VIA MEDICA

RESEARCH LETTER

Cardiology Journal
2024, Vol. 31, No. 1, 177–178
DOI: 10.5603/cj.95563

ISSN 1897–5593
eISSN 1898–018X

Bicuspid aortic valve in transplanted hearts. Systematic study

Maria Sudomir[1], Maciej Michałowski[1], Anna Drohomirecka[2], Justyna Gruczek[3], Piotr Kołsut[4], Ilona Michałowska[5], Adam Witkowski[1], Tomasz Zieliński[2], Paweł Tyczyński[1]

[1]Department of Interventional Cardiology and Angiology, National Institute of Cardiology, Warsaw, Poland
[2]Department of Heart Failure and Transplantology, National Institute of Cardiology, Warsaw, Poland
[3]Information Technology, National Institute of Cardiology, Warsaw, Poland
[4]Department of Cardiac Surgery and Transplantology, National Institute of Cardiology, Warsaw, Poland
[5]Department of Radiology, National Institute of Cardiology, Warsaw, Poland

Bicuspid aortic valve (BAV) is the most frequent congenital cardiac anomaly, affecting 1–2% of the population [1]. The abnormality is followed by an increased risk of many potentially severe complications, such as BAV degeneration, dilatation of the ascending aorta (AAo) and others [2]. The recently published International Consensus Statement on nomenclature and classification of BAV distinguishes three types of BAV with specific phenotypes [3]. Although in many cases BAV may have a severe clinical outcome, in other patients it can go undetected. Orthotopic heart transplantation (OHT) remains the gold standard treatment of end-stage heart failure (HF). The presence of BAV in a donor-heart is not a contraindication for OHT, unless BAV is severely diseased. According to available research, only single reports of BAV in transplanted hearts have been reported to date.

Discharge summaries were retrospectively screened and the reports of transthoracic echocardiography (TTE) of 623 OHT-recipients in the electronic database from a tertiary high-volume heart center for the presence of BAV were included in the study. Key word: "bicuspid aortic valve" with its abbreviations and grammatical variations were used during screening. This database contains discharge summaries from January 2008 to October 2023. Nonetheless, some of these patients underwent OHT before 2008 (some of them are from another cardiovascular center, who were then followed-up at the present institution). All OHT-patients underwent typical of follow-up procedure and every patient underwent several echocardiographic assessments after OHT. All post-OHT TTE patients were screened for the presence of BAV. Only the last TTE was included in the results of the identified OHT-patients with BAV.

Six OHT-recipients (5 males) with BAV in transplanted hearts were identified. They ranged from 12 to 65 years of age. The follow-up was from 1.0 year to 21.0 years (Table 1). The prevalence of BAV among transplanted (donor) hearts was 0.96% (6/623).

Patient 1. He underwent OHT due to ischemic cardiomyopathy. Risk factors included male sex, diabetes mellitus and hypertension. Recent TTE (21 years after OHT) revealed degenerated and calcified BAV-fusion of the right coronary cusp (RCC) with the left coronary cusp (LCC), aortic peak pressure gradient of 17 mmHg, dilated AAo 39 mm (without any change in diameter over the last 7 years), moderate/significant tricuspid regurgitation and hypertrophied left ventricle (LV). Otherwise, systolic function of both ventricles was preserved.

Patient 2. He underwent OHT due to congenital heart defects. His recent TTE (3 years after OHT) showed BAV-fusion of the non-coronary cusp (NCC) with RCC, negligible aortic regurgitation (AR), discrete AAo dilatation 37 mm (without any change in diameter over the last 12 years), and impaired systolic function of the right ventricle (RV). Otherwise, LV systolic function was preserved.

Address for correspondence: Dr. Paweł Tyczyński, Department of Interventional Cardiology and Angiology, National Institute of Cardiology, ul. Alpejska 42, 02–462 Warszawa, tel: +48 22 3434272, e-mail: medykpol@wp.pl

Received: 15.05.2023 Accepted: 23.12.2023 Early publication date: 29.01.2024

Table 1. Summary of patients with bicuspid aortic valve in transplantaed (donor) heart.

Patient	Sex	Age at OHT, years	Reason for OHT	Time of follow-up after OHT, years	Type of BAV (fusion)
1	M	49	Ischemic cardiomyopathy	21	RCC-LCC
2	M	29	CHD	3	NCC-RCC
3	M	48	Ischemic cardiomyopathy	15	RCC-LCC
4	F	12	Acute myocarditis	1.6	RCC-LCC
5	M	28	CHD	3.2	NCC-RCC
6	M	65	DCM	1.0	RCC-LCC

BAV — bicuspid aortic valve; CHD — congenital heart defect; DCM — dilated cardiomyopathy; F — female; LCC — left coronary cusp; M — male; NCC — non-coronary cusp; OHT — ortothopic heart transplantation; RCC — right coronary cusp

Patient 3. He underwent OHT due to ischemic cardiomyopathy. His recent TTE (15 years after OHT) revealed BAV-fusion of RCC-LCC, with mild to moderate AR, LV hypertrophy (14 mm). Sinus of Valsalva was mildly dilated (46 mm) and AAo was normal (without any change in diameter over the last 10 years). Systolic function of both ventricles was mildly impaired.

Patient 4. She underwent OHT due to acute myocarditis complicated by cardiogenic shock, requiring 4-months support by an LV assist device while on the OHT waiting list. During that period, she experienced two ischemic strokes. Post-OHT course was complicated by rupture of AAo, requiring two re-do operations with supra-aortic prosthesis of AAo. Her recent TTE (1.6 years after OHT) revealed BAV-fusion of RCC-LCC with negligible AR, impaired RV systolic function and supra-valvular aortic gradient at the prosthesis (41 mmHg). Otherwise, LV systolic function was preserved.

Patient 5. He underwent OHT due to congenital heart defects. His recent TTE (3.2 years after OHT) revealed BAV-fusion of NCC-RCC with dilated AAo (41 mm without any change in diameter over 3.2 years). Systolic function of RV was impaired, while LV systolic function was preserved.

Patient 6. He underwent OHT due to dilated cardiomyopathy. His recent TTE (1 year after OHT) revealed BAV-fusion of RCC-LCC, mild to moderate AR, dilated AAo (44 mm without any change over 1 year) and impaired RV systolic function. Otherwise, LV systolic function was preserved.

Very few papers have shed light on the natural history of BAV in transplanted hearts. Some points are worth emphasizing, namely:

— Type of BAV in transplanted heart. All presented patients showed two types of BAV: fusion of NCC-RCC and fusion of RCC-LCC;
— Degeneration of BAV in transplanted heart. Given the rarity of reported BAV in transplanted heart, its natural history and the impact of immunosuppressive drugs on BAV remains largely unknown. The first patient had already developed BAV degeneration (although insignificant) during 21-years of observation. When there is significant BAV degeneration, re-do intervention may be needed. Although successful surgical BAV replacements in OHT-recipients were previously reported [4, 5], nonetheless transcatheter aortic valve replacement (TAVI) seems to be a treatment of choice for degenerated BAV in transplanted heart [6]. Of note, mid-, and long-term results of TAVI in OHT recipients remain unknown;
— Dilatation of AAo in OHT-recipients of donor-heart with BAV. 4 patients (1, 2, 5, and 6) presented with modest AAo dilatation diagnosed before OHT, without any change during follow-up after OHT. To date nothing is known about AAo diameter over time in recipients of donor-heart with BAV;
— What is done when donor-heart presents with already degenerated BAV (before OHT)? Elde et al. [7] published an interesting report of an OHT-recipient. During peri-OHT work-up degenerated BAV was discovered and replaced by a bioprosthetic valve. Then the donor-heart with new bioprosthesis was implanted to the patient, who remained healthy for 12 months after OHT [7]. Similarly, Saito et al. [8] reported on a patient who received a donor-heart with a mechanical aortic valve (implanted after harvesting the organ). In these both cases the long-term follow-up is unknown.

The retrospective nature of the study has inherent limitations. This study did not specifically analyze every TTE, but instead specific keywords were searched for (BAV with its grammatical variations and abbreviations) in post-OHT TTE reports.

Conflict of interest: None declared