

Our Experience of Surgical Aortic Reconstruction in Patient with Bicuspid Aortic Valve in Japan

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ABSTRACT

OBJECTIVES: We present the classification of bicuspid aortic valve (BAV) and morphometric analysis of the ascending aorta in patients with BAV.

METHODS: From 2003 to 2018, 102 patients underwent aortic valve replacement for BAV in our institution. Surgical treatment for aorta reconstruction was performed in 39 patients. All patients were evaluated by Sievers classification of the BAV and Fazel Cluster classification of the aortic form.

RESULTS: Sievers classification of the BAV showed 8 patients for type 0, 30 cases for type 1, and 1 case for type 2. Additionally, type 0 subtype included one patient of ap-type and seven patients of lat-type. In type 1, there are 18 patients as LR fusion type, 9 patients as RN fusion type, and 3 patients as LN fusion type. The mean diameter

of the ascending aorta of 39 BAV patients who underwent aortic surgery was 47.8 ± 6.1 mm. Fazel Cluster classification showed eight patients in type 1, 28 patients in type 2 and 3, and three patients in type 4.

CONCLUSION: The frequency of Sievers classification was the same as that reported previously. All BAV types may be associated with a risk of the enlargement of ascending and arch aorta.

Key words: Aortic Dilatation; Bicuspid Aortic Valve

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INTRODUCTION

Bicuspid aortic valve (BAV) is a connective tissue disease that can be diagnosed at any stage during a lifetime and affects 0.5 to 2.0% of the population^[1-5]. BAV is thought to be associated with the dilatation of the aortic root and ascending aorta, aortic valve dysfunction, and aortic coarctation^[6-8]. Problematically, the guidelines for the concomitant surgical treatments for BAV patients with ascending aorta dilatation change yearly^[9,10]. In our institution, prophylactic aorta surgical reconstruction recommended by our recent guideline (JCS 2012) was performed when the ascending aortic diameter was ≥ 45 mm and if concomitant aortic valve replacement (AVR) is performed^[11]. Nonetheless, given that the blood vessels can be vulnerable, ascending and arch replacement is performed when diameter is less than 45mm. Here, we demonstrate a method of surgical management for the enlargement of ascending aorta in patients with BAV through the classification of BAV and morphometric analysis of the ascending aorta.

PATIENTS AND METHODS

Study population and surgical procedure

In total, 693 patients underwent AVR between January 2003 and April 2018. Out of the 693 patients, 102 patients were diagnosed with BAV. Additionally, 39 of the BAV patients underwent a concomitant surgical operation of the aorta. We retrospectively reviewed and analyzed the clinical information of these 39 patients. All patients gave informed consent, and this study protocol was approved by the ethics committee of Teikyo University. In this cohort, 24 patients were male and 15 patients were female, with an average age of 63.9 ± 13.8 years (range: 29-82 years) (Table 1). The average diameter of the ascending aorta was 47.8 ± 6.1 mm. 37 patients had symmetric form of the ascending aorta, while 2 had asymmetric form. Sievers classification and Fazel Cluster classification (FC type) were applied to classify the type of BAV (Figure 1A) and the morphological type of aorta (Figure 1B)^[12].

The operation was performed under general anesthesia. The typical surgical procedure used is described as follows. All procedures were performed using a total median sternotomy. Standard cardiopulmonary bypass techniques were used including cannulation of the ascending aorta and right atrium. The target blood and rectal temperature were 28°C. Circulatory arrest (CA) with retrograde and antegrade cerebral perfusion were performed on patients requiring ascending aorta replacement and aortic arch replacement, respectively. To patients who required aortic root replacement, aortic cross-clamp under mild hypothermia without CA was used.

RESULTS

Sievers classification showed 8 patients in type 0 (20.5%), 30

patients in type 1 (76.9%), and 1 patient in type 2 (2.6%). Out of the eight patients with type 0, there was one patient for ap-type (12.5%) and seven patients for lat-type (87.5%). The thirty patients in type 1 consisted of eight patients for LR fusion type (60.0%), nine patients for RN fusion type (30.0%), and three patients for LN fusion type (10.0%). Fazel Cluster classification showed 8 FC type I (20.5%), 28 FC type II and III (71.8%), and 3 FC type IV (7.7%) (Table 1). The concomitant aortic procedure included ascending aorta replacement in 28 patients (71.8%), ascending aortic arch replacement in 1 patient (2.6%), aortic root replacement in 8 patients (20.5%) and ascending aorta and aortic root replacement in 2 patients (5.1%). Each rate of valve fusion classified by Sievers classification was shown in Table 2. In all types of Sievers classification, FC type 2 and 3 was the most frequent. In-hospital mortality was observed in one patient with multiple organ failure. Additionally, one patient died due to cancer and another due to the rupture of descending aorta.

DISCUSSION

BAV is at risk for the dilatation of aortic root and ascending aorta, aortic valve dysfunction, and aortic coarctation^[6-8], suggesting that there is a great need for further discussion on the validity of concomitant aortic reconstruction with AVR. Generally, AVR alone is thought to restrain the rate of aortic dilatation^[13]; however, recent studies showed that AVR did not necessarily eradicate the progression of thoracic aortic aneurysm (TAA)^[14,15], which may result in the outbreak of aortic dissection^[16]. Progressive surgical intervention tend to be acceptable considering the risk of re-

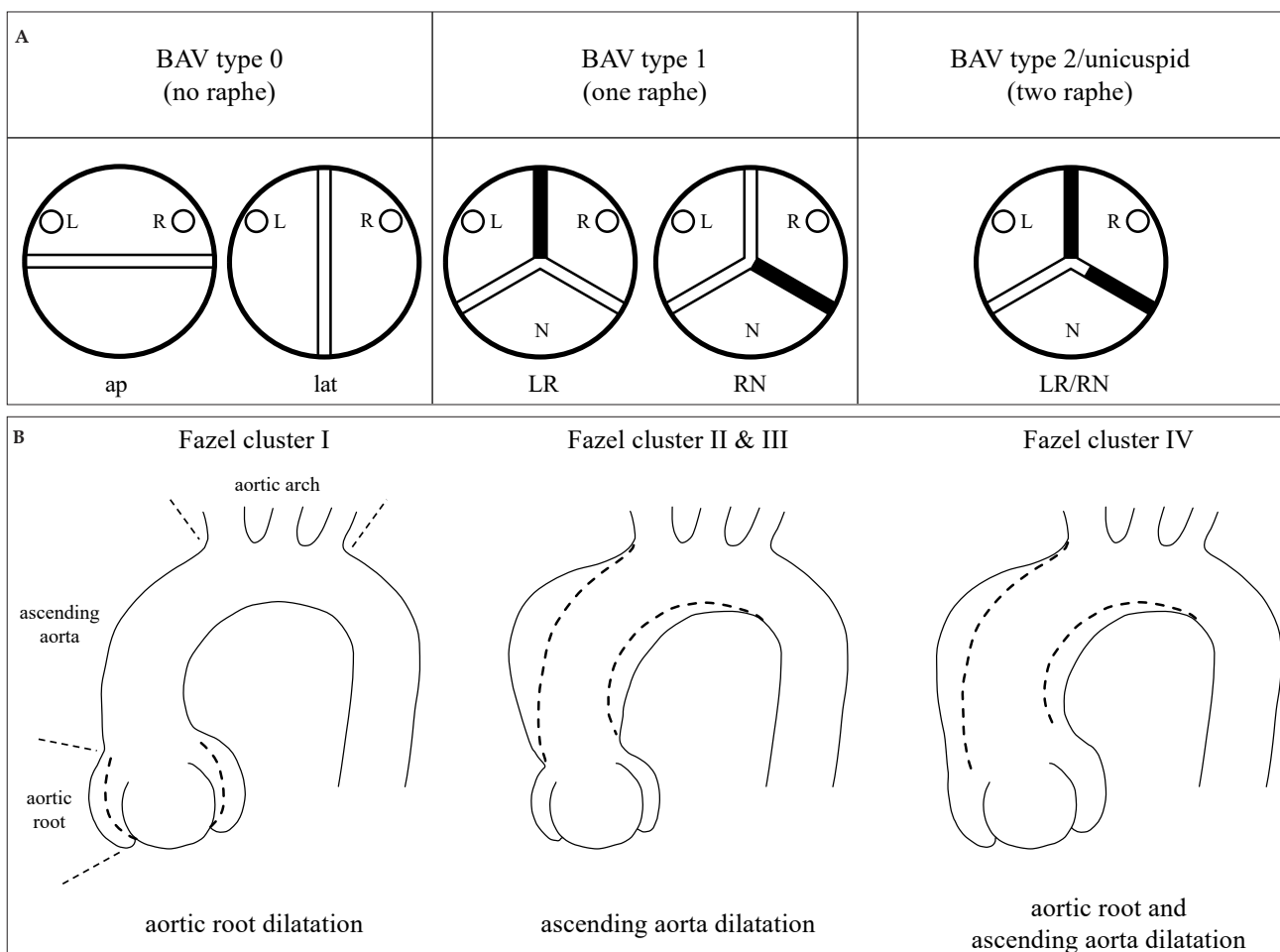


Figure 1 (A) Sievers classification of bicuspid aortic valve. (B) Fazel cluster classification of the morphological type of aorta.

operation in the near future. In our institution, concomitant aortic reconstruction in BAV patients accounted for 38.2%. Conversely, concomitant aortic reconstruction in tricuspid aortic valve (TAV) patients accounted for 12.0%. Although a direct comparison cannot be made between the concomitant aortic reconstruction rate in BAV patients with that in TAV patients, these findings suggest that BAV may be one of the trigger factors of aortic dilatation and that the aorta in BAV patients is likely to require surgical intervention. Surgical guidelines for concomitant aortic reconstruction with BAV has changed yearly. New 2015 AHA/ACC and 2017 EACTS guidelines recommend the concomitant aortic reconstruction with AVR when the ascending aorta diameter is ≥ 55 mm (Class I) and ≥ 45 mm (Class IIa)^[9,10]. On the other hand, 2012 JCS guidelines recommend aorta surgical reconstruction when the ascending aortic diameter is ≥ 45 mm (Class I) and ≥ 45 mm (Class IIa) and if concomitant AVR is being performed^[11]. In 2003, the rate of the maximal aortic cross-sectional area in square centimeters divided by the patient's height in meters was proposed as a new parameter, in which a rate of ≥ 10 recommends more aggressive surgical treatments^[17]. However, it is difficult to apply the same formula to Japanese patients due to smaller stature on average compared to Westerners. Another report for Japanese patients with BAV demonstrated that aortic diameter of ≥ 40 mm was a predictor of aortic dilatation risk after AVR^[18], and concomitant aortic reconstruction was recommended regardless of aortic dilatation^[19]. In our experience, eight patients with vulnerable or perishable vessel walls underwent concomitant aortic reconstruction with AVR even if the ascending aortic diameter was 40 to 45 mm. As shown in Figure 2, the BAV patient with severe aortic valve stenosis (AS) with an ascending aorta and aortic arch dilatation of 43mm was scheduled to undergo AVR before operation. Instead however, concomitant aortic reconstruction with

AVR was performed because intraoperative findings revealed that the ascending aorta was vulnerable and easily bled during cannulation. Unfortunately, although there have been numerous case reports and retrospective studies, many were non-randomized trials and make it difficult to make any definitive conclusions on the timing of precautionary surgical intervention for any thoracic aneurysmal diseases. Thus, concomitant aortic reconstruction to BAV patients with aortic dilatation around 45 mm will remain a standard surgical treatment considering the future risks.

Many studies have demonstrated that fluid hemodynamic-induced stress and shear stress in the BAV aorta could contribute to aortic dilatation. Notably, LR fusion type in BAV patients is thought to have idiosyncratic systemic blood flow to the right front which could affect the ascending aorta dilatation, while RN fusion type is thought to have eccentric systemic blood flow to the rear which could result in the dilatation of the proximal part in the arch^[20]. Moreover, in another study, BAV patients had a higher incidence of TAA compared to TAV patients with comparable AS^[21]. Each valve fusion type rate in our BAV patients indicated the same trend as demonstrated in the previous report as shown in Figure 3^[22]. However, in our BAV patients, a similar trend in typical morphological changes of the aorta as shown in the previous studies was not detected (Table 2). Thus, it is evident that a correlation between BAV dysfunction and TAA formation exists. Nonetheless, mechanisms excluding juvenile and congenital types, such as gene mutations, remains to be elucidated. The post-AVR patients without the ascending aorta replacement deserve further study, both to confirm these findings and to elucidate the mechanisms.

This present study was subject to limitations. The decision on the concomitant surgical intervention with AVR was carefully considered on the specificities of each patient. Moreover, the size of the study

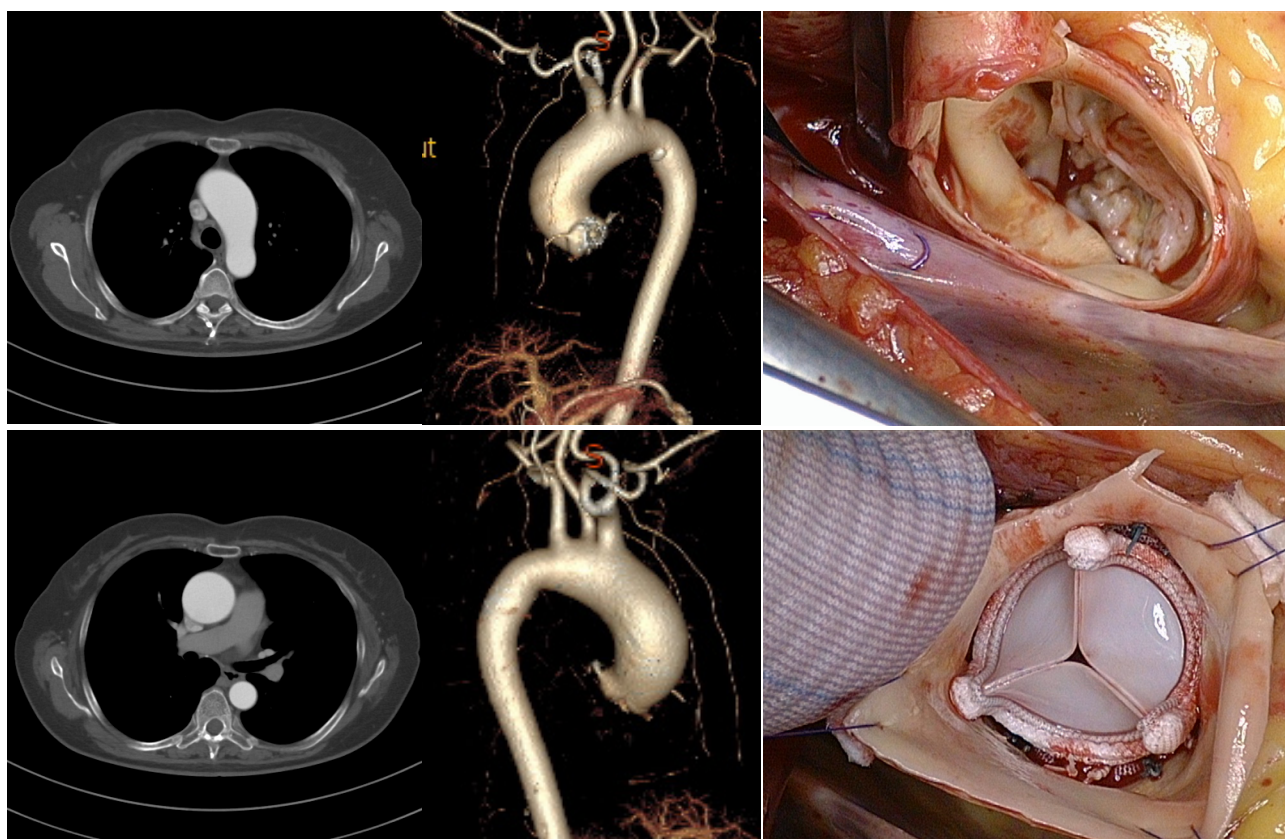


Figure 2 Actual case presentation. (A, B) Computed tomography (CT) (A) and three dimensional CT (B) shows the ascending aorta and aortic arch dilatation (43mm). (C) Intraoperative findings shows severe aortic valve stenosis and attenuated aortic wall (upper) and post-aortic valve replacement (lower).

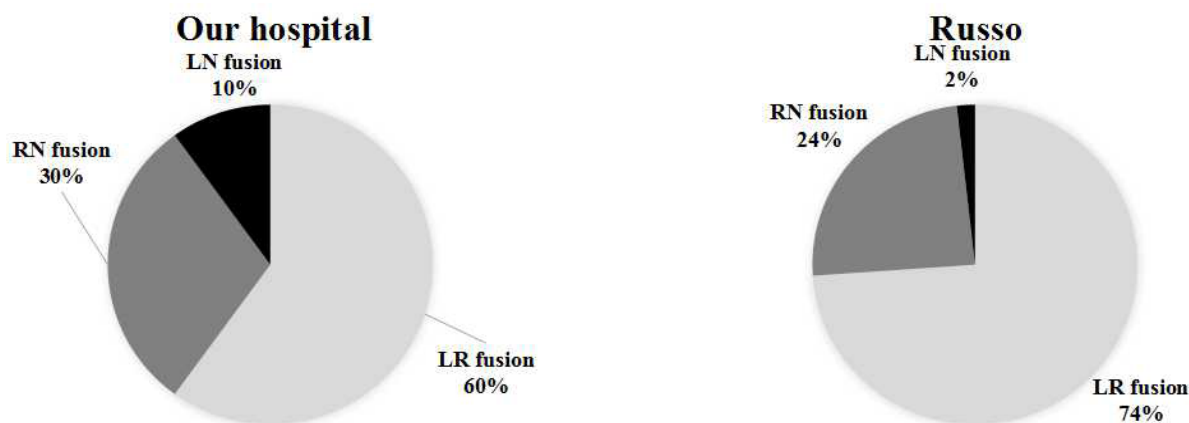


Figure 3 Comparison of type 1 in Sievers classification. Data of Russo et al. was cited from J Thorac Cardiovasc Surg 2008;136:937-42.

Table 1 Patient characteristics.

Variables	n = 39
Age, years, mean (± SD)	63.9 (± 13.8)
Female gender, n (%)	15 (38.4%)
Form of ascending aorta, n (%)	
Symmetric form	37 (94.9%)
Asymmetric form	2 (5.1%)
Average aortic diameter, mm, (± SD)	47.8 (± 6.1)
Sievers classification, n (%)	
Type 0	8 (20.5%)
ap-type	1 (12.5%)
lat-type	7 (87.5%)
Type 1	30 (76.9%)
LR fusion type	18 (60.0%)
RN fusion type	9 (30.0%)
LN fusion type	3 (10.0%)
Type 2	1 (2.6%)
Fazel Cluster classification, n (%)	
FC type I	8 (20.5%)
FC type II&III	28 (71.8%)
FC type IV	3 (7.7%)

AVR: aortic valve replacement, BAV: bicuspid aortic valve, FC: Fazel Cluster, SD: standard deviation.

was quite small (39 patients) and also there is no follow-up time for the post-AVR patients without the ascending aorta replacement. Therefore, although surgical outcomes at our single center might not be representative of a general patient cohort, we intend to continue to investigate these findings in future cases.

CONCLUSION

The frequency of Sievers classification in 39 BAV patients was similar to that reported previously; however, the fusion type was unlikely to affect the aortic dilatation type in BAV patients. All fusion types may be associated with a risk of the enlargement of the ascending and arch aorta.

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Contributorship

Dr. Mitsuru Iida had full access to all of the data in this study and

Table 2 Operative characteristics.

Sievers classification, n (%)		FC classification, n (%)	
Type 0	8 (20.5%)	FC type I	1 (12.5%)
		FC type II&III	7 (87.5%)
		FC type IV	0 (0%)
Type 1	30 (76.9%)		
LR fusion type	18 (60.0%)	FC type I	4 (22.2%)
		FC type II&III	11 (61.1%)
		FC type IV	3 (16.7%)
RN fusion type	9 (30.0%)	FC type I	2 (22.2%)
		FC type II&III	7 (77.8%)
		FC type IV	0 (0%)
LN fusion type	3 (10.0%)	FC type I	0 (0%)
		FC type II&III	3 (100%)
		FC type IV	0 (0%)
Type 2	1 (2.6%)	FC type I	0 (0%)
		FC type II&III	1 (100%)
		FC type IV	0 (0%)

takes responsibility for this integrity of the data and the accuracy of the data analysis.

Study concept and design: Mitsuru Iida, Masateru Uchiyama, Tomohiro Imazuru, Tomoki Shimokawa.

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