ORIGINAL ARTICLE

Coronary Artery Anomalies: A New Facet of Bicuspid Aortic Valve?

Anomalías coronarias: ¿Una nueva arista de la válvula aórtica bicúspide?

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ABSTRACT

Background: Bicuspid aortic valve (BAV) is conceived as a valvuloaortopathy, but its association with coronary artery anomalies has not been deeply investigated.
Objective: The aim of this study was to identify the association between coronary artery anomalies and BAV in adult patients using transthoracic echocardiography (TTE) and cardiac computed tomography (CCT) scan compared with patients with tricuspid aortic valve (TAV).
Methods: Consecutive patients with BAV undergoing CCT scan between 2018 and 2019 were analyzed and compared with patients with TAV matched for age and gender. All patients underwent TTE. Aortic diameters, aortic valve calcium score and coronary calcium score were estimated and the presence of fibrosis of the aortic valve was evaluated.
Results: A total of 87 patients were included in the study (42 in the BAV group and 45 controls with TAV). Mean age was 46.8 ± 16.8 years and 74.7% were men. Coronary artery anomalies were identified by CCT scan in 11 patients (26.1%) with BAV and in 1 patient with TAV (2.2%), p = 0.001.
High take-off of the left main coronary artery from the ascending aorta was the most common anomaly (4 cases). Two patients presented interarterial course. High take-off was not associated with greater aortic root diameters.
Conclusions: Coronary artery anomalies were mostly benign and more common in patients with BAV than with TAV. Their correct diagnosis is important to achieve an adequate surgical planning.

Key words: Bicuspid aortic valve – Coronary anomalies- Cardiac computed tomography

RESUMEN

Introducción: La válvula aórtica bicúspide (VAB) es concebida como una válvulo-aortopatía. Su asociación con anomalías coronarias no ha sido estudiada en profundidad.
Objetivo: Reconocer la asociación de anomalías coronarias con VABen pacientes adultos con ecocardiograma transtorácico (ETT)y tomografía cardíaca (TC) en comparación con pacientes con válvula aórtica trivalva (VAT).
Material y métodos: Se incluyeron pacientes con VAB consecutivos a los que se les realizó TC entre 2018 y 2019 y se los comparó con pacientes con VAT de edad y género similares. En todos se realizó también ETT. Se realizaron mediciones de aorta, score de calcio valvular aórtico y coronario, e identificación de fibrosis aórtica.
Resultados: Se incluyeron 87 pacientes (42 del grupo con VAB y 45 controles con VAT), edad media 46,8 ± 16,8 años, un 74,7% hombres. La TC permitió identificar anomalías coronarias en 11 pacientes (26,1%) con VAB y en 1 paciente con VAT (2,2%), p = 0,001. El origen alto del tronco de la coronaria izquierda al nivel de la aorta ascendente fue la anomalía más frecuente (4 casos). Dos pacientes mostraron trayecto interarterial. El origen alto de arterias coronarias no se asoció a mayores diámetros de la raíz aórtica.
Conclusiones: Las anomalías coronarias (en su mayoría benignas) fueron más frecuentes en pacientes con VAB que en VAT. Su correcto diagnóstico tiene importancia a la hora de lograr una correcta planificación quirúrgica.

Palabras clave: Válvula aórtica bicúspide - Anomalías coronarias - Tomografía cardíaca.

Abbreviations

BAV	Bicuspid aortic valve	**RCA**	Right coronary artery
CCT	Cardiac computed tomography	**TTE**	Transthoracic echocardiography
IQR	Interqueartile range	**TAV**	Tricuspid aortic valve
LCA	Left coronary artery	**TAVR**	Transcatheter aortic valve replacement
NC	Noncoronary cusp		

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INTRODUCTION

Bicuspid aortic valve (BAV) is the most common congenital heart defect, affecting 1%-2% of the population. (1) Aortic valve dysfunction due to progressive valve calcification with development of aortic stenosis, and aortopathy, are the most common complications. (2-5) The anomalous origin of the coronary arteries is rare. The association of this defect with BAV has not been emphasized and there has been little information on the possible clinical consequences of this combination of congenital defects. (6) The differences in the distribution of coronary arteries in patients with BAV have been occasionally reported with specific surgical strategies to treat them. (3, 7) However, no systematic data analysis has been performed on the prevalence of coronary artery anomalies in this population and on the frequency of specific variations of coronary artery circulation in patients with BAV compared with tricuspid aortic valve (TAV).

The information about the association of coronary artery anomalies and BAV is controversial. Some reports suggest that BAV may coexist with a coronary artery anomaly. (3, 7, 8) The incidence of left dominance has been reported to be higher in patients with BAV compared with those with TAV. (9) A genetic association between BAV and coronary artery anomalies has not been established. (10) Naito et al. reported a significantly increased prevalence of coronary artery anomalies in BAV patients when compared with their TAV counterparts. (11) In addition, patients with diagnosed coronary artery anomalies had more postoperative ischemic cardiac events. On the other hand, Michalowska et al. reported that the frequency of coronary artery anomalies in BAV patients was similar when compared with their TAV counterparts. (9)

Contrast-enhanced cardiac computed tomography (CCT) scan offers excellent spatial resolution and identifies most anomalies of the coronary arteries in their origin and course, in addition to its superiority over echocardiography for measuring the aorta and determining the calcium score. (9, 12, 13) As many patients with BAV will undergo surgical or transcatheter aortic valve replacement (TAVR), there is a potential risk of coronary artery injury during the procedure. Nowadays, most patients with BAV undergo CCT scan or cardiac magnetic resonance imaging (MRI) before surgery. This multimodality imaging approach can add useful information about aortic dimensions and aortic valve anatomy in relation to the coronary arteries. (2)

In patients with newly diagnosed BAV, it is advisable to perform a three-dimensional imaging technique (MRI or CCT scan) to rule out a significant underestimation of the aortic diameter. (12, 14)

We designed this prospective and comparative study to identify the association between BAV and coronary artery anomalies, compared with TAV.

METHODS

Population

We designed a prospective comparative study of consecutive patients with BAV who underwent transthoracic echocardiography (TTE) and CCT scan to evaluate the aortic valve, coronary arteries, and the aorta. The maximal time interval between both tests was 6 months. The control group included patients with TAV matched for age and sex, who underwent a CCT scan during the same period for evaluation of the pulmonary veins before catheter ablation of arrhythmias.

All patients were being followed up in our Health Network (ICSI, Sanatorio Las Lomas and Centro Médico Nordelta, 2 secondary and tertiary community-based health care centers, respectively).

Patients >18 years who signed the informed consent form were included in the study. Exclusion criteria were: history of cardiac surgery, complex congenital heart defects and genetic aortic diseases (Marfan syndrome, Loeys-Dietz syndrome, etc.), nephropathy (serum creatinine level >150 mmol/L), known hypersensitivity to iodinated contrast media, and untreated hyperthyroidism.

Study protocol and outcomes

The study protocol was approved by the Institutional Ethics Committee, which granted access to the health records and imaging data. The following data were recorded: demographic and anthropometric data, cardiovascular risk factors (hypertension, diabetes mellitus, smoking habits or dyslipidemia), history of cardiac procedures, BAV in first-degree relatives, cardiovascular symptoms (dyspnea, syncope, and angina), cardiac medication, blood pressure, auscultatory findings and echocardiographic parameters. Body surface area was calculated using the DuBois method [BSA (m2) = 0.007184 × height (cm) 0.725 × weight (Kg) 0.425]. (15)

Transthoracic echocardiography

All patients underwent complete transthoracic echocardiography (TTE) using Vivid S5 (GE® Vingmed Ultrasound, Israel or T8; GE® Vingmed Ultrasound, China) ultrasound machines. The diagnosis of BAV was based on a systolic elliptical valvular opening and existence of only 2 commissures delimiting only 2 aortic valve leaflets, (4, 16) In all the patients, BAV was confirmed by CCT scan. The Schaefer classification was used. (16) Aortic stenosis and aortic regurgitation were classified based on standard criteria of current international guidelines. (17, 18)

Aortic valve prolapse was defined when one or more aortic cusps showed backward bowing towards the left ventricle >1 mm. (19, 20) Aortic diameters were measured according to current recommendations. (21) Aortic aneurysm was defined as an aortic diameter ≥45 mm. (4,5) Aortopathy was defined according to the classification of Verma and Siu. (2)

Cardiac computed tomography scan

All patients underwent contrast-enhanced CT scan of the thoracic aorta with a dual-source 128-row system (Somatom Definition, Siemens Healthcare) using a retrospective electrocardiogram-gated protocol. Before the contrast agent was administered, calcium scoring was determined in 3-mm contiguous axial slices between the aortic arch and the diaphragm. (22) A non-ionic iodinated contrast material was injected intravenously, and the bolus-tracking program was used to start the scan.

To evaluate aortic valve morphology, reconstructions were created from 0 to 90 % (10% to 40% for the systolic

window and 60% to 80% for the diastolic phase). All the images were post-processed and analyzed using syngo.via (Siemens®) imaging software. (9)

Definition of coronary artery anomalies

Coronary artery anomalies were defined using the definition by Naito et al. (11)

- Separate ostium: Absence of the left main coronary artery and separate ostia of the left anterior descending artery and the left circumflex artery.
- Anomalous origin: Origin of the right coronary artery (RCA) from the left coronary sinus or the left coronary artery (LCA) from the right coronary artery sinus, as well as anomalous coronary ostium location outside the sinus of Valsalva, including the left ventricle, ascending aorta (high take-off) and aortic arch.
- Anomalous course: When the course of the coronary artery is different from the normal course, particularly if this is an interarterial course (between the aorta and the pulmonary artery).

Statistical analysis

Continuous variables are expressed as mean and standard deviation, or median and interquartile range (IQR), according to their distribution. Discrete variables are expressed as frequencies and percentages. Continuous variables were compared using a two-tailed Student's t test or the Mann-Whitney U test, as applicable. Categorical variables were compared using the chi-square test with Yates's correction or Fisher's exact test, as applicable according to the distribution and number of patients.

The correlations between continuous variables were evaluated using Pearson's r or Spearman's rho correlation coefficient. A p value < 0.05 was considered statistically significant.

All the statistical calculations were performed using R Project software package under Windows operating system.

Ethical considerations

Patient identity was preserved ensuring privacy and confidentiality of the information. The study protocol was approved by the institutional Teaching and Research Committee. All the patients signed an informed consent form. The study was conducted following the recommendations of the Declaration of Helsinki and Good Clinical Practice Guidelines.

RESULTS

Population

Between January 2018 and November 2019, 87 patients with mean age 46.8 ± 16.8 years and 74.7% men were included in the study. Forty-two patients had BAV (Figure 1) and 45 belonged to the control group (TAV). The demographic data of both groups (BAV and TAV) are shown in Table 1.

The mean time interval between the CCT scan and TTE was 4 ± 2.6 months. In the BAV group, 7 (16.7%) patients presented significant aortic stenosis and 18 (42.8%) had some degree of aortic regurgitation. There were no differences in the severity of valvular dysfunction between men and women.

Coronary artery anomalies

Coronary artery anomalies were identified by CCT scan in 11 patients (26.1%) with BAV and in 1 patient with TAV (2.2%), p = 0.001. The distribution of the type of coronary artery anomalies are described in Figure 2. Anomalous origin of the LCA from the ascending aorta (high take-off of the LCA) was the most common anomaly, observed in 4 patients (9.5%) (Figure 3). Three patients (7.1%) presented anomalous origin of the RCA ostium arising from the ascending aorta (high take-off of the RCA) (Figure 4). Two patients (4.8%) had anomalous origin and course, with ectopic RCA originating from the left sinus of Valsalva as a separate vessel with interarterial course. The anatomic characteristics of all the coronary artery anomalies diagnosed are shown in Table 2 and Figures 3 and 4.

We did not find a significant association between BAV phenotype and coronary artery anomalies. To rule out that the high take-off could be due to an aneurysm or aortic root dilation, the aortic root diameters were compared in patients with and without high take-off. There were no differences in the aortic root diameter (36.1 mm ± 4.6 mm in the group of patients with high take-off vs. 35.9 mm ± 4.4 mm in the rest of the patients, p = 0.58). Only one patient with high take-off of coronary arteries had aortic root diameter indexed to body surface area >21 mm/m^2.

Right coronary dominance was observed in 24 (80.9%) patients in the BAV group and in 36 (80%) in the TAV group, p = 0.5. In the BAV group, left dominance occurred in 7 (16.7%) patients and 1 patient (2.3%) presented codominance. There were no differences in coronary dominance according to the BAV phenotype (right dominance 80.7% en type I vs. 85.1%

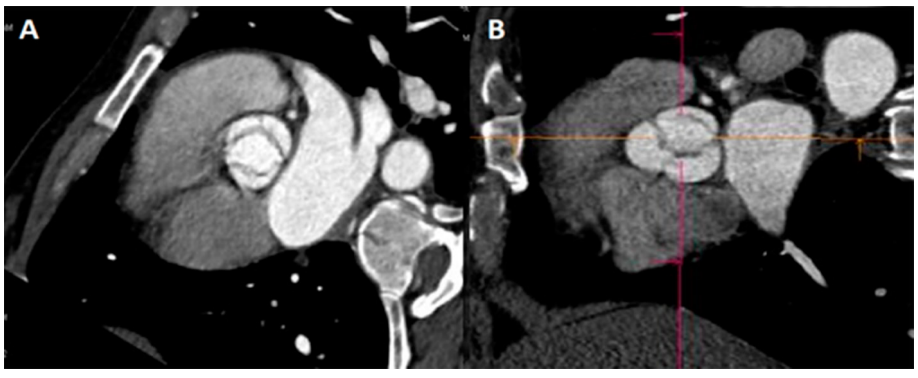

Fig. 1. Cross sectional computed tomography reconstruction through the aortic valve plane showing the elliptical opening of 2 bicuspid aortic valves (BAV). **A:** BAV with thick free edges and fusion of the right and left coronary cusps with raphe; **B:** BAV with fibrosis and thickened free edges corresponding to fusion of the right coronary cusp and noncoronary cusp with raphe

	BAV n = 42	TAV n = 45	p
Age (years)	43.9 ± 15.1	51.5 ± 17.1	0.07
Men (%)	30 (71.4%)	35 (77.7%)	0.6
BSA Dubois (m^2)	1.93 ± 0.25	1.96 ± 0.27	0.1
LVEF (%)	64.2 ± 5.9	63.2 ± 6.3	0.1
LA volume index (mL/m^2)	27.4 ± 5.2	26.9 ± 5.9	0.07
Age of BAV diagnosis (years)	35.7 ± 8.1		
Without CV risk factors, n (%)	28 (66.6%)	28 (62.2%)	0.8
Hypertension, n (%)	7 (16.6%)	10 (22.2%)	0.59
Smoking habits, n (%)	7 (16.6%)	11 (24.4%)	0.43
Dyslipidemia, n (%)	6 (14.2%)	10 (22.2%)	0.41
DLP (radiation)	776 ± 106	798 ± 129	0.08
Aortic valve calcium score (AU)	0 (0-457)	0 (0-68)	0.5
Coronary artery calcium score (AU)	0 (0-0)	0 (0-82)	0.07
Aortic diameter >21 mm/m^2	15 (35.7%)	4 (8.8%)	0.003
Aortic aneurysm (>45mm)	4 (9.5%)	0 (0%)	0.05

BAV: bicuspid aortic valve; TAV: tricuspid aortic valve; BSA; body surface area; LVEF: left ventricular ejection fraction; LA: Left atrial; CV: cardiovascular; DLP: dose-length product; AU: Agatston units

Table 1. Baseline population characteristics

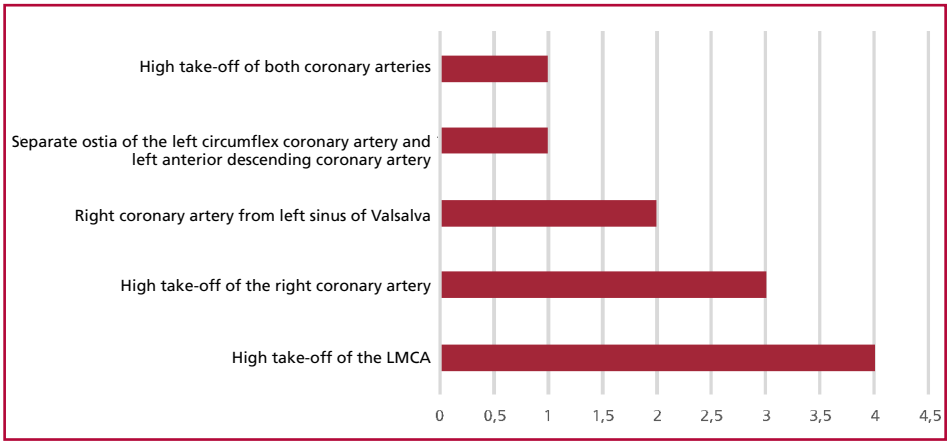

Fig. 2. Frequency (in absolute numbers) of coronary artery anomalies detected by computed tomography scan. High take-off of the left main coronary artery (LMCA) was the most common coronary anomaly detected

in type II, p = NS). Left dominance was observed in 3 patients with coronary artery anomalies (27.2%) but was not statistically more common than in patients without coronary artery anomalies.

The surgical strategy was modified in 2 patients with coronary artery anomalies. One patient with symptomatic severe aortic regurgitation and anomalous origin and course of the RCA underwent aortic valve replacement with ligation of the proximal RCA and bypass graft from the right internal thoracic artery to the RCA. Selective angiography of the RCA was not possible in another patient with dyspnea and severe aortic regurgitation with high take-off of the RCA. Computed tomography angiography helped guide and plan the strategy, thus avoiding the section of the RCA and reinsertion of the coronary ostium.

Additional information provided by CCT scan

In the BAV group, the aortic valve calcium score was 0 AU in 25 patients (59.5%). As expected, this score significantly correlated with age (r = 0.47, p = 0.0002). Mean aortic valve thickness was greater in patients with BAV (0.25 ± 0.05 cm vs. 0.11 ± 0.03 cm, p < 0.001).

Aortic dilation

As expected, aortic dilatation was more common in patients with BAV (35.7% vs. 8.8%, p = 0.003). The phenotypes of aortic dilation were mixed dilation (involving the ascending tubular aorta and the aortic root) in 8 patients (19%), isolated aortic root dilation in 5 patients (11.9%) and isolated ascending tubular aorta dilation in 2 patients (4.7%). Four patients presented aneurysm of the ascending aorta (diameter >45 mm), more common in BAV with raphe (p = 0.006). There were no significant differences in the prevalence or types of aortic dilation between BAV phenotypes (fusion of the right and left coronary cusps or fusion of the right coronary and noncoronary cusps).

DISCUSSION

Our study showed a high prevalence of coronary artery anomalies in patients with BAV compared with patients with TAV matched for sex and age. Most cas-

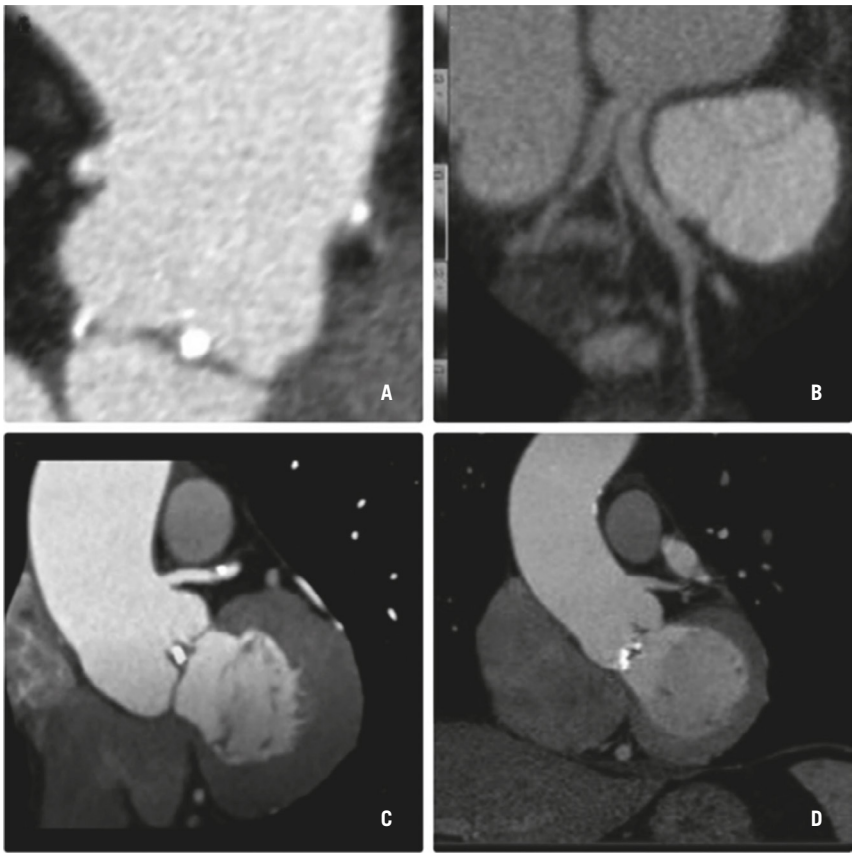

Fig. 3. Coronary artery anomalies detected by computed tomography scan in patients with bicuspid aortic valve. From left to right, top to bottom: **A:** High take-off of the left main coronary artery and right coronary artery above the sinotubular junction. **B:** Separate origin of the left anterior descending coronary artery and left circumflex coronary artery. **C** and **D:** High take-off of the left main coronary artery in patients with calcification of the bicuspid aortic valve.

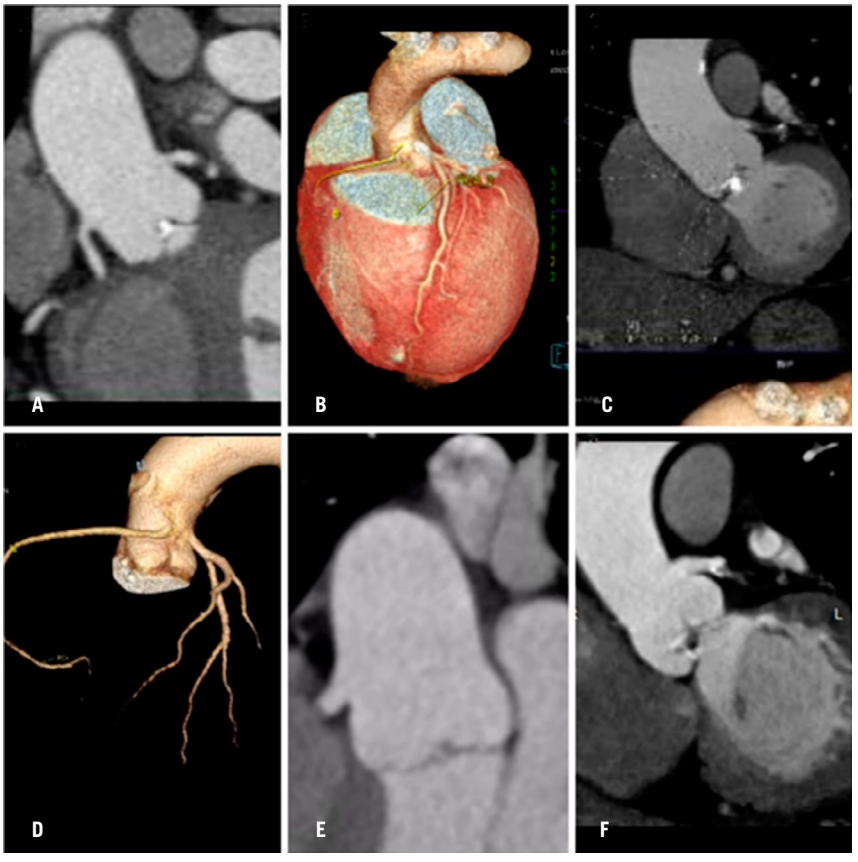

Fig. 4. Coronary artery anomalies detected by CT in patients with BAV. From left to right, top to bottom: **A:** High take-off of the right coronary artery and left main coronary artery with calcified bicuspid aortic valve. **B:** Three-dimensional reconstruction of the aorta and coronary arteries showing a right coronary artery emerging anteromedial to the left coronary sinus as an independent vessel with interarterial course. **C:** High and angular take-off of the left main coronary artery. **D:** Reconstruction with volume rendering showing a right coronary artery emerging from the left sinus of Valsalva with interarterial course. **E:** High take-off of the right coronary artery. **F:** High take-off of the left main coronary artery. Abbreviations: CT: Computed tomography; BAV: bicuspid aortic valve.

Table 2. Coronary artery anomalies identified in 12 patients (11 with BAV and 1 with TAV)

	Age	Sex	Type of BAV	Aortic stenosis (Mod/S)	Aortic regurgitation (Mod/S	Coronary artery calcium score (AU)	Aortic valve calcium score (AU)	Aortic valve thickness (cm)	Sinuses of Valsalva (mm)	Ascending aorta (mm)	Coarctation of the aorta	Raphe	Coronary artery anomaly	Coronary dominance
#1	60	F	I (R-L)	Yes	No	0	2062	0.29	35	38.7	No	Yes	Separate ostium LAD and CX	Left
#2	65	M	I (R-L)	Yes	No	124	2016	0.26	38.3	39.5	No	Yes	High take-off of the LMCA	Right
#3	27	M	I (R-L)	No	Yes	0.6	0	0.23	36.6	33	No	Yes	High take-off of RCA	Right
#4	75	M	II (R-NC)	No	Yes	0	2145	0.27	31.2	32.7	No	Yes	High take-off of the LMCA	Left
#5	55	M	I (R-L)	No	No	0	48	0.29	40.4	40	Yes	Yes	High take-off of RCA	Right
#6	48	F	I (R-L)	No	Yes	0	3.5	0.25	32.9	43.1	No	Yes	High take-off of LCA	Right
#7	20	M	I (R-L)	No	No	0	0	0.17	29.6	25	No	Yes	Origin and course: RCA originating from the left sinus of Valsalva with interarterial course	Right
#8	42	M	I (R-L)	No	No	0	259.6	0.30	31.8	34.2	No	Yes	High take-off of LMCA and RCA	Right
#9	34	M	I (R-L)	Yes	Yes	0	3097	0.29	34.5	37.5	No	Yes	Origin and course: RCA originating from the left sinus of Valsalva with interarterial course	Codominance
#10	49	F	I (R-L)	No	Yes	0	0	0.24	30.6	33.1	No	Yes	High take-off of RCA	Left
#11	68	M	II (R-NC)	Yes	No	605	4353	0.32	41.3	39.6	No	Yes	High take-off of the LMCA	Right
#12	48	M	TAV	No	No	0	0	0.11	34.7	33.1	No	-	Separate ostium LAD and CX	Right

Abbreviations: CX: circumflex; LAD: left anterior descending, RCA: Right Coronary Artery; LMCA: Left main coronary artery; R-L: fusion of the right and left coronary cusps; R-NC: fusion of the right coronary cusp and noncoronary cusp; F: female; M: male; Mod: moderate; S: severe; AU: Agatston units; BAV: bicuspid aortic valve; TAV: tricuspid aortic valve.

es were benign anomalies, with relevance for planning procedures, but with no significant clinical impact. Furthermore, aortic valve fibrosis was prevalent in patients with BAV, even before significant valve dysfunction developed.

Bicuspid aortic valves are the result of abnormal aortic cusp formation during valvulogenesis and of a complex developmental process rather than the simple fusion of 2 normal cusps. (3) Thus, heart defects often affect not only the valve, but also the aorta and eventually the coronary arteries.

A coronary anomaly should be defined as any coronary pattern with a feature "rarely" encountered in the general population. When these anomalies occur, they are usually an ectopic origin of the right or left coronary artery from the aorta. Most coronary artery anomalies are benign and, accordingly, are only identified as incidental findings during diagnostic tests for other reasons. Coronary artery anomalies can rarely lead to poor myocardial perfusion with devastating clinical consequences. According to the literature, they can be found incidentally in 1% of the general population. This percentage is obtained from coronary angiographies performed for suspected coronary artery disease. (23) The incidence varies and depends on the methods used for detection (autopsy, surgery, coronary angiography, CCT scan) and ranges from 0.7% to 6.6% in CT coronary angiography. (24) Angelini et al. evaluated 1950 coronary angiographies and found an incidence of coronary artery anomalies in 5.6% of cases. (23)

There is little information about the association between coronary artery anomalies and BAV. In our population, coronary artery anomalies were significantly more common than in the normal population. Our results are consistent with those reported by Naito et al. (11) Most cases showed benign anomalies, as high take-off and rotation. However, Michalowska et al. have reported a prevalence of coronary artery anomalies similar to that of the general population. (9) High take-off and separate ostia of the coronary arteries should be considered when aortic valve replacement, aortic surgery or TAVR is required in patients with BAV, for adequate planning of the procedure. This frequent association could suggest that this valvuloaortopathy could also affect the origin of the coronary arteries.

Naito et al. reported that patients with BAV and coronary artery anomalies had more postoperative ischemic cardiac events after aortic valve surgery. (11) Nowadays, as most patients with BAV will undergo CCT scan or MRI before surgery, this multimodality imaging approach could help to identify coronary artery anomalies and surgery or TAVR planning.

In our study population, the incidence of right dominance and left dominance was 80.9% and 16.7%, respectively. These results are comparable to those reported by Michalowska et al. (9) Nevertheless, some authors have found left dominance in 57% of patients with BAV.

Despite the combined multimodality imaging approach represents the standard of care for patients with BAV in developed countries, in less privileged settings, such as our country, not all patients with BAV are evaluated with CCT scanning. Besides providing the evaluation of the coronary arteries, CCT scan allows the examination of the aorta in segments where TTE has limitations, such as the distal tubular portion. In our study, the diagnosis of aortic dilation was made by CCT scan in 3 cases that were not identified by TTE. Since CCT scan involves radiation and requires infusion of iodinated contrast, these aspects should be considered.

Although our study has several limitations, it provides evidence that not only the aortic valve and aorta may be involved in patients with BAV, but also the coronary arteries. This association could correspond to specific genetic abnormalities and opens an interesting path for further studies, including genetic tests. In addition to coronary anatomy, CCT can provide useful information about leaflet fibrosis with aortic valve calcium score, valve thickness, and aortic valve anatomy (valvular phenotype). (22)

Study limitations

Some limitations of this study should be mentioned. Firstly, the number of patients included. Secondly, the time interval between TTE and CCT scan was 4 months and, ideally, both studies should be performed within a shorter interval; however, for the purpose of the diagnosis of coronary artery anomalies, this interval does not affect the result. Thirdly, CCT scan involves exposure to radiation that should be considered. Finally, the "control" population was made up of patients undergoing CCT scanning for some underlying cardiac condition that required the imaging test; thus. it is difficult to find a completely normal population to constitute the control group and compare the findings.

CONCLUSIONS

In our study, coronary artery anomalies were more common in patients with BAV than with TAV. Although in most cases these anomalies were benign, their correct diagnosis is important achieve an adequate surgical planning. Moreover, CCT adds complementary information to echocardiography not only for the evaluation of the coronary arteries, but also in relation to valvular fibrosis/calcification and aortic diameters. Cardiac computed tomography scan is recommended for patients with BAV, although radiation and the need for contrast agent should be considered.

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Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/ Supplementary material)

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