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Case Report

SUCCESSFUL ABLATION OF CONCEALED ACCESSORY PATHWAY MEDIATED SUPRAVENTRICULAR TACHYCARDIA IN A 5-YEAR-OLD CHILD WITH EBSTEIN ANOMALY

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SUMMARY – Here we report a case of a 5-year-old boy with Ebstein anomaly and supraventricular tachycardia. He was diagnosed with Ebstein anomaly at the age of 3 months during the workup for afebrile convulsions, and has been followed by a pediatric cardiologist since. Electrocardiography recorded a small Rr` pattern in V1 on a few occasions, without other abnormalities. At the age of 2.5 years, the first episode of supraventricular tachycardia was recorded and stopped with adenosine. He was started on atenolol and was asymptomatic until a year later when he presented with the second recorded supraventricular tachycardia. He was referred to our institution for an electrophysiology study with ablation. Tachycardia was entrained and all the maneuvers were in accordance with atrioventricular reentry tachycardia. Tricuspid annulus was mapped during tachycardia. The earliest atrial signals appeared in the posteroseptal region of the valve, mechanical blocking of the tachycardia was recorded in that region, and early fractionated signal was present during ventricular retrograde pacing. The pathway was successfully ablated. Basal electrocardiogram was without change after ablation and the patient did not experience recurrence of tachycardia during follow-up. This case shows the efficacy and importance of ablation in Ebstein anomaly, without x-ray or intracardiac echocardiography.

Key words: *Ebstein anomaly; Tachycardia, supraventricular; Electrophysiology*

Introduction

Ebstein anomaly[1] is a congenital deformity of the tricuspid valve, in which the valve orifice is displaced apically to the right ventricle with subsequent 'atrialization' of the ventricle. Depending on the portion of the ventricle affected, symptoms vary from non-existent to serious and can appear even in the neonatal age[2] with right heart failure and cyanosis. Another burden that these patients carry are arrhythmias, and most of them are attributable to accessory pathways, frequently multiple, on the right side of the heart[3,4]. Except for supraventricular tachycardia, accessory pathways can be responsible for sudden cardiac death due to conducting atrial fibrillation at a fast rate to ventricles and provoking ventricular fibrillation[5-7]. In these cases, Ebstein anomaly could be an independent risk factor for cardiac arrest[8]. This is why electrophysiology study with ablation is recommended to these patients, especially before surgical correction of the anomaly[3,8].

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Case Report

A 5-year-old boy was admitted to our hospital for a planned electrophysiology study with ablation. He was born from an unremarkable pregnancy in the 40th week of gestation, with birth weight 3800 g and birth length 56 cm. His first contact with a pediatric cardiologist was at the age of 3 months when he was admitted to another institution due to afebrile convulsions. The electrocardiogram (ECG) showed a rightward axis without other abnormalities. Echocardiography showed an Ebstein anomaly. Neurological workup was normal except for muscle hypertonia. He was followed-up by a pediatric neurologist and physiatrist with a good clinical outcome.

Regular echocardiography checkups did not show any progression of structural changes or hemodynamic consequences of the anomaly. Preexcitation or classical right bundle branch block (RBBB) were never recorded on the ECG, while a small Rr` pattern with right ventricular conduction delay in V1 was noted on a few occasions. At the age of 2.5 years, he visited the endocrinology outpatient clinic because of suspected undescended testicles. Heart rate of 225/min was measured and the ECG showed supraventricular tachycardia. He was administered adenosine and tachycardia stopped. The patient was started on atenolol and did not complain of any symptoms until a year later when he presented with a second recorded supraventricular tachycardia, which was terminated again with adenosine and amiodarone.

Three months later, he was admitted to our hospital for electrophysiology study and ablation. One week before the procedure, atenolol was discontinued. One day before the procedure, ECG was normal except for rightward axis and mild right ventricular conduction delay seen in V1 (Fig. 1). Electrophysiology procedure was done with the EnSite Precision™ Cardiac Mapping System (Abbott, Illinois, USA). Diagnostic electro catheters were placed through the right femoral and right jugular vein into the area of His, the right ventricle and the coronary sinus.

Basic measurements of the conduction system were normal (AH 76 ms, HV 44 ms, PQ 144 ms, QT 326 ms). Tachycardia started after a mechanically induced supraventricular extrasystole and later by programmed stimulation of the atria. During anterograde conduction, there was no evidence for an accessory pathway, but retrograde conduction was eccentric, earliest atrial signal was on electrodes CS 7-8 (at the ostium of coronary sinus), before the His electrode. Tachycardia was entrained and all the maneuvers were in accordance with atrioventricular reentry tachycardia (VAV response, difference between the postpacing interval and tachycardia cycle length was 100 ms, difference between the stimulus to atrium and ventricle to atrium was 66 ms). His synchronous premature ventricular complex delayed atrial activation. A long steerable introducer and non-irrigated ablation catheter were inserted.

Tricuspid annulus was mapped during tachycardia. The earliest atrial signals were present in the posteroseptal region of the annulus and mechanical block of the tachycardia was recorded in that region on two occasions. During retrograde pacing from the ventricle on that spot, an early fractionated signal appeared (Fig. 2). Tachycardia was induced and temperature-controlled radiofrequency energy (45-50 W, 52-60 °C) was applied. After 1 second, the tachycardia was terminated and ablation of the pathway was successful (Figs. 3 and 4). After 30 minutes of waiting, the accessory pathway did not recur and tachycardia could not be induced. ECG showed no change after ablation. At outpatient clinic visits, 2 and 6 months later, the patient was well, without any symptoms, ECG and cardiac ultrasound were unchanged.

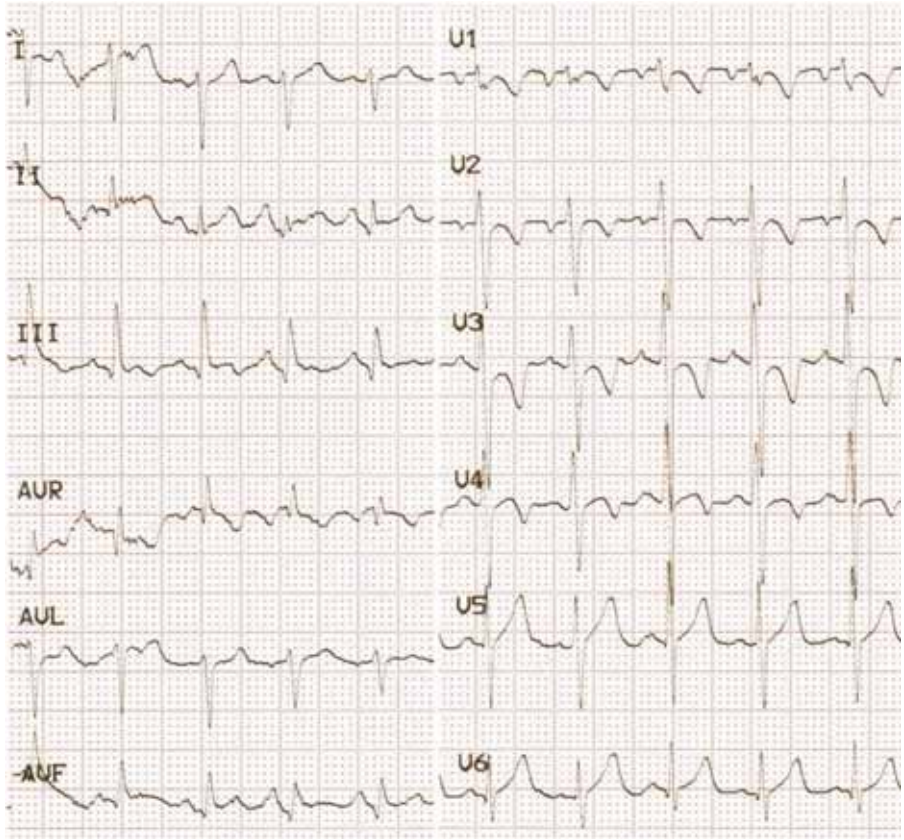

Fig. 1. A 12-lead ECG before electrophysiology procedure: right ventricular conduction delay in V1.

Fig. 2. Intracardiac ECG at the site of successful ablation during ventricular pacing. An early atrial fractionated signal is visible on the ablation catheter.

I, II, III, V1, V6 = standard ECG; ABLd = distal pole of ablation catheter; ABLp = proximal pole of ablation catheter; CS 1-2 to CS 9-10 = catheter in coronary sinus; RVd = distal pole of ventricular catheter

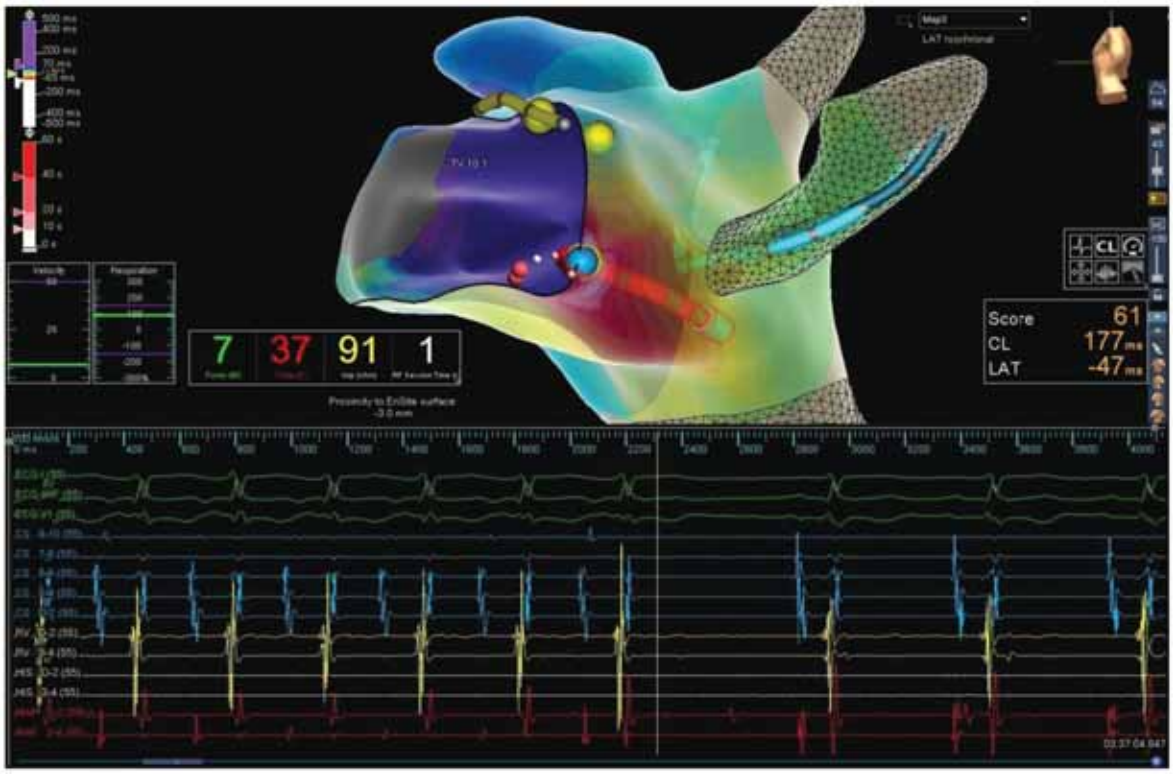

Fig. 3. Position of ablation catheter at the time of successful termination of tachycardia during radiofrequency ablation (EnSite Precision™ Cardiac Mapping System, Abbott).

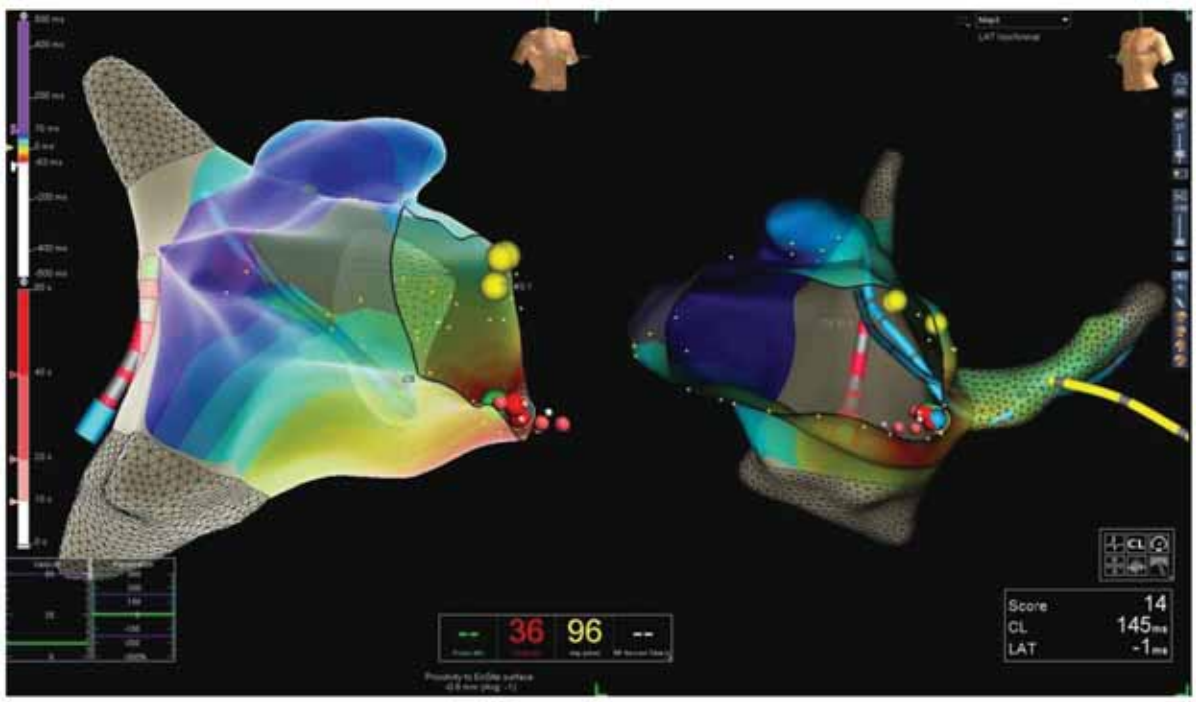

Fig. 4. Right anterior oblique (on the left side) and left anterior oblique (on the right side) view of the successful ablation point – blue dot (EnSite Precision™ Cardiac Mapping System, Abbott).

Discussion

In children, the lack of symptoms does not exclude tachycardia since children are more likely to get used to the sensation, considering it normal and even ignoring it at an older age. Anterogradely conducting pathways and preexcitation in Ebstein anomaly are not obvious in some cases, perhaps because atrial depolarization is too slow to activate the atrial side of the accessory pathways[9], therefore the lack of visible RBBB in Ebstein patients can be a sign of preexcitation. Abnormal development of the tricuspid annulus predisposes to slower activation of the right ventricle through the right bundle showing RBBB in the ECG, but if a right-sided accessory pathway is present, conduction to the right ventricle appears normal on the ECG[10]. In addition, prolonged intra-atrial conduction through the enlarged right atrium combined with early conduction through the accessory pathways may result in a normal or sometimes even prolonged PR interval[10], which must not be considered as the lack of preexcitation in these patients.

Catheter ablation in small children with Ebstein anomaly can be extremely challenging. Defining the target site of accessory pathways in the atrialized right ventricle presents an impediment due to challenging mapping of the dysplastic tricuspid annulus. Nevertheless, in our case, the pathway was ablated, and the ECG did not change proving that this pathway did not have any anterograde conducting abilities. This case shows that even in Ebstein anomaly, ablation can be done safely and successfully without x-ray or intracardiac echocardiography, using 3D electroanatomic mapping. Also, this case is consistent with the hypothesis that not all patients with Ebstein anomaly have typical features of an accessory pathway in their ECG, indicating that regular checkups and ECG monitoring are crucial in their follow-up, along with electrophysiology study performed at the right time.

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Sažetak

USPJEŠNA ABLACIJA SKRIVENOG AKCESORNOG PUTA U DJETETA S EBSTEINOVOM ANOMALIJOM I SUPRAVENTRIKULSKOM TAHIKARDIJOM

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U ovom prikazu opisan je slučaj dječaka u dobi od 5 godina s Ebsteinovom anomalijom i supraventrikulskom tahikardijom. U dobi od 3 mjeseca, tijekom obrade afebrilnih konvulzija, ustanovljena mu je navedena anomalija te se od tada prati kod dječjeg kardiologa. Elektrokardiografski mu je u nekoliko navrata zabilježen uzorak Rr` u V1, bez drugih abnormalnosti. U dobi od 2,5 godine zabilježena je prva epizoda supraventrikulske tahikardije koja je prekinuta adenozinom te je dječak od tada na terapiji atenololom. Nije imao tegoba sve do godinu dana kasnije, kada mu je drugi puta zabilježena supraventrikulska tahikardija, zbog čega je upućen u našu ustanovu na elektrofiziološku studiju s ablacijom. Učinjenim manevrima inducirana je atrioventrikulska kružna tahikardija. Trikuspidni prsten je mapiran tijekom tahikardije. Najraniji signali pojavili su se u posteroseptalnom području trikuspidne valvule, gdje je također zabilježeno mehaničko prekidanje tahikardije, a rani frakcionirani signal bio je prisutan tijekom ventrikulske retrogradne stimulacije. Akcesorni put je uspješno abliran, elektrokardiogram je bio bez promjena nakon zahvata, a tahikardija se tijekom praćenja nije ponavljala. Ovaj slučaj naglašava učinkovitost i važnost ablacije u bolesnika s Ebsteinovom anomalijom, bez korištenja rendgena ili intrakardijalne ehokardiografije.

Ključne riječi: *Ebsteinova anomalija; Supraventrikulska tahikardija; Elektrofiziologija*