Mapping of accessory pathways in pediatric patients with Ebstein anomaly using open-window mapping

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Introduction

In patients with Ebstein anomaly, accessory pathways (APs) are common and are predominantly right-sided.[1,2] Mapping and ablation of APs in these patients can be challenging owing to altered anatomy and abnormal fractionated local electrograms. In addition, APs in patients with Ebstein anomaly are located in the true atrioventricular groove, which can be distorted by the massive atrial dilation. The combination of fractionated electrograms, atrial distortion, and an abnormal tricuspid valve can make ablation of APs in these patients challenging, resulting in a lower acute and long-term ablation success rate. The open-window mapping (OWM) technique allows rapid high-density point collection, which can help visualize conduction through APs. It is particularly helpful in patients with structurally abnormal hearts because it does not rely on identifying the origin of local electrograms. In this case series, we present 3 cases of pediatric patients with Ebstein anomaly who underwent mapping and ablation of APs using OWM.

KEY TEACHING POINTS

- Mapping accessory pathways (APs) in patients with Ebstein anomaly can be challenging owing to abnormal local electrograms in the atrialized right ventricle and difficulty localizing the true anatomic right atrioventricular annulus.
- The open-window mapping (OWM) technique does not rely on identifying the origin of local electrograms. Therefore, it is particularly helpful in patients with structurally abnormal hearts.
- High-density mapping using the OWM technique is effective at localizing APs in patients with Ebstein anomaly.

Case report

Case 1

A 5-year-old male child with anatomically severe Ebstein anomaly and cyanotic heart disease was referred for surgical repair. His medical history was notable for supraventricular tachycardia (SVT), which was medically managed with atenolol. He also had evidence of manifest preexcitation consistent with antegrade conduction through a right posterolateral AP (Figure 1A). High-density 3-dimensional mapping was performed during sinus rhythm using the Advisor HD Grid mapping catheter (Abbott Laboratories, Abbott Park, IL) and the EnSite X electroanatomic mapping system (Abbott Laboratories). We localized the pathway within the right posterolateral tricuspid annulus using OWM (Figure 1B). Ablation was performed with the TactiCath DF, Sensor Enabled catheter (Abbott Laboratories) at the site of pathway breakout, which successfully eliminated the pathway conduction (Figure 1C).

Postablation, there was still evidence of manifest preexcitation, which became more pronounced after adenosine administration. This was consistent with antegrade conduction through a second AP. Mapping of the AP was done as described above. We localized the second pathway within the right posteroseptal tricuspid annulus (Figure 1D). Ablation in this area successfully eliminated the pathway conduction (Figure 1E). In follow-up, the patient has remained free of SVT with no evidence of preexcitation on electrocardiograms (ECGs) (Figure 1F).

Case 2

A 5-year-old female child presented with symptomatic SVT refractory to medical therapy. Her medical history was significant for anatomically and functionally severe Ebstein anomaly diagnosed shortly after birth. As she got older, she became more symptomatic during SVT, with episodes of desaturation. A decision was then made to consider surgical repair and perform an electrophysiology study (EPS) and ablation before surgery. Her baseline ECG showed no

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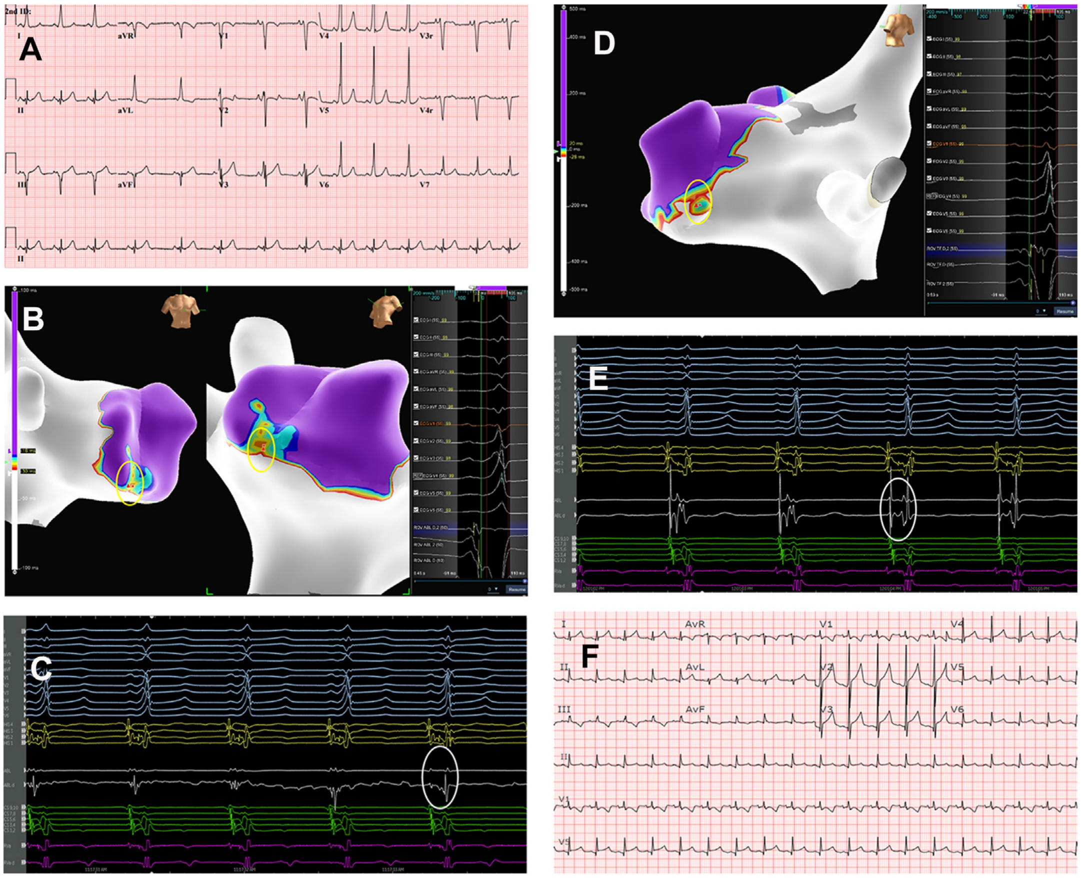

Figure 1 **A:** Baseline electrocardiogram showing manifest preexcitation. **B:** Local activation map was created in preexcited sinus rhythm using open-window mapping localizing an accessory pathway (AP) to the posterolateral tricuspid annulus (*yellow circle*). The intracardiac electrogram is shown to the right of the respective map. Because of the atrial distortion and enlargement, the true annulus position is displaced apically. **C:** Local electrograms at the site of successful AP ablation. **D:** HD Grid (Abbott Laboratories, Abbott Park, IL) local activation map was created in preexcited sinus rhythm localizing an AP to the posteroseptal tricuspid annulus (*yellow circle*). The intracardiac electrogram is shown to the right of the respective map. **E:** Local electrograms at the site of successful AP ablation. **F:** Postablation electrocardiogram.

preexcitation (Figure 2A), while an ECG during tachycardia revealed a narrow complex mid-RP tachycardia (Figure 2B).

EPS was diagnostic of an AP with retrograde conduction only. With atrial pacing, orthodromic reciprocating tachycardia using a right-sided AP was induced. A 4-mm-tip Safire radiofrequency ablation catheter (Abbott Laboratories) was then used for mapping. Using the EnSite X 3-dimensional mapping system (Abbott Laboratories) and an OWM technique during ventricular pacing, the AP was mapped to the right posterior tricuspid annulus (Figure 2C). Ablation was performed at this location during ventricular pacing, which resulted in loss of AP conduction (Figure 2D). Postablation, there was no evidence of AP conduction or inducible tachycardia.

Case 3

A 5-year-old male child with Ebstein anomaly and SVT was referred for tricuspid valve repair and closure of an atrial septal defect. He had undergone a bidirectional Glenn procedure prior to presentation. A diagnostic EPS was performed prior to consideration for surgery. On baseline ECG, there was no evidence of preexcitation (Figure 3A). EPS was diagnostic of a right posterolateral AP with retrograde conduction only. Orthodromic reciprocating tachycardia was easily inducible during the EPS. Using a 4-mm-tip Safire radiofrequency ablation catheter (Abbott Laboratories), sequential mapping of the basal right ventricle and right atrium was done during tachycardia using OWM. The pathway was localized at the right posteroseptal tricuspid annulus (Figure 3B). Ablation at this location resulted in successful pathway elimination (Figure 3C). Postablation, there was no evidence of AP conduction or inducible tachycardia.

Discussion

Ebstein anomaly is a rare congenital heart malformation with a prevalence of 1 in 20,000 live births and <1% in patients with adult congenital heart disease.[1] It is characterized by failure of delamination and apical displacement of the septal and posterior tricuspid valve leaflets. There is associated redundancy and tethering of the anterior tricuspid valve leaflet. This results in atrialization and thinning of the right ventricle, tricuspid regurgitation, and right atrial enlargement.[1] Patients may present with cyanosis from associated

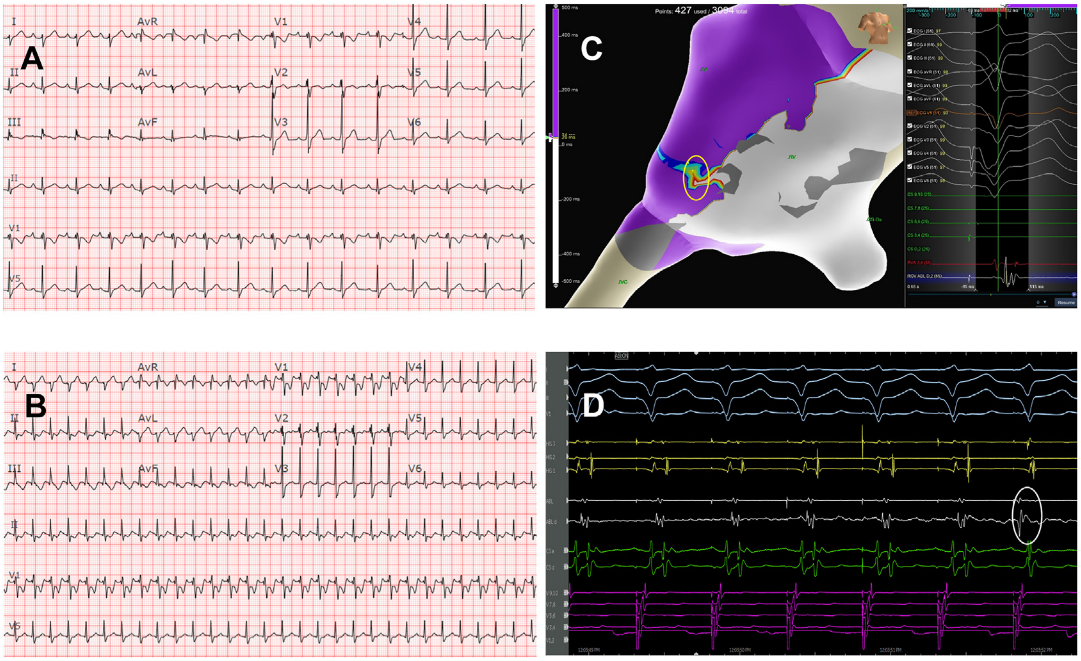

Figure 2 **A:** Baseline electrocardiogram with no evidence of preexcitation. **B:** Electrocardiogram during tachycardia showing mid-RP narrow complex tachycardia. **C:** Local activation map was created during ventricular pacing rhythm using open-window mapping localizing an accessory pathway (AP) to the posterolateral tricuspid annulus (*yellow circle*). The intracardiac electrogram is shown to the right of the respective map. **D:** Local electrograms at the site of successful AP ablation. Note the relative distance between the atrial and ventricular signal marking slower conduction in an Ebstein AP compared to a traditional AP.

atrial septal defect and right-to-left shunting, right heart failure, arrhythmias, and sudden cardiac death.[1]

Both supraventricular and ventricular tachycardias have been described in patients with Ebstein anomaly. While focal atrial tachycardia, atrial flutter, and atrial fibrillation are commonly seen in postsurgical patients with Ebstein anomaly, AP-mediated tachycardias predominate in the unrepaired cohort of patients.[1,2] APs are reported to occur in 10%–30% of patients with Ebstein anomaly and are most commonly located on the deformed tricuspid annulus.[2] This is thought to be owing to disruption of the annulus fibrosis, which can lead to AP formation. Patients can have more than 1 AP, and in addition to atrioventricular pathways, atriofascicular, nodofascicular, and fasciculoventricular tracts have also been reported.[2]

AP mapping and ablation in patients with Ebstein anomaly can pose several challenges. Atrialization of the right ventricle creates areas within the atrium that carry the electrical and morphologic properties of the ventricle.[1] This, along with tricuspid valve displacement, leads to areas of delayed conduction and can result in fractionated and low-amplitude electrograms. This may result in difficulty identifying and isolating the AP potentials. Moreover, the distortion of the tricuspid annulus that results from tricuspid leaflet displacement can make it problematic to distinguish the anatomic annulus from the functional annulus.[2] Previous mapping of these pathways could be aided by placing a small electrode wire directly in the right coronary artery (that marks the location of the true annulus) to localize the pathway. The catheters used for this purpose are no longer being manufactured. Therefore, it may be necessary to use a right coronary artery angiogram to identify the true annulus, but this involves additional time and risk and only shows the tricuspid valve position and not the location of the AP(s).[3] Intraprocedural intracardiac echocardiography imaging can be useful to localize the true annulus and facilitate mapping and ablation of APs in patients with Ebstein anomaly.[4,5]

In addition to the challenges faced with identifying the true annulus, catheter stability can often be an issue owing to the presence of atrioventricular ridges, hypermobility and dilatation of the tricuspid annulus, and tricuspid regurgitation. The unusual shape of the tricuspid valve annulus in Ebstein anomaly makes performing a complete activation sequence with point-by-point mapping difficult, and the APs are sometimes slanted or have areas of slow conduction. Finally, the presence of multiple pathways and pathways with multiple broad insertion sites can create a barrier to successful ablation.[5] It is because of these limitations that AP ablation in Ebstein anomaly carries a lower acute success rate (∼88%) and a higher recurrence rate (19%) when compared to patients with other congenital heart disease or structurally normal hearts.[6]

OWM is a mapping technique that has been used in AP ablation. It differs from conventional mapping strategies, which rely on point-by-point mapping of the earliest atrial and ventricular signals, or direct identification of pathway potentials. These strategies often require careful analysis of acquired signals by an experienced operator, which can lead to longer mapping times. In addition, errors in pathway localization may occur, especially when insufficient points are

Figure 3 **A:** Baseline electrocardiogram with no evidence of preexcitation. **B:** Local activation map was created during tachycardia using open-window mapping localizing an accessory pathway (AP) to the posteroseptal tricuspid annulus (*yellow circle*). The intracardiac electrogram is shown to the right of the respective map. **C:** Local electrograms at the site of successful AP ablation.

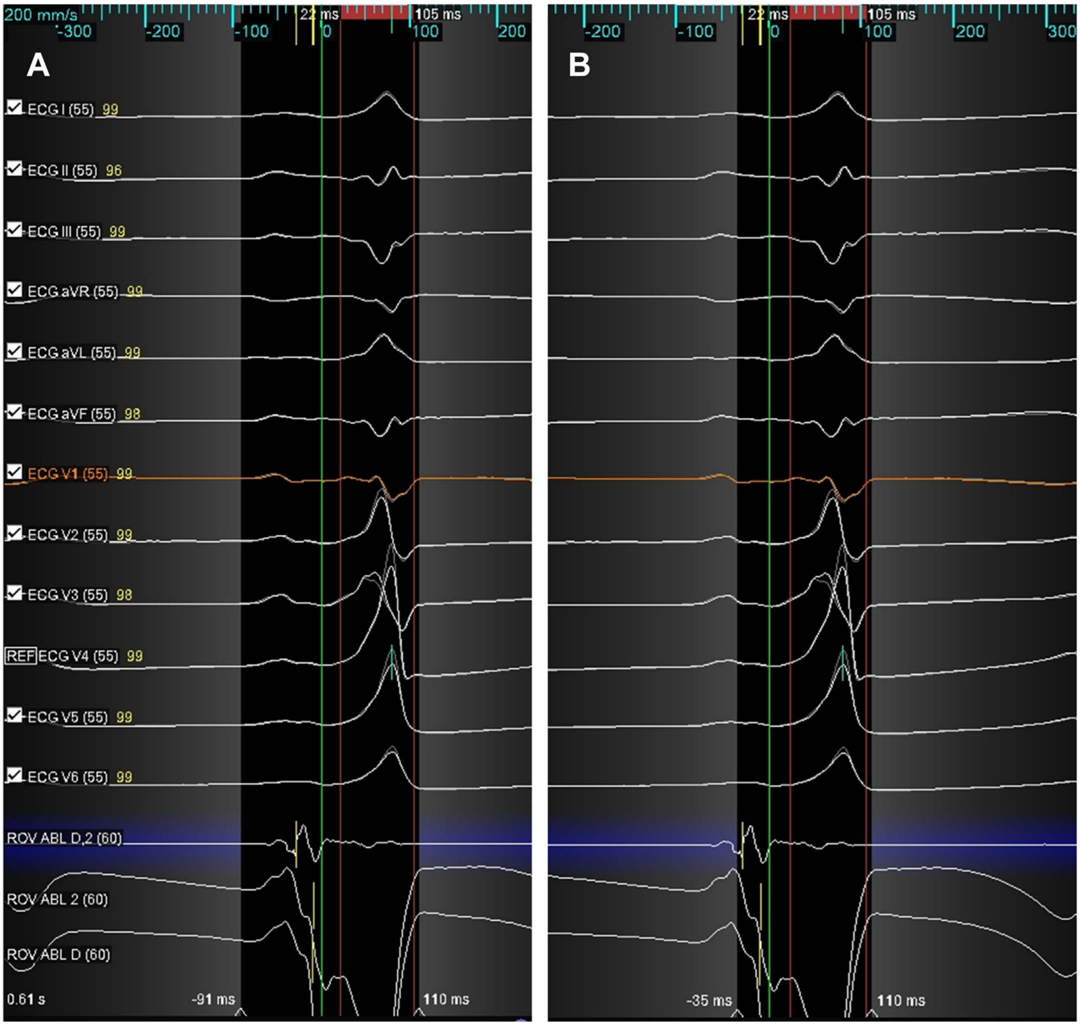

Figure 4 Changes applied to the window of interest (WOI) when performing open-window mapping **(A)** as compared to traditional mapping of the ventricular insertion site of the accessory pathway in sinus rhythm **(B).** In open-window mapping, the WOI is expanded to include any atrial and ventricular signals.

available or points are incorrectly annotated to the wrong cardiac chamber. OWM is a high-density, automated mapping tool that relies not on the origin of the signal, but rather on the sharpest signal at each point. It involves expanding the acquisition window to fully include any atrial and ventricular signals (Figure 4). The mapping system then automatically annotates the sharpest local signal with the maximum rate of voltage change (dV/dt), disregarding whether it originates from the atrium, ventricle, or AP. This allows for the visualization of activation patterns as well as potential breakout points, defined as the earliest sharp signal in the chamber where activation exits from the pathway, or pathway insertion site, which is crucial for a successful ablation.[7–10] OWM is 100% effective in identifying pathway locations with accuracy in structurally normal hearts.[7]

To our knowledge, this is the first report of OWM for mapping APs in pediatric patients with Ebstein anomaly, in whom conventional mapping is challenging owing to altered anatomy, abnormal local signals, and the potential for multiple APs. OWM's ability to capture both atrial and ventricular signals simultaneously, and its focus on maximum dV/dt for annotation without requiring specific annotation of signal origins, can be advantageous in these patients. Using the Grid catheter, it is possible to quickly and accurately map activation sequences to determine the best location for targeting an ablation. It is also possible to potentially see multiple APs or pathways with unusual conducting properties in real time, which may reduce the time needed for a successful ablation as well as increase the success rate for patients with Ebstein or other anatomic abnormalities that distort the valve anatomy.

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