1.4%
Ascending aortic aneurysm	16	22.5%
Ascending aortic aneurysm with AS and/or AR	9	12.7%
Endocarditis	2	2.8%

	All patients (N=245)	All patients (N=245)
Endocarditis with AR and/or AS	3	4.2%
Thoracic aneurysm	1	1.4%
Aortic disease	2	2.8%
No data	3	4.2%
Need for a second surgery	1	1.4%
Overall mortality	2	2.8%

AS=aortic stenosis, AR=aortic regurgitation, AVR=aortic valve replacement, BAV=bicuspid aortic valve, CT=computerized tomography, echo=echocardiogram, cMRI=cardiac magnetic resonance imaging, Stdev or SD=standard deviation, TAVR=transcatheter aortic valve replacement

Figure Legends:

Figure 1: Follow-up patterns in patients diagnosed with bicuspid aortic valve by echocardiogram

Flow-chart of patient cardiology follow-up and echocardiogram studies following initial ECHO (2013-2018). Follow-up was determined if there was at least one recorded appointment with outpatient cardiology or outpatient cardiothoracic surgery following the initial ECHO in this study. Recorded follow-up lasted until January 2020. Unconfirmed BAV diagnosis was determined if the ECHO described the aortic valve as "cannot rule out bicuspid valve", "possible bicuspid valve", or "unclear if bicuspid" and further ECHO studies did not clarify. BAV reversal was determined if a follow-up echocardiogram stated "tricuspid aortic valve".