Cone repair after tricuspid valve replacement in Ebstein anomaly

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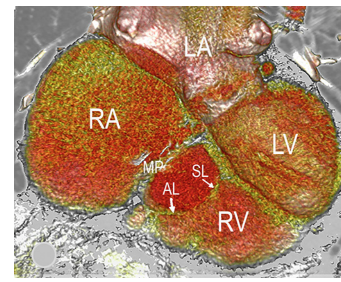


Cardiac computed tomography identifying native tricuspid valve leaflet tissues.

CENTRAL MESSAGE

The Cone repair is viable in selected Ebstein anomaly patients after the tricuspid valve replacement , encouraging the preservation of leaflet tissues at the time of prosthesis implantation.

▶ Video clip is available online.

Since its creation in 1993, the cone repair has been applied to all anatomical variations of Ebstein anomaly (EA), achieving durable tricuspid valve (TV) competency.[1-3] Although the cone repair has been described following certain TV repair procedures,[4,5] the literature has not addressed the cone repair after TV replacement in EA. We present 2 cases of TV prosthesis removal and native valve repair using the cone technique.

CASES

Since 2010, 2 of 5 patients with EA with previous TV prosthesis implants done at other institutions were reoperated on using the cone repair. The patients provided signed consent for medical data publication, and the institutional review board at the University of Pittsburgh approved this study (#20080084) on June 1, 2022.

The first patient was a 22-year-old man who presented with venous congestion and poor exercise tolerance 12 years after a TV bioprosthesis implant. The echocardiogram and the magnetic resonance imaging (MRI) of the chest demonstrated normal right ventricle (RV) function, a calcified TV prosthesis with severe stenosis, and regurgitation (Figure 1, *A*). We were also able to appreciate his native TV leaflets on his cardiac MRI. We removed the calcified prosthesis at the operation, finding preserved native TV leaflets downwardly displaced inside the RV. We conducted a TV repair using the cone technique, achieving a good valve performance (Figure 1, *B*). Currently, the patient is 34 years old, asymptomatic, and using metoprolol to treat systemic hypertension. His most recent echocardiogram, 12 years following repair, demonstrated normal RV function, mild RV and right atrium enlargement, and trivial TV regurgitation without stenosis (Figure 1, *C*).

The second patient was a 47-year-old woman who had undergone 4 previous open-heart operations: TV repair as a neonate, atrial septal defect closure at 5 years of age, mechanical TV implantation with concomitant homograft pulmonary valve replacement at 24 years of age, and mechanical valve re-replacement and maze procedure at 34 years of age. She was referred for surgical reintervention due to recurrent tachyarrhythmias, fatigue, and lower-extremity edema. Her computed tomography images demonstrated the presence of viable TV tissues for the cone procedure (Figure 1, *D*). The transesophageal echocardiogram demonstrated TV prosthesis thrombosis with

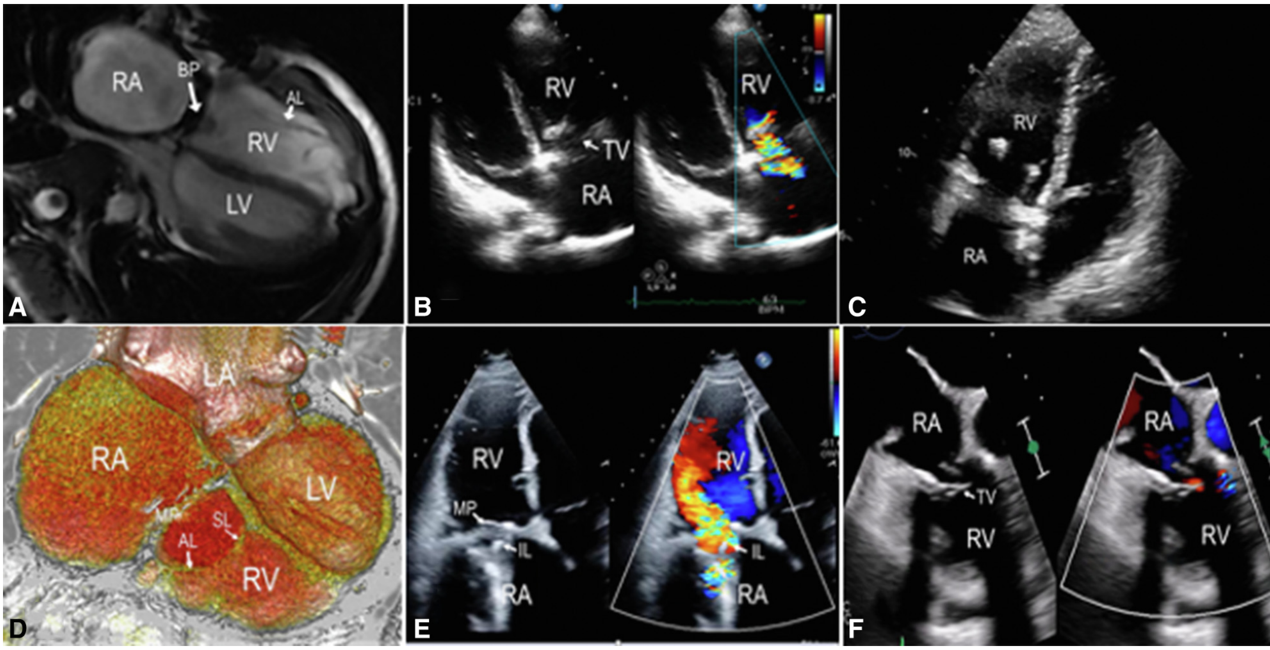


FIGURE 1. Pre- and postoperative diagnostic images of patients 1 (A-C) and 2 (D-F). A, Preoperative magnetic resonance image shows severe *right* atrial enlargement and the presence of tricuspid valve anterior and inferior leaflets tissues underneath the bioprosthesis. B, At 1 year after the repair, echocardiogram showed mild/moderate tricuspid regurgitation and mild *right* atrium and *right* ventricle dilation. C, Twelve years after the cone repair, the echocardiogram shows good tricuspid valve function and nearly normal-sized *right* heart chambers. D, Computed tomography with contrast shows a severe *right* atrial dilation and the presence of a tricuspid valve mechanical prosthesis. It reveals native tricuspid valve leaflet tissues inside the *right* ventricle. E, Prerepair transesophageal echocardiogram shows an immobile prosthetic leaflet causing severe tricuspid stenosis (mean diastolic gradient = 15 mm Hg). F, Post-cone repair transesophageal echocardiogram shows mild tricuspid valve regurgitation. *RA*, Right atrium; *BP*, biological prosthesis; *AL*, anterior leaflet; *RV*, right ventricle; *LV*, left ventricle; *TV*, tricuspid valve; *LA*, left atrium; *MP*, mechanical prosthesis; *SL*, septal leaflet; *IL*, immobile leaflet.

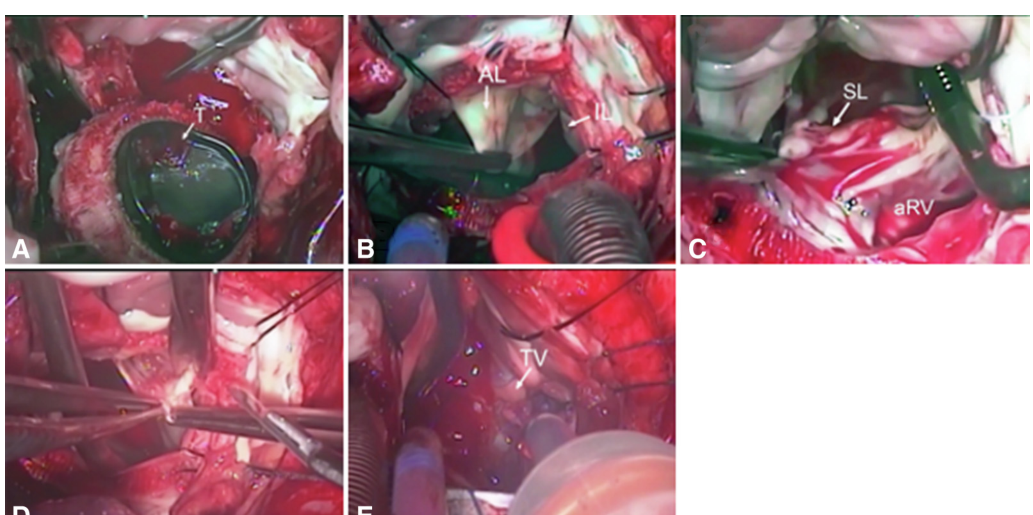
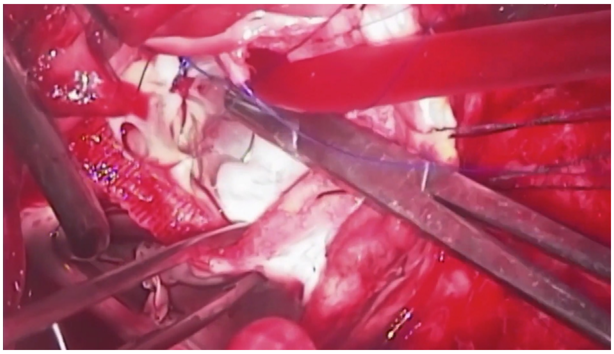


FIGURE 2. Intraoperative images on patient 2 show (A) the thrombosed mechanical tricuspid valve prosthesis extraction; (B) the exposure of the large anterior leaflet of the tricuspid valve and the displaced inferior leaflet; (C) the displaced hypoplastic septal leaflet; and (D) the tricuspid valve after an incision made at the anterior leaflet proximal attachment at the 2-o'clock position and extended towards the inferior leaflet. The anterior and inferior leaflets were completely mobilized by cutting all the abnormal attachments to the *right* ventricular wall. Another incision was made at the anteroseptal annulus, mobilizing the median edge of the anterior leaflet and the whole septal leaflet; (E) The saline test demonstrated good coaptation of tricuspid valve leaflets, without regurgitation. Other surgical details are demonstrated in Video 1. *T*, Thrombus; *AL*, anterior leaflet; *IL*, inferior leaflet; *SL*, septal leaflet; *aRV*, atrialized right ventricle; *TV*, tricuspid valve.



VIDEO 1. The surgical video shows the operative details on patient 2. It illustrates the operative steps consisting of the mechanical tricuspid valve extraction, the underlining native tricuspid valve exposure, the mobilization of the tricuspid valve leaflets, and the cone construction. The cone reconstruction of the tricuspid valve is done by placing a vertical suture uniting the anterior leaflet medial edge with the septal leaflet superior edge. A second vertical suture unites the inferior leaflet to the septal leaflet inferior edge, constructing a cone-like structure. These vertical sutures are done using a 5-0 polypropylene interrupted suture technique. Some proximal holes in the leaflets are closed. A vertical plication excluding a small thin atrialized area of the right ventricle inferior wall is carried out with 4-0 polypropylene interrupted suture technique. The cone is reattached to the normal atrioventricular junction after a small tricuspid annulus reduction. The saline test shows a competent tricuspid valve. In addition, this video shows some aspects of the biatrial maze procedure and the pulmonic valve replacement with a biological valve prosthesis. *T*, Thrombus; *AL*, anterior leaflet; *IL*, inferior leaflet; *SL*, septal leaflet; *aRV*, atrialized right ventricle; *TV*, tricuspid valve. Video available at: https://www.jtcvs.org/article/S2666-2507(23)00136-0/fulltext.

moderate regurgitation and severe stenosis (Figure 1, *E*). In addition, her homograft pulmonic valve had moderate regurgitation. We proceeded with mechanical valve extraction, TV cone repair, pulmonic valve replacement with a bioprosthesis, and redo bi-atrial maze ablation. The postoperative echocardiogram (Figure 1, *F*) showed no TV regurgitation or stenosis, with a 2.7-mm Hg mean diastolic TV gradient. She recovered well and was discharged home on postoperative day 6. At a 7-month echocardiographic follow-up, the TV had no stenosis and trivial regurgitation. We detail this operation in Figure 2 and Video 1.

DISCUSSION

The cone procedure has proven suitable in multiple situations and variable spectrums of EA anatomical presentations.[2] We describe these 2 cases to highlight a previously untapped EA population that can benefit from the cone repair. Echocardiogram, MRI, and operative reports of previous procedures can indicate the presence of TV leaflets underneath the tricuspid prosthesis, raising the possibility of valve repair.

Our decision to repair the TV instead of a prosthetic replacement was based on feasibility and to avoid multiple reinterventions, particularly in young patients. Our first patient excellent clinical condition and normal TV function at a 12 years' follow-up support our approach. Even 2 previous mechanical prosthesis implantations in our second patient did not preclude the viability of the cone procedure, achieving a competent valve.

Considering the option of future valve repair, the surgeons should keep the valvar tissues as much as possible while ensuring no obstruction to the RV inflow or outflow tracts during TV replacement in EA. Any valvar tissue that could cause obstruction or interfere with the prosthesis mechanism should be folded, relocated, or incised without resection. Implanting the prosthesis in a slightly supravalvar position may prevent heart block and facilitate the prosthesis extraction and a future valve repair. The sutures should be placed proximally to the anterior and inferior annulus, avoiding leaflets incorporation. At the septal area, the sutures are placed at the base of the coronary sinus and in the Todaro tendon, away from the atrioventricular node. The Thebesian valve may require an incision to keep the coronary sinus widely opened into the right atrium.

We observed that the RV function and structure were preserved in both patients, suggesting that TV replacement may help preserve the RV function or promote reverse remodeling of a dilated and dysfunctional RV in preparation for the cone procedure.

CONCLUSIONS

In conclusion, cone repair after the TV prosthesis implant is feasible, encouraging the preservation of most leaflet tissues during TV replacement in EA.

References

1. Da Silva JP, Baumgratz JF, Da Fonseca L, Franchi SM, Lopes LM, Tavares GMP, et al. The cone reconstruction of the tricuspid valve in Ebstein's anomaly. The operation: early and midterm results. *J Thorac Cardiovasc Surg*. 2007;133:215-23.
2. Da Silva JP, Da Fonseca Da Silva L. Ebstein's anomaly of the tricuspid valve: the cone repair. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu*. 2012;15:38-45.
3. Burri M, Agua KM, Cleuziou J, Beran E, Nagdyman N, Kuhn A, et al. Cone versus conventional repair for Ebstein's anomaly. *J Thorac Cardiovasc Surg*. 2020;160:1545-53.
4. Dearani JA, Said SM, Burkhart HM, Pike RB, O'Leary PW, Cetta F. Strategies for tricuspid valve re-repair in Ebstein's malformation using the Cone technique. *Ann Thorac Surg*. 2013;96:202-8.
5. Da Silva JP, Viegas M, Castro-Medina M, Da Fonseca Da Silva L. Da Silva Cone operation after the Starnes procedure for Ebstein's anomaly: new surgical strategy and initial results. *J Thoracic Cardiovasc Surg Tech*. 2020;3:281-3.