(Almost) All Nonstenotic Bicuspid Aortic Valves Should Be Preserved or Repaired

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Valve preservation or repair of the regurgitant bicuspid aortic valve (BAV) minimizes valve-related complications. The purpose of the current analysis was to determine the proportion of BAVs than can be repaired. Between 2000 and 9/2018, 1168 operations for BAV without stenotic pathology were performed. Of these, 103 underwent isolated aortic valve replacement, the remaining 1055 individuals underwent valve repair. Reasons for replacement were analyzed. The proportion of repaired BAVs remained constant at 85% over the years. The most frequent reasons for replacement were cusp calcification or active endocarditis. In only 2 instances, replacement was performed as intraoperative conversion after failed repair. The vast majority of BAVs can be repaired with good durability as long as cusp substance is preserved.

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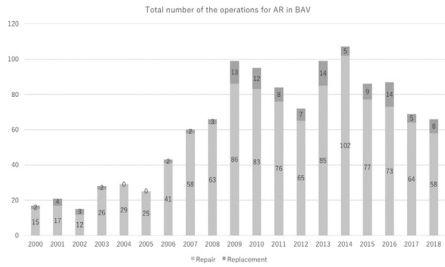

Number of the patients undergoing aortic valve surgery.

Central Message

With current techniques, the vast majority of BAVs can be repaired with good durability, which is better than that of bioprostheses. The incidence of valve-related complications is low.

INTRODUCTION

With a prevalence of 1–2% bicuspid anatomy of the aortic valve (BAV) is the most frequent congenital cardiovascular anomaly. Approximately half of the individuals with BAV will develop aneurysm during their lifetime and are thus potentially exposed to the occurrence of aortic complications.[1–3] In addition, the BAV makes them prone to aortic regurgitation (AR), and development of stenosis.[4,5] A relevant proportion of patients with aneurysm formation and the majority of individuals with AR will require surgery before the sixth decade of life.[5,6] This implies that surgery will traditionally consist of mechanical aortic valve replacement, with or without concomitant aortic replacement. The typical complications are thromboembolic events or hemorrhage due to anticoagulation.[7] In addition, there is valve-related mortality which is not insignificant.[8] While the majority of BAVs require treatment for stenosis,[9] clinical observations indicate that some BAVs may take a relatively benign course, in which a normally functioning BAV can be found as late as the eighth decade of life.[1] Finally, it is increasingly realized that the BAV includes a continuum of anatomic variations[10]; it is unclear whether certain forms of BAV may have better preservation of valve function during the natural course.

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Repair of BAVs was initially started by Cosgrove et al.[11] Midterm results of his and other groups were disappointing with a high rate of valve failure.[12] We and others could identify specific scenarios that were particularly prone to failure, such as commissural orientation, annular dilatation, persistence of prolapse, or the use of pericardium in augmentation or defect closure for cusp pathology.[13,14] Specific technical approaches were developed in order to aid decision-making for repair and conduct of the repair procedure.

Intraoperative measurement of geometric height allows for objective detection of cusp retraction.[15] The measurement of effective height aids in detection and correction of prolapse.[16] Annular reduction and stabilization have led to drastic improvement of valve stability.[17] Modification of commissural orientation has not only improved durability but also improved systolic valve function.[18,19] Results of BAV repair in the second decade have shown good functional durability, at least in the context of root replacement.[20]

In essence, we are in a time in which the results of BAV repair are beginning to enter the third decade, and repair tapered to the known predictors for failure repair yields excellent results. The hypothesis thus is that all BAVs should be repairable unless they exhibit established predictors of failure. The purpose of the current analysis was to analyze the reasons for replacement as well as the frequency of repairable BAVs.

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METHODS

Between January 2000 and September 2018, 3473 patients underwent surgery for aortic aneurysm in the absence of aortic stenosis or aortic valve surgery for relevant AR. The patients were identified through the departmental database, and all operative reports were reviewed to ascertain correctness of data, in particular aortic valve anatomy and correctness of data. 392 were excluded because of combined aortic valve dysfunction. A total of 1913 cases were excluded because of nonbicuspid anatomy of the aortic valve.

The remaining 1168 individuals were the subject of this investigation. Of these, 31 underwent combined replacement of aortic valve and root, 82 isolated aortic valve replacement. During the same time, 394 individuals underwent valve sparing root replacement in the form of root remodeling; 661 were treated by isolated aortic valve repair, in 194 cases in conjunction with tubular replacement of the ascending aorta.

If the valve was replaced as part of the procedure, the details of valve pathology were analyzed, specifying the reason for replacement. The resulting data were analyzed to 1. Define the proportion of repair and 2. The reasons for replacement over time. The analysis and subsequent publication were approved by the regional ethics committee, and individual informed consent was waived for analysis and publication in anonymized fashion.

RESULTS

Different reasons for replacement were found with cusp calcification being the most frequent (44%; Table 1). Interestingly, in only 2 instances, a replacement was performed as intraoperative conversion after failed repair. Long-term freedom from reoperation in the cohort of all BAV repair and root remodeling was 80.1 ± 2.6% and 87.5 ± 2.8% at 15 years, respectively, with the longest follow-up of 18 years (Fig. 1A and B).

The proportion of replaced vs repaired BAVs remained constant over the years (Fig. 2). When comparing the patients with repaired vs replaced BAVs, patient age was significantly younger in repair group (Table 2; repair: 46.6 ± 13.7 years vs replacement: 54.9 ± 12.9 years, $P < 0.0001$). Interestingly, there was a higher proportion of root replacement in the repair cohort.

Table 1. Basic Characteristics and Concomitant Procedures

	Repair	Replacement	*P*
Age (y)	46.6	54.9	<0.0001
Female (%)	5.1	16.8	<0.0001
Concomitant procedures (%)			
Root replacement	37.4	27.4	0.034
Tubular ascending aortic replacement	55.8	30.9	<0.0001
Partial arch replacement	9.8	14.2	0.17
Total arch replacement	1.2	0	0.1
Mitral valve repair	3.5	8.6	0.015
Mitral valve replacement	0	0.88	0.031
Tricuspid valve repair	1.3	0	0.09
MAZE	4	0	0.0032

DISCUSSION

The prevalence of BAVs and the associated aortopathy makes it a relevant burden to health care. Of the different complications that are associated with BAV anatomy, aortic stenosis is the most frequent.[9] The second most important complication is aortopathy, which either leads to the dreaded aortic complications or the need for prophylactic aortic replacement.[2] In addition, AR will require surgical treatment also in a relevant proportion of cases. While aortic stenosis commonly develops in the more advanced age groups, proximal aortic replacement and surgical treatment of AR are commonly necessary in younger individuals. This is supported by the patient age in our series.

Even though aortic valve function is not infrequently preserved in aneurysm of the tubular ascending aorta, aortic replacement will often lead to distortion of the intercommissural distance of the valve, and postoperative aggravation of AR may be the consequence. Thus, the patient group requiring surgery on the aortic valve for a nonstenotic dysfunction is actually larger than the pure population with AR. Our patient cohort reflects these considerations; only 45% required surgery for isolated AR, and combined treatment of valve and aorta was necessary in the majority of cases.

Treatment of AR has traditionally consisted of valve replacement. Aortic replacement for aneurysm of the root phenotype often will also require concomitant surgery on the aortic valve, traditionally by valve replacement. This has been supported by the hypothesis that all BAVs are restrictive by nature and will become stenotic.[9,21] For valve replacement, either a biologic or mechanical prosthesis has commonly been used. Both variants are associated with a relevant incidence of valve-related complications, including thromboembolism, hemorrhage, structural failure, and prosthetic valve endocarditis.[22,23] In addition, there is an excess mortality associated with aortic valve replacement, which seems to be higher after treatment of AR[8] and particularly involves the younger patients. Interestingly, this excess mortality involves both bioprosthesis and mechanical valves.[24] While replacement with a pulmonary autograft is an option for young patients, it has yielded inferior durability in the subset of individuals with BAV and AR. Thus, replacement has clear limitations, in particular for young individuals.

Repair of BAVs, either because of AR or in the context of aneurysm, has become an alternative in the past 20 years. It has been associated with minimal valve-related complications if a durable repair result could be achieved; in fact, in our experience, failure of the reconstructed BAV has constituted more than 90% of all valve-related complications. Despite its obvious ease in repair because of only 1 coaptation line, durability of BAV repair has been limited in the past. Different causes of failure have been identified in the past, such as persistent prolapse or annular dilatation.[13] Using objective criteria, such as geometric height for selection of adequate repair substrates[15] or measurement of effective height to guide detection and correction of

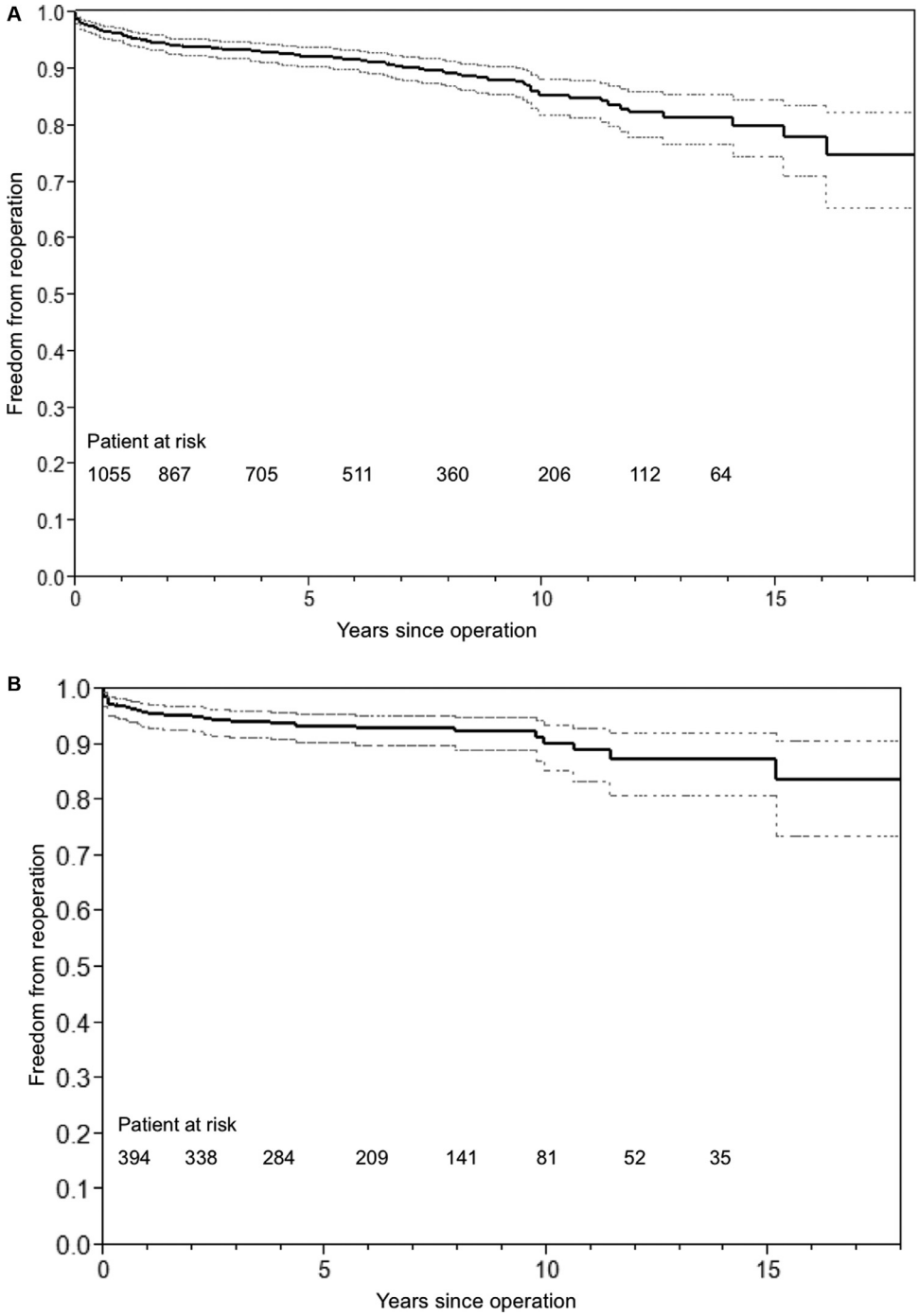

Figure 1. (A) Kaplan-Meier curve with 95% confidence interval indicating freedom from reoperation on aortic valve in patients who underwent valve preserving surgery. (B) Kaplan-Meier curve with 95% confidence interval indicating freedom from reoperation on aortic valve in patients who underwent aortic root remodeling.

prolapse,[14] repair has become more reproducible and results more predictable. The importance of annular dilatation on repair durability has been recognized[14] and annuloplasty has been shown to drastically improve the durability of isolated BAV repair. In addition, the anatomic variability and the relevance of commissural orientation have been recognized.[14] The introduction of systematic modification of commissural orientation has been shown to likewise markedly improve systolic function of the repaired BAV and its durability.[14,19] Finally, repair has also been shown to improve survival compared to replacement, albeit in a very limited cohort of patients.[25] Durability reaching and exceeding 20 years has been documented[20]; the transition from repaired BAV to stenosis has been rare, and already at 18 years, a freedom from reoperation of more than 70% in a patient

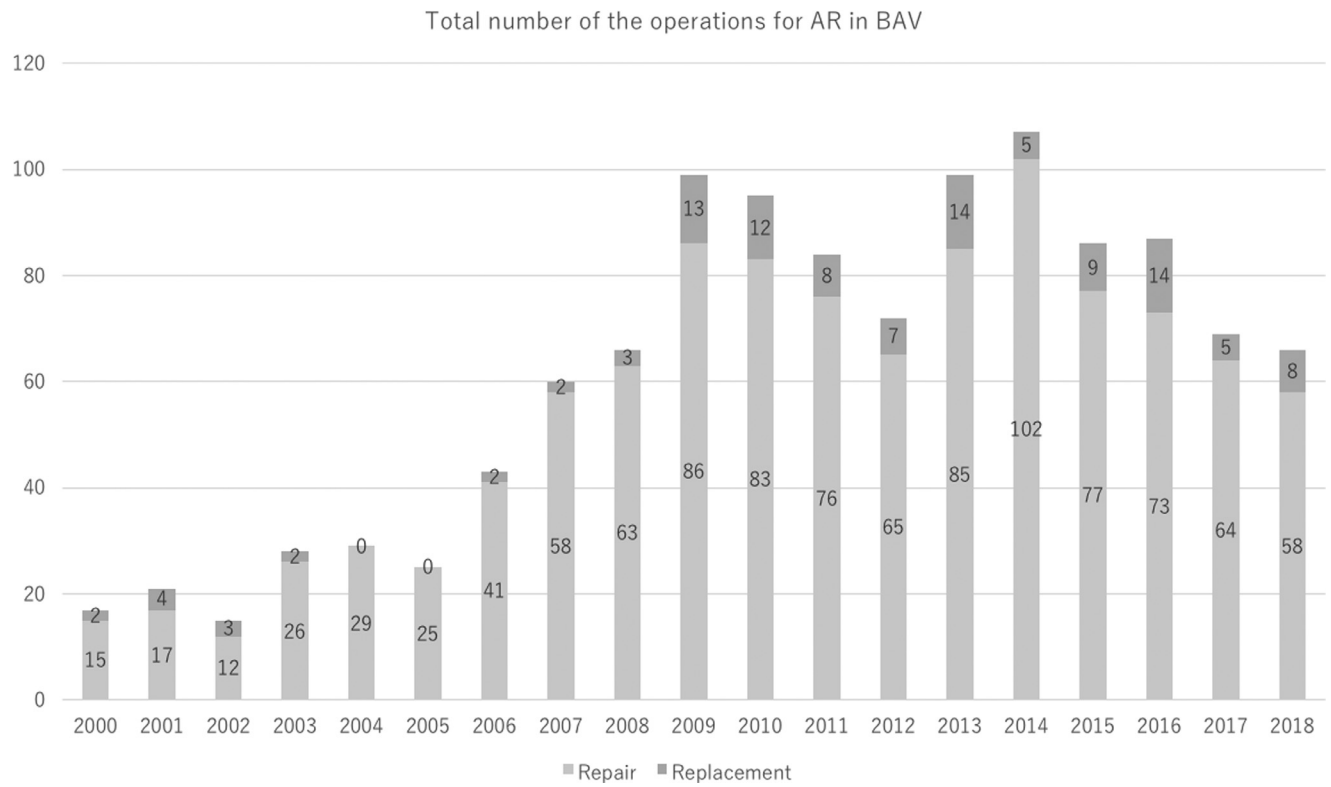

Figure 2. Annual number of the patients undergoing aortic valve surgery for pure aortic regurgitation with bicuspid pathology. The proportion of replacements has been low over the years. AR, aortic regurgitation; BAV, bicuspid aortic valve.

Table 2. Reasons for Aortic Valve Replacement

Reasons for Replacement	*n*	%
Calcification	50	44
Infective endocarditis	29	26
Retraction	15	13
Fenestration	9	8
Commissural orientation	4	3
Perforation	3	3
Repair failure	2	2
Diffuse fibroelastoma	1	1

cohort with a mean age of 47 years can be considered as superior to bioprosthesis (Fig. 1A).

In our experience, we have deliberately tested repair for different scenarios. It has become apparent that most BAVs in the context of AR and/or aneurysm are be repairable if cusp substance or structure is preserved, which is not always the case. Calcification requires specific treatment, such as excision and partial cusp replacement as do cusp defects in acute infective endocarditis; retracted cusp tissue requires augmentation with cusp replacement material. Even though autologous pericardium has yielded good results in one series,[26] any insertion of pericardial patches has consistently been a risk factor for repair failure in our hands.[27] Until better and more durable cusp replacement material is identified, these pathologies should be probably best treated by valve replacement. It is, however, as yet unknown which proportion of valves will thus not be repairable.

Considering the obstacles to a durable repair, we have been relatively selective in our activity. Nonetheless, we have been able to repair approximately more than 90% of BAVs when the indication for surgery was regurgitation or aneurysm. Calcification (which would have required excision and patch insertion) were the 2 most frequent reasons for replacement. The operative strategy (repair vs replacement) probably also explains the higher proportion of root replacement in the repair cohort. Since root replacement has been shown to have a stabilizing effect on a repaired BAV,[28] we have had a lower threshold to replace the root if the valve was to be repaired.

While the current results support the hypothesis that the vast majority of BAVs should be repaired, there are limitations to its generalization. The main limitation of repairability has been the absence of preserved cusp tissue. Most of our patients come from central Europe with a very low prevalence of rheumatic or other inflammatory heart valve disease. The prevalence of retraction and calcification may be higher in other areas, and this will influence repairability. More importantly, we have learnt that repair concepts have to address the anatomic characteristics of the BAV, in particular with respect to annular dilatation and commissural orientation. Repair has become very predictable and reproducible with a very systematic approach, including the concepts of geometric and effective height for selection and guidance during the procedure

(operative techniques). Our concepts and results may thus not be transferable to a surgical practice in which the surgical intervention is solely based on "judgment" and improvisation.

In conclusion, aortic valve replacement with either biological or mechanical prostheses is associated with not only a relevant incidence of valve-related complications but also an increased mortality. Using systematic repair concepts already the vast majority of BAVs can be repaired with good durability as long as cusp substance is preserved. Further extension of repairability will depend on the development of better cusp replacement material.

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