The concept of cone creation to treat isolated tricuspid valve dysplasia and the case of a double-orifice tricuspid valve

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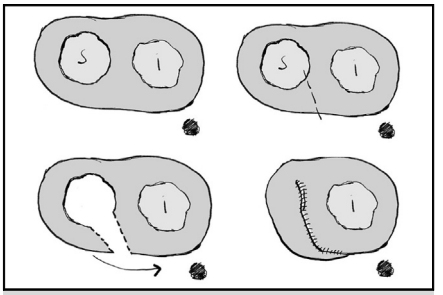
ABSTRACT

Objectives: Isolated tricuspid valve dysplasia is a rare disease characterized by a wide spectrum of possible anomalies. We describe the use of the Cone concept to treat a patient with a double-orifice tricuspid valve with massive regurgitation and severe deficit of coaptation.

Methods: Three adult patients with congenital non-Ebstein tricuspid valve anomaly characterized by severe coaptation deficiency underwent tricuspid valve repair applying the Cone technique. In particular, we describe the case of a symptomatic 21-year-old woman with a double-orifice tricuspid valve, with massive regurgitation and severe right ventricular dilatation. The tricuspid valve was transformed from a double-orifice valve into a single-orifice valve. The most superior orifice was opened, and the tissue surrounding the orifice was used to extend the leaflet of the inferior orifice. A Cone was created, and a ring annuloplasty was used to stabilize the result.

Results: The patient was discharged home after 7 days with trivial residual tricuspid regurgitation and no significant antegrade gradient. The final coaptation height was 2.8 cm. The cardiothoracic ratio decreased from 0.77 to 0.59 after 2 months, and symptoms promptly improved.

Conclusions: Over the past 2 years, we have applied the Cone creation concept to patients with a severely dysplastic tricuspid valve with excellent early results. One patient had a double-orifice tricuspid valve, and a Cone repair concept was adopted anyway. One orifice was sacrificed, and surrounding tissue was used to augment the leaflets of the other orifice. A Cone was created to improve central coaptation with a good initial result. (JTCVS Techniques 2023;20:71-8)



Transformation of a double-orifice into a single-orifice valve.

CENTRAL MESSAGE

Cone repair is a standardized and reproducible technique that should also be considered outside of EA. A double-orifice valve is no exception.

PERSPECTIVE

Isolated TV dysplasia is a rare disease with a wide spectrum of presentations. Cone repair could provide an excellent solution to encompass anatomic difficulties in this unique group of patients.

Video clip is available online.

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Congenital tricuspid valve (TV) disease is a rare malformation, accounting for approximately 1% of all the congenital cardiac anomalies. Among congenital TV diseases, Ebstein's anomaly (EA) represents 40% of the tricuspid malformations.[1,2] The remaining malformations, the so-called non-Ebstein, are mostly represented by TV diseases associated with other congenital anomalies: tetralogy of Fallot, right ventricular (RV) outflow tract obstruction, left to right

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Abbreviations and Acronyms
EA = Ebstein's anomaly
RV = right ventricular
TR = tricuspid regurgitation
TV = tricuspid valve

shunt (atrial septal defect, ventricular septal defect, atrioventricular septal defect, coronary fistula), and pulmonary stenosis/atresia.[2] The true isolated congenital TV dysplasia is the rarest form of TV disease and is characterized by a wide spectrum of possible anatomic conditions. The main features are absence of downward displacement of the leaflets,[3] involvement of the leaflets and chordae, and annular dilatation. Because the competence of the TV depends on the perfect interaction among leaflets, annulus, and subvalvar apparatus, TV dysplasia is frequently associated with regurgitation. For many decades, regurgitant TV has been tolerated as a kind of mildly dangerous patient companion; however, recent reports[4,5] have emphasized that this companion is far from benign and affects the long-term survival of patients. Between 2021 and 2022, we surgically repaired isolated non-Ebstein TV in 3 symptomatic patients with severe tricuspid regurgitation (TR) characterized by a large coaptation deficit using the Cone technique.[6] In particular, we describe the case of a patient with a congenital double-orifice TV that was successfully repaired by transforming the valve from a double-orifice opening into a single cone.

The aim of this article is to consider the use of the Cone technique in patients with isolated non-Ebstein TR characterized by a severe coaptation deficit, because it is a standardizable, reliable, and effective technique.

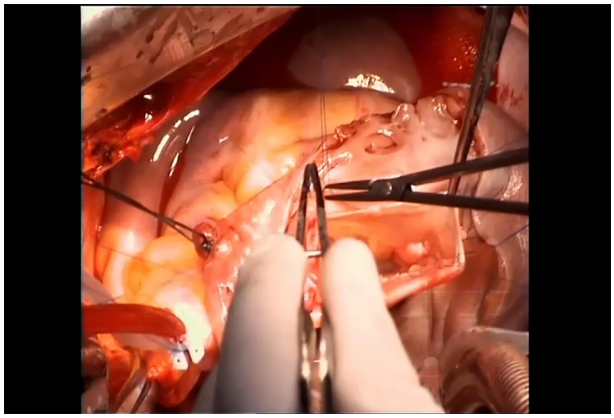
MATERIAL AND METHODS

The authors obtained written informed consent for the publication of data, photographs, and videos. Per institution, Institutional Review Board approval was not required.

Three patients with isolated TV dysplasia underwent TV repair applying the Cone creation concept. Regardless of the anatomic differences between patients, the primary reason for choosing the Cone technique was that a severe coaptation deficit was observed (mean coaptation deficit 2.8 cm), mainly due to shortened chordae with multiple attachments, leaflet deficit, and RV dilation causing severe tethering. In particular, we describe the case of a 21-year-old woman (52 kg, 168 cm, 1.57 m^2) presenting a double-orifice TV with massive regurgitation from both orifices and severe right atrial and RV dilatation. The patient presented dyspnea with minimal effort. The electrocardiogram showed sinus rhythm with right bundle branch block. A Holter electrocardiogram failed to detect arrythmias of any kind. A transthoracic echocardiogram showed massive TR with double-orifice TV, severe right atrial dilatation with an indexed volume of 277/mL/m^2, severe RV dilatation with shortening fraction of 31%, and normal-sized left ventricle with normal ejection fraction. Magnetic resonance imaging was performed to evaluate RV volume and function. The RV was found to have 417 mL/m^2 total volume and 40% ejection fraction. The right atrium had a volume of 256 mL/m^2. The left ventricle was found to be in the lower limit in size with an indexed end-diastolic volume of 47 mL/m^2 and normal ejection fraction. Although the surgical indication was already evident, a cardiopulmonary exercise test was performed, and a severe reduction in cardiopulmonary capacity was found with oxygen consumption of 22 mL/min/kg (56% of the expected value) and a minimal deoxygenation under strenuous effort. Surgical repair was deemed necessary, and the patient underwent TV repair.

Surgical Technique

The heart was accessed through a full sternotomy (Video 1). Cardiopulmonary bypass was conducted at 32 °C. After infusion of del Nido cardioplegic solution into the aortic root, the right atrium was opened with an incision parallel to the atrioventricular groove. A reduction of the right atrium was performed. The TV was exposed and revealed the 2 orifices, 1 superior and 1 inferior. The 2 orifices appeared to be guarded by short leaflets with multiple secondary chordae and fibrous attachments. In a saline water test, a complete lack of central coaptation was observed on both sides. The decision was to create a single-orifice valve to facilitate repair. The leaflets were detached from the annulus at 360°, and the superior orifice was opened, and all its subvalvar apparatus was removed. The secondary chordae and fibrous attachments on the leaflets guarding the inferior orifice were dissected to maximize tissue mobility. Residual tissue from the superior orifice was rotated counterclockwise and sutured to the septal portion of the inferior orifice (Figure 1). At this point, it was evident that both the anterior and septal leaflets were too short to guarantee a high coaptation height, and lengthening with fresh autologous pericardium was considered essential. The fenestrations were sutured directly if small or with an autologous pericardial patch if larger than 5 mm. In this patient, there were no anatomic commissures, and a cone was created to just increase the height of the already existing leaflets. The size of the cone was tested with an appropriate size Hegar dilator. The annulus was plicated with a mattress suture over the inferior portion and then reduced with a suture applied directly on the annular margins. Finally, the cone was reinserted onto the annulus with 4 intertwined sutures to avoid further shrinking of the TV orifice (Figure 2). At the level of the septum, the suture was displaced downward to avoid the atrioventricular node and possible atrioventricular block. A prosthetic ring (36 Contour 3D, Medtronic) was then placed to stabilize the surgical result. The water test was satisfactory, and the right atrium was closed. After the crossclamp was removed, the



VIDEO 1. Step-by-step procedure of transforming a double-orifice TV into a single-orifice TV, resulting in the creation of a single cone. Video available at: https://www.jtcvs.org/article/S2666-2507(23)00139-6/fulltext.

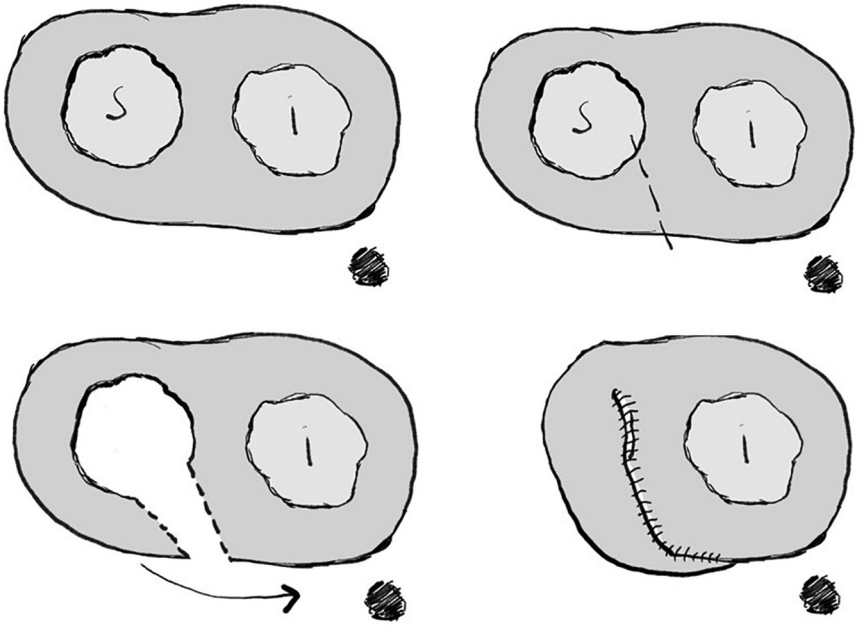


FIGURE 1. Sketch showing the transformation of a double-orifice TV into a single-orifice valve. S: superior orifice; I: Inferior orifice.

heart resumed sinus rhythm and the cardiopulmonary bypass was easily discontinued. A transesophageal echocardiographic evaluation showed no residual regurgitation with a large coaptation height and no significant antegrade gradient.

In the other 2 patients, although different surgical techniques were adopted, the concept of creating a cone was maintained. In 1 patient, whose anterior leaflet was deficient, all leaflets were detached and freed from the secondary chordae. The anterior leaflet was augmented with autologous

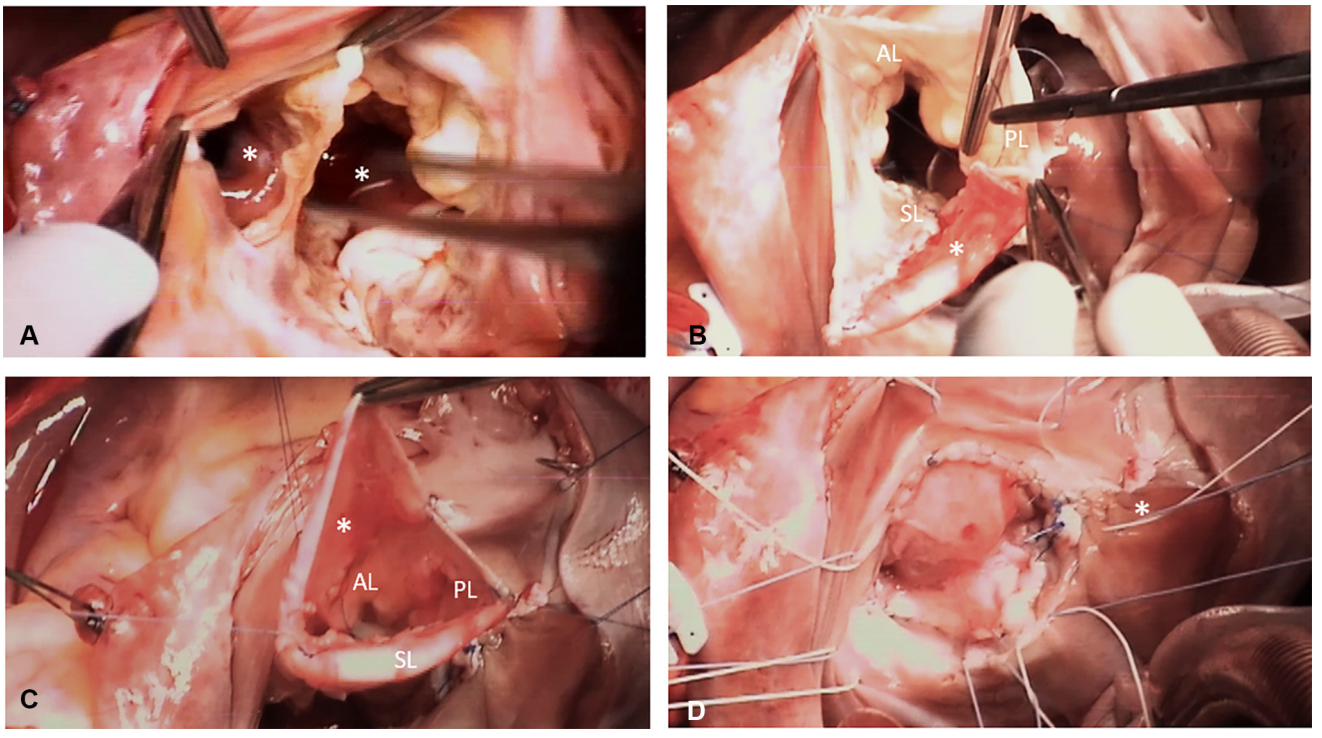


FIGURE 2. Technique of a construction of a single cone starting from a double-orifice TV. A, Double orifices are evidenced. B, Septal leaflet is elongated with autologous pericardium marked with *. C, Anterior leaflet is elongated with autologous pericardium marked with *. D, Final appearance of the cone reimplanted on a reduced annulus, marked with *. *AL*, Anterior leaflet; *PL*, posterior leaflet; *SL*, septal leaflet.

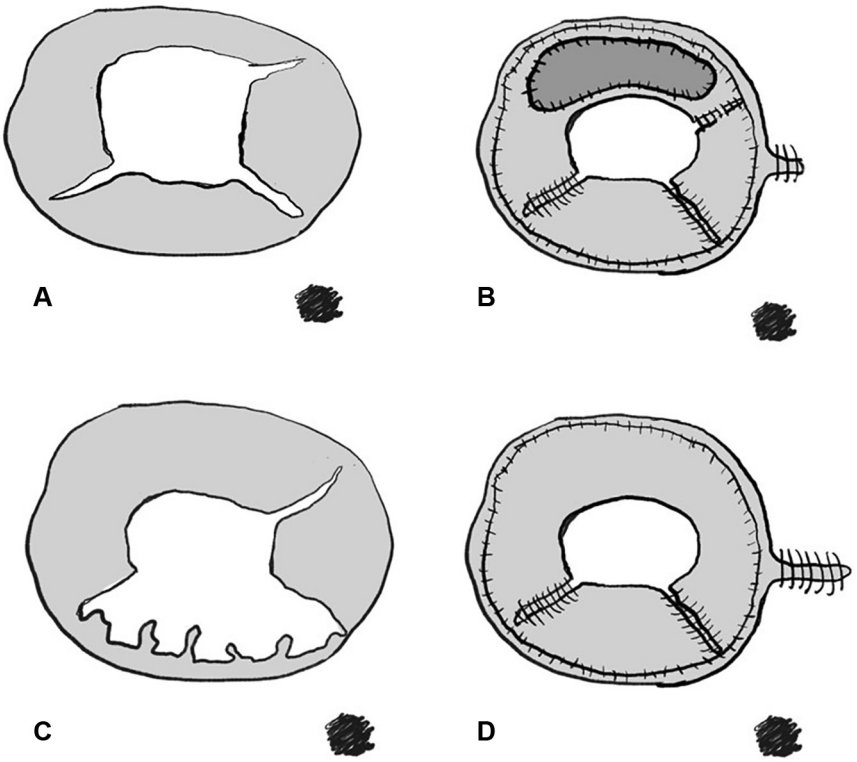


FIGURE 3. Schematic diagram showing the surgical technique adopted in the other 2 patients. A and B, Preoperative and postoperative view of a patient with a short anterior leaflet. C and D, Preoperative and postoperative view of a patient with a rudimental septal leaflet.

pericardium, the commissures were sutured, the annulus was reduced, and the cone was reinserted in the annular plane (Figure 3, *A* and *B*).

A third patient showed a rudimentary septal leaflet. In this situation, the anterior and posterior leaflets have been detached and freed from all secondary chordae. The annulus was then reduced, the anteroposterior commissure was sutured, and after a clockwise rotation the posterior leaflet was used to cover the septal portion of the annulus and was sutured to the anterior leaflet (Figure 3, *C* and *D*).

RESULTS

At 13 months of mean follow-up, the 3 patients had less than moderate TR, no significant antegrade gradient, and high coaptation height. The RV and right atrial volume decreased by 26% and 43%, respectively, and symptoms improved significantly (Table 1).

The patient whose double orifice was transformed into a single cone showed marked improvement in functional capacity and is currently in New York Heart Association class I at 8 months follow-up. The RV volume was reduced to 286 mL and right atrium to 117 mL; however, the RV shortening fraction was reduced to 23%. This finding was also observed in the other 2 patients whose massive TR was repaired. The coaptation height was particularly satisfactory and measured 2.8 cm (Figure 4). The cardiothoracic ratio improved significantly from a preoperative value of 0.77 to a postoperative value of 0.59.

DISCUSSION

Isolated TV dysplasia is a rare anomaly characterized by the absence of downward displacement of the leaflets. Common anatomic findings are dilatation of the annulus, shortened chordae, and rolled up leaflets all contributing to TR. The 3 leaflets are present and delaminated, but the shortened chordae could create tethering and apical regurgitation, mimicking the EA. For many decades, isolated TR was considered almost a benign finding, particularly when compared with other single heart valve lesions. However, several recent articles have highlighted that TR significantly influences morbidity and mortality in patients with acquired and congenital heart diseases.[4,7]

There is a lack of clear indications for the treatment of isolated congenital TR, and surgeons refer to the indications used in TV diseases associated with acquired heart disease. In our practice, we indicate surgery in patients with dilating right heart chambers or in patients with symptoms or evidence of severely reduced cardiopulmonary function.

An additional problem is related to the mortality in adult patients receiving isolated TV surgery, which has been shown to be higher compared with other single-valve procedures. TV repair induces a geometric readjustment of the RV that is difficult to manage in the postoperative period compared with the left ventricle. Furthermore, TV leaflets

TABLE 1. Preoperative and postoperative details of the 3 patients with isolated tricuspid valve dysplasia treated with the Cone creation

	Anatomic details	Surgical technique	Right atrial volume reduction (%)	TV annulus		TV coaptation		TR		RV volume reduction (%)	RV FAC		Cardiothoracic ratio	
				PRE	POST	PRE coaptation deficit	POST coaptation height	PRE	POST		PRE	POST	PRE	POST
Pt 1	Double-orifice TV valve	Cone creation on a single orifice and anterior and septal leaflet augmentation	58%	60 mm	28 mm	30 mm	28 mm	severe	mild to moderate	24%	31%	23%	0.77	0.59
Pt 2	Severe hypo-represented septal leaflet	Cone creation and septal leaflet exclusion	37%	42 mm	26 mm	26 mm	23 mm	severe	mild	33%	40%	34%	0.52	0.46
Pt 3	Retracted anterior leaflet	Cone creation and anterior leaflet augmentation	34%	40 mm	28 mm	28 mm	24 mm	severe	mild to moderate	22%	30%	27%	0.48	0.45

TV, Tricuspid valve; *TR*, tricuspid regurgitation; *RV*, right ventricular; *FAC*, fractional area change.

are very fragile, and this, in association with the presence of a severely dilated RV, can make it difficult to obtain adequate coaptation.

The main mechanism underlying regurgitation in these patients is poor central coaptation.[8,9] In a previously published article, we described our experience in the use of autologous pericardium to increase leaflet size and coaptation height.[10] However, over the past 2 years, our experience using the Cone technique in EA has led to its use in isolated non-Ebstein TV disease as well. A long cone acts like a Heimlich valve and ensures an incredibly high coaptation margin.

The Cone technique differs from previous techniques used to address EA primarily in that it creates a perfectly guarded TV anulus over a 360° angle. The results on the Ebstein-type valve proved to be excellent with regard to the absence of recurrent regurgitation and reoperation in the short to medium term. The Mayo Clinic, which had the greatest experience with the Cone technique, describes a surgical mortality close to 0% and an early reoperation rate of 4.9%.[11] At medium-term follow-up, Holst and colleagues[12] described a freedom from late reoperation of 97.9% and a progressive and persistent decline in RV size.

Outside EA, TV valve repair in adults with congenital heart disease has shown worse short- and medium-term results. Multiple articles[5,8,13] evidenced that, in this population, recurrence of TR is a common finding from discharge through medium-term follow-up, with greater than moderate TR occurring in approximately 1 in 5 patients over 3 years after repair. These data led us to use the Cone technique in non-Ebstein (Figure 5).

Among the 3 patients with isolated TV dysplasia treated with the Cone technique, the double-orifice TV was the most difficult to repair. TV duplication is rarer than mitral valve duplication and is usually associated with other anatomic defects.[14,15] Hartmann[16] and Sanchez Cascos[17] described and classified the double-orifice atrioventricular valve into 3 subtypes: *commissural type*, in which an accessory orifice is located within the commissure; *central type*, with 2 symmetrical orifices and a single or double tensor apparatus; *hole type*, characterized by a hole into 1 leaflet. The patient had a central type with 2 different tension apparatuses. Among the possible options, valve replacement was not considered because, in our experience, we always tend to repair the TV unless the valve has already undergone extensive repair surgery or in case of regurgitation of a systemic TV.

Functionally, both orifices were regurgitant and a severe coaptation deficit was present on both sides. Because the inferior orifice was adequately sized for the patient, it was decided to eliminate the superior orifice to create a single conical orifice valve. The idea of repairing both orifices was not considered, as was the idea of plugging 1 orifice with a patch and repairing the remaining orifice. Instead,

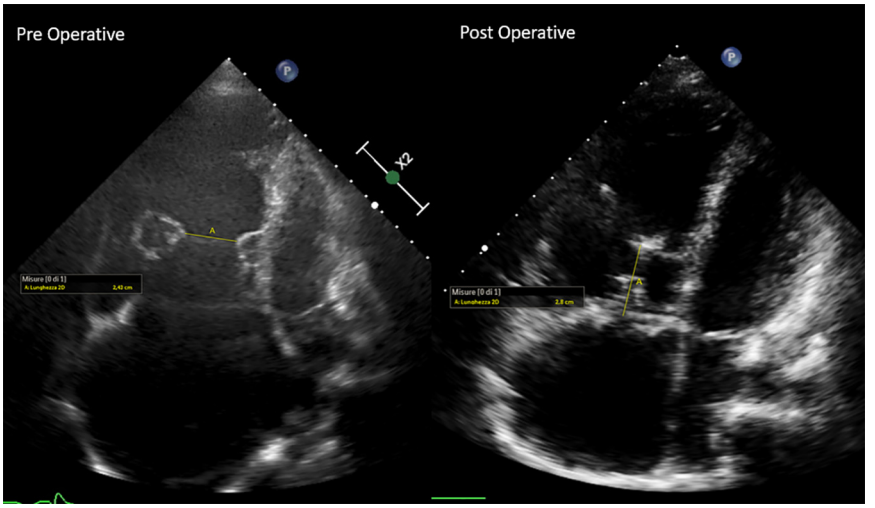


FIGURE 4. Comparison of preoperative and postoperative coaptation height as evidenced by a transthoracic echocardiogram.

tissue from 1 orifice was used to enlarge the second orifice. We were forced to completely detach the valve leaflets from the annulus to open the superior orifice. However, this is also our preferred approach in EA because it allows for better visualization of the leaflets and subvalvar apparatus. Furthermore, the newly created cone could be reattached

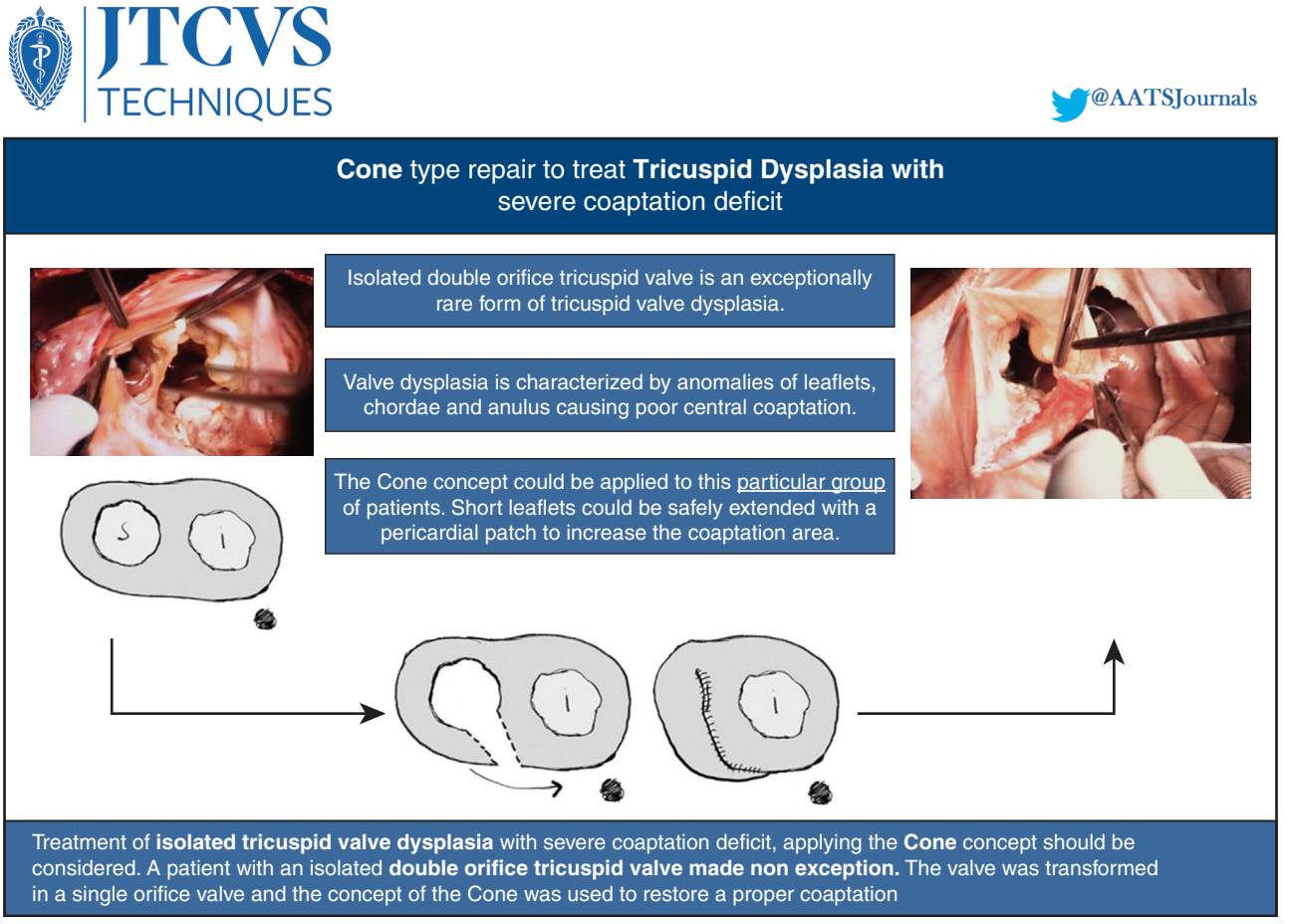


FIGURE 5. Graphical Abstract showing the application of the Cone concept to create a competent valve in the setting of a double orifice tricuspid valve.

with reduced twisting of the chordae. After dissection of the fibrous attachments and the secondary chordae, the leaflets were lengthened with autologous pericardium because they appeared too short to ensure good central coaptation. This technique is widely adopted to address short leaflets in tethered TV[10] and is also described by Dearani[18] during cone creation as a way to bring the edge of the anterior leaflet closer to the septal leaflet. In our patient, the septal leaflet was also very short and required lengthening with a pericardial patch. In 1 other patient with a short anterior leaflet, it was necessary to augment the leaflet with autologous pericardium. We do not adopt any equation to size the patch, but pulling on the free wall of the RV could mimic the filling of the ventricle, making evident how much length the anterior leaflet lacks to have good mobility. It should be noted that the extensive use of pericardial patches in pediatric patients may reduce the duration of repair and the growth potential; however, in adult patients, this problem is less significant.

The annuloplasty was performed with a mattress suture reinforced with pledgets on the entire inferior portion of the annulus to distribute the plication and avoid possible kinking of the coronary artery. The decision was made to stabilize the annuloplasty with a 36 Contour 3D ring (Medtronic) to avoid future dilatation and reduce tension on the reinserted leaflets. The size was chosen on the basis of the size of the repaired valve without having the intention of proceeding with a further reduction of the ring.

Unlike EA, there was no need for RV plication because there was no atrialized RV. Furthermore, none of the patients had an arrhythmic substrate because in isolated dysplastic TV there is no myopathy, and the complete delamination of the leaflets and the absence of muscular attachments decrease the presence of accessory conduction pathways. In fact, none of the patients had heart rhythm disturbances before and after the surgery.

A final consideration must be made with regard to the ventricles. In EA, the left ventricle is usually reduced in size and volume, and the RV may be severely enlarged and dysfunctional, potentially requiring a cavopulmonary anastomosis. In isolated TV dysplasia, the 2 ventricles are more balanced and the cavopulmonary connection probably is of reduced usefulness.

Postoperative management was conducted as in patients with EA: reduced carbon dioxide, avoidance of volume overload, inotropic support, afterload reduction, and pacing if bradycardia was a concern.

The immediate result was satisfactory, and the RV volume decreased consistently since discharge. The shortening fraction decreased as seen for the left ventricle after mitral valve repair; however, we expect improvement along follow-up. Although the shortening fraction has decreased, the antegrade flow through the pulmonary valve may be thought to have increased anyway. Although magnetic resonance imaging has not yet been performed to highlight these data, the improvement of the functional class demonstrates a better hemodynamic performance.

CONCLUSIONS

Isolated TV dysplasia is a rare congenital disorder characterized by the absence of downward displacement of the leaflets and severe TR primarily related to leaflets tethering. TR has been erroneously considered benign for a long time; however, recent reports have highlighted that its presence could adversely affect the long-term survival of patients. As the Cone technique to treat EA is increasingly adopted worldwide because it is reproducible and reliable, we used this technique to also treat isolated non-Ebstein TV dysplasia. In particular, we describe the case of a girl with a rare, isolated double-orifice TV. To make it possible to use the Cone technique in this patient, an orifice was sacrificed, the valve was turned into a single orifice, and a cone was created. The result was satisfactory with no residual regurgitation or stenosis.

Webcast

You can watch a Webcast of this AATS meeting presentation by going to: https://www.aats.org/resources/the-concept-of-cone-creation-to-treat-severe-coaptation-deficit-in-double-orifice-tricuspid-valve.

Conflict of Interest Statement

The 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.

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Key Words: cone repair, congenital heart disease, tricuspid dysplasia, tricuspid regurgitation