Successful catheter ablation for ventricular tachycardia after cone procedure in Ebstein anomaly

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Introduction

Ebstein anomaly (EA) is a rare congenital heart disorder characterized by delaminated tricuspid valve (TV). This condition serves as an arrhythmic substrate contributing to atrial and/or ventricular arrhythmia and is notably associated with a high incidence of arrhythmias.[1,2] The cone reconstruction (CR) approach, which was first described by da Silva[3] in 2007, has advanced with time and is widely applied to patients with EA.[4] Despite the good outcomes of CR,[5,6] arrhythmic complications can occur after the procedure. We report a case of EA with early postoperative sustained ventricular tachycardia (VT) originating from the site of vertical plication after CR, which was successfully treated using radiofrequency (RF) catheter ablation.

KEY TEACHING POINTS

- Ebstein anomaly is a rare congenital heart disease that is often associated with a supraventricular or ventricular arrhythmia.
- The ventricular plication site of the cone procedure in Ebstein anomaly may provide the focus of ventricular tachycardia (VT).
- The radiofrequency ablation for VT can be applied safely and effectively in the early postoperative period of the cone procedure.

Case report

A 13-year-old boy was diagnosed with EA 2 months before hospitalization during a school health screening program. He complained of mild limitation of physical activity. Walking to and from school resulted in fatigue and dyspnea on exertion (NYHA class II). At the school health screening, chest radiography revealed significant cardiomegaly; thus he was referred to our hospital for further evaluation.

Transthoracic echocardiography revealed an atrialized right ventricle and apical displacement of the septal and posterior leaflets of the TV (Figure 1A), with severe tricuspid regurgitation. Twelve-lead electrocardiogram (ECG) revealed right atrial enlargement and an incomplete right bundle branch block (RBBB) pattern, with no definite ventricular pre-excitation. Because there were no documented arrhythmic events and no symptoms of arrhythmia, a preoperative electrophysiological study (EPS) was not conducted.

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CR was performed with vertical plication of the atrialized right ventricle and reduction plasty of the right atrium (Figure 1B). The degree of TV regurgitation improved, and there was no evidence of coronary insufficiency after CR. Three days postoperatively, junctional tachycardia was detected (heart rate, 130 beats/min), which was controlled by discontinuing inotropic drugs and cooling the patient. Five days postoperatively, monomorphic VT (Figure 2A) was observed while the patient defecated. The QRS complex of VT exhibited a superior axis with RBBB pattern. Tachycardia demonstrated ventriculoatrial dissociation. This refractory VT was resistant to overdrive pacing, direct current cardioversion, and antiarrhythmic drugs including amiodarone and lidocaine. Overdrive pacing and direct current cardioversion resulted in no change in the VT rate, while the administration of amiodarone decreased the VT rate (from 155 to 140 beats/min).

Owing to the persistent VT that was refractory to medical management, a transcatheter ablation was performed. Because VT demonstrated a superior axis with an RBBB pattern and QRS with transition at V_2, a left ventricular approach was performed. Because VT was not affected by burst left ventricular pacing, it was assumed to be automatic. However, the earliest activation site, the left interventricular

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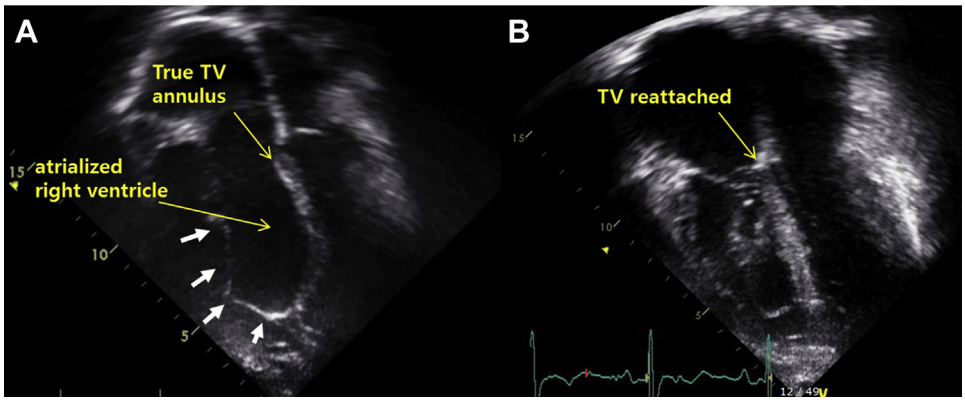

Figure 1 **A:** Preoperative 2-dimensional echocardiography image of apical 4-chamber view demonstrated characteristic displaced tricuspid valve (TV) anterior and septal leaflets (*arrow*). **B:** Postoperative 2-dimensional echocardiography image of apical 4-chamber view revealed a reattached TV leaflet at true atrioventricular valve annulus by cone reconstruction.

septal surface, was wide and late (2 ms later than the QRS complex). Finally, the right ventricle was explored, and right ventriculography was performed (Figure 3A and B). There was a recess at the right ventricular (RV) apical septum left by vertical plication during CR. During the early postoperative period, catheter maneuvers were extremely difficult and may have been dangerous owing to the fragile TV tissue and newly plicated RV wall. To avoid further damage during catheter manipulation, a long sheath was inserted into the right ventricle. During prudent exploration of the RV septal area, the earliest signal was detected 26 ms earlier than the QRS complex (Figure 2B). The origin of automatic VT was approached via the recess at the immediate apical side of the ventricular plication (Figure 3A and B). Full electroanatomical mapping was not performed owing to the patient's tenuous hemodynamic status and difficult catheter manipulation within the right ventricle. Owing to the softer nature of conventional RF ablation catheters, a temperature-controlled conventional RF catheter was used first. Multiple trials of ablation at VT foci failed, owing to very low power (2–3 W), which was delivered using the temperature-controlled mode (70°C). An irrigated-tip ablation catheter was introduced and carefully advanced into the recess (Figure 3C and D). Eventually, after 12 failures with conventional catheter, the irrigated-tip catheter was used to conduct conveyance of appropriate energy to the substrate, and VT was terminated at the end of the 13th attempt. RF ablation energy was delivered using the 30 W setting, an average temperature of 37°C, and average impedance of 79 ohms (maximum 103 ohms) for a total of 160 seconds. In addition, 3 more consolidation lesions were placed. There were no complications, and cardiomegaly exhibited marked improvement. TV function remained unchanged after the procedure. The patient has not complained of symptoms, and the ECG, including Holter monitoring, has been free of any VT during 3 years of follow-up.

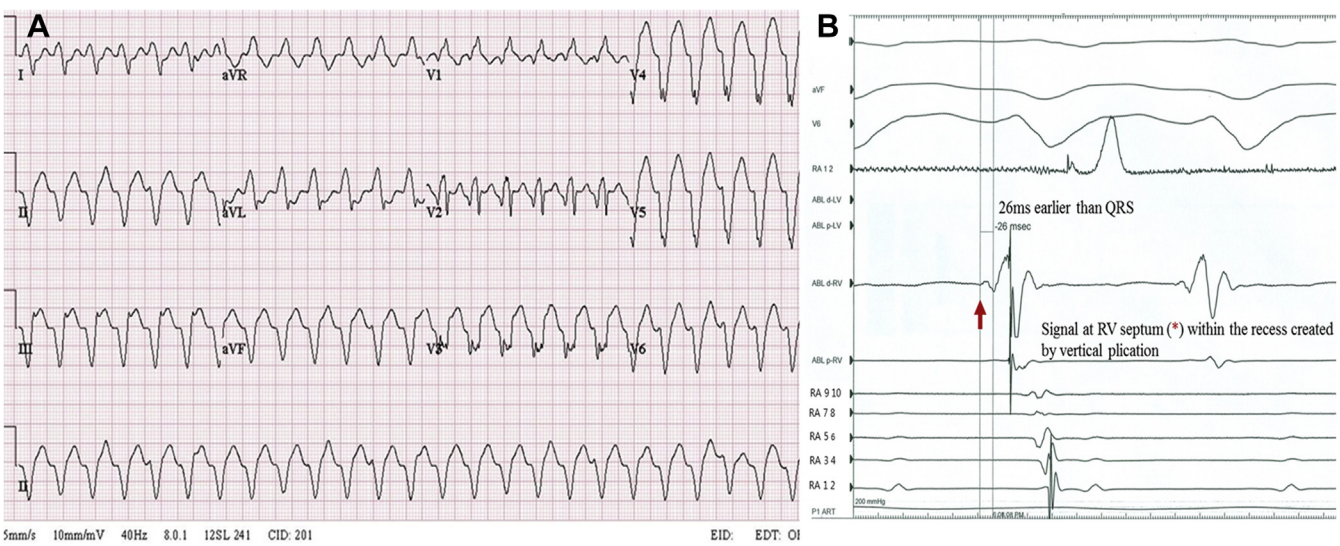

Figure 2 **A:** The electrocardiogram at postoperative day 5 showed ventricular tachycardia with superior axis, right bundle branch block pattern QRS complexes and ventriculoatrial dissociation. **B:** The signal coming from the right ventricular (RV) septum was 26 ms earlier than the beginning of the QRS complex (*arrow*).

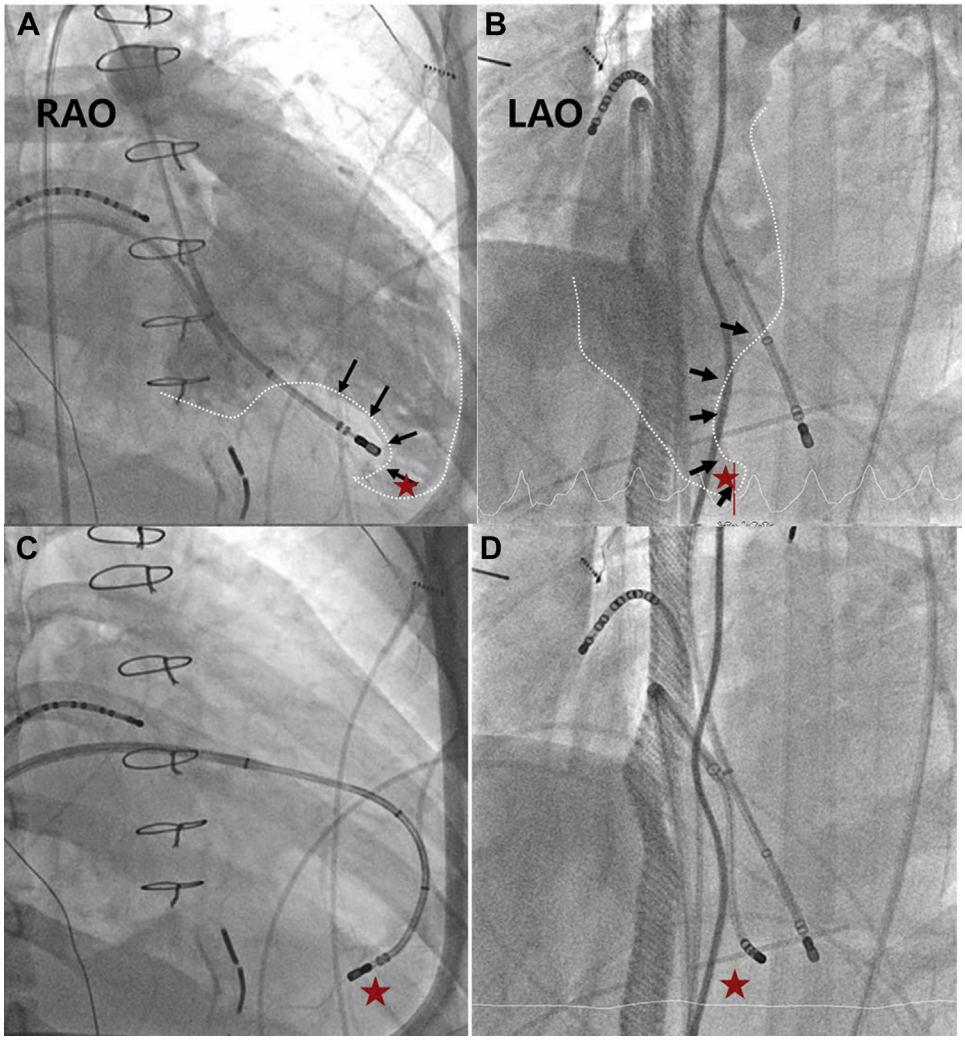

Figure 3 **A:** Right anterior oblique (RAO) and **B:** left anterior oblique (LAO) projection of the ventricular plication site (*arrow*) and the recess. **C,D:** Using the irrigation-tip ablation catheter, the ventricular tachycardia was successfully terminated just at the apical side of the ventricular plication site (*asterisk*).

Discussion

EA is a rare disease, occurring in 1–5 per 200,000 live births.[7] Clinical manifestations are diverse depending on the extent of TV regurgitation, dysfunction of the RV myocardium, and dysrhythmias at variable presenting ages.[8,9] Components of the arrhythmic substrate include atrioventricular accessory pathways, distended and damaged atrial and atrialized RV myocardium, and postsurgical scarring.[10] The CR procedure involves mobilization of the TV and reattachment to the true atrioventricular valve annulus after rotation. During this procedure, the atrialized right ventricle is longitudinally plicated to exclude its thin part.[3,4,11] Although CR can be associated with low morbidity and mortality, several cardiac arrhythmias may persist after the procedure.[5,12] We described the case of a 13-year-old EA patient who underwent CR, which was complicated by early postoperative VT. Preoperative assessment of arrhythmia using EPS may be considered to investigate arrhythmic substrates in patients. However, it is debatable whether preoperative EPS should be performed in all patients or if it is needed only in selected patients with Wolff-Parkinson-White syndrome, previously documented arrhythmia, or suspected arrhythmia(s) based on symptoms.[12,13] In assessing ventricular arrhythmias in EA patients with and without surgery, the substrates may be macroreentrant or focal.[14] After surgical repair of EA, macroreentrant VT may occur along regions of scarring, involving the anterior or inferior free walls. A recent multicenter study investigating RF catheter ablation for VT in EA demonstrated that focal VT after surgical repair was related to the anterior septum near the resected leaflet tissue, or the diseased Purkinje tissue.[14]

Because we did not perform EPS in this patient before surgery, we cannot deny the possibility of the preexisting nature of VT. However, VT in this patient may have been a complication of surgery, because the VT originated from the very end of the ventricular plication suture. The VT ECG of this patient exhibited an RBBB pattern in the QRS wave with a superior axis; however, the earliest activation site was located at the apical septal side of the RV plication. This atypical QRS pattern of VT may be caused by an enlarged RV volume and distortion of normal geometry. Eventually, VT arose from the plicated right ventricle, which may be a unique

nidus for ventricular arrhythmia. Considering the pathology of EA, the culprit may possibly be hidden at the plicated ventricular myocardium. We thoroughly assessed the right ventricle owing to the possibility of immediate postoperative vulnerability and limited catheter manipulations after CR.[9] To avoid further mechanical injury, we did not use a stiffer and larger-caliber 3-dimensional mapping catheter. However, because vertical plication of the atrialized right ventricle resulted in a recess and the exact origin of the VT was within the plicated bulk of the myocardium, a conventional temperature-controlled RF ablation catheter failed to deliver adequate energy. Ultimately, an irrigated-tip ablation catheter enabled transmittance of adequate energy to the VT foci within the recess by creating a deeper lesion.

Conclusions

Arrhythmia can be a critical factor responsible for early morbidity and mortality after cardiac surgery. To our knowledge, we reported the first case of RF ablation from within a plicated right ventricle for early postoperative sustained VT following CR. RF ablation was highly effective, even during the very early period of valvuloplasty and ventriculoplasty for EA.

References

1. Dearani JA, Danielson GK. Congenital Heart Surgery Nomenclature and Database Project: Ebstein's anomaly and tricuspid valve disease. Ann Thorac Surg 2000;69:S106–S117.
2. Chauvaud SM, Brancaccio G, Carpentier AF. Cardiac arrhythmia in patients undergoing surgical repair of Ebstein's anomaly. Ann Thorac Surg 2001; 71:1547–1552.
3. da Silva JP, Baumgratz JF, da Fonseca L, et al. The cone reconstruction of the tricuspid valve in Ebstein's anomaly. The operation: early and midterm results. J Thorac Cardiovasc Surg 2007;133:215–223.
4. Dearani JA, Said SM, O'Leary PW, Burkhart HM, Barnes RD, Cetta F. Anatomic repair of Ebstein's malformation: lessons learned with cone reconstruction. Ann Thorac Surg 2013;95:220–226. discussion 226–228.
5. Holst KA, Dearani JA, Said S, et al. Improving results of surgery for Ebstein anomaly: where are we after 235 cone repairs? Ann Thorac Surg 2018; 105:160–168.
6. Anderson HN, Dearani JA, Said SM, et al. Cone reconstruction in children with Ebstein anomaly: the Mayo Clinic experience. Congenit Heart Dis 2014; 9:266–271.
7. Silversides CK, Kiess M, Beauchesne L, et al. Canadian Cardiovascular Society 2009 Consensus Conference on the management of adults with congenital heart disease: outflow tract obstruction, coarctation of the aorta, tetralogy of Fallot, Ebstein anomaly and Marfan's syndrome. Can J Cardiol 2010;26:e80–e97.
8. Geerdink LM, Kapusta L. Dealing with Ebstein's anomaly. Cardiol Young 2014; 24:191–200.
9. Sherwin ED, Abrams DJ. Ebstein anomaly. Card Electrophysiol Clin 2017; 9:245–254.
10. Paranon S, Acar P. Ebstein's anomaly of the tricuspid valve: from fetus to adult: congenital heart disease. Heart 2008;94:237–243.
11. Oh JK, Holmes DR Jr, Hayes DL, Porter CB, Danielson GK. Cardiac arrhythmias in patients with surgical repair of Ebstein's anomaly. J Am Coll Cardiol 1985; 6:1351–1357.
12. Wackel P, Cannon B, Dearani J, et al. Arrhythmia after cone repair for Ebstein anomaly: the Mayo Clinic experience in 143 young patients. Congenit Heart Dis 2018;13:26–30.
13. Shivapour JK, Sherwin ED, Alexander ME, et al. Utility of preoperative electrophysiologic studies in patients with Ebstein's anomaly undergoing the Cone procedure. Heart Rhythm 2014;11:182–186.
14. Moore JP, Shannon KM, Gallotti RG, et al. Catheter ablation of ventricular arrhythmia for Ebstein's anomaly in unoperated and post-surgical patients. JACC Clin Electrophysiol 2018;4:1300–1307.