Congenital Heart Disease

Tech Science Press

Doi:10.32604/chd.2025.064675

REVIEW

Bicuspid Aortic Valve Disease, the Dilated Proximal Aorta, and the Surgical Treatment Options: A Narrative Review

Wilhelm Mistiaen*

Department of Rehabilitation Sciences, Faculty of Medicine and Health Science, University of Antwerp, Antwerp, 2610, Belgium
*Corresponding Author: Wilhelm Mistiaen. Email: wilhelm.mistiaen@uantwerpen.be

Received: 21 February 2025; Accepted: 20 June 2025; Published: 11 July 2025

ABSTRACT: The presence of a bicuspid aortic valve (BAV) is the most common congenital heart anomaly, which can remain asymptomatic for decades, if it is not a part of a syndrome, such as Turner syndrome or genetic connective tissue disorders. There are several classifications for BAV, each with its advantages and drawbacks. The condition can lead to valvular malfunction such as regurgitation and stenosis, but is often associated with dilatation of the aortic root, the ascending aorta, the aortic arch, or a combination. Altered flow patterns due to the valve dysfunction as well as the breakdown of elastin in the aortic wall could be responsible for this development. Published surgical series are usually small and research designs vary, which makes the formulation of universal recommendations for treatment difficult. This narrative review provides data from the most recent series in this respect. Often, the condition becomes symptomatic in patients who are about 10 years younger compared to those with a diseased tricuspid aortic valve. The timing of surgery of the valve depends on the degree of its dysfunction. Repair of a dysfunctional BAV should be attempted whenever possible because of the patient's age. The options for valve repair are summarized, including the need for the use as the effective geometric height. The use of a pericardial patch and the presence of calcified areas of the valve can be predictors for an increased need for reintervention. In those cases, a valve replacement should be preferred. If a dilatation of the ascending aorta or the aortic root is present, this should also be addressed surgically, but the threshold for such a procedure varies. Several techniques are available to treat a dilated ascending aorta and root. For the latter, remodeling and reimplantation can be applied. Depending on the patient's characteristics, the size of the dilatation, and the affected part, the most appropriate technique should be selected. This requires surgical expertise, which can only be obtained in high-volume centers.

KEYWORDS: Bicuspid aortic valve; aortic valve repair; aortic root aneurysm; ascending aorta dilatation

1 Introduction

A bicuspid aortic valve (BAV) is the most common congenital heart disease with a prevalence of 1% to 2% [1–7]. It is a heterogeneous condition with respect to the valve itself, the aortic root, and the ascending aorta [8]. More than 95% of BAV cases are sporadic cases and are not part of a recognizable syndrome [8]. The existence of BAV becomes usually evident later in life because of progressive valve dysfunction, such as aortic stenosis (AS), aortic regurgitation (AR), or mixed disease [2,4,5,9,10]. However, the condition might remain silent during the entire life [11]. BAV dysfunction can occur at an earlier age compared to the degeneration of a native tricuspid aortic

valve (TAV). In stenotic BAV specimens, increased collagen deposition and signs of inflammation can be found [12]. Patients with BAV and AR are younger compared to those with AS [4,5]. AR could be the consequence of dilatation of the ventricular-aortic junction (VAJ), sinuses of Valsalva (SOV), the sinotubular junction (STJ), and ascending aorta, prolapse of a leaflet as a result of excessive cusp tissue or of commissural disruption and retraction of a cusp due to fibrosis or calcification [13]. Patients with BAV are also at risk for infective endocarditis (IE), which might be eleven times higher than that of the general population [14]. The rate of endocarditis in patients with BAV seems to decrease gradually with age, from 4.4% in patients younger than 44 years to 1.4% in patients 75 years and older [15]. Early detection of BAV is important to prevent IE and provide aggressive surgical treatment should IE occur in these cases [11].

In the majority of patients, BAV is the result of a fusion of two functional cusps conjoined by a raphe or fibrous ridge, and with three distinguishable sinuses [8,16]. This is also known as the Sievers type 1 BAV. The non-fused cusp with its corresponding sinus is often enlarged. The left-right (L-R) fusion is the most commonly observed type [6,8]. Other types are the left-non-coronary (L-NC) and R-NC fusions. A second consideration is the retraction of the conjoined cusp as well as the sizes of the cusps, leading to different degrees of asymmetry, which is of critical importance for planning valve repair in the case of AR [8]. There is a varying degree of asymmetry based on the orientation of the commissures with angulations between 120°–139°, 140°–159° and 160°–180° [13]. The two-sinus BAV has two cusps and two sinuses of about the same size, a 180° angle of the commissures, and the absence of a raphe. The more common phenotype is the laterolateral location of the cusps. This is also known as the Sievers 0 type. In a rarer Sievers 2 type, there are two raphes, conjoining two of the three cusps, mostly L-R and right-noncoronary (R-NC) [8,16]. A partial-fusion BAV has an unknown prevalence and shows a variable and partial fusion with a mini-raphe. It resembles a TAV, but its presence could be suspected on transthoracic and transesophageal echocardiography (TTE/TEE) and be confirmed by computer tomography and magnetic resonance imaging (CT/MRI) or during surgery. This type of BAV shows three developed cusps and a triangular opening at the systole. The commissures have an angle of 120°. This anatomy is responsible for an eccentric flow with vortices and a higher prevalence of aortic dilatation [8].

BAV is associated with progressive dilatation of the root, the ascending aorta [17], and sometimes of the aortic arch [18]. There is a reported prevalence of 20–40% [16], and in some populations over 50%, with a male predominance. However, no clear age-related increase in dilatation of the aortic root has been observed [17]. Abnormal flow patterns, such as vortices and asymmetric flow [19] caused by BAV, could promote aneurysmal degeneration of the proximal aorta, but genetic disorders were also implicated [3]. These anomalies involve fragmentation of elastin fibers, and interlamellar degeneration with mucoid replacement. However, there is no consensus about their degree of involvement [6]. Aortic dilatation carries a risk of intimal tears and dissection [2,20]. Once such aneurysmal dilatation develops, circumferential and longitudinal wall stress increases at the level of the aortic root. This seems to be of more importance in patients with BAV, compared to those with a native TAV. There are several types of BAV-related aortic dilatation. The dilatation of the ascending aorta is more common in patients with predominant AS, and the dilatation of the root in patients with predominant AR [15,21]. The dilatation could also develop from the root into the ascending aorta. These differences in types of aneurysms might be explained by flow patterns detected by 4D-cardiac MRI. The R-NC fusion and AS are more associated with

dilatation with the ascending aorta and arch and less with dilatation of the root. The R-L fusion could cause a higher wall shear stress, and dominance of the root phenotype [8].

Accurate medical imaging is pivotal for the diagnosis and choice of surgical approach in patients with BAV. It also allows the assessment of the severity of valve dysfunction. Classic TTE might be insufficient to distinguish between the different types of BAV, or show discrepancies of aortic sizes. CT and Gadolinium-contrast based cardiac MRI have a high spatial resolution and allow for adequate imaging of the subtypes of BAV and the entire aortic root [8]. A 4D-flow Cardiac MRI (CMR) allows for the visualization of altered blood flow characteristics, its dependence on BAV phenotypes as well as wall shear stress, which is implicated in vascular remodeling [8]. In R-L fusion BAV, the flow impinges on the outer curvature of the proximal ascending aorta and could cause root dilatation. An R-NC fusion displays a posteriorly directed flow jet directed towards the proximal ascending aorta and the outer wall of the distal ascending aorta, causing a more distal dilatation [8]. Volume change of the root over the cardiac cycle was comparatively greater in BAV. Aortic root dynamics were most significantly different at the level of the SOV in BAVs with moderate to severe regurgitation than in competent TAVs and BAVs [22].

The the American Heart Association/American College of Cardiology (AHA/ACC) guidelines [23], recommend TTE as the first-line imaging mode for BAV, the aortic root, and ascending aorta with a class of recommendation (COR) 1 and level of expertise (LOE) B. Cardiac CT and MRI are alternatives if TTE is insufficient, with a COR 1 and LOE C. in presence of an aortic size of 40 mm or more, a lifelong evaluation is considered as necessary, with intervals depending on growth rate. A comparable policy is recommended after surgical aortic valve replacement (SAVR) or after aortic valve repair, with a COR of 1 and LOE C. The timing and type of surgery for the replacement of the aorta are dependent on the anatomy of the aorta, patient characteristics, and institutional expertise. For patients with an asymptomatic BAV with an aortic size of at least 55 mm, aortic surgery is recommended with COR 2a and LOE B. The risk for an acute dissection is very low if routine surveillance of the aorta is performed. If the aortic size is between 50 and 54 mm and a risk factor for future dissection is present (a family history of acute dissection, coarctation, growth rate of over >5 mm/year), performing aortic surgery is reasonable, with COR 2a and LOE B. In patients with a dysfunctional valve, for whom a SAVR is indicated, and the aortic size is at least 45 mm, aortic surgery is a reasonable option (COR 2a and LOE B). Without performing additional aortic surgery, the risk for future dilatation and acute dissection is unknown. In patients with a well-functioning BAV and criteria for replacement of SOV, the performance of a valve-sparing root replacement (VSRR) is a reasonable option (COR 2b and LOE C). In patients with asymptomatic BAV, a low risk and an aortic size between 50 and 54 mm and without additional risk for acute dissection performing VSRR is a reasonable option (COR 2b and LOE B). The indications for the timing of aortic valve intervention in patients with a dysfunctional BAV are similar to those for TAV. The choice of prosthetic valve type in patients with a BAV is similar to that for patients with TAV. In patients with BAV and severe AR, a repair can be considered (COR 2b and LOE C). In patients with BAV and severe AS, a TAVI procedure might performed (COR 2b and LOE B), but the relatively young age of BAV patients should be considered. While both the 2020 AHA/ACC and 2021 European Society of Cardiology/European Association for Cardio-Thoracic Surgery (ESC/EACTS) guidelines recognize the increased risk of aortic dilation in BAV, they differ in timing and criteria for surgical intervention. The ESC emphasizes aortic diameter thresholds >50 mm in the presence

of risk factors, whereas AHA guidelines are slightly more conservative, recommending surgery at 55 mm unless high-risk features are present. These differences reflect the ongoing debate around optimal surgical timing in asymptomatic BAV patients.

This narrative review deals with the treatment option of a dysfunctional BAV and, if affected, the aorta. For this reason, a search for the last 5 years was performed through Web of Science, with the MeSH terms: "bicuspid aortic valve AND surgical aortic valve replacement NOT transcatheter". Manuscripts dealing with aortic valve repair, VSRR, repair of the ascending aorta, hemi-arch, and aortic arch replacement were included. Pediatric series and series dealing with acute complications were excluded. Also excluded were studies of BAV with other major cardiac and aortic congenital malformations and genetic connective tissue diseases were also excluded. These conditions could affect the prognosis to a higher degree compared to BAV and aortopathy itself, and could therefore confound the postoperative outcomes.

2 Literature Overview

A variety of documents were identified concerning the surgical approach of the valve and the proximal aorta (the root as well as the ascending aorta), and functional imaging depicting the flow abnormalities in the aorta.

2.1 Aortic Valve Regurgitation and Its Option for Repair

There are three types of aortic valve AR. The first and most common type is the result of a dilatation of the aortic root. The mobility of the leaflets in these patients remains unaffected. The second type of AR is the result of a leaflet prolapse, which is defined as an effective height of less than 9 mm [24], resulting in an eccentric jet. The third type of AR is the result of restriction of the leaflet by a degenerative or rheumatic valvular disease with thickening, fibrosis, and calcification. This is accompanied by a peak gradient of over 20 mm Hg and is a marker for poor durability of its repair [13]. Cusp retraction and a combination of cusp and commissural anomalies are less common [25]. A preoperative distinction of these types by imaging is an important issue for the choice to replace or repair the valve. Aortic valve repair could be performed in the majority of patients with BAV, but this has a higher risk for a need of reintervention [4]. To keep this risk as low as possible, repair of the valve and VSRR should be a part of the armamentarium of the cardiac surgeon. SAVR as treatment for a dysfunctional BAV is more simple and reliable but carries the risks of valve-related complications and the need for anticoagulation [21]. These complications have to be taken into account since BAV patients are younger and have better survival [26].

The main goal of the repair of the regurgitating valve is the restoration of an adequate coaptation zone that remains effective and durable. The introduction of the anatomic repair concept for the valve, with the aim to achieve an effective height of 9–10 mm, has reduced the incidence of reoperation rate threefold, from 24% to 8% [27]. The effective height of the affected cusp and free margin length should thereby be measured during the operation [24,28,29] since these data are critical in the performance of the repair. The need for expertise is illustrated by the improvements in long-term results over time [4]. The different techniques are presented in Table 1. Plication sutures of the cusp can be used in single, conjoined, and bi-leaflet structures but also to close defects [4,22]. Pericardial patches are used to augment the cusps, and reconstruct commissures, as attempts to "tricuspidize" a BAV [24,25], close perforations or treat retractions of

raphes [23,26]. The use of pericardium and decalcification during repair can be associated with a higher need for reoperations [24,27]. However, resecting and shaving of a raphe is protective against the development of AS [30]. "Bicuspidization" results in an almost straight line of coaptation a more physiologic flow and lower wall shear stress at the level of the STJ. From a hemodynamic point of view, symmetric valve geometry should be attempted in most BAV repairs [8,31]. A more challenging "tricuspidation" should be attempted in cases with a higher degree of asymmetry and with retraction of the raphe [8]. A higher degree of asymmetry could also affect the time to reoperate [27]. An annuloplasty reduces valve regurgitation in the long term [28]. In patients with a suture annuloplasty, a second band at the STJ, would not be needed if the ascending aorta is replaced, because the proximal anastomosis of the ascending graft would stabilize the STJ [8]. During valve-sparing root procedures, CV-0 sutures are routinely employed to reapproximate cusp edges or reinforce the annulus, providing precise tension control and minimizing tissue stress. Their use is particularly beneficial in BAV repair where cusp asymmetry necessitates delicate realignment.

The evaluation of the surgical result can only be reliably performed during the operation, although TEE before installation of the CPB might be helpful in measuring the angle and planning the repair [8]. A failure to attempt to repair the valve in the first session might be a predictor of the future development of AS [30]. Since the use of a pericardial patch in cases of calcified zones increases the risk for reoperation [28], SAVR seems preferable over the use of a pericardial patch in cases with calcification [27]. In the presence of calcifications, immobile cusps, severe fenestration, or an inadequate geometric height, SAVR should be performed. If an aortic dilatation is present, a Bentall procedure is the best option [32].

2.2 Options for the Root and the Ascending Aorta

The decision for valve surgery in BAV cannot be disentangled from the problems posed by an affected aortic root [32], since aortic valve regurgitation can be a consequence of the aortic dilatation. Dissection is a major complication of aortic dilatation [8,33]. This complication is potentially fatal and can be prevented by timely surgery. The relationship of BAV with aortic complications is illustrated by the observation that patients with an acute type A aortic dissection have a higher association with BAV, compared to the general population, especially when AR is present [33–35]. Root replacement is often required in patients with BAV [10]. However, it is still not well established which is the best surgical strategy and when a specific technique should be preferred in root surgery, especially regarding BAV anatomy [6]. For a dilatation limited to the ascending aorta, an intervention (AAI) is indicated, but there is a significant variation in the timing for such a procedure [36]. The decision to perform surgery does not only depend on the size of the aorta but also on the risk factors for dissection such as AR and hypertension. Without these risk factors, the proposed threshold is at 55 mm [5,8]. However, the value of 55 mm might fail to prevent complications [33]. In the presence of risk factors, this threshold is set at 50 mm [8,37]. For specific genetic syndromes, which carry an increased risk for the presence of BAV, separate guidelines have been proposed [23]. In patients with symptomatic AR and ascending aortic diameter >45 mm, a replacement was advocated [5], and more than one-third of patients with a BAV underwent AAI at aortic diameters <45 mm. Patients with higher age and the presence of aortic valve stenosis were more likely to receive SAVR without AAI, but the risk-benefit ratio of

routine aortic interventions at smaller diameters needs to be determined [36]. This is in line with the observation that the aortic root does not dilate over time when not replaced. Adherence to the 45 mm size threshold for AAI protects against adverse events after SAVR. The cutoff for AAI might be lowered to 45 mm, without additional short-term risks [38]. Moreover, if the aortic dilatation was a consequence of altered hemodynamics and not by genetic factors, the risk for complications might disappear after valve surgery [35]. Nevertheless, even patients with roots smaller than 35 mm with RL-BAV would require more surveillance after AAI [6]. The balance between the risk for dissection and potential postoperative complications should be based on the patient's characteristics and also on the available surgical expertise [33]. The absolute size as a parameter might be insufficient to determine the need for aortic intervention. Some authors advocate the use of indexed sizes of the SOV, STJ, and ascending aorta using the patients' height for indexing. The existence of abnormal indexed sizes with absolute values between 45 and 50 mm holds a significant risk [33]. Moreover, there is the gender effect: female patients with BAV referred for surgery were older, and had a more significantly diseased aortic wall with larger indexed sizes and higher risks for complications. These findings indicated that women should be monitored more closely [39]. All these findings made reproducible serial imaging to monitor the progress of an aortopathy all the more important [8]. There was no consensus to extend the repair into the aortic arch to avoid future risks for arch-related complications. This more complex and longer procedure involved a circulatory arrest. Its necessity was not established, since there is no documented clinical benefit with respect to in-hospital complications, long-term survival, or reintervention rate [16]. A more extensive resection into the arch for patients with non-syndromic BAV seemed generally not justified when the ascending aorta was replaced [8].

Table 1: Options for valve repair.

Reference	Problem	Option	Comment
Froede et al., 2022 [28] Miyahara et al., 2020 [29] Patel et al., 2021 [5] Yokawa et al., 2020 [13] Levine et al., 2023 [4]	Prolapse of the cusps	Correction by centrally placed plication suture	Can be used in single and bi-leaflet structures
Saku et al., 2024 [24]	Closing defects	Suture	
Spadaccio et al., 2022 [30]	Need to reinforce cusp	PTFE free-edge running suture	Risk for future aortic valve stenosis
Froede et al., 2022 [28]	Non-aligning of fused cusp with unaffected cusp	Surgical aligning	
Froede et al., 2022 [28] Patel et al., 2021 [5] Kari et al., 2022 [40]	Fibrosis/calcification of raphe	Triangular resection of raphe	Only in minority of cases
Spadaccio et al., 2022 [30]	Fibrosis/calcification of raphe	Resecting or shaving of raphe	Protects against future aortic valve stenosis
Karliova et al., 2020 [25] Froede et al., 2022 [28] Kari et al., 2022 [40] Yokawa et al., 2020 [13] Saku et al., 2024 [24]	Perforation; retraction; need to augment cusp	Autologous glutaraldehyde fixed pericardial patch	Used less frequently, low durability in BAV compared to TAV, risk for reoperation risk for higher transvalvular gradients

Table 1: *Cont.*

Reference	Problem	Option	Comment
Yokawa et al., 2020 [13] Levine et al., 2023 [4]	Repair of calcified areas	Decalcification	Less in use, risk for reoperation
Yokawa et al., 2020 [13] Levine et al., 2023 [4] Patel et al., 2021 [5] Kari et al., 2022 [40] Karliova et al., 2020 [25]	Need to reconstruct commissures	Commissuroplasty or commissurotomy in a lesser degree	Less in use, the risk for reoperation, especially with a patch
Michelena et al., 2021 [8] Petersen et al., 2021 [31]	Bicuspidization to obtain a more physiologic flow	Plication of aortic wall to reduce the circumference of the annulus	
Karliova et al., 2020 [25] Saku et al., 2024 [24]	High degree of asymmetry	Tricuspidation	Is very challenging; might involve a patch
Froede et al., 2022 [28] Miyahara et al., 2020 [29]	Unstable annulus	Suture annuloplasty at the basal level of the root	Use of Hegar dilator as sizer; reduces risk for AR

The technique for VSRR requires an inspection of the valve cusps for calcification, perforations, fenestrations, elongation of the free margin, and prolapse. The findings determine the repairability of the valve. During a VSRR, the root should be completely dissected free from the surrounding structures, with excision of all abnormal parts of the sinus, leaving a 4–5 mm rim of aortic tissue along with the annulus. The coronary buttons are dissected free and mobilized [5]. Several options are available: (1) a straight tube graft can be reimplanted, without creation of neo-sinuses; (2) a remodeling according to Yacoub, with a three-tongued graft sewn at the annulus and small remnants of the sinuses, (3) with or without annular support and (4) reimplantation by creating a neo-STJ and neo-sinuses by plication of the graft or using separate grafts, the proximal one being the larger [40]. Care should be taken to maintain an optimal coaptation height by an appropriate suturing of the commissures and respecting the native anatomy. Once these positions are finalized, the commissural sutures are tied to the graft, and the coronary buttons are reimplanted. The root is fully inspected to see if any adjunctive cusp repair is needed [5].

In cases with a severe AR and a normal-sized aortic root but a dilatation of an ascending aorta, a valve repair could be performed with an STJ remodeling by using a tube graft and external sutures at the basal ring around a Hegar dilator [24]. Remodeling of the root has become the primary treatment for root enlargement and a noncalcified aortic valve, irrespective of the degree of AR. The remodeling reduces the annular diameter and improves the coaptation height. A suture annuloplasty might improve the durability of the repair. Measurement of an effective height allows the detection of any prolapse. One should be aware of aggravated prolapse by reducing the distance between the commissures [29]. In case of an asymmetrically dilated root with an R-L type BAV, a supra-coronary replacement was proposed with an extension for the replacement of the NC-cusp. Results were good in patients with small roots and only a mild residual AR [6].

The “Florida sleeve” technique was proposed for AR due to the dilatation of the root. This technique involves four sub-annular anchoring sutures placed two to three mm below the nadir of the leaflets. Three sutures are in line with the commissures and the fourth is placed under the noncoronary cusp. The keyhole for the coronary arteries is cut after the sleeve is temporarily seated. After the definitive seating of the sleeve, the slits in the graft below the coronary keyholes are

sutured. The running horizontal mattress suture both suspends the aorta and orients the posts of the commissures. Redundant aortic wall at the STJ should be made to overlap with small pleats. The procedure seems safe and offers durable results [41].

A reduction aortoplasty is not recommended as a first-line procedure but seems applicable in patients of higher age who are unfit for extended aortic surgery. Connective tissue disorders are clear contraindications. A longitudinal aortotomy just proximal from the clamp to the NC sinus is followed by a wedge-shaped excision and a double-layered closure, which can be reinforced with a pericardial strip. This technique preserves the Windkessel-effect but there is a risk for recurrent dilatation, especially with high preoperative aortic diameters or with underlying histopathologic aortic wall changes associated with BAV. Support by an external wrapping with a dacron prosthesis aims for stabilization but with a risk for prosthesis dislocation and aortic tissue lesions. The procedure itself is safe, with a low complication rate and a good durability and survival, irrespective of the valve morphology. An adequate pre- and intraoperative tissue quality assessment and patient selection are crucial [37]. Extravascular wrapping of a mildly dilated ascending aorta as a sole procedure in patients with BAV-related aortopathy is also safe and easy to perform. The wrapping excludes the SOV and the coronary arteries. Comparison of SAVR with and wrapping versus ascending aorta replacement showed longer CPB and ACC times for replacement but without difference in adverse postoperative events and good mid-to-long-term outcomes. Wrapping in these cases could be considered as a prophylactic measure [21].

If a dysfunctional valve is repairable, VSRR for aortic dilatation is the preferable option [4,5,42]. This might be better compared to a SAVR or a composite graft, at least in experienced centers [4,5]. The preservation of a native valve avoids potential valve-related complications and the need for anticoagulation [3]. In the presence of calcification, immobile cusps, severe fenestration, or geometric height below 18 mm, a Bentall procedure should be performed [32,42]. A conversion from valve repair to SAVR might be needed in a high degree of asymmetry of BAV and of the root with a higher severity of AR in BAV Sievers type 1, other than L-R fusion [40], although "tricuspidation" could be attempted in these cases [28,29]. The postoperative mortality is lower in VSRR compared to Bentall, but the rate of recurrent AR is higher [42]. Although VSRR with valve repair in young patients with a regurgitating BAV has a good 15-year survival rate, a high reoperation rate of 31% was observed [27]. With proper patient selection, VSRR in BAV is considered a durable option [5]. Selected patients with a diameter of the aorta below 55 mm, undergoing SAVR showed a low rate of rupture, dissection, or sudden death. This seemingly contradicts the guidelines using 45 mm as a threshold. This observation, however, needs confirmation by future studies [35].

2.3 Postoperative Outcome

The durability of valve and aortic operations has its importance because of the relatively young age of patients with BAV, with a consequently longer life expectancy. In patients with BAV and mild-to-moderate AR, valve dysfunction will not worsen after isolated replacement of the ascending aorta. The need for reintervention was low at 12 years [7]. The 10-year reoperation rate after VSRR was also low [5]. After ARR, the 15-year reoperation rate is increased, especially for the repaired valve. The survival rate was comparable to that of the age and gender-matched general population [10]. Recurrent root dilation after ascending aorta replacement with or without replacement of the NC-sinus in the long term was predicted by an aortic root diameter at discharge

> 35 mm, a preoperative AR labeled as more than mild, and no SAVR. Age and replacement of the NC sinus had a borderline effect. Reoperation for aortic root dissection or dilation was extremely rare at follow-up, so full replacement of the aortic root when this was only mildly dilated still does not appear justified [6]. A VSRR of the David V type which creates neo-sinuses showed a good 12-year survival with low reoperation rate in patients with BAV as well as TAV [4]. Conformance to the 45 mm size cutoff for preemptive ascending aortic replacement during SAVR in BAV patients was not associated with an increased risk for early adverse events [38]. A valvuloplasty of BAV showed higher transvalvular gradients compared to a repair of a TAV. These higher gradients did not affect rates of regurgitation and reoperation [24].

The fate of the remaining aorta (i.e., the SOV and remaining distal ascending aorta) after combined SAVR with ascending aorta surgery for BAV after excluding patients with isolated SAVR, aortic root replacement (ARR), aortic arch surgery or connective tissue disease was also a matter for debate. Most patients under study had AS. A mixed AS-AR and pure AR were less common. One-quarter of the patients underwent surgery with circulatory arrest. Serial CT exams showed that the remaining SOV diameters increased hardly, while there was a significant increase in the remaining distal ascending aorta. Only 1 patient required surgery for proximal pseudoaneurysm, and none for dilatation of the remaining distal aorta. Long-term survival was high. Rapid dilatation of the residual aorta rarely occurred in patients with a BAV who underwent SAVR and grafting of the ascending aorta in the mid-term follow-up. For selected patients with a surgical indication for ascending aortic dilatation, simple SAVR and graft replacement of the ascending aorta may be sufficient surgical options [17]. In selected patients with a BAV with a >45 mm ascending aorta and normal roots and arches, the replacement of the ascending aorta by a graft without concomitant root and arch procedure produced excellent survival rates, freedom from MACCEs, and fewer reoperation and aorta-related events in the mid-term follow-up. Rapid dilatation of the residual aorta rarely occurred in patients with a BAV who had undergone SAVR and grafting of the ascending aorta. For selected patients with a surgical indication for ascending aortic dilatation, simple AVR and grafting of the ascending aorta may be sufficient surgical options [17].

2.4 Outcome in BAV versus TAV Patients

Table 2 shows the comparison between the demographic, operative, and postoperative results (survival, reoperation rate) of patients with BAV versus TAV. The included articles had a very different design, which makes a comparative analysis difficult. In all series, the BAV patients were at least 10 years younger, compared to their TAV counterparts. This was also the case in two series before a propensity score match (PSM) analysis was performed. The mean ages were 57 years versus 68 years [43] and 44 years versus 54 years [4]. In these series, patients with BAV had a higher rate of aortic arch procedures. This younger age in patients with BAV translated also in lower mean Euroscores II of 1.8% versus 2.7% [44] and 1.6% versus 2.6% [45]. A low Society of Thoracic Surgeons risk score of about 0.7 was observed in other single-arm series studying BAV patients with mean ages below 40 years [31], while in another small series, mean Euroscores of 2.1 for Sievers type and 2.9 for Sievers type 0 were observed. These patients, however, had mean ages of 62 and 64.5 years [46]. In all series 30-day complication and mortality rates were low. The age of BAV patients also allows for a long follow-up with respect to reoperation rate and survival. The results in Table 2 show that for patients with a TAV morphology, long-term survival varies widely.

In three comparative series, patients were treated with SAVR instead of a repair. Long-term survival was lower in TAV patients, which could be attributed to the age difference. Reoperation rate differed in one series, where a stentless device was implanted. In one series, where a propensity score match analysis was performed between patients with BAV and TAV, the presence of BAV was identified as an independent predictor in favor of long-term survival. Because of the research design, preoperative and operative characteristics were comparable for both patient groups. The reoperation rate for BAV patients was higher, but a higher mortality rate in TAV patients, as a competing event, could contribute to this observation [43]. Biological valve prostheses should be used with caution in these patients. A Resilia valve, which is designed to prevent or at least delay calcification by capping residual aldehyde groups, showed encouraging results with respect to durability [44]. The use of rapid deployment valves such as Intuity seemed less favorable in patients with BAV [43] and was more appropriate in older and higher-risk patients because of the shorter operation times. The presence of BAV in patients with infective endocarditis was also identified as an independent protective predictor for short- and long-term mortality. These patients were also younger and healthier compared to their TAV counterparts since infective endocarditis occurs generally sooner in patients with BAV [11].

Comparative series concerning repair with pericardial tissue showed worse results, especially in patients with BAV [25]. Aortic valvuloplasty with annuloplasty and a tube graft showed better early postoperative results in patients with TAV compared to those with BAV. Patients with a repaired BAV have a higher transvalvular gradient, but this did not affect mid-term outcomes nor the reverse remodeling of the left ventricle [24]. However, VSRR for patients with BAV, such as the Florida-sleeve technique [41], a tube graft [29] with external annuloplasty and STJ remodeling [24], a Valsalva graft with the commissural resuspension and other necessary valve repair procedures [4], showed acceptable outcomes.

Table 2: Series comparing BAV and TAV patients.

Author	Valve	*n*	Proceed.	Aim	Mean Age (year)	Mort	Survival (%)	Re-Op Rate (%)
Aalei et al., 2020 [41]	BAV TAV	18 159	Florida sleeve	Durability Safety	47.8 ± 11.2 49.6 ± 15.8 NS	0 1.9 NS	100 91 NS at 1 year	7 1 $p = 0.042$ at 8 year
Bavaria et al., 2024 [44]	BAV TAV	214 458	SAVR SAVR	Resilia	59.8 ± 12.4 70.2 ± 9.5 $p < 0.001$	0.9 1.3 NS	95.9 (93.0–98.7) 86.3 (83.0–89.7) <0.001 at 5 year	98.5 (96.8–100.0) 98.8 (97.7–99.8) NS at 5 year
Brown et al., 2022 [43]	BAV TAV	330 330	SAVR SAVR	Stentless	62 (52–71) 63 (52–72) After PSM	1.8 1.0 NS	46 33 $p = 0.002$ at 15 year	15 11 $p = 0.048$ at 15 year
Coti et al., 2022 [45]	BAV TAV	107 690	SAVR SAVR	Intuity	67.8 ± 8.4 74.4 ± 7.0 <0.001	0.0 0.6 NS	88 50 (estimated) $p < 0.001$ at 10 year	2.8 1.9 NS
Karliova et al., 2022 [25]	BAV TAV	136 139	Repair Repair	Pericardial patch	53 ± 14 (all pts)	0.4 (all pts)	56 (all pts) - NS at 15 year	64 31 $p < 0.001$ at 10 year
Le et al., 2021 [11]	BAV TAV	51 159	SAVR SAVR	Infective endoc	54 (44–65) 42 (30–56) <0.001	2.0 5.0	63.8 45.5 $p = 0.019$ at 10 year	5.7 4.5 $p = 0.045$

Table 2: *Cont.*

Author	Valve	*n*	Proceed.	Aim	Mean Age (year)	Mort	Survival (%)	Re-Op Rate (%)
Levine et al., 2023 [4]	BAV TAV	156 156	VSRR VSRR	Durability of repair	45 (38–55) 46 (36–53) After PSM	0.0 0.8	92.0 89.9 NS at 12 year	15.7 5.7 NS at 12 year
Miyahara et al., 2020 [29]	BAV TAV	414 589	VSRR VSRR	Durability and Survival	54 ± 15 (all pts)	1.6 - (all)	86.1 - At 15 year (all pts)	20 6 0.003 at 15 year
Saku et al., 2024 [24]	BAV TAV	41 67	Repair Repair	Hemodyn	40 (26–49) 57 (50–66) $p < 0.001$	0.0 0.0	95.2 - At 5 year (all pts)	94.2 - At 5 year (all pts)

BAV: bicuspid aortic valve; endoc: endocarditis; NS: not significant; pts: patients; PSM: propensity score match; proced: procedure; TAV: tricuspid aortic valve; y.

2.5 Effect of Surgery on the LA and the LV

The existence of BAV has also consequences beyond the valve and the aorta. The eccentric opening of a BAV and the altered elasticity of the aorta can contribute to a chronically elevated afterload imposed on the left ventricle (LV). This leads to the observation that an impaired LV function is more common in patients with BAV, compared to age and sex-matched controls with dysfunctional TAV [47,48]. Especially adolescents and young adults with a BAV have early echocardiographic signs of LV remodeling [49]. In patients with predominantly AR, an eccentric LV hypertrophy was observed [1]. Patients with isolated severe AS had, compared to TAV counterparts, a higher degree of ventricular systolic and diastolic dysfunction [50].

There were also other differences between patients with a dysfunctional TAV and BAV. Table 2 shows that patients with BAV dysfunction are mostly younger compared to those with TAV disease and have fewer comorbid conditions. The increased LV compliance in younger BAV patients could lead to a more prominent dilatation of the LV. This is an important observation since about half of the operations on the aortic valve are based on an increased end-diastolic diameter of the LV [41–54]. Patients with BAV show also a higher risk for early congestive heart failure [55,56]. If a severely reduced left ventricular ejection fraction (LVEF) is present in patients with AR, the risk for persisting lower LVEF is higher after corrective surgery [57]. Early valve surgery should be recommended since survival is reduced with a decreased preoperative LVEF [58].

The extent of postoperative reverse remodeling of the LV depends on the preoperative size of the LV [59]. Early surgery should occur because of its beneficial effect [47,59]. It should be noted that many patients with LV end-diastolic dimensions below the current surgical thresholds for surgery, did not experience a normalization LV size. Compared to patients with isolated AS in patients with TAV, those with BAV had a lower preoperative LV function and higher LV mass index. Patients with BAV required significantly more postoperative inotropic support [60]. They also had a higher cumulative incidence of postoperative admissions for congestive heart failure compared with patients with TAV. However, survival was not different between these groups. Patients who had BAV with AS might benefit from closer surveillance and possibly earlier intervention. For these reasons, patients with BAV and TAV with isolated severe AS should not be regarded as one entity [60].

BAV disease also affects LA remodeling and function to a stronger degree than a diseased TAV. Aortic valve morphology is associated with preoperative LA reservoir function [48,58]. Even

with an earlier intervention a more severely affected LA function in patients with BAV, has the consequence of a higher risk for postoperative atrial fibrillation. Ischemic neurologic events during follow-up are more common in AS patients with BAV morphology, probably due to the effect of a diseased BAV on the LA function [61].

3 Discussion

BAV is a heterogeneous condition and its classification systems vary considerably [62–67]. Series is based on anatomopathological specimens [64] echocardiographic evaluation [63,66,68], CT-angiography [62,65], and cardiac MRI [67]. Some of these imaging reports included a clock face description and an addition of the concept of partial fusion and asymmetry of the leaflets [67]. These descriptions show varying emphasis on the orientation of the cusps, the fusions of the cusps, and the presence of raphes. The heterogeneity in classification is illustrated by the introduction of several attempts using numbers and letters [63,64]. Some classifications were very simplistic and dichotomous [66] others were complicated and included also the aortic phenotypes [62]. The Sievers classification is widely adopted, but it has some shortcomings. First, this classification does not take into account the division into the angle between the commissures which can be graded as symmetrical (between 160° and 180°), asymmetrical (between 140° and 159°), and very asymmetrical (between 120° and 139°). Second, the unicuspid and quadricuspid aortic valves were not explicitly described in this classification [64]. Third, the potential absence of a raphe in fused cusps, the concept of incomplete fusion, and the aortic phenotypes were not incorporated in the Sievers classification. These features are an important aspect in the repair of regurgitating BAV. A repair of BAV aiming for a commissural angle of 180° shows a physiological hemodynamic and results in better long-term durability. Attempts to achieve a tricuspid morphology are only recommended for repair of very asymmetrical valves, in order to avoid postoperative stenosis [58]. The concept of symmetry of the BAV is a critical element in the surgical repair of BAV [69,70]. Symmetry can be achieved by plication of the fused sinuses from the outside or from the inside in their basal circumference by Teflon-pledget sutures. A peak tricuspid valve gradient (TVG) after such repair should be kept below 20 mmHg for reasons of durability. A comparable concept was applied by other authors in a repair-oriented approach for aortic valve regurgitation [13]. Recent progress has achieved similar reoperation rates between BAV and TAV valve repair. To achieve such an outcome, a uniform and consistent system is desirable, taking into account the hemodynamic and clinical effects of the observed differences in morphologic valve abnormalities. It should provide for a common language between geneticists, interventional cardiologists, surgeons, and specialists in echocardiography, CT, and MRI scans, needed for optimal communication with respect to morphology and functionality in diagnostic and therapeutic procedures [71,72].

The measurement of the geometric height during the repair of a dysfunctional BAV has become an important concept [58]. The length of the free margin of the cusp, the commissural height, the number and the location of fenestrations, fibrosis/calcification patterns, and completeness of raphe in BAV are also considerations for the feasibility of valve repair [40]. Adherence to these principles during surgery lowered the transvalvular gradient [24], lowered recurrent prolapse [24,29], and improved the outcome of valve repair. Although VSSR in patients with BAV is as safe as a Bentall procedure, extensive decalcification and the use of a reparative and augmenting pericardial patch were avoided since its use could increase the transvalvular gradient [24], lower its durability [25,28],

and worsen outcome [3], with an increased reoperation rate [28]. The results of a cusp repair in BAV were worse compared to TAV [25,29] because of the uneven distribution of biomechanical stress on patch material. Moreover, the presence of BAV, which could serve as a surrogate for younger age is associated with faster degeneration.

Separate from the repairability of a dysfunctional BAV is the approach of the dilatation of the proximal aorta. The threshold to perform a procedure of any kind on the proximal aorta is still a matter of debate. In patients undergoing SAVR for a dysfunctional BAV, a "preemptive" AAI at 45 mm or more did not increase the risk for adverse events and could improve short-term outcomes [38]. This policy rarely resulted in postoperative further dilation [17], but this was doubted with respect to the risk-benefit ratio [36]. Currently, there are 2 widely used approaches for VSSR: the aortic root reimplantation (the David procedure) and aortic root the remodeling (the Yacoub procedure). In the reimplantation technique, the native aortic valve is sutured in a straight vascular graft, with and without neo-sinuses [58]. The entire valve is supported to a level below the nadir of each cusp. The technique is applied in 55% of the large German Aortic Root Repair Registry. In another 39% of the cases, neo-sinuses were created with modifications, but only in 2% by a smaller separate graft for the ascending aorta [40]. The technique can be used in patients with of proximal aneurysm [4,32]. Reimplantation with Valsalva or straight tube graft (±plication by pleating) after removal of aneurysmal SOV tissue in BAV patients resulted in good outcomes [5]. The David V reimplantation for BAV anomalies had good outcomes, but in later years, there was a higher need for reintervention compared to TAV aneurysms [4].

In the more challenging remodeling approach, a scalloped graft is sutured to the rim of a remnant aortic wall on the aortic valve. The aortic annulus is not supported within the graft. To prevent future annular dilatation, a polytetrafluoroethylene suture or an external annuloplasty ring could be used [40,58]. The procedure was applied in 5% of the patients included in this registry [40]. The use of an external subannular ring was applied in patients with a size over 28 mm, while in patients with smaller annuli, a subcommisural annuloplasty was performed [32]. In many instances of BAV, two symmetric tongues with a 180° orientation were created. The graft was sutured to the cusp insertion lines [29]. Tube grafts and grafts with neosinuses were applied [58].

Some variations were introduced to treat aortic aneurysms limited to the ascending aorta. The more recently introduced "Florida sleeve" technique places the graft around the aortic root, as external support at the annular level. The graft leaves keyhole-shaped fenestrations for the coronary arteries, while the supracoronary aorta is replaced with a graft [41]. The Personalized External Aortic Root Support (PEARS) technique uses a customized graft to wrap around the aorta, which is secured at the annular level. The prosthesis is entirely extravascular, which would make the use of a cardiopulmonary bypass unnecessary. This would limit its use to good functioning BAV [73]. An ascending aorta graft can be performed with extension into the NC-cusp if indicated. The risk for future dilation is higher in patients with a moderate or severe preoperative AR, and a root larger than 35 mm at discharge. Although a higher rate of surveillance is recommended, a reoperation is rarely needed [6]. A reduction aortoplasty is safe [2,37], even if the aortic wall is not reinforced, since there is a low rate of re-dilatation. The presence of a BAV has no effect on this outcome [37]. Wrapping of the ascending aorta with a size between 40 and 45 mm in patients undergoing SAVR for a dysfunctional BAV had shorter surgical times compared to AAI. The long-term outcomes were good [21]. BAV is associated with intrinsic abnormalities in the aortic media, including smooth

muscle cell apoptosis and matrix metalloproteinase overexpression, which contribute to progressive aortic dilation. These structural defects often precede hemodynamic changes, underscoring the importance of early imaging surveillance. Clinically, the variability in cusp fusion patterns and valve orientation can influence progression to stenosis or regurgitation and should be factored into surgical planning.

The main limitations of the included series are the retrospective and observational nature, and the lack of randomization. Another issue is the variability in the presentation of BAV and the involvement of the aortic root and the ascending aorta. The adoption of a universally recognized and exhaustive classification of BAV pathology should be followed by establishing a multicentric international prospective registry of patients undergoing repair of BAV pathology and the proximal aorta, with a precise description of the surgical procedures, the immediate postoperative clinical and hemodynamic data and a long-term follow-up with respect to survival, durability and reintervention rate. This registry should address questions with respect to:

- The reparation of defects in the cusps after decalcification.
- The need for reserving asymmetry to avoid the tension of the non-fused cusp.
- The threshold for repair of the aortic root and ascending aorta in patients with and without additional risk factors for acute dissection.
- The effect of the normalization of the blood flow after repair of a dysfunctional BAV on future expansion of a mild or moderately dilated aorta.
- The timing for the repair of an asymptomatic but regurgitation BAV with respect to left ventricular dysfunction and its postoperative reverse remodeling.

The limitations of the current series are the exclusion of syndromic BAV and acute complications such as dissection of the aneurysms. Because of the different designs of the included series, a meta-analysis with respect to the main outcomes was not possible. The inclusion was limited to the last 5 years in order to include only the most relevant series with respect to the recent innovation such as the intraoperative measurement of effective height.

4 Conclusions

Non-syndromic BAV is the most frequently present congenital heart disease with an incidence of about 2%. There are different types of BAV, according to several classification systems. This condition can progress towards valve dysfunction such as valve regurgitation and stenosis. Due to the associated fragmentation of elastin in the largest vessels or to altered flow patterns in the proximal aorta, a dilatation could develop at the ascending aorta, the root, or as a combination. This could lead to catastrophic events such as a dissection but also contribute to valve regurgitation, because of the relatively young age of BAV patients, valve repair, and if necessary root replacement is preferable. This should be done through anatomical-based procedures in order to restore an adequate coaptation zone. The use of pericardial patches proved to be less durable. There is still disagreement about the threshold for treating the dilated root or ascending aorta. Most repair techniques lead to high survival rates and acceptable reoperation rates in this rather young population. Nevertheless, a lower rate of reverse remodeling of the left ventricle and reversal of atrial dysfunction with possible postoperative atrial fibrillation should be taken into account.

Acknowledgement: Not applicable.

Funding Statement: The author received no specific funding for this study.

Availability of Data and Materials: Not applicable.

Ethics Approval: Not applicable.

Conflicts of Interest: The author declares no conflicts of interest to report regarding the present study.

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