

# Cavotricuspid isthmus ablation using multimodality imaging in Ebstein anomaly with a mechanical tricuspid valve replacement

Sang Hyun Lee, MD,\* Hyung Oh Choi, MD,<sup>†</sup> Ki Won Hwang, MD, FHRS\*

From the \*Division of Cardiology, Department of Internal Medicine, Pusan National University Yangsan Hospital, Pusan National University of Medicine, Yangsan, Republic of Korea, and <sup>†</sup>Division of Cardiology, Department of Internal Medicine, Soonchunhyang University Hospital, Soonchunhyang University College of Medicine, Bucheon, Republic of Korea.

## Introduction

Ebstein anomaly (EA) is a congenital malformation of the tricuspid valve (TV) and is characterized by an atrialized right ventricle with apical displacement of the septal and posterior leaflets of the TV.<sup>1</sup> The TV may be replaced in some patients because of unfavorable anatomy or failure of a previous TV repair.<sup>2,3</sup> After standard repair or prosthetic valve replacement in patients with EA, cavotricuspid isthmus (CTI)-dependent atrial flutter (AFL) could develop owing to scarring/fibrosis, postoperative scar, and slow conduction, which is associated with tricuspid regurgitation or concomitant malformation.<sup>4,5</sup> However, CTI ablation is challenging because of the anomalous anatomy of this area and the possibility of prosthetic valve-related risks if the procedure is performed using conventional fluoroscopic guidance.<sup>6</sup> Here, we report the successful ablation of typical CTI-dependent AFL using a 3-dimensional mapping system and intracardiac echocardiography (ICE) in a patient with a mechanical TV replacement (TVR).

### **Case report**

A 26-year-old man with EA had a history of an initial prosthetic TVR in 2005 (#33 Hancock porcine valve, Medtronic, Minneapolis, MN) and redo TVR in 2009 (#31 Regent valve, St. Jude Medical, St. Paul, MN) owing to valve dysfunction. His electrocardiogram showed incessant AFL after redo TVR. AFL recurred despite biphasic direct current cardioversion with concomitant administration of an antiarrhythmic

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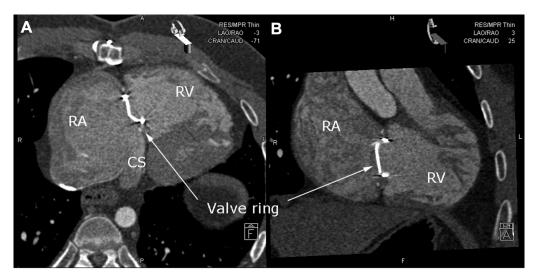
## **KEY TEACHING POINTS**

- Cavotricuspid isthmus-dependent atrial flutter occurs in patients with Ebstein anomaly after a tricuspid valve replacement.
- There are anatomical obstacles that interrupt the creation of a transmural lesion in patients with Ebstein anomaly after a mechanical valve replacement when delivering radiofrequency energy using conventional fluoroscopy.
- Multimodality imaging before and during the procedure may result in a high success rate with a low recurrence and low complication rate.

agent. Thus, he was referred to our hospital for radiofrequency catheter ablation.

Multislice contrast-enhanced computed tomography (MDCT) was performed before the procedure to identify the anatomical relationship between the prosthetic valve and the true atrioventricular (AV) annulus (Figure 1). The coronary sinus (CS) ostium was situated on the atrial side of the prosthetic valve ring with a dilated CS draining into the right ventricle. Electroanatomic mapping of the right atrium (RA) was performed using a PentaRay catheter (Biosense Webster, Diamond Bar, CA) and a 3.5-mm irrigated-tip ablation catheter (ThermoCool, Biosense Webster) via a nonsteerable long sheath (SR0) under the guidance of a 3-dimensional mapping system (CARTO 3, Biosense Webster). The flutter waves showed a negative deflection in the precordial lead V1 and a positive deflection in the inferior leads. The tachycardia cycle length of AFL was 260 ms. During activation mapping, a clockwise activation pattern around the tricuspid annulus was seen (Figure 2). CTI-dependent AFL was confirmed using entrainment

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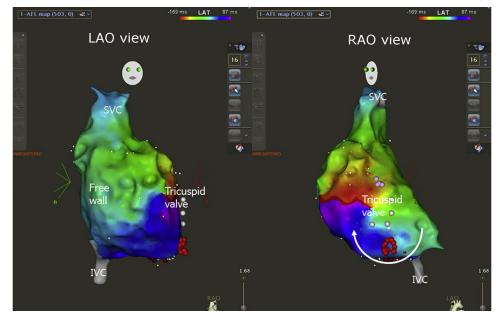
**Figure 1** Multislice contrast-enhanced computed tomography scan. A: The coronary sinus drains into the right atrium under the ring of the mechanical valve (*white arrow*). B: The right oblique view of the computed tomography scan revealed that the ring of the mechanical valve is located on the true atrioventricular annulus. CS = coronary sinus; RA = right atrium; RV = right ventricle.

maneuvers. An ICE-guided ablation catheter (ACUSON AcuNav, Siemens, Erlangen, Germany) was positioned at the ventricular end of the CTI below the mechanical TV (Figure 3). Radiofrequency energy was applied using a point-by-point approach. Power was titrated up to 40 W with a temperature limit set at 35°C. A series of ablation lesions (103 radiofrequency applications; a total duration of 4487 seconds) were delivered along the CTI. Despite termination of AFL, a CTI block could not be achieved, necessitating the delivery of further lesions on the ventricular side of the CTI. A bidirectional block of the CTI was confirmed using pacing maneuvers.

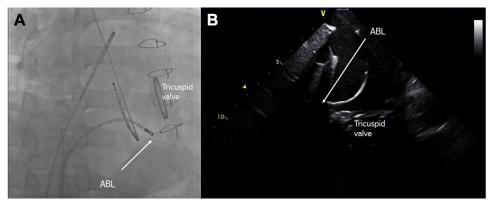
One year after the procedure, the patient remains free from atrial tachyarrhythmias without the use of any antiarrhythmic agents.

#### Discussion

CTI ablation is commonly performed using 3-dimensional electroanatomic mapping system in most centers, although for cost efficiency there are centers using only fluoroscopy. However, CTI ablation has rarely been reported in patients with EA after mechanical TVR.<sup>7</sup> Several obstacles prevent successful CTI ablation in patients with EA after prosthetic TVR.



**Figure 2** A 3-dimensional geometry was created using a PentaRay catheter. An activation map of 503 points was acquired. The activation map of atrial flutter showing the clockwise activation pattern around the tricuspid annulus in the right anterior oblique (LAO) and left anterior oblique (RAO) views. IVC = inferior vena cava; SVC = superior vena cava.



**Figure 3** A: The fluoroscopic aspect in the right oblique view during radiofrequency catheter ablation. B: Intracardiac echocardiography showing good contact of the ablation catheter (*white arrow*) at the ventricular end of the cavotricuspid isthmus. ABL = ablation.

When TVR is performed, the valve suture line depends on the distance between the CS and the AV node.<sup>1,3</sup> To avoid injury to the AV node, the prosthetic valve is generally placed on the atrial side of the AV node and the membranous septum. When the distance between the CS and the AV node is sufficient, the CS is left to drain into the RA. The sewing ring of the valve can obscure the isthmus, and alternative techniques need to be used to achieve a block across the CTI.<sup>8</sup> MDCT is a useful modality that plans an ablation procedure in the EA or another congenital heart disease. The location of the CS ostium is important in that it provides information on the relationship between the prosthetic valve and the true AV annulus. If the CS is located on the ventricular side in patients with EA, part of the CTI is located on the ventricular side of the valve ring in EA. Right ventricular access may become impossible in the presence of a mechanical valve. Previous cases have reported that the anatomical relationship between the artificial valve and the CS ostium was the landmark that determined whether it should be approached from either the atrial or the ventricular aspect for the successful catheter ablation of the CTI.<sup>9</sup> In our case, the CS drained into the RA. It is likely to be situated at the ventricular side on the level of the true AV annulus.<sup>1</sup> A successful bidirectional block of the CTI was obtained with ablation performed only at the atrial aspect.

Radiofrequency catheter ablation using conventional fluoroscopic guidance is difficult and time-consuming in a patient with a prosthetic valve.<sup>6,7,10</sup> Previous cases have reported anatomical obstacles such as a pouch or an area of protected atrial myocardium associated with TVR.<sup>11,12</sup> It is difficult to create a continuous and transmural lesion with conventional techniques. The 3-dimensional mapping system for ablation helps to reduce the risk of procedurerelated complications and increase the success rate.<sup>13</sup> Electroanatomic mapping combined with ICE offers accurate information on the anatomical relationship of the CTI and the precise distance between the ablation catheter and the mechanical TV.<sup>9</sup> When delivering the radiofrequency energy close to the prosthetic valve, it presents real-time imaging for close monitoring of catheter contact and stability during the ablation procedure.

#### Conclusion

This case demonstrates a successful ablation of CTI-dependent AFL in a patient with EA undergoing mechanical TVR. Preemptive MDCT is useful to identify the accurate anatomical relationship before the procedure, and ICE served as a tool for monitoring catheter contact and stability, thereby minimizing the risk of prosthetic valvular damage. These newer technologies including multi-modality imaging help effectively plan the ablation procedure and increase the success rates and decrease the procedure-related complication rates in patients with EA.

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