Congenital Heart Disease

Tech Science Press

DOI: 10.32604/CHD.2021.013180

ARTICLE

High-Risk Congenital Coronary Abnormalities in Patients with Bicuspid Aortic Valve

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Received: 28 July 2020 Accepted: 10 October 2020

ABSTRACT

Objective: Abnormal coronary artery origin (ACAO) from the opposite sinus with inter-arterial course of the ectopic proximal vessel is associated with the greatest potential for clinical manifestations, specifically sudden death. Data remain limited regarding the association between bicuspid aortic valve (BAV) and this potentially dangerous coronary variant reported in up to 0.6% in the general population. We investigated the frequency of this high-risk ACAO with inter-arterial course in our surgical series of BAV patients. **Methods and Results:** We conducted a retrospective study to identify BAV patients with ACAO and inter-arterial course who underwent elective aortic valve/root surgery between 2010 and 2019 in our tertiary center. A total of 279 consecutive patients with BAV were identified. Among these, four patients (1.4%) had ACAO with inter-arterial course. Three patients had abnormal right coronary artery arising from the theoretical opposite coronary sinus with intramural course. The fourth patient presented an abnormal left coronary artery from a single coronary artery arising from the theoretical right coronary sinus with a long inter-arterial course. **Conclusion:** Our results from a single-center experience support high-risk ACAOS may be more frequently diagnosed in BAV patients, especially in surgical series, justifying a careful preoperative assessment.

KEYWORDS

Congenital heart disease; coronary artery; bicuspid aortic valve

1 Introduction

Bicuspid aortic valve (BAV) is the most common congenital heart defect, with a prevalence estimated between 0.5% and 2% [1]. Alterations in coronary anatomy and distribution have been described in patients with BAV including higher incidence of separate left coronary ostia or left dominant system [2], as well as abnormal coronary artery origin (ACAO) [3]. Among the group of congenital coronary anomalies, ACAOs with an inter-arterial course of the ectopic proximal vessel represent a subgroup associated with the greatest potential for clinical consequences, specifically, sudden death in teenagers and young adults [4]. The prevalence of this high-risk ACAO in the BAV population is unknown. We aimed to investigate the frequency of this potentially dangerous coronary variant in our surgical series of BAV patients.

2 Methods

We retrospectively identified the consecutive patients who were referred to the cardiac surgery unit of the tertiary Caen University Hospital Center between June 2010 and June 2019 for the following interventions: (1) Aortic valve replacement or plasty, (2) Aortic root replacement. After reviewing the medical and surgical reports of the patients, we identified those with BAV associated with a high-risk ACAO defined by an inter-arterial course of the proximal ectopic vessel. Anomalous origin of the coronary artery was defined as an anomalous offspring of either the right coronary artery from the theoretical position of the left coronary sinus or the left coronary artery from the theoretical position of the right coronary sinus. The ACAO had to be associated with an inter-arterial course pre-operatively identified by computed tomography (CT) scan and confirmed by the intra operative surgeon's description. BAV type was defined according to the classification of Sievers [5]. The final decision on the BAV diagnosis and type was made based on the intraoperative description by the surgeon. The study protocol was approved by our ethical committee (reference code: 915619) and informed consent was obtained.

3 Results

During the study period, 1652 patients underwent aortic valve replacement, plasty or aortic root replacement in our center. Among them, 1373 had tricuspid aortic valve and 279 (17%) had BAV. Among BAV patients, four patients (1.4%) were identified with high-risk ACAO (Fig. 1). Besides ACAO, absence of left main coronary artery with separate origins of the left artery descending and the left circumflex artery was observed in 3 cases (1%) and origin of the left circumflex artery from the proximal part of the RCA was found in 2 (0.7%) patients with BAV. Among patients with high-risk ACAO, 3 had abnormal right coronary artery (ARCA) arising from the theoretical left coronary sinus with intramural course. Among them, one had a type 1 R-L BAV (Figure, patient 1), a second had a type 1 L-NC BAV and the third had a type 2 BAV. The fourth patient presented an abnormal left coronary artery (ALCA) from a single coronary artery arising from the theoretical right coronary sinus with a long inter-arterial course, associated with a type 1 R-L BAV (Figure, patient 4). The 3 patients with ARCA (patients 1, 2 and 3) presented dyspnea attributed to their aortic valve disease. None of them had history of arrhythmia, chest pain neither syncope. The fourth patient with ALCA was free of symptoms even during strenuous exercise. ACAO was firstly identified by trans thoracic echocardiogram for Patients 1, 3 and 4. No patients with ACAO had an adverse surgical event. In the 3 patients with ARCA, reimplantation of the RCA was performed. ARCA was dissected at the takeoff from the intra mural course, and then translocated and reimplanted into the appropriate right sinus of Valsalva (for patients 1 and 2), or in the conduit graft (for patient 3). Characteristics of these four patients are detailed in Tab. 1.

4 Discussion

Congenital coronary artery anomalies are a heterogenous group of congenital disorders whose pathophysiological mechanisms and clinical manifestations are highly variable. Although many congenital coronary anomalies have a benign outcome and do not require specific treatment, some of them, including ACAO, can have dramatic complications [6]. We focused on ACAO associated with inter-arterial course as this anatomical feature, so called 'malignant variant' [4], has been associated with an increased risk for myocardial ischemia, ventricular arrhythmias, and sudden cardiac death during strenuous exertion [7]. Prevalence estimates of this high-risk ACAO in the general population range from 0.1% to 0.6% [8]. In a coronary CT angiographic study 5,634 adults were evaluated, of these, 36 patients (prevalence: 0.64%) had an ACAO with an inter-arterial course [9]. A recent study screened 5,169 adolescents with cardiac MRI and showed a prevalence of high-risk ACAO of 0.44% [10]. It should be noted that these two studies did not mention the presence of BAV in their ACAO population. In a study from our center, after the trans-thoracic echocardiographic screening of 3,227 patients, we found 9 patients with a malignant form of ACAO (prevalence 0.27%), including 2 patients with BAV

[11]. The presence of BAV was also reported in other previous case series of ACAO with inter-arterial course [12] and several case reports [13].

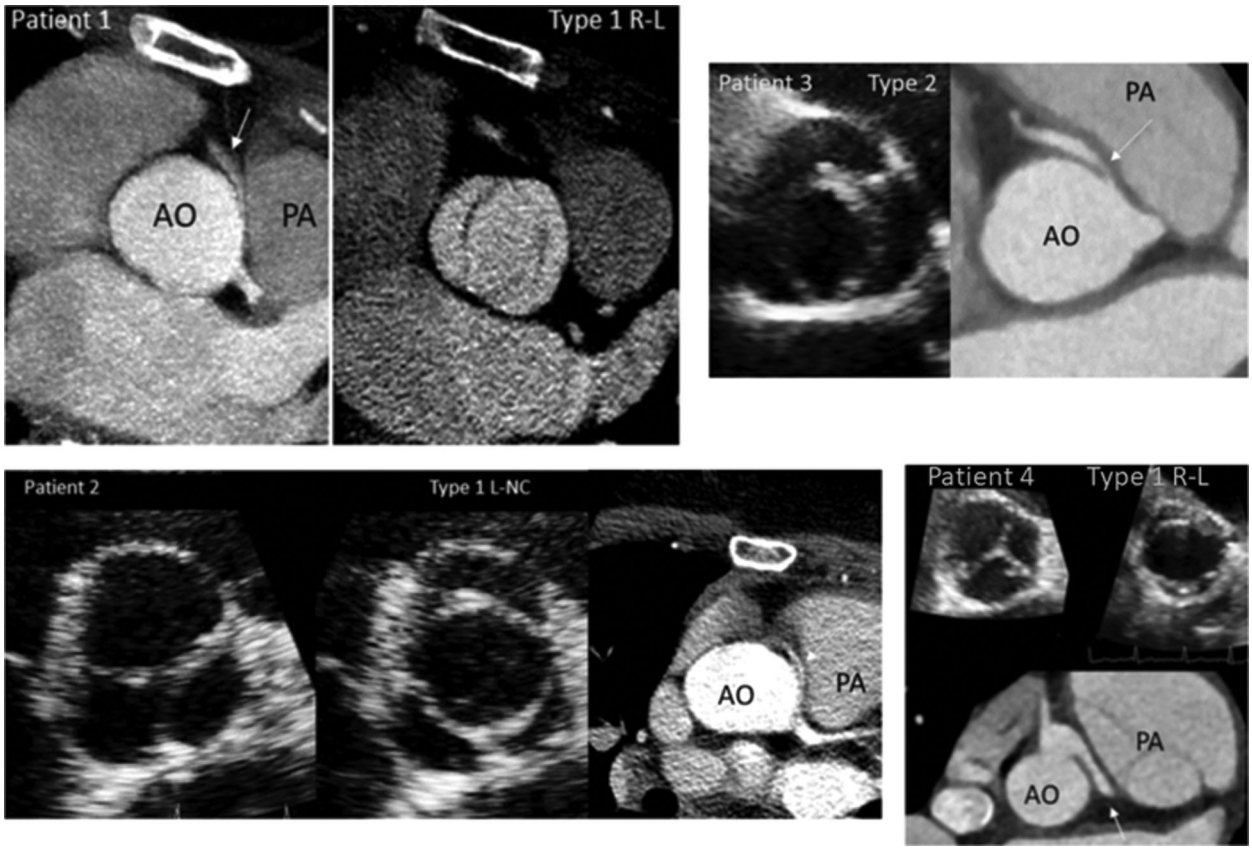

Figure 1: Abnormal coronary artery origin with inter arterial course is indicated by the white arrow. PA: Pulmonary artery; AO: Aorta; R: Right; L: Left; NC: Non-Coronary; *Type of bicuspid aortic valve was defined according to the classification by Sievers

In our surgical series of BAV patients, high-risk ACAOs were found in 1.4% of patients. Our findings suggest that high-risk ACAO may be more frequent in the BAV population compared to general population. Recently, Michałowska et al. [14] performed a multislice CT scan study in BAV patients and found a significantly higher prevalence of separate left anterior descending coronary artery/circumflex ostia in BAV patients, when compared to tricuspid aortic valve patients. This study also reported shorter left main coronary artery and a higher prevalence of left coronary dominance. Koenraadt et al. [2] reported the same findings further suggesting that peculiar anatomy of left coronary artery could be influenced by the type of BAV. Although well described, these anatomical studies only focused on benign forms of congenital coronary anomaly that were without clinical significance. Previous report regarding the association between ACAO and BAV did not specify if the anomalous vessel had a benign or malignant course [15]. Naito et al. [15] found 21 patients with anomalous origin of the right (n = 15) and left (n = 6) coronary artery in a surgical population of 345 BAV patients (prevalence: 6%). The Authors did not specify if the anomalous/ectopic vessel had an inter arterial course, which may explain the unexpected high prevalence of ACAO in their BAV population.

Table 1: Patient characteristics, anatomic diagnoses, and surgical technique

Patients	Age	Gender	Type BAV*	Coronary anomaly	Associated heart lesion	Operative indication	Surgical technique
1	44	M	1 R-L	ARCA with intramural course	None	Aortic regurgitation	Aortic valve replacement with RCA reimplantation
2	36	F	1 L-NC	ARCA with intramural course	None	Aortic regurgitation	Aortic valve plasty with RCA reimplantation
3	38	M	2	ARCA with intramural course	Aortic aneurysm	Aortic stenosis	Bentall with RCA reimplantation
4	21	F	1 R-L	ALCA from a single coronary artery arising from the 'right coronary sinus' with inter arterial course	Aortic coarctation/ VSD	Endocarditis, ALCA	Unroofing procedure, VSD closure

ALCA: Abnormal left coronary artery; ARCA: abnormal right coronary artery; BAV: bicuspid aortic valve; VSD: ventricular septal defect; RCA: right coronary artery; R: right; L: Left; NC: non-coronary. *Type of bicuspid aortic valve was defined according to the classification by Sievers.

Even though much remains unknown about the coronary artery embryonic development, anomalous behaviors of cells from both the cardiac neural crest [16] and second heart field were postulated as the common underlying morphogenetic defect for ACAO in animal models. In humans, defects of neural crest cells have been suggested to produce BAV [17], and also play a key role in the organization, so called "patterning", of the coronary artery [18]. In addition, the second heart field is important in defining the connections of the embryonic coronary stems to the sinuses of Valsalva [19].

In BAV patients, ACAO with malignant course should be actively searched, especially during transthoracic echocardiograms, which remain as the first line of examination for the diagnosis and follow-up of these patients [11]. ACAO are associated with an increased risk of coronary complication and myocardial ischemia during surgical treatments on the aortic valve and/or aortic root (as Ross procedure for example) [20]. Many of BAV patients will be potential candidates for aortic valve/root interventions, including trans-aortic valve implantation procedure, throughout their life. In addition to preventing potentially life-threatening complications caused by compression of the abnormal vessel during exercise, an appropriate definition of the coronary anatomy may allow for the development of better treatment strategies and would prevent peri-procedural complications

5 Limitations

Our study has some limitations. As we report a surgical series, we are aware we cannot extrapolate our findings to the entire BAV population. The inter-arterial course between the great vessels may be erroneously suspected by TTE or CT scan, in particular when the course of the ectopic vessel is close to the edge of the anterior commissure. For this reason, we focused on a surgical series for which BAV with high-risk ACAOs were confirmed by surgical exploration. Although, three different BAV types were associated with high-risk ACAO in our patients, furthers studies are needed to precise the potential association between the cusp-fusion type and this high-risk coronary anomaly.

6 Conclusion

Our results from a surgical series suggest high-risk ACAO may be more frequently diagnosed in patients with BAV compared to general population and should always be considered when diagnosing this frequent congenital cardiac defect.

Author Contributions: Félix Collard: Data collection, data interpretation, statistics, drafting article, approval of article. Pascale Maragnes: data collection, data analysis, approval of article. Dimitrios Buklas: critical revision and approval of article. Fabien Labombarda: concept, data interpretation, critical revision and approval of article.

Availability of Data and Materials: Data are available on request to corresponding author.

Funding Statement: The author(s) received no specific funding for this study.

Conflicts of Interest: The authors declare that they have no conflicts of interest to report regarding the present study.

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