Valve-preserving root replacement in bicuspid aortic valves

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Objective: Bicuspid aortic valve anatomy is associated with aortic root aneurysm in a relevant proportion of patients. These patients require root replacement for prognostic reasons, and the valve may be preserved. The objective of this analysis is to analyze the early and late outcomes of root remodeling for bicuspid aortic valve.

Methods: Between November 1995 and December 2009, 153 patients (133 male) were treated by root remodeling in the presence of a bicuspid aortic valve. Acute dissection was present in 6 individuals. In 137 instances, additional correction of cusp pathology was achieved by plication ($n = 119$), triangular resection ($n = 59$), and implantation of a pericardial patch ($n = 27$). Follow-up ranges from 3 months to 14.5 years (mean, 4.9 ± 3.8 years; cumulative, 757 years) and is complete in 99.3%.

Results: One patient died of intracranial hemorrhage in the hospital (mortality 0.7%). Survival at 5 and 10 years was 99% and 91%, respectively. Seven patients required reoperation for stenosis ($n = 1$) or recurrent aortic insufficiency ($n = 6$) between 1 month and 11 years postoperatively. The aortic valve was re-repaired in 2 cases. Freedom from reoperation at 5 and 10 years was 95%; freedom from valve replacement was 97%. Freedom from valve-related complications was 91% at 5 and 10 years.

Conclusions: Root remodeling for aortic root aneurysm in the presence of a bicuspid aortic valve can be performed with a low morbidity and mortality. The long-term stability of the reconstructed aortic valve is excellent if normal valve configuration is achieved. The occurrence of late stenosis seems to be rare, and freedom from valve-related complications is high. (*J Thorac Cardiovasc Surg* 2010;140:S36-40)

Dilatation of the proximal aorta occurs in up to 60% of individuals born with bicuspid aortic valve (BAV) morphology.^{1,2} Different patterns of aortic dilatation have been observed. Dilatation may spare the root and primarily involve the ascending aorta, possibly extending into the arch.^{3,4} The optimal treatment for these conditions is controversial, but the root may be spared in many instances. In a relevant proportion of patients, aortic dilatation primarily involves the root.⁴ These patients obviously require a form of root replacement to eliminate the aortic pathology.

Root replacement implies a choice of composite replacement or valve-preserving root replacement. Preservation of the valve obviously avoids long-term anticoagulation and the valve-related complications of mechanical substitutes.^{5,6} There are, however, only few reports on long-term stability of the preserved aortic valve. In addition, most reports on valve-preserving aortic replacement deal with tricuspid aortic valves.⁷⁻⁹ Limited information is available regarding long-term results of reconstruction of BAVs. We previously

reported good mid-term results comparing root remodeling for tricuspid aortic valves versus BAVs.¹⁰

During a 14-year period, we have consistently performed root remodeling, with preservation or repair of a BAV whenever the root was dilated and cusp tissue appeared pliable.¹¹ The objective of this analysis is to update the early and analyze the late outcome of this approach.

PATIENTS AND METHODS

Between November 1995 and December 2009, 155 patients with a regurgitant BAV underwent operation for aortic root dilatation or dissection at the University Hospital of Saarland. Root remodeling was performed in 153 cases, and these are the focus of the current analysis. An additional 2 patients with a BAV were treated with valve reimplantation because there was only limited cusp fusion and the commissures had an approximately 120-degree orientation, thus making the valves almost tricuspid in morphology.

The majority of patients ($n = 133$) were men; the age ranged from 25 to 77 years, with a mean of 51 ± 12 years.

The preoperative degree of aortic regurgitation ranged from 0 to 4+ (mean, 2.6 ± 0.8). The primary indication for surgery was aortic aneurysm ($n = 73$), severe aortic regurgitation with clinical or echocardiographic signs of left ventricular overload ($n = 72$), acute aortic dissection ($n = 6$), or active endocarditis ($n = 2$).

All patients had a maximum sinus diameter of more than 45 mm as measured by transesophageal echocardiography (mean, 48 ± 3 mm). The maximum diameter of the supracommissural ascending aorta ranged from 4.5 to 8 cm (mean, 5.3 ± 0.6 cm). The sinotubular junction measured under cardioplegic arrest was enlarged in all patients (mean, 38 ± 5 mm); the size of the aortoventricular junction ranged from 25 to 35 mm (mean, 29 ± 3 mm). The patterns of fusion were right/left in 139 cases (91%), right/noncoronary in 11 cases (7%), and left/noncoronary in 3 cases (2%). Complete fusion was present in 96 instances and partial fusion in 57 instances.

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Abbreviations and Acronyms

BAV = bicuspid aortic valve
BSA = body surface area

Concomitant cardiovascular disease was present in 44 individuals (29%), that is, coronary artery disease ($n = 15$), mitral regurgitation ($n = 1$), arch aneurysm ($n = 25$), or permanent atrial fibrillation ($n = 4$).

Operative Technique

The technique has been described in detail.¹⁰ In all operations, the chest was opened by a median sternotomy. In acute dissection, either the right femoral ($n = 2$) or right axillary artery ($n = 4$) was used for arterial inflow. The aorta was completely transected above the commissures and mobilized. Graft size was chosen smaller than the aortoventricular diameter, taking the body surface area (BSA) of the patient into consideration. Generally, a 26-mm graft was chosen for a BSA less than 2 m^2 , and a 28-mm graft was chosen for larger patients. The graft was tailored to accommodate the asymmetry of the root, with a large tongue for the sinus of the nonfused cusp and 2 smaller tongues for the rudimentary sinuses. In the last 2 years, we have tended to create 2 almost symmetric tongues for a 170- to 180-degree orientation of the 2 normal commissures of the nonfused cusp (Figure 1). The sinuses were excised, and the graft was inserted, taking care to maintain sufficient height of the commissures.

In 16 instances, no apparent cusp pathology was observed, and no further procedure on the cusps was undertaken. These patients had no or only minimal aortic regurgitation preoperatively. In the majority of instances, prolapse of 1 ($n = 41$) or both cusps ($n = 96$) was recognized and corrected.

For correction of cusp pathology, the large normal cusp (commonly the noncoronary) was used as reference. The edges of the normal and fused cusps were aligned before performing root remodeling, and prolapse of the fused cusps was corrected by placing central plicating sutures on the free margin. In June 2004, we realized that root replacement could induce prolapse by reducing intercommissural distance; we also recognized that preexisting prolapse of the noncoronary cusp could be present, and we therefore modified the procedure. Since then, root replacement has been performed first to create the final root configuration as a prerequisite for assessment of cusp configuration. After completion of remodeling, the graft was cut 10 to 20 mm above the commissures, and the aortic valve was inspected carefully. The effective height of the normal cusp was measured with radial and upward tension on the commissures¹²; the effective height was increased to 10 mm by central plication sutures on the free margin.¹² The free margins of the nonfused and the fused cusps were then aligned. Redundancy of the free margin of the fused cusps was eliminated by central plication ($n = 119$) or triangular resection, and readaptation ($n = 59$). If more extensive calcifications were present in the raphe, the tissue was excised and replaced by a patch of autologous pericardium ($n = 27$). Similarly, endocarditic defects were closed with a pericardial patch ($n = 2$). In 3 patients, limited calcific plaques were removed without replacement of the involved cusp tissue. If more extensive aortic replacement (ie, arch) was required, it was performed with a second graft after completion of the valve and root procedure.

Additional coronary artery bypass grafting was performed in 15 patients, and concomitant mitral repair was performed in 1 patient. For partial ($n = 22$) or total ($n = 3$) replacement of the arch, hypothermic circulatory arrest was used (nasopharyngeal temperature 18°C – 21°C).

Echocardiographic Assessment

All patients underwent intraoperative transesophageal echocardiography for analysis of the aortic valve. Transthoracic echocardiography was performed postoperatively on a routine basis before discharge; at 3, 6, and

12 months postoperatively; and yearly thereafter. The degree of aortic regurgitation was quantified according to current guidelines.¹³ In addition, peak and mean systolic gradients were determined by transthoracic echocardiography using continuous-wave Doppler. An effort was made to follow the patients in our own institution. In 25 patients, this was not possible for geographic reasons, and for the current analysis we assessed the echocardiographic examinations performed by distant cardiologists and sent to us.

Statistical Analysis

All data are presented as mean $\pm$ standard deviation. Kaplan–Meier curves were calculated for freedom of relevant regurgitation, freedom from reoperation, and freedom from valve replacement using a commercially available software package (Prism, GraphPad Inc, San Diego, CA).

RESULTS

Mean myocardial ischemic time was 75 ± 14 minutes, and duration of extracorporeal circulation was 100 ± 26 minutes. For isolated remodeling without a concomitant procedure, mean ischemic time was 70 ± 7 minutes.

One 38-year-old patient died in the hospital, for a mortality of 0.7%. He had undergone elective surgery with extubation after 3 hours and a normal neurologic status. On postoperative day 1, an intracranial hemorrhage developed from a previously undiagnosed cerebral aneurysm and he died the same day. Early morbidity included reexploration for bleeding ($n = 4$) and atrioventricular block requiring implantation of a pacemaker ($n = 1$).

During follow-up, 4 patients died between 8 months and 9 years postoperatively. All had shown stable valve function during scheduled follow-up visits, and the cause of death could not be determined. Survival was 99% and 91% at 5 and 10 years postoperatively, respectively.

Increasing aortic regurgitation developed in 10 patients during follow-up. Six patients required reoperation for clinical or echocardiographic signs of left ventricular volume overload. At reoperation, the cause of failure was symmetric prolapse ($n = 2$), suture line dehiscence after plication ($n = 1$), or partial cusp replacement ($n = 3$). The valve was re-repaired in 2 patients and replaced in the other 4 patients. The remaining 4 patients have remained stable and asymptomatic. Freedom from aortic regurgitation grade 2+ or higher is 95% at 5 years and 90% at 10 years. Aortic stenosis was observed in 1 patient 11 years after the initial procedure, which had included removal of a calcific plaque from the noncoronary cusp. He underwent uneventful reoperation and valve replacement.

Freedom from reoperation is 95% at both 5 and 10 years (Figure 2); freedom from valve replacement is 97% at 5 and 10 years postoperatively (Figure 3).

Four thromboembolic events were observed between 3 weeks and 3 months postoperatively. In 2 events, this was apparently related to paroxysmal atrial fibrillation, and in the other 2 events, no additional disease could be found. There were no episodes of endocarditis throughout the follow-up period. Freedom from all valve-related

FIGURE 1. Computed tomography of an aortic root with BAV preoperatively (A) and postoperatively (B). The commissural orientation of the commissures of the nonfused cusps (dotted lines).

complications is 91% at both 5 and 10 years postoperatively (Figure 4).

DISCUSSION

The occurrence of aortic dilatation in conjunction with bicuspid anatomy of the aortic valve has been studied more extensively in the past decade. Aortic aneurysm has been shown to occur in approximately 60% of individuals with this anatomic variant irrespective of the presence or type of hemodynamic dysfunction of the aortic valve.^{14,15} Different patterns of aortic dilatation have been described,^{3,16} and the patterns have been linked to the fusion type of the aortic valve.^{4,17} Notably, the distribution of fusion pattern in our patients was similar to that published for BAVs in general.¹⁸ Thus, we cannot confirm the proposed relationship between aortic dilatation and fusion pattern.⁴

The prognostic impact of aortic dilatation has been increasingly recognized¹⁹ and has resulted in the lowering of thresholds for prophylactic surgery compared with

aneurysms and tricuspid aortic valves.^{20,21} It is, however, unclear whether the diameter of the root should be considered different from that of the tubular ascending aorta. We believe that replacement of a moderately enlarged root helps to stabilize the aortic valve if repair is performed. Thus, we consider a root diameter of more than 40 to 44 mm, depending on the BSA of the patient, as a cutoff in favor of root replacement. Further information will be necessary to guide surgical decision-making.

The conventional standard treatment for a diseased BAV and aortic dilatation has been composite replacement of the valve and root. A large series treated this way was published recently.²² Our current population seems generally comparable, with a similar mean age (51 vs 53 years) and similar follow-up (mean 4.9 vs 5.9 years). The current 10-year survival is similar to that reported. The authors described a stroke rate of 0.7% per patient year and hemorrhagic complications of 0.5% per patient year. Cumulative freedom from valve-related complications was not stated, and complete follow-up was available in only 88% of the patients.²²

FIGURE 2. Actuarial freedom from reoperation after root remodeling in patients with BAV.

FIGURE 3. Actuarial freedom from valve replacement after root remodeling in patients with BAV.

FIGURE 4. Actuarial freedom from all valve-related complications after root remodeling in BAVs.

Our current data compare favorably with this large series. Even though a learning curve is included in our series, freedom from reoperation is also identical with that reported in another large series with 2 replacement strategies in a less selected patient population.²³

Both forms of valve-preserving aortic replacement have been used in the context of BAVs: root remodeling and valve reimplantation.^{7,11,24} At this time there is no specific published information regarding long-term results of valve reimplantation in the setting of bicuspid anatomy. We have consistently chosen root remodeling for this scenario,¹¹ and the current results seem to support this approach. We cannot, however, determine the superiority of one approach over the other. We have not seen progressive dilatation of the aortoventricular junction as risk factor for valve failure or reoperation, indicating sufficient stabilization of that structure by remodeling. An important cause for failure was iatrogenic prolapse induced by reduction of root size,¹¹ which has largely been avoided since the introduction of the effective height concept.¹² Limited calcifications of the aortic cusps may be seen as another risk factor. Simple removal of the calcium may be associated with late development of stenosis, as seen in 1 of our patients. We have also observed the need for reoperation in 3 of 27 individuals in whom the calcified cusp tissue was replaced with a pericardial patch. Thus, with longer follow-up and more experience, we may have to reconsider our previous observations on the safety of patch implantation in the setting of BAV.¹¹

The limitations of the current analysis are primarily due to the lack of a matched control group. Because most of our patients are referred for valve repair, generation of a prospective control, ideally in the form of randomization, has been and will be psychologically difficult. Despite the size of the patient cohort studied, the number of individuals is still small compared with large valve trials, and only a limited proportion of our patients have exceeded 10 years of

follow-up. Finally, at this time there is no information on “very long” follow-up, that is, exceeding 20 years. This will be important to determine the true long-term role of preservation of BAVs.

CONCLUSIONS

Root remodeling for root aneurysm in the presence of a BAV can be performed with low morbidity and mortality. Long-term stability of the reconstructed aortic valve is excellent if normal valve configuration is achieved. The occurrence of late stenosis seems to be rare, and freedom from valve-related complications is high. Valve preservation for the combination of regurgitant BAV and root aneurysm seems to be at least equivalent to composite replacement.

References

1. Nkomo VT, Enriquez-Sarano M, Ammass NM, Melton LJ III, Bailey KR, Desjardins V, et al. Bicuspid aortic valve associated with aortic dilatation: a community-based study. *Arterioscler Thromb Vasc Biol.* 2003;23:351-6.
2. Nistri S, Sorbo MD, Marin M, Palisi M, Scognamiglio R, Thiene G. Aortic root dilatation in young men with normally functioning bicuspid aortic valves. *Heart.* 1999;82:19-22.
3. Fazel SS, Mallidi HR, Lee RS, Sheehan MP, Liang D, Fleischman D, et al. The aortopathy of bicuspid aortic valve disease has distinctive patterns and usually involves the transverse aortic arch. *J Thorac Cardiovasc Surg.* 2008;135:901-7.
4. Schaefer BM, Lewin MB, Stout KK, Gill E, Prueitt A, Byers PH, et al. The bicuspid aortic valve: an integrated phenotypic classification of leaflet morphology and aortic root shape. *Heart.* 2008;94:1634-8.
5. Ikonidis JS, Kratz JM, Crumley AJ III, Stroud MR, Bradley SM, Sade RM, et al. Twenty-year experience with the St Jude Medical mechanical valve prosthesis. *J Thorac Cardiovasc Surg.* 2003;126:2022-31.
6. Hammermeister K, Sethi GK, Henderson WG, Grover FL, Oprian C, Rahimtoola SH. Outcomes 15 years after valve replacement with a mechanical versus a bioprosthetic valve: final report of the Veterans Affairs randomized trial. *J Am Coll Cardiol.* 2000;36:1152-8.
7. David TE, Feindel CM, Webb GD, Colman JM, Armstrong S, Maganti M. Long-term results of aortic valve-sparing operations for aortic root aneurysm. *J Thorac Cardiovasc Surg.* 2006;132:347-54.
8. Yacoub M, Gehle P, Chandrasekaran V, Birks E, Child A, Radley-Smith R. Late results of a valve-preserving operation in patients with aneurysms of the ascending aorta and root. *J Thorac Cardiovasc Surg.* 1998;115:1080-4.
9. Aicher D, Fries R, Rodionicheva S, Schmidt K, Langer F, Schäfers HJ. Aortic valve repair leads to a low incidence of valve-related complications. *Eur J Cardiothorac Surg.* 2010;37:127-32. Epub 2009 Jul 29.
10. Aicher D, Langer F, Kissinger A, Lausberg H, Fries R, Schäfers HJ. Valve-sparing aortic root replacement in bicuspid aortic valves: a reasonable option? *J Thorac Cardiovasc Surg.* 2004;128:662-8.
11. Schäfers HJ, Aicher D, Langer F, Lausberg HF. Preservation of the bicuspid aortic valve. *Ann Thorac Surg.* 2007;83:S740-5.
12. Schäfers HJ, Bierbach B, Aicher D. A new approach to the assessment of aortic cusp geometry. *J Thorac Cardiovasc Surg.* 2006;132:436-8.
13. Zoghbi WA, Enriquez-Sarano M, Foster E, Grayburn PA, Kraft CD, Levine RA, et al. Recommendations for evaluation of the severity of native valvular regurgitation with two-dimensional and Doppler echocardiography. *J Am Soc Echocardiogr.* 2003;16:777-802.
14. Keane MG, Wiegers SE, Plappert T, Pochettino A, Bavaria JE, Sutton MG. Bicuspid aortic valves are associated with aortic dilatation out of proportion to coexistent valvular lesions. *Circulation.* 2000;102(19 Suppl. 3):III35-9.
15. Hahn RT, Roman MJ, Mogtader AH, Devereux RB. Association of aortic dilation with regurgitant, stenotic and functionally normal bicuspid aortic valves. *J Am Coll Cardiol.* 1992;19:283-8.
16. Novaro GM, Tiong IY, Pearce GL, Grimm RA, Smedira N, Griffin BP. Features and predictors of ascending aortic dilatation in association with a congenital bicuspid aortic valve. *Am J Cardiol.* 2003;92:99-101.

17. Fernandes SM, Sanders SP, Khairy P, Jenkins KJ, Gauvreau K, Lang P, et al. Morphology of bicuspid aortic valve in children and adolescents. *J Am Coll Cardiol*. 2004;44:1648-51.
18. Sabet HY, Edwards WD, Tazelaar HD, Daly RC. Congenitally bicuspid aortic valves: a surgical pathology study of 542 cases (1991 through 1996) and a literature review of 2,715 additional cases. *Mayo Clin Proc*. 1999;74:14-26.
19. Russo CF, Mazzetti S, Garatti A, Ribera E, Milazzo A, Bruschi G, et al. Aortic complications after bicuspid aortic valve replacement: long-term results. *Ann Thorac Surg*. 2002;74:S1773-6.
20. Vahanian A, Baumgartner H, Bax J, Butchart E, Dion R, Filippatos G, et al. Guidelines on the management of valvular heart disease: The Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology. *Eur Heart J*. 2007;28:230-68.
21. Bonow RO, Carabello BA, Kanu C, de Leon ACJ, Faxon DP, Freed MD, et al. ACC/AHA 2006 guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to revise the 1998 Guidelines for the Management of Patients With Valvular Heart Disease): developed in collaboration with the Society of Cardiovascular Anesthesiologists: endorsed by the Society for Cardiovascular Angiography and Interventions and the Society of Thoracic Surgeons. *Circulation*. 2006;114:e84-231.
22. Etz CD, Homann TM, Silovitz D, Spielvogel D, Bodian CA, Luehr M, et al. Long-term survival after the Bentall procedure in 206 patients with bicuspid aortic valve. *Ann Thorac Surg*. 2007;84:1186-93.
23. Sioris T, David TE, Ivanov J, Armstrong S, Feindel CM. Clinical outcomes after separate and composite replacement of the aortic valve and ascending aorta. *J Thorac Cardiovasc Surg*. 2004;128:260-5.
24. El Khoury G, Vanoverschelde JL, Glineur D, Pierard F, Verhelst RR, Rubay J, et al. Repair of bicuspid aortic valves in patients with aortic regurgitation. *Circulation*. 2006;114(1 Suppl):I610-6.