Peer Review File

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Reviewer #1

Comment 1: Please check the entire manuscript for typing and style errors and unify (e.g., line 103, 149, 177, 185,...).

The manuscript has been reviewed for typing and style errors.

Reviewer #2

I thank the authors on their achievement in this manuscript. However, I feel there are some points that can be improved.

Comment 1: Line 87-88

Please refer to the previous studies which compared aortic valve repair for bicuspid aortic valve to aortic valve replacement.

We added references to the following papers:

- Svensson LG, Batizy LH, Blackstone EH, et al. Results of matching valve and root repair to aortic valve and root pathology. *J Thorac Cardiovasc Surg.* 2011;142(6):1491-1498.e7.
- Ashikhmina E, Sundt TM, Dearani JA, Connolly HM, Li Z, Schaff HV. Repair of the bicuspid aortic valve: a viable alternative to replacement with a bioprosthesis. *J Thorac Cardiovasc Surg.* 2010;139(6):1395-1401.
- Lansac E, Bouchot O, Arnaud Crozat E, et al. Standardized approach to valve repair using an expansible aortic ring versus mechanical Bentall: early outcomes of the CAVIAAR multicentric prospective cohort study. J Thorac Cardiovasc Surg. 2015;149(2 Suppl):S37-45.

As well as referenced our group's prior work:



 Vallabhajosyula P, Szeto WY, Habertheuer A, et al. Bicuspid Aortic Insufficiency With Aortic Root Aneurysm: Root Reimplantation Versus Bentall Root Replacement. Ann Thorac Surg. 2016;102(4):1221-8.

Comment 2: Line 204-206 Please show references.

These lines describe our group's (Dr. Bavaria and Dr. Desai) method for sizing the external subannular ring and the expected reduction in annular diameter. This technique was previously described in the following paper by Ko and colleagues:

 Ko H, Bavaria JE, Habertheuer A, et al. Functional Outcomes of Type I Bicuspid Aortic Valve Repair With Annular Stabilization: Subcommissural Annuloplasty Versus External Subannular Aortic Ring. Ann Thorac Surg. 2019;107(1):68-75.

Comment 3: You referred the studies effectiveness ESAR for aortic valve repair. Please specify the novelty of the manuscript.

This case report and accompanying surgical video demonstrates our step-by-step process in performing a bicuspid aortic valve repair and ESAR with emphasis on our thought process of sizing and positioning the ring. Our group has not previously published a video of our technique of performing ESAR.

Reviewer #3

This case report entitled "Bicuspid Aortic Valve Repair with External Subannular Ring: A Case Report", is submitted by a well-known team who are renowned for their expertise in aortic valve repair. This is a well written manuscript with a well-made surgical video. I think that, because aortic valve repair is still an evolving field, this video is useful for surgeons who are not familiar with these techniques and adds to the available resources that are out there to educate. However, the technique in itself is not novel as it has been described before by other groups, but the authors showcase their particular approach and present their treatment rationale and approach in regard to annular and root dimension, which is certainly very valuable.



A few comments:

Comment 1: In the abstract and conclusion section the authors comment that Annular stabilization is a key component of BAV repair to prevent annular dilatation. But the authors don't clearly mention that it also treats the pre-existing annular dilatation. By down-sizing the annulus, leaflet coaptation is enhanced. This should be added to the manuscript, since the authors are not only preventing future dilatation, but are treating a severe annular enlargement of 34mm.

Thank you for your insightful comment. We agree that down-sizing the annulus to improve leaflet coaptation is an essential part of a successful BAV repair, which, as you mentioned, was illustrated by the 34 mm annulus in our case report. For this reason, we have modified the abstract, introduction, and conclusion sections of the case report to mention that annuloplasty both reduces the annular diameter to improve leaflet coaptation and stabilizes the annulus to prevent future dilation and recurrent AI (lines 54-56, 95-98, and 534-535).

Comment 2: What was the mechanism of the eccentric jet on Echo? Was there a prolapse of the fused cusp, in addition to the restriction? Since, there was excessive free margin length, it appears that there must have been a prolapse. If so, it should be added, or the mechanism of eccentric regurgitation should be explained otherwise.

The mechanism of the eccentric jet on echocardiogram was a severe prolapse of the conjoined leaflet. This was added to lines 310-311.

Comment 3: How much of the calcium was removed? On the postoperative Echo the cusp still appears calcified.

Over 75% of the calcium was removed. There was still a mild amount of residual calcium in the area where the raphe meets the annulus.

Comment 4: Is there billowing of the cusps on the postoperative Echo? Can the authors comment on that and the potential long-term effects and repair durability?



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In certain TEE views with relatively symmetric bicuspid valves (160-180° initial angle configuration), there can be the appearance of "billowing" as the annulus has deep nadirs. It is unknown whether billowing with an excellent coaptation height, especially in a more symmetric valve such as the one presented in our video, is associated with long-term durability. These are obviously complex concepts that require further study.

Comment 5: The authors mention a coaptation height of 8.5mm. The terminology can be confusing at times to clinicians who don't deal with it on a regular basis. In order to shed more light on coaptation height, can the authors describe how it was determined, and what the goal coaptation height is? Also, in the Discussion, how is the geometric height determined.

Coaptation height is defined as the zone of cusp apposition during diastole [1,2]. We determine coaptation height using 3D TEE. The goal coaptation height is >7-8 mm. This was added (lines 348-349). As described by Schäfers and colleagues, the geometric height of each leaflet is measured intraoperatively using a ruler from the aortic insertion in the nadir of each sinus to the corresponding nodule of Arantius [3]. This was added to the discussion as well (lines 372-374).

- Berrebi A, Monin J-L, Lansac E. Systematic echocardiographic assessment of aortic regurgitation-what should the surgeon know for aortic valve repair? *Ann Cardiothorac Surg.* 2019;8(3):331-341.
- Vanoverschelde J-L, van Dyck M, Gerber B, et al. The role of echocardiography in aortic valve repair. *Ann Cardiothorac Surg*. 2013;2(1):65-72.
- Schäfers HJ, Schmied W, Marom G, Aicher D. Cusp height in aortic valves. J Thorac Cardiovasc Surg. 2013;146(2):269-74.

6. Some groups use a Hegar dilator to tie down the annuloplasty sutures around it. Why did the authors do it without, and how can they be certain that the annuloplasty sutures decrease the annulus consistently by 4-5mm? How tight do the authors tie their sutures? Does it have any effect on the annuloplasty amount? And why do the authors use 6, or 7 mattress sutures in this case? Does the annuloplasty amount change with using more or less sutures?



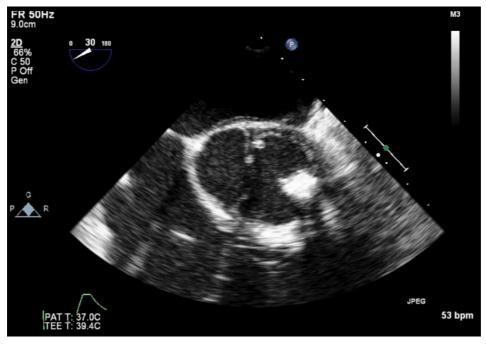
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In this case, the annulus was so large that Dr. Bavaria did not feel it was necessary to use a Hegar dilator. The sutures are tied down firm but not too tight. In our experience, the annulus is decreased approximately 6 mm smaller than the chosen graft diameter. This is an empirical result based on experience. For smaller annuli, we do use a Hegar dilator when tying down the annuloplasty sutures.

For a bicuspid valve-sparing root reimplantation, Dr. Bavaria usually uses 8 subannular sutures for the primary suture line (4 per cusp). For an ESAR, Dr. Bavaria usually uses 6 mattress sutures, eliminating the two sutures at the locations of the left main and right coronary arteries.

Comment 6: In figure 1 legend, can you please comment on the cusp fusion anatomy (right/left)? That should be clear without reading the entire manuscript and watching the video.

Intra-operative TEE showed a Sievers Type 1 bicuspid valve with fusion of the right and left coronary cusps and a calcified raphe with a mobile component. This was added to the legend for Figure 1. We also replaced Figure 1 with a better image from the TEE:





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Reviewer #4

Good job! I strongly believe that aortic valve repair should become a routine procedure, especially for bicuspid incontinent aortic valves, so I appreciated your paper. I have only two points to focus on:

Comment 1: why did you choose 27 mm diameter as a threshold for the treatment of annular dilation? As you mentioned in the Discussion, Lansac et al use the Coroneo ring when the annular dilation is > or = 25 mm diameter, as a prevention for further dilation. Could you please comment this aspect?

Thank you for your insightful question. The decision to use > 27 mm as our threshold for performing ESAR instead of SCA is based on our group's prior study of SCA, in which we found that SCA performed in an annulus \leq 27 mm was stable over time with minimal midterm AI, whereas SCA in a preoperative annulus \geq 28 was predictive of recurrent AI grade > 1+ and associated with significantly lower freedom from AI grade >1+ at five years [1]. For this reason, our group uses 27 mm annular diameter in our algorithm for deciding between SCA and ESAR when performing a BAV repair.

 Vallabhajosyula P, Komlo CM, Szeto WY, et al. Aortic Annulus Diameter Affects Durability of the Repaired Bicuspid Aortic Valve. J Heart Valve Dis. 2015;24(4):412-9.

Comment 2: as you write at line 90, several techniques have been described, for aortic valve repair... I think that "teaching papers" should be added to your references, such as:

Youssefi P, Brega C, Shraer N, Zacek P, Debauchez M, Lansac E. Isolated Bicuspid Aortic Valve Repair With Double Annuloplasty: How I Teach It. Ann Thorac Surg. 2019 Dec;108(6):1596-1604. doi: 10.1016/j.athoracsur.2019.09.016. Epub 2019 Oct 10. PMID: 31606519.

Carlotta B, Zakkar M, Zacek P, Palladino M, Lansac E. Bicuspid aortic valve repair with hemi-remodeling technique and external ring annuloplasty. J Card Surg. 2020 Jan;35(1):146-150. doi: 10.1111/jocs.14334. Epub 2019 Nov 9. PMID: 31705828.



Thank you for your insightful comment. We have added the teaching papers you suggest to our references, as well as the following papers:

- El Khoury G, Vanoverschelde J-L, Glineur D, et al. Repair of bicuspid aortic valves in patients with aortic regurgitation. *Circulation*. 2006;114(1 Suppl):I610-616.
- Youssefi P, Zacek P, Debauchez M, Lansac E. Valve-sparing aortic root replacement using the remodeling technique with aortic annuloplasty: bicuspid valves with repair of specific lesion sets: how I teach it. *Ann Thorac Surg.* 2019;108(2):324-333.
- Sultan I, Komlo CM, Bavaria JE. How I teach a valve-sparing root replacement. *Ann Thorac Surg.* 2016;101(2):422-425.

