

Geometric ring annuloplasty for bicuspid aortic valve repair in a child



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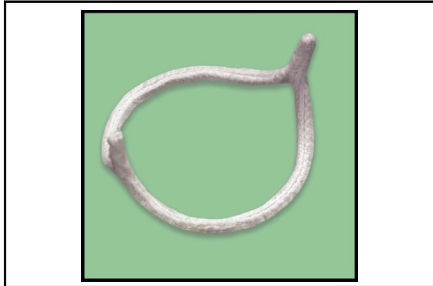
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Geometric bicuspid annuloplasty ring used in this procedure.

Central Message

An insufficient pediatric bicuspid valve was repaired by geometric ring annuloplasty and leaflet reconstruction, remodeling commissures to 180° and recruiting leaflet centrally to promote coaptation.

See Commentaries on pages e139 and e141.

Ring annuloplasty has been used routinely for surgical repair of tricuspid and mitral valves for more than 35 years, and more recently, a similar ring has been developed for bicuspid aortic valve (BAV) repair.¹ Annuloplasty rings restore normal size and geometry of the valve annulus, bring the native leaflets into better coaptation, and prevent future annular dilatation. In this article, the first case of a pediatric patient with a BAV and severe aortic insufficiency (AI) managed with internal ring annuloplasty is presented.

CASE REPORT

A murmur was discovered during a routine physical examination of an active, symptom-free 12-year-old boy (height, 159.6 cm; weight, 58.7 kg, body surface area, 1.6 m²). Cardiac Magnetic resonance imaging demonstrated AI with a 40% regurgitation fraction and severe left ventricular hypertrophy. A preoperative transthoracic echocardiogram showed a typical Sievers type 1 BAV,² with right-left commissural fusion, leaflet prolapse, and a large central coaptation gap.

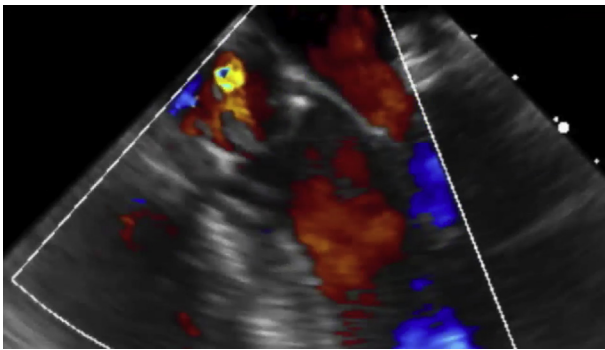
At surgery, the dilated valve annulus sized to a 21-mm diameter, and the noncoronary leaflet sized to a 19-mm bicuspid ring (HAART 200 Aortic Annuloplasty Device; BioStable Science and Engineering, Austin Tex; Figure 1). The validated sizing strategy measured the non-fused leaflet free edge length with special ball sizers, and

the length divided by 1.8 equaled the diameter of ring required for leaflet competence. The posts of the ring first were sutured to each subcommissural space with pledgeted horizontal mattress sutures (Video 1). Then, 2 transannular



FIGURE 1. Bicuspid annuloplasty ring used in this patient.

CONG



VIDEO 1. Repair of a bicuspid aortic valve with a geometric ring in a child. This video was presented at the European Congenital Heart Surgeons Association meeting, Lisbon, Portugal, Friday, June 1, 2018. Video available at: [https://www.jtcvs.org/article/S0022-5223\(19\)31340-6/fulltext](https://www.jtcvs.org/article/S0022-5223(19)31340-6/fulltext).

horizontal mattress sutures were placed through the sinus aspect of each leaflet annulus and looped around the ring to obtain a subannular position.

After the annuloplasty, the commissures were aligned to 180°, and both leaflets were moved centrally. Both leaflets still prolapsed as a result of unequal leaflet free-edge lengths, however, and the Schäfers leaflet reconstruction was used.³ Leaflet plication stitches were placed in the redundant noncoronary leaflet to raise it to a reference effective height of 8 mm (Video 1). Then, the cleft in the fused leaflet was closed with 3 simple sutures. This process brought the 2 leaflets into coaptation with equivalent free-edge lengths and effective heights, attaining good valve competence. After repair, transesophageal echocardiography showed good leaflet mobility and opening, no residual leak, and a 15-mm Hg mean systolic gradient (Figure 2). The patient’s postoperative course was unremarkable, and he was discharged on the third postoperative day. He continues to do well, with no symptoms and were fully functional more than 1 year after surgery. Magnetic resonance imaging at 6 months showed mild AI (12% regurgitant fraction) and mild aortic stenosis (peak velocity, 284 cm/s).

Pre-Repair

Post-Repair

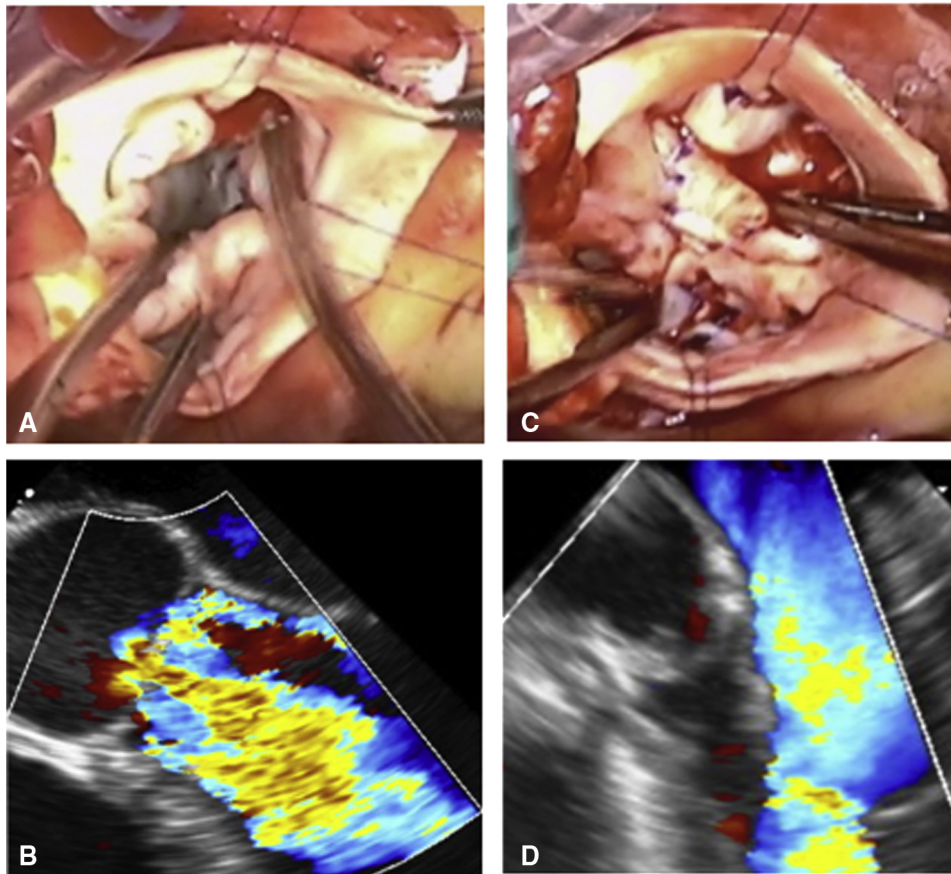


FIGURE 2. A, Prerepair appearance of type 1 bicuspid valve showing dysplastic incompetent leaflets. B, Prerepair echocardiogram showing severe aortic insufficiency. C, Postrepair valve appearance with good leaflet coaptation. D, Postrepair echocardiogram with a fully competent valve.

COMMENT

Annuloplasty is well established for mitral and tricuspid reconstruction, and a new internal geometric annuloplasty ring has shown promise for BAV repair in adults.¹ This case represents the first application of this technology to the pediatric population, but it seemed that repair principles were similar to those observed in adult valves. Objective reduction of annular diameter that is based on nonfused leaflet size is an advantage, and creation of 180° commissures facilitates leaflet reconstruction. In adults, ring annuloplasty has been applicable to all types of bicuspid anatomy, and intermediate-term results have been excellent.⁴ Effectively recruiting the patient's leaflets to the midline for coaptation and primarily relying on native leaflet tissue are advantages,⁵ because all types of pericardial leaflet substitutes have significant failure rates. The results obtained in this initial pediatric application seem promising, but more experience and follow-up will be required for full validation. Pediatric aortic valve disorders are composed primarily of BAV disease and its variants, so this topic is of prime importance.

Postoperative results associated with valve replacement in children have been suboptimal.⁶ Reports of outcomes after bicuspid valve repair have suggested better results, but reintervention rates could be significant, sometimes as a result of late annular dilatation,⁷ which could be prevented by an annuloplasty ring. In patients undergoing BAV repair at a mean age of 9 years, freedoms from reintervention and replacement at 7 years were 80% and 81%, respectively. At 7-years, however, more than a third of survivors had significant AI or valve stenosis.⁷ In the report by Siddiqui and colleagues,⁸ 35 of 146 patients undergoing BAV repair required subsequent reintervention at a mean follow-up of

8 years, and 30 more had significant valve dysfunction without reoperation. Of the remaining patients, only 54% had freedom from more than moderate valvular stenosis at 10 years, with the number as low as 36% at 18 years.⁷ The 10-year freedom from any significant event was 60% for those without use of patch material, whereas nearly all those with pericardial patch repair had an adverse event—again emphasizing the need to use primarily native leaflet tissue in the repair. Ring annuloplasty thus may be a viable alternative for BAV repair in pediatric patients, in an attempt to improve on current results; however, experience with more types of defects and more follow-up will be required to establish long-term clinical efficacy more completely.

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