Concepts and techniques of bicuspid aortic valve repair

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Background: The bicuspid aortic valve (BAV) frequently requires surgical intervention for aortic regurgitation and/or aneurysm. Valve-preserving surgery and valve repair have been increasingly utilized as alternatives to replacement to avoid prosthesis-related complications.

Anatomy and Methods: Apart from aortic dilatation, prolapse of the fused cusp is an almost constant finding in the regurgitant BAV. Valve-preserving surgery can be applied, and correcting of the prolapsing fused cusp appears as obvious. Prolapse of the nonfused cusp, annular dilatation in the presence of preserved root dimensions, the use of pericardium for cusp repair, and asymmetric orientation have been found to be the most important anatomic predictors of repair failure. They can be treated by measuring effective height of the nonfused cusp, annuloplasty, and creation of a symmetric configuration whenever possible.

Results: If the defined anatomic features are addressed systematically, survival and the incidence of valve-related complications is low. Freedom from reoperation at 10 years is more than 90%.

Conclusion: Repair of the BAV has evolved over the past 2 decades. By addressing specific anatomic features excellent and stable results can be obtained.

Keywords: Bicuspid aortic valve (BAV); aortic valve repair; aortic aneurysm; valve-sparing aortic surgery

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Introduction

The bicuspid aortic valve (BAV) is the most common cardiac anomaly with an incidence of 0.5–2% (1,2). The BAV has 2 functional commissures of normal height, making it a bicommissural valve. A third, rudimentary commissure is almost always present and varies in height. Regurgitation develops at the age of ~30 years (3,4), in addition the patients may require surgery due to aneurysmal dilatation as early as the 3rd decade of life.

Repair techniques for significant aortic regurgitation (AR) and/or aneurysm related to BAV have evolved over the past 25 years. Initially, a number of reoperations were required at mid-term follow-up despite excellent short-term results after valve repair (5). Over the years different mechanisms for failure have been identified (6); by specifically addressing them at the time of surgery repair durability could be improved (7-9). The correction of concomitant aortic dilatation has contributed to improved long-term results (9). We review the current principles and outcomes of BAV repair.

Anatomy and pathophysiology of BAV and aortopathy

There are various classifications of BAV, with the classification proposed by Sievers most widely adopted (10). In this classification the BAV phenotype has been divided into 3 types based on the number of raphes, type 0, type 1, and type 2 (10). Type 0 BAV is rare, it has 2 almost equal cusps and no or only a minimal raphe with symmetrical commissural orientation (10). While this classification is frequently used, it is of limited use in the context of BAV repair. Type 1 encompasses all different circumferential orientations of a BAV, and type 2 is not a BAV, but a unicuspid aortic valve (11), which has a different pathophysiology, natural history and pattern of aortic dilatation (12).

The different fusion patterns noted in BAV are right-left, right-non, and left-non (13). From a surgical standpoint this variability is of limited relevance, since it has no influence on the type of procedure which only distinguishes fused *vs.* non-fused cusps. The existing variability in commissural orientation is less frequently appreciated, though it has recently been shown to have a strong effect on repair durability (14). The orientation of the 2 functional commissures may vary from 180° (i.e., a symmetric configuration) to 120–140° (an almost tricuspid configuration) (13,14). There is also variability in the degree of fusion, which seems to relate to commissural orientation (14). Symmetric commissures correlate with complete fusion, while the more asymmetric the commissural orientation, the less the fusion. In those instances, the rudimentary commissure may be of almost normal height. A new classification has recently been proposed which takes these different anatomic patterns into consideration (14).

The mechanism of the development of AR has common components in all phenotypes. Prolapse of the fused cusp is almost always present in regurgitant valves (6,15). With lesser degrees of cusp fusion, prolapse may primarily be present in the rudimentary right cusp. In a proportion of cases, the non-fused cusp may also exhibit prolapse (6), possibly as a result of long-standing regurgitation. Annular dilatation is frequently observed in regurgitant BAVs and its presence seems to impact repair durability if left uncorrected (6,7,16,17).

BAV-related aortopathy

Aortic dilatation is often present in patients with BAVs and can be a contributing factor to the pathophysiology of AR (3,4). In addition, continued dilatation of the aorta despite BAV repair can result in recurrent AR during follow-up. BAV is associated with dilatation of the proximal aorta independent of valvular dysfunction in roughly 50% of patients (3). Patients with BAV aortopathy have a 6 to 9 fold increased risk of dissection and aortic rupture compared with the general population (18,19). This appears to be related to the increased occurrence of dilatation per se rather than the BAV being a specific risk factor (18,19). The BAV associated aortopathy has been classified into different categories based on the site of dilatation (20,21). For practical purposes, it is reasonable to distinguish between aortopathy of the root type and the tubular type.

Why repair?

The native aortic valve has excellent hemodynamics, absence of thrombogenicity, resistance to infection, and maintained coronary flow reserve. It seems reasonable to assume that, similar to the mitral valve, repair of the native valve should translate into better and long-term clinically relevant outcomes.

Aortic valve replacement (AVR) with mechanical or biologic prostheses has long been the standard of care. Although it is an effective procedure, there are important long-term drawbacks, particularly in a young population. Mechanical prostheses are associated with reoperation rates in the range of 1% per patient year. In addition, survival in non-elderly adults following mechanical AVR is significantly lower than the age- and sex-matched general population (22,23) with a mortality rate of ~1% per year (22-24). Finally, the need for anticoagulation has a strong impact on quality of life (25) and is associated with risks of major hemorrhagic or thromboembolic events (26). Patients receiving biologic substitutes are at risk of structural valve degeneration, thromboembolism, patient-prosthesis mismatch and reoperation. In addition, excess long-term mortality in non-elderly adults has also been observed after biologic AVR (27).

Over the past 20 years, BAV repair has become a seemingly better alternative to AVR with favourable hemodynamics and survival (28-30). The incidence of valve-related complications is low (31-33), with repair failure being the most frequent complication. The risk of reoperation has been low. With careful patient selection and adequate repair and valve-preserving techniques, durability of >20 years has been documented (34).

History of BAV repair

An initial series of BAV repair was reported by Fraser and coworkers (35), consisting of free margin plication or triangular resection of the prolapsing fused cusp tissue. Subcommissural plication was added (36) to increase leaflet coaptation (35). Early results were promising; intermediate results, however, showed freedom from reoperation of only 87% at 5 years (5). Repair failure was due to progressive stenosis or recurrent regurgitation (5). Proposed risk factors for failure were triangular resection and dilatation of the ascending aorta (5,37).

Determinants of BAV repair durability

Effective height

In view of these observations we proposed a liberal use of aortic replacement in order to stabilize the repair (37). Subsequently we realized that symmetric prolapse or annular dilatation were still associated with repair failure (6). The presence of prolapse led to the concept of effective height, i.e., the distance between the central free margin and the annluar plane (38). In normal aortic root anatomy the central margins of the cusps are 9 to 10 mm above the annular plane (39). A caliper was developed for objective assessment of cusp effective height as configuration parameter intraoperatively (38). Since then, we systematically aim for an effective height of >8 mm (40). This has led to excellent results with marked improvements in durability (17,40).

Geometric height

It also became evident that the ability to correct cusp prolapse was related to the amount of existing cusp tissue. This was termed “geometric height”, i.e., the distance from the nadir of the cusp to the central free margin (41). In BAVs, geometric height of the nonfused cusp was found to be ≥20 mm in 95% of individuals with a mean of 24 mm. This cut-off value was then introduced as a surrogate parameter for the detection of cusp retraction. Using geometric height for selection of adequate repair substrates and measurement of effective height to guide correction of prolapse, repair has become reproducible and results more predictable (6,38).

Annular dilatation

The importance of annular dilatation on repair durability was later recognized (6,7,16). Annular dilatation (i.e., >25–27 mm) is present in the majority of patients with severe AR. It has been demonstrated that annular dilatation is an independent risk factor for recurrence (6,7,16). Stabilizing and/or reducing the aortic annulus at the time of surgery has been shown to significantly improve the durability of BAV repair (7,16,17).

Commissural orientation

In parallel, the relevance of commissural orientation became apparent (6). The best durability (6) and flow characteristics across the aortic valve (42) are seen with a symmetric configuration, i.e., a commissural angle of 160° to 180°. Standard repair techniques in patients with a commissural angle of <160° led to relevant systolic gradients and poor durability (6). The introduction of systematic modification of commissural orientation was shown to markedly improve systolic valve function and repair durability (8). Alternatively, liberal root replacement in patients with moderately dilated sinuses has been proposed in order to reposition the commissures at 180° (43).

Patch reconstruction of the cusp

Various studies have shown that use of pericardium as partial cusp replacement is an independent risk factor for early failure (6,9,44). Thus, whenever patch reconstruction is required for BAV repair, this should be balanced against durability concerns.

Current concepts of BAV repair

Cusp repair

Cusp repair is invariably required in isolated BAV repair for AR since cusp pathology is a key component. The most frequent cusp pathology is prolapse of the fused cusp. There may also be prolapse or retraction of the non-fused cusp. Correction of cusp configuration will also often be necessary in valve-preserving aortic surgery since the reduction of intercommissural distance will frequently result in relevant cusp prolapse (40).

At the beginning of the procedure, a detailed assessment of cusp morphology, fusion pattern, and circumferential orientation of the commissures has to be performed. Stay sutures are placed in the commissures and kept under tension to provide adequate exposure and mimic a pressurized aortic root (45). Annular dimensions are measured on transesophageal echocardiography and re-assessed by direct intubation using Hegar dilators. An annular diameter of more than 26 mm is considered as

Video 1 Bicuspid aortic valve repair (50).

dilated. Geometric height and effective height of the non-fused cusp (reference cusp) are measured (45).

Selection of adequate substrates for repair is important component of the procedure. The cusp tissue should be pliable and without calcification, there must be an adequate amount of cusp tissue. This is best determined by measuring geometric height in the non-fused cusp. A geometric height of ≥20 mm will be sufficient for repair (41). In order to identify prolapse reproducibly, intraoperative measurement of effective height with a caliper has shown to improve assessment of cusp configuration (38). In BAVs the level of aortic insertion of the 2 components of the valve varies. Effective height can only be reliably measured in the non-fused cusp, and this is then used as reference for the fused cusp. An effective height of 9–10 mm of the non-fused cusp has consistently led to stable results unless other complicating pathology was present. Induced prolapse due to reduction of intercommissural distance is a frequent finding if associated aneurysm is treated by aortic replacement involving the sinotubular junction. In such cases, it is important to perform cusp assessment after aortic replacement, i.e., when intercommissural distance has been reduced.

Central plication of the cusp free margin has proven to be the most reproducible technique for correcting cusp prolapse (35,46,47). Sutures placed in the pericommissural areas of the cusp can easily tear because this portion of the root is under highest stress (48,49). The plication is primarily done at the level of the free margin; depending on the extent of prolapse it may be extended into the belly of the cusp to limit postoperative billowing. Prolapse of the fused cusp is essentially always present, so the plication will be applied to this cusp regularly. If there is additional prolapse of the non-fused cusp, both cusps can be corrected without impacting the results negatively.

Not infrequently, dense fibrosis or limited calcification may be present in the raphe. If central plication is difficult because of these tissue changes, a triangular resection may be performed (45). We use fine interrupted polyethylene sutures (5-0) to re-approximate cusp tissue and avoid a continuous suture in order to minimize the chance of cusp retraction. If cusp calcification extends beyond the raphe, BAV repair should be reconsidered because of a high probability of limited durability.

Commissural orientation is an important part of BAV anatomy and must be taken into consideration when choosing the repair strategy. A commissural angle of 160–180° can be left as such (*Video 1*). If the angle is 140–160°, its modification has shown to decrease postoperative systolic gradients and improve mobility of the fused cusp and durability (8,9). This can be achieved by plication of the fused sinus from he base and to the level of the sinotubular junction (8). Alternatively it may be performed through root replacement and change of the configuration inside a graft for reimplantation (51) or also through appropriate configuration of the graft tongues in root remodeling (9). If the commissural angle is <140°, the valve is probably best treated in analogy to tricuspid aortic valves (46). In this setting, prolapse of the individual cusps is treated separately.

Concomitant aortic replacement

Patients undergoing cardiac surgery with an ascending aortic diameter of >45 mm can be managed with concomitant repair of ascending aorta/root for prognostic reasons (52). However, the recommendation does not differentiate between valve repair and replacement. Indeed, if the valve is repaired, concomitant aortic replacement (in mildly dilated aortas) has been associated with improved durability (6,51).

In deciding for or against root replacement we take into consideration the increased complexity of the procedure against the improved durability of repair after root replacement. Whenever root dimensions are enlarged (≥42 mm), ascending aorta and root are replaced if repair is performed in our routine. The graft size must accommodate the size of the patient as well as downstream aorta. In root remodeling the commissural orientation of the graft is placed at 180° (9). Two symmetric tongues are created to accommodate both fused and nonfused sinus (45). It is important to ensure that the commissures are placed high to avoid commissural restriction. Therefore, we propose suturing the graft from the nadir to the commissures, extending the graft incision as dictated by the patient's anatomy. Recently some data have been published using reimplantation for the same purpose. Early results have been comparable (53,54), while late results are not yet available.

If root dimensions are <43 mm (depending on age and body surface area of the patient), we refrain from replacing the root. Mid- to long-term observations have shown stability of the aortic root with such an approach (7,8,29).

Aortic annuloplasty

Annular dilatation is almost always present in regurgitant BAVs and has been associated with poor durability (6). Our definition of annular dilatation currently is a diameter ≥26–27 mm. We and others (40,47) had used subcommissural plication sutures previously. It was found that they did not stabilize the annulus sufficiently (16) and were actually associated with repair failure (6).

In choosing an annuloplasty technique the anatomy of the aortic annulus must be considered. The structure to be stabilized is not the true anatomic annulus, best represented by the crown-shaped fibrous structure of the cusp insertion lines (55). It is rather the caudal border of the root, which connects the cusp nadirs along a horizontal plane (also known as the basal ring). This should be considered as the functional annulus because it determines valve geometry (55). This basal plane can differ from the aortoventricular junction, with the aortoventricular junction lying more cranial than the virtual basal ring (55). In normal, tricuspid aortic valves, this difference is generally limited (56,57) while in BAVs a more pronounced distance (reaching up to 10–15 mm in height) may be encountered, particularly in the right coronary sinus (14,56,57). This can limit the extent to which an external ring can be brought down to the level of the basal plane. Therefore we favour addressing annular dilatation by performing a suture annuloplasty using expanded polytetrafluorethylene (PTFE; CV-0, Gore-Tex CV-0; WL Gore and Associates, Munich, Germany). This material has proven to be good for this purpose (7). The suture is placed circumferentially along the basal ring and tied around a Hegar dilator at the desired annular diameter. We commonly take a 25 mm Hegar for larger patients (>2 m^2 body surface area) and 23 mm for smaller individuals.

The annuloplasty can be used in isolated BAV repair or as an adjunct to valve-sparing root remodeling. Some early complications with initial use of different suture materials (7) could be largely eliminated with use of the PTFE suture (Gore-Tex CV-0; WL Gore and Associates, Munich, Germany). Regardless of any annuloplasty technique, careful attention to the specific anatomy of the left coronary artery is critical to avoid interference with the left main or the circumflex coronary. As yet, the addition of an annuloplasty to remodeling in BAV has resulted in a higher proportion of completely competent aortic valves (9,58); however, an improvement in freedom from reoperation has not yet been demonstrated (9).

When not to repair?

Limitations of repair are related to the morphology of the cusps and commissures. Currently, the need for cusp augmentation or partial replacement of cusps using patch material is associated with poor durability. This is related to early degeneration of autologous pericardium currently used for cusp replacement. We have also observed progressive calcification of the whole valve in patients who required partial cusp replacement after calcium excision. Thus, cusp retraction (geometric height <20 mm) or calcification of the raphe that cannot be treated by excision and direct approximation of cusp tissue are currently better treated by replacement. The presence of active endocarditis falls into the same category according to current knowledge.

Unfavorable commissural orientation will increase the complexity of the repair and decrease the durability (14). Although asymmetric or very asymmetric BAVs can be repaired, they represent a higher technical challenge to the surgeon. Closure of fenestrations in BAVs and the creation of a tricuspid design by a commissural reconstruction are associated with decreased durability in our experience. In such scenarios the threshold for replacement should be low in light of shorter expected durability.

Conclusions

Aortic valve repair has evolved over the past 2 decades. The techniques have become standardized and reproducible and offer a tailored and predictable approach to the majority of patients with non-calcified BAVs (59). Currently, BAV repair remains underused in the majority of centers, including high-volume aortic centers. Improved understanding of the mechanisms of regurgitation, predictors of repair durability, and surgical techniques will hopefully translate into wider adoption of this approach, similar to mitral valve repair. A new classification system that includes the anatomic features of the BAV in the context of surgical repair has been proposed (14). This will further improve the clarity and comparability of different surgical approaches in the future.

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Footnote

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