Bicuspid aortic valve: differences in the phenotypic continuum affect the repair technique

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Abstract

Objective: Bicuspid aortic valve (BAV) may be considered in the context of a phenotypic continuum of the aortic valve configuration, ranging from severe forms of unicuspid valve to rare forms of quadricuspid valve. In this article, we report our results with BAV repair using surgical techniques tailored to the specific features of the BAV phenotypic continuum.

Patients and methods: Between September 2003 and May 2009, 31 patients with BAV (mean age 49.9 ± 17.3 years; five female; 26 male) were prospectively enrolled for aortic valve repair. The surgical strategy was tailored depending on the anatomical and structural characteristic of the BAV continuum: type 1 real bicuspid (five patients), type 2 bicuspid with raphe (24 patients) and type 3 clefted bicuspid (two patients). An echocardiographic analysis was performed preoperatively, postoperatively, at discharge and every 6 months during follow-up. Kaplan—Meier analysis was used to assess the freedom from re-operation.

Results: One patient died in the intensive care unit due to aortic wall rupture. After BAV repair there was an increase of leaflet coaptation length (from 2.0 ± 0.5 mm to 8.2 ± 1.9 mm, \( p < 0.01 \)) and a decrease of the diameters of the virtual basal ring (from 24.6 ± 3.6 mm to 21.5 ± 0.71 mm, \( p < 0.01 \)), of the aortic root (from 43.12 ± 13.23 mm to 31.0 ± 3.2 mm, \( p < 0.01 \)) and of the sino-tubular junction (from 40.1 ± 6.95 mm to 33.5 ± 9.19 mm, \( p < 0.05 \)). The aortic regurgitation echocardiographic grade II was found preoperatively in 30 patients and postoperatively in one patient (\( p < 0.01 \)). Six patients underwent an associated aortic valve-sparing procedure (three re-implantation and three remodelling). Kaplan—Meier analysis showed a 96.6% freedom from re-operation at 5 years, with a single new repair procedure and a 100% freedom from aortic valve replacement.

Conclusions: BAV repair provides a feasible therapeutic option in selected patients with good medium-term results. The surgical techniques should be applied after a careful analysis of the BAV considered in the context of the phenotypic continuum.

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Keywords: Bicuspid aortic valve; Aortic valve repair; Aortic root; Aortic valve

1. Introduction

Bicuspid aortic valve (BAV) is the most common congenital anomaly of the aorta found in the adult, occurring in about 0.7—2.0% [1]. Since the availability of prosthetic valves, aortic valve replacement has been the standard surgical approach for treating stenosis and regurgitation in patients with BAV. Good medium-term results obtained with aortic valve repair [2,3] have prompted the application of tricuspid valve repair procedures to bicuspid valves. In this regard, several approaches have been followed so far. While some authors have proposed maintaining the two-leaflet structure [4,5], others have suggested the reshaping to a crown-like annulus with the tricuspidisation of the valve [6]. However, the choice of the repair technique has never been focussed on the different types of BAV. The historical anatomical classification of BAV is based on the position of the coronary ostia. Only recently, Sievers and Schmidtke [7] have proposed a BAV classification based on the different morphologic phenotypes that can be related to different functional patterns.

In this article, we propose a novel approach to BAV repair based on the integration of anatomical and functional data that come up from the assumption that BAV could be part of a phenotypic continuum ranging from unicuspid valves to rare quadricuspid forms.

2. Materials and methods

2.1. Inclusion criteria

Between September 2003 and May 2009, we prospectively enrolled a total of 31 patients with a diagnosis of aortic valve regurgitation in the setting of BAV. The mean age was...
49.9 ± 17.3 years. In this cohort, 26 patients were male and five female. All subjects were diagnosed based on echocardiography criteria and scheduled for surgery using aortic valve repair techniques after signing an informative paper on the repair procedures and their results from literature. These procedures represented approximately 22.1% of all interventions for valve leaflet repair (146 patients) performed in our division during the study period.

2.2. Exclusion criteria

Patients were excluded in presence of valve stenosis with complete fusion and calcification of the two leaflets, regardless of the ventriculo-aortic gradient.

2.3. Preoperative assessment

Routine blood chemistry and thyroid function tests were performed in all subjects. In addition, permeability of the carotid artery was assessed. Coronary catheterisation was performed in patients aged above 45 years or in the presence of significant surgical risk factors. Preoperative transthoracic echocardiography (TTE) was performed to assess aortic valve pathology, left ventricular function and to rule out the presence of associated mitral or tricuspid disease. The degree of valvular regurgitation was evaluated as grade 0—IV. Before cardiopulmonary bypass (CPB), transesophageal echocardiography (TEE) was performed to assess the mechanism of regurgitation according to the El Khoury classification[8] and to measure the Aortic Functional Unit features as described by Anderson [9]: virtual basal ring (VBR), aortic root, sino-tubular junction (STJ) and ascending aorta (AA).

2.4. Repair technique

The chest was opened through a median sternotomy incision and the patient was placed on CPB with cannulation of the ascending aorta and the right atrium appendage or, if a mitral or tricuspid repair was necessary, both venae cavae. After induction of cardiac arrest by infusion of cold crystalloid cardioplegia (St. Thomas Solution®, Monica S.P.A Via ponte di Pietra 7 Venezia) into the coronary ostia, the aortic valve and root were inspected to confirm the adequacy of the repair procedures and their results from literature. These procedures represented approximately 22.1% of all interventions for valve leaflet repair (146 patients) performed in our division during the study period.

(1) Valve analysis: Transverse aortotomy was performed larger than 75% of the aorta circumference to allow a complete visualisation. Two commissural stay sutures (4/0 polypropylene) were placed to achieve an adequate exposure and analysis of the aortic root. Leaflet distortion due to inadequate traction of these sutures was carefully avoided. A 6/0 polypropylene suture was passed through the two Aranzius nodules to precisely measure the relative length and height of each leaflet.

Based on the phenotypical BAV continuum, bicuspid aortic valve disease was divided into three different forms (Fig. 1):

- **Type 1**: real bicuspid valve with two complete leaflets (usually of the same length) and two sinuses but without raphe. Regurgitation mechanism: prolapse of one leaflet causing an eccentric jet.
- **Type 2**: bicuspid valve with a fibrotic or calcified raphe usually hampering normal leaflet motion or retracting the free edge towards the annulus in the middle portion of the leaflet. Presence of two or three sinuses. Regurgitation mechanism: pseudo-prolapse of the opposite leaflet with an eccentric jet or incomplete coaptation due to retraction of the calcified raphe generating a central jet.
- **Type 3**: pseudo-bicuspid valve with one big leaflet and two additional small leaflets with a commissure between them and three sinuses or a bicuspid valve with two sinuses and a cleft. Regurgitation mechanism: fibrotic free-edge degeneration of the two small leaflets creating a central coaptation defect with a central jet.

(2) Leaflet repair: The main purpose of the procedure was to restore a complete motility of both leaflets achieving optimal coaptation length and coaptation height (Fig. 2).

In the presence of a thickened or calcified free margin, excision of the diseased tissue was performed by a shaving technique followed by different repair procedures according to the type of BAV:

- **Type 1**: triangular resection or leaflet plicature with a 6/0 polypropylene was pursued in the presence of leaflet prolapse. In case of a small prolapse, correction was pursued by free margin remodelling using a Gore-Tex® CV-7 locked suture. Free-edge elevation of prolapsing leaflets could be also achieved by weavering an appropriate length of the Gore-Tex® suture to reach the middle height of the Valsalva sinus.

**Fig. 2.** Transesophageal echocardiography long-axis view measures.
Type 2: in the presence of calcified or fibrotic tissue that might hamper valve leaflet motion, a complete excision of the diseased tissue was performed. A pericardial patch was always implanted to fill the gap. The patch shape was dependent on the anatomical findings of the raphe (Fig. 3): (1) a triangular or trapezoidal patch was used in presence of a calcified raphe with good free edges in a non-prolapsing leaflet; (2) a lens patch was used in presence of a calcified raphe with good free edges in a prolapsing leaflet to reduce the free-edge length; (3) a Fish Tail patch was used in presence of extensively calcified free edges to remove all the calcified tissue, increasing motility and coaptation capability.

Type 3: extensive leaflet shaving was adopted to reduce the irregular diastolic coaptation of the stiff free edges. In presence of a residual prolapse, a type 1 repair was added.

3. Results

3.1. Operative results

No patient underwent emergency surgery or underwent valve replacement following the first repair attempt. No patient refused repair procedures. One patient died within 30 days from the procedure (operative mortality: 3.2%) due to aortic wall rupture in the incannulation site. No patient had perioperative myocardial infarction or required prolonged ventilatory support. BAV type 1 was found in five patients (16.1%), type 2 in 24 patients (77.4%) and type 3 in two patients (6.4%). In six patients (19.3%), two with grade I and four with grade II aortic regurgitation, an aortic-sparing procedure for severe dilatation of the aortic root was performed before leaflet repair (three remodelling and three re-implantations) (Table 1). The indication for bicuspid repair in the patients with grade I aortic regurgitation after sparing procedures was the anatomical modifications induced by re-implanting huge leaflets into a smaller tubular prosthesis. For was aimed to avoid further dilatation generating valve regurgitation.

The patients were operated by four different surgeons after an ‘Aortic Valve Repair Knowledge Transfer’ programme, which included a 2-day wet-lab to standardise procedures.

2.5. Postoperative assessment

Immediately after the procedure, TEE was performed to assess the anatomical and functional characteristics of the repaired valve. The following variables were examined: (1) valve competence; (2) the extent of leaflet coaptation (coaptation length); (3) the level of coaptation (coaptation height); and (4) any residual leaflet prolapse.

2.6. Short-term follow-up assessment

All patients were scheduled for an outpatient visit 3 months after surgery. A TTE examination was performed at 6 months and annually thereafter. All patients underwent a telephone interview during May 2009.

2.7. Data analysis

Statistical analysis of data was performed using SPSS® 13.0 software (SPSS Inc., Chicago, IL, USA). Normal distribution was tested using both the Kolmogorov—Smirnov statistics with a Lilliefors significance level and the Shapiro—Wilk test. Continuous data are presented as mean ± standard deviations. The Student’s paired t-test was used after evidence of normality. Nominal data are presented as absolute frequencies or percentages. Analysis of categorical variables was performed by chi-square ($\chi^2$) test or Fisher’s exact test where appropriate. Freedom from re-operation was determined by Kaplan—Meier analysis. A two-tailed p value <0.05 was considered statistically significant.
patch repair we used fresh autologous pericardium in six patients and treated eight patients with heterologous pericardium. Associated procedures included coronary artery bypass grafting (one patient), mitral valve repair (one patient) and tricuspid valve repair (one patient). The mean CPB time was 138.8 ± 39.1 min, while the mean aortic cross-clamp time was 111.8 ± 34.7 min.

After the repair procedure there was an increase of coaptation length (from 2.0 ± 0.5 mm to 8.2 ± 1.9 mm, \( p < 0.01 \)) and a decrease of the diameters of the VBR (from 24.6 ± 3.6 mm to 21.5 ± 0.71 mm, \( p < 0.01 \)), of the aortic root (from 43.12 ± 13.23 mm to 31.0 ± 3.2 mm, \( p < 0.01 \)) and of the STJ (from 40.1 ± 6.95 mm to 33.5 ± 9.19 mm, \( p < 0.05 \)). The aortic regurgitation echocardiographic grade II was found preoperatively in 30 patients and at discharge in one patient (\( \chi^2 = 49.5, p < 0.01 \)) (Fig. 4). The mean and maximum gradient between the left ventricle and the aorta did not change significantly (\( D_{\text{max}} \) from 23.6 ± 9.81 mmHg to 22.8 ± 9.6 mmHg, \( p = \text{ns} \); \( D_{\text{mean}} \) from 11.8 ± 5.21 mmHg to 12.8 ± 4.9 mmHg, \( p = \text{ns} \)). The left ventricular end-diastolic volume decreased from 149 ± 56.9 ml to 136.8 ± 58.8 ml (\( p = \text{ns} \)) and the left ventricular end-diastolic diameter changed from 56.6 ± 8.1 mm to 53.7 ± 10.7 mm (\( p = \text{ns} \)). The mean intensive care unit stay was 18.2 ± 3 h. The mean postoperative hospital stay was 8.5 ± 2.5 days.

### 3.2. Follow-up

Mean echocardiographic follow-up time was 836.3 ± 624.8 days (range: 1–1950 days). All patients treated with repair procedures underwent TTE at 6 months (Fig. 4). There was no progression of aortic regurgitation, the only exception being one patient who was re-admitted for severe aortic regurgitation due to the dehiscence of pericardial patch suture. This patient underwent re-operation with a new double suture using the same tailored patch.

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**Table 1**

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Fig. 4. Echocardiographic results of bicuspid aortic valve repair. AR: aortic regurgitation; TTE: transthoracic echocardiography; and TEE: transesophageal echocardiography.
compares to 0.8% of all other forms of congenital cardiac
occurs in approximately 1—2% of the population. This

4. Discussion

BAV is a common congenital abnormality found in adults. It
occurs in approximately 1—2% of the population. This
compares to 0.8% of all other forms of congenital cardiac
disease combined [10].

The semilunar valves are derived from valve swellings of
subendocardial tissue located near the luminar surface of the
aortopulmonary trunk, a process that involves tissue re-
absorption and remodelling, ultimately leading to the
formation of the thin-walled semilunar cusps. BAV is thought
to arise from a disruption of this development process,
although the exact mechanism triggering its development
remains unclear. An anomalous behaviour of cells derived
from the neural crest has been suggested to play a role in the
association of BAV with congenital malformations of the
aortic arch and other neural-crest-derived systems [11,12].
Lee et al. [13] have reported that mice lacking endothelial
nitric oxide synthase have an increased susceptibility to BAV,
thereby suggesting that abnormalities in this enzyme may
lead to disruption of signalling pathways required for proper
valvulogenesis in the mammalian heart.

The anatomy of the most common BAV (type 2 in our
classification) includes unequal cusp size (due to fusion of
two cusps leading to a larger cusp), the presence of a central
raphe (usually in the centre of the larger of the two cusps)
and smooth cusp margins. The raphe or fibrous ridge is the
site of congenital fusion of the two components of the
conjoined cusps and is identifiable in most BAV patients.
Notably, pathologic examination of the raphe has shown that
they do not contain valve tissue [14]. A review of the
echocardiograms of 1135 children with BAV [15] revealed that
right-coronary and left-coronary leaflets fusion was the most
common morphologic variant. Sabet et al. [16] confirmed
the same data in a surgical pathology study on 542 cases
associated with a literature review of 2715 patients: left and
right leaflets fusion occurred in 86% of patients, right and
non-coronary fusion in 12% of patients, and left and non-
coronary fusion in 3%. This classification has predictive value
in BAV patients since the fusion of right and non-coronary
cusp may be related to an increased risk of aortic stenosis and
aortic valve regurgitation than other anatomical patterns
[15].

Robicsek et al. [17] described in a recent study, based on
mock-loop (left heart simulator using conventional and 500
frames per second cinematography) and digital models’
experience, the systo-diastolic BAV behaviour. Specifically,
the authors reported that BAV requires several compensatory
mechanisms to function properly:

(a) the anatomical length of the leaflet edges is constant but
their ‘functional’ length must change to match the
geometry of full closure and opening, by a gradual
folding and unfolding action persisting throughout the
cardiac cycle. These folds are sharper and shaped in
more acute angles as opposed to the normal tricuspid valve;

(b) nature and time tend to increase the coaptation area to
compensate the leaflet length discrepancy by an
increased bulge (doming) of the leaflet bellies;

(c) the asymmetrical approximation of the leaflets (their
line of coaptation is laterally displaced) permits the
closure of the BAV until it behaves as a unicuspid valve;

(d) the elliptical shape of the orifice and the inability to fully
open to match the area bordered by the annulus
enhances all the other compensatory mechanisms.

BAV undergoes a certain degree of remodelling, and with
time all compensatory mechanisms tend to fail due to the
presence of calcific tissue hampering the normal leaflet
motion. Sans-Coma et al. [18] have investigated isolated BAV
in Syrian hamsters subjected to systematic inbreeding. These
authors have suggested that congenital BAV with coronary
leaflets fusion (ventral orientation) is the expression of a
trait; the variation of which takes the form of a continuous
phenotypic spectrum, ranging from a tricuspid aortic valve
with no fusion of the ventral commissure to a bicuspid aortic
valve devoid of any raphe. As suggested by Robicsek et al.,
a detailed analysis of the anatomical aspects of the BAV
phenotypic continuum is warranted to re-create an adequate
compensatory mechanism. In addition, efforts should be
made to modify the valve to re-create two leaflets of about
the same length and ensure their correct folding and
unfolding during the cardiac cycle. Notably, a symmetric
coaptation in the middle part of the orifice and an adequate
coopation length (>6 mm) are desirable. Based on these
studies, we propose a classification aiming to guide the
surgeon to proper repair procedures. It is important to
emphasise that this classification divides the phenotypic
continuum of BAV into three arbitrary patterns. However,
many other forms of BAV can be found in surgical practice.
Mancuso et al. [12] have reported the occurrence of a BAV
with a median cleft in the ventral leaflet alongside with a
rudimental fenestrated raphe that does not reach the free
margin of the leaflet. Thus, this specific form of BAV may
be considered as an intermediate between type 2 and type 3.
The choice to replace the raphe with a pericardial patch or to perform an attempt to obtain a complete motility of the two pseudo-leaflets is dependent on the anatomy of the aortic root. The presence of three sinuses and/or of a real inter-leaflet triangle may lead to classify a BAV as type 3; by contrast, the presence of two sinuses without an inter-leaflet triangle may lead to a type 2 classification.

The different shape of the pericardial patch in type 2 repair techniques may be relevant to achieve suitable results. A triangular shape patch with the base on the free margin and the apex on the VAJ is used to replace the calcified or fibrotic tissue without modifying the cusp length. In one patient, we performed a triangular patch leaving the free edge intact. This allowed the preservation of its length in the presence of a non-prolapsing leaflet. This procedure could be especially useful in the presence of non-calcified free margins. A lens shape may allow to replace the hampering tissue and to reduce the free margin length, such as a plication or a triangular resection. The Fish Tail shape allows the removal of the hampering tissue from the free edge and the contiguous tissue. In a BAV, the free margin and the raphe are the most common regions undergoing calcification and fibrosis. In this context, the association of free-edge shaving with the use of one of the patch shape described may restore motility and coaptation capability.

Robicsek et al. [19] have reported that in normal tricuspid aortic valve — besides the eddy currents — the aortic sinuses modulate valve function by pulling the commissures inward during diastole for the transmission of tension on the coapted leaflets, followed by a release due to the increase in both left ventricular and intra-aortic pressures with the creation of a stellate orifice by the tangential tension on the leaflets. These authors analysed this commissural ‘pull-and-release’ mechanism using a finite element model. They found that an additional flexion stress exists on the cusps in the presence of abnormal commissures. Mercer et al. [20] and Sutton et al. [21] have reported that the parabolic shape of the aortic valve may form a suitable support similar to a suspension bridge, both at the attachment of the leaflets to the aortic wall and along the thickened coapting portions of the free edge. In BAV, this pull-and-release mechanism related to the commissure motion may have a greater importance than in a tricuspid valve. The absence of the third commissure leaves the valve without an important support and transfer to that mechanism the folding and unfolding control to be able to start opening the valve while the pressure in aorta is still higher than in the ventricle and close the valve while the blood is still streaming from the ventricle. Optimal length and tension of the Gore-Tex® CV-7 suture on the free edge may re-create this mechanism transferring the diastolic stress from the leaflet to the commissures and to the sinus-tubular ridge. Moreover, an STJ plicature in the middle of the Valsalva sinus can increase the motility of the corresponding leaflet by drawing up the commissures and lowering the coaptation line.

In conclusion, BAV repair provides a feasible therapeutic option in selected patients with good medium-term results at the Kaplan–Meier analysis but with a small cohort of ‘patients at risk’ in the past years. The surgical techniques suggested in this article should be applied after a careful analysis of the BAV considered in the context of the phenotypic continuum. Long-term clinical and echocardiographic follow-up studies are warranted to broaden the acceptance of this methodology.