Upsized ring annuloplasty and autologous leaflet augmentation: A new paradigm for pediatric aortic valve repair

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The institutional review board at our university does not require review for isolated retrospective case reports; the patient's parents provided written consent for this publication.

Presented at the 60th Annual Meeting of the Society of Thoracic Surgeons, San Antonio, Tex, January 29, 2024.

Received for publication March 13, 2024; revisions received July 5, 2024; accepted for publication July 16, 2024; available ahead of print July 23, 2024.

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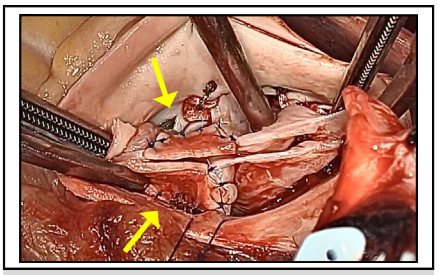
JTCVS Techniques 2024;27:135-7

2666-2507

https://doi.org/10.1016/j.xjtc.2024.07.009



Hypoplastic unicuspid valve repaired with upsized ring annuloplasty and 2 aortic wall patches (*yellow arrows*).

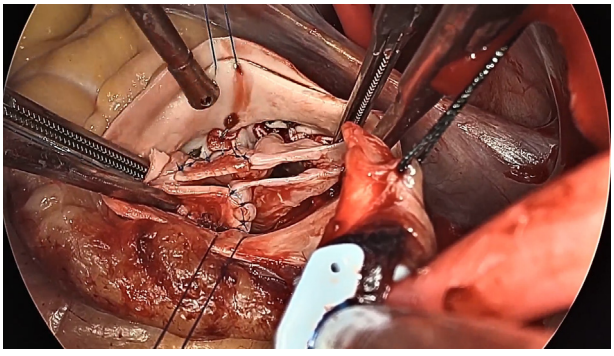
CENTRAL MESSAGE

Using upsized aortic ring annuloplasty and autologous aortic wall patches, hypoplastic unicuspid aortic valves can achieve stable repair to adult-sized configurations.

Video clip is available online.

Unicuspid aortic valves typically have a major fusion and clefting of the right-left coronary commissure, a minor fusion of the right-noncoronary commissure, and a normal left-noncoronary commissure. Referred to as Sievers type 2 bicuspid valve, this configuration accounts for 95% of the unicuspid aortic valves with the remainder having the major fusion at the right-noncoronary commissure[1] or other rare variations. Often exhibiting aortic stenosis and aortic insufficiency, these valves have been difficult to repair because of the 2 fusions, frequent inconsistency in sinus size, annular hypoplasia, leaflet tissue deficiency, and frequent damage secondary to balloon valvuloplasty. We present a successful case of complex unicuspid aortic valve repair utilizing an upsized bicuspid annuloplasty ring along with 2 patches of autologous ascending aorta to reconstruct a hypoplastic unicuspid aortic valve into an adult size and configuration. The institutional review board at our university does not require review for isolated retrospective case reports; the patient's parents provided written consent for this publication.

A 14-year-old boy with a unicuspid aortic valve, severe aortic insufficiency, moderate aortic stenosis, and normal left ventricular function was referred for valve repair versus a Ross procedure. On surgical inspection (Video 1), the annulus was somewhat hypoplastic at 17 mm, and all cusps were small, with the largest left cusp sizing only to a 19-mm ring, too small for an adult man. The aortic valve had a



VIDEO 1. Upsized ring annuloplasty and autologous leaflet augmentation: A new paradigm for pediatric aortic valve repair. Video available at: https://www.jtcvs.org/article/S2666-2507(24)00318-3/fulltext.

major fusion with dysplastic clefting at the right-noncoronary commissure, a minor fusion with clefting at the right-left commissure, and mild fusion at the left-noncoronary commissure, manifesting rare unicuspid aortic valve anatomy. Dysplastic clefting of both fusions may have contributed to leaflet and annular hypoplasia. When inspecting the right-left fused commissure, a sizable fenestration and multiple chords were noted. Given the fenestration, we planned to augment the left cusp with a patch, thereby increasing the free-edge length to match a larger 21-mm adult-sized annuloplasty ring.

The right-left commissural fusion was opened. A 21-mm bicuspid, internal geometric annuloplasty ring (HAART 200; BioStable Science and Engineering) was utilized with the 2 posts being sutured into the subcommissural triangles at a 180° orientation, and the ring was gently insinuated into the narrower subannular position with some difficulty. The rigid ring upsized the annulus from 17 to 21 mm (2 sizes). The ring then was sutured beneath the annulus using 3-0 coated braided polyester looping sutures, placed up from below the valve, 2-mm deep to the leaflet-aortic junction and avoiding gaps. All sutures were tied and fixed laterally to prevent leaflet contact. A 15-mm triangular patch of ascending aorta was excised from the distal aortotomy and positioned to fill the gap and lengthen the left cusp free-edge length to 33 mm (half the circumference of a 21-mm ring). With the patch positioned so that the intima faced the coaptation, the patch was attached to the cusp and commissure with interrupted 6-0 Prolene sutures, leaving the knots toward the distal aorta. At the top of the commissure, a vertical mattress suture was placed to the outside of the aorta. For reinforcement, an alignment suture was placed in both leaflets at the top of the commissure and tied on the outside of the aorta.

If the cleft in the right-nonfused cusp was closed, the free-edge was only 30 mm (again it needed to be 33 mm), and the geometric height was only 15 mm (needed three-fourths of the free-edge length, or 24 mm), making this cusp inadequate for the larger ring. Therefore, a similar sized second aortic wall patch was utilized to fill in the cleft centrally and augment the right-noncusp appropriately. After repair, both leaflet free-edge lengths approximated 33 mm, with excellent and equal coaptation heights (Figure 1). The distal aortic defects were patched with glutaraldehyde-fixed bovine pericardium. Intraoperative echocardiography postrepair demonstrated no residual aortic valve leak and a mean pressure gradient of 19 mm Hg. The operation was concluded, and the patient recovered uneventfully. At 6-month follow-up, the patient had resumed normal activity, and the valve remained fully competent with a 14 mm Hg mean gradient.

Unicuspid aortic valves can account for up to half of pediatric aortic valve defects, and historically, have been difficult to repair for a variety of reasons. Si and colleagues[2] reported a multi-institutional unicuspid repair experience utilizing the HAART bicuspid annuloplasty ring, and in 20 patients, they reported successful and reproducible bileaflet repair with annular remodeling to 180° commissures that equalized the annular circumferences of the fused and nonfused cusps. In these patients with usually dilated annuli, the annuloplasty rings reduced the annular diameter appropriate to leaflet size and resulted in better leaflet

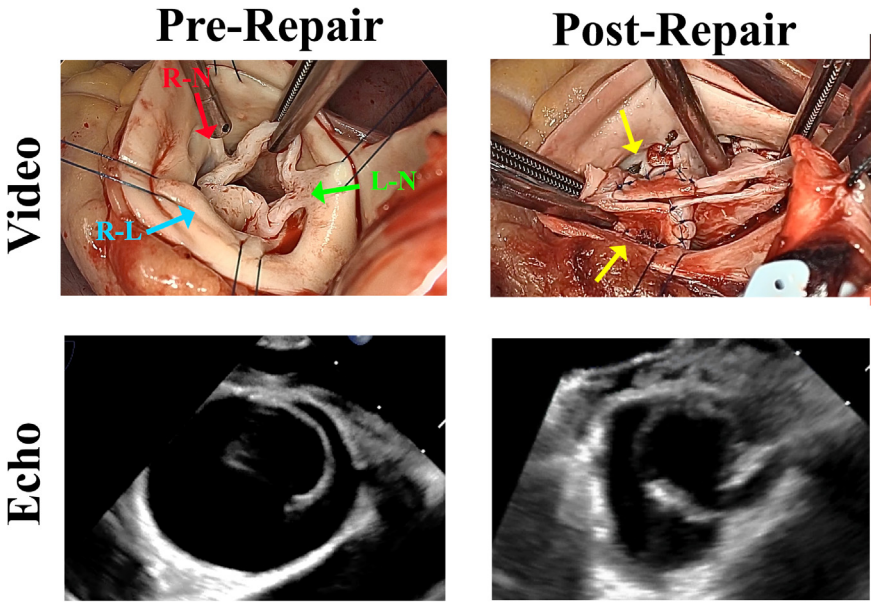


FIGURE 1. Video and echocardiograph views of a hypoplastic unicuspid valve before and after upsized repair. *Red*, *blue*, and *green arrows* denote the various commissures, and the *yellow arrows* indicate the aortic wall patches.

coaptation with excellent late valve competency. Subsequent midterm outcomes in this and other bicuspid ring repair series have been excellent.[3] In the current patient, given the hypoplastic valve and smaller annular diameter, we could not reduce the valve further but, in fact, needed to enlarge it by 2 sizes to create an adult-sized valve. We opted to also augment the cusps with aortic wall patches to increase the leaflet area appropriate for the larger annuloplasty ring. Myers and colleagues[4] recently reported using autologous ascending aortic patches in aortic valve repair with encouraging results. Now in more than 40 patients followed beyond 2 years of maximal follow-up (unpublished data), outcomes have been excellent. Being able to upsize smaller valves to an adult-sized ring, and then augment leaflets with living autologous aortic wall to the same adult size could extend valve repair into smaller children and allow routine adult-sized repairs that potentially could last into later life. Using simple geometric algorithms, entire leaflets can even be replaced, as required.[5] This approach could extend repair into smaller valves and obviate many of the problems observed with previous techniques.[6]

CONCLUSIONS

In summary, unicuspid aortic valves can be challenging to repair, especially when hypoplastic. We present a pediatric case with a small deformed valve managed by upsizing to an adult annuloplasty ring and then augmenting the cusps with autologous aortic wall to obtain a competent adult-sized valve. This paradigm offers the potential for extending autologous valve repair into most bicuspid-unicuspid defects, but larger series and longer follow-up will be required for full validation.

Conflict of Interest Statement

Dr Rankin has been a consultant for BioStable Science and Engineering. All other authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

1. Sievers HH, Schmidtke C. A classification system for the bicuspid aortic valve from 304 surgical specimens. *J Thorac Cardiovasc Surg*. 2007;133:1226-1233.
2. Si MS, Conte JV, Romano JC, et al. Unicuspid aortic valve repair using geometric ring annuloplasty. *Ann Thorac Surg*. 2021;111:1359-1366.
3. Gerdisch MW, Reece BT, Emerson D, et al. Early results of geometric ring annuloplasty for bicuspid aortic valve repair during aortic aneurysm surgery. *J Thorac Cardiovasc Surg Tech*. 2022;14:55-65.
4. Myers JL, Clark JB, James TW, et al. Use of aortic wall patches as leaflet replacement material during aortic valve repair. *J Thorac Cardiovasc Surg Tech*. 2023;19:30-37.
5. James TW, Mehaffey JH, Wei LM, Voeller RK, Badhwar V, Rankin JS. Repair of calcified bicuspid aortic valves using living autologous aortic wall leaflets. *J Thorac Cardiovasc Surg Tech*. 2024;25:48-51.
6. Aicher D, Bewarder M, Kindermann M, Abdul-Khalique H, Schafers HJ. Aortic valve function after bicuspidization of the unicuspid aortic valve. *Ann Thorac Surg*. 2013;95:1545-1550.