RADIOFREQUENCY CURRENT CATHETER ABLATION OF ACCESSORY PATHWAY IN EBSTEIN'S ANOMALY AND A REVIEW OF THE LITERATURE

M. Hakan DİNÇKAL, MD, Vedat DAVUTOĞLU, MD, Serdar SOYDİNÇ, MD, Yusuf SEZEN, MD

Gaziantep Üniversitesi Tıp Fakültesi Kardiyoloji Anabilim Dalı, Gaziantep

SUMMARY

A 16-year-old female with Ebstein's anomaly admitted to the hospital due to paroxysmal supraventricular tachycardia (SVT), which is refractory to propafenone. A surface ECG showed a short PR interval and a right posteroseptal preexcitation confirming the diagnosis of Wolff-Parkinson-White's (WPW) syndrome. Orthodromic-, antidromic atrioventricular reentrant tachycardia (AVRT) and atrial fibrillation were induced by programmed electrical stimulation. Radiofrequency (RF) catheter ablation was performed successfully on posteroseptal site above the septal valve at the tricuspid annulus. The patient has been free from arrhythmias during a follow-up period of 9 months. RF catheter ablation of accessory pathway is a curative therapy with its special difficulties in Ebstein's anomaly. Related literature was reviewed. ***Türk Kardiyol Dern Arş 2003;31:239-243***

Key words: *Ebstein's anomaly, Wolff-Parkinson-White syndrome, radiofrequency catheter ablation*

ÖZET

Ebstein anomalisinde aksesuar yolun radyofrekans kateter ablasyonu ve ilgili literatürün gözden geçirilmesi

Ebstein Anomalili 16 yaşındakadın hasta, propafenon'a cevapsız paroksismal supraventriküler taşikardi (SVT) nedeni ile başvurdu. Yüzey EKG'sinde posteroseptal geçişli Wolff Parkinson White (WPW) paterni görüldü. Elektrofizyolojik çalışmada ortodromik ve antidromik atrioventriküler reentran taşikardiile atriyal fibrilasyon indüklendi. Radyofrekans kateter ablasyonu septal yaprak üzerinde posteroseptal anulusta başarıyla gerçekleştirildi. Dokuz aylık izlemde hastada aritmi gözlenmedi. Ebstein anomalisinde aksesuar yolun radyofrekans kateter ablasyonu kendine has zorluklar içermekle beraber küratif bir tedavidir. Bu konu ile ilgili literatur gözden geçirildi. ***Arch Turk Soc Cardiol 2003;31:239-243***

Anahtar kelimeler: *Ebstein anomalisi, Wolff Parkinson White sendromu, radyofrekans kateter ablasyonu*

Ebstein's anomaly is an uncommon congenital lesion affecting approximately 1% of patients with congenital heart disease[1]. It is characterized by downward displacement of the tricuspid valve with the septal and posterior leaflets of the tricuspid valve adherent to the right ventricular septal wall. Ebstein's anomaly is often associated with supraventricular tachycardia, with an incidence as high as 42%[1]. Wolff-Parkinson-White syndrome has been estimated to affect up to 30% of this patient group, and sudden death has occurred in 20% of older patients and adults[2]. We present a successful radiofrequency catheter ablation of accessory pathway in Ebstein's anomaly.

Yazışma Adresi: Dr. M. Hakan Dinçkal. Gazi Mahallesi Karacaoğlan Cad. No:26/B-1 Şükrü Ercan Apt. Şehitkamil 27090 Gaziantep
Tel: 0342 335 2998 / Faks: 0342 360 1617
E-posta: mhdinckal@gantep.edu.tr
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CASE REPORT

A 16-year-old female presented with one year history of palpitation. She had documented narrow QRS tachycardias, which increased in frequency despite propafenone therapy for one month. She had no past history of cardiac diseases and family history. There was a 2/6 systolic murmur in left sternal localization, otherwise her physical examination was unremarkable. Her electrocardiogram (ECG) was notable for the following: sinus rhythm, PR interval 0,08 sec, Q formation in leads D2, D3, aVF, and maximum delta waves in leads D1, aVL. Negative delta waves in leads D2, D3, aVF and V1, and positive delta waves in aVL and D1 supported right posteroseptal accessory pathway (Figure 1). Chest X-ray showed normal cardiothoracic index with minimal right atrial enlargement. There was no pulmonary congestion findings. The diagnosis of Ebstein's anomaly (EA) was based on the presence of septal tricuspid displacement below the true annulus 1,8 cm into the right ventricle with mild to moderate tricuspid regurgitation in the echocardiography. Because of preexcitation and frequent palpitations, electrophysiologic study (EPS) was performed. In the EPS, orthodromic and antidromic atrioventricular reentrant tachycardia were induced by programmed electrical stimulation. During the burst atrial pacing to overdrive the tachycardia it converted into atrial fibrillation (Figure 2). Afterwards it was terminated spontaneously. Considering the absence of surgery indication for tricuspid valve replacement, ablation therapy was concerned. We had difficulty in keeping the catheter in a stable position placed at the AV annulus rather than the displaced anatomical AV groove and used a long sheath (SAFL; St. Jude Medical, Inc. - Daig Division, Minnetonka; USA) to stabilize it. A maximum energy (50 Watts) has been applied to achieve an effective temperature (55°C) and successful ablation of the accessory pathway (Figure 3). The patient has been free from arrhythmias during a follow-up period of 9 months.

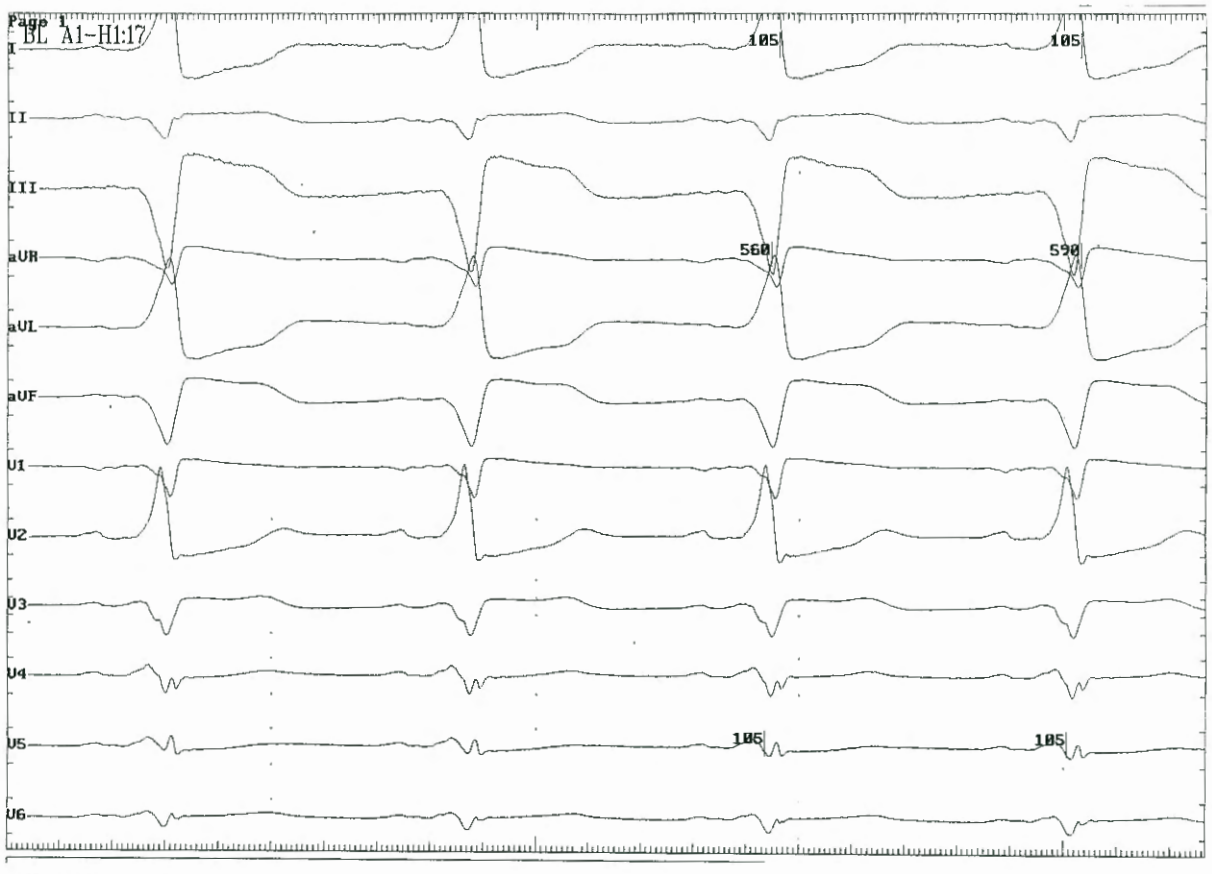

Figure 1: *Surface ECG of the patient. Scale: 100 mm/sec; 1 mV*

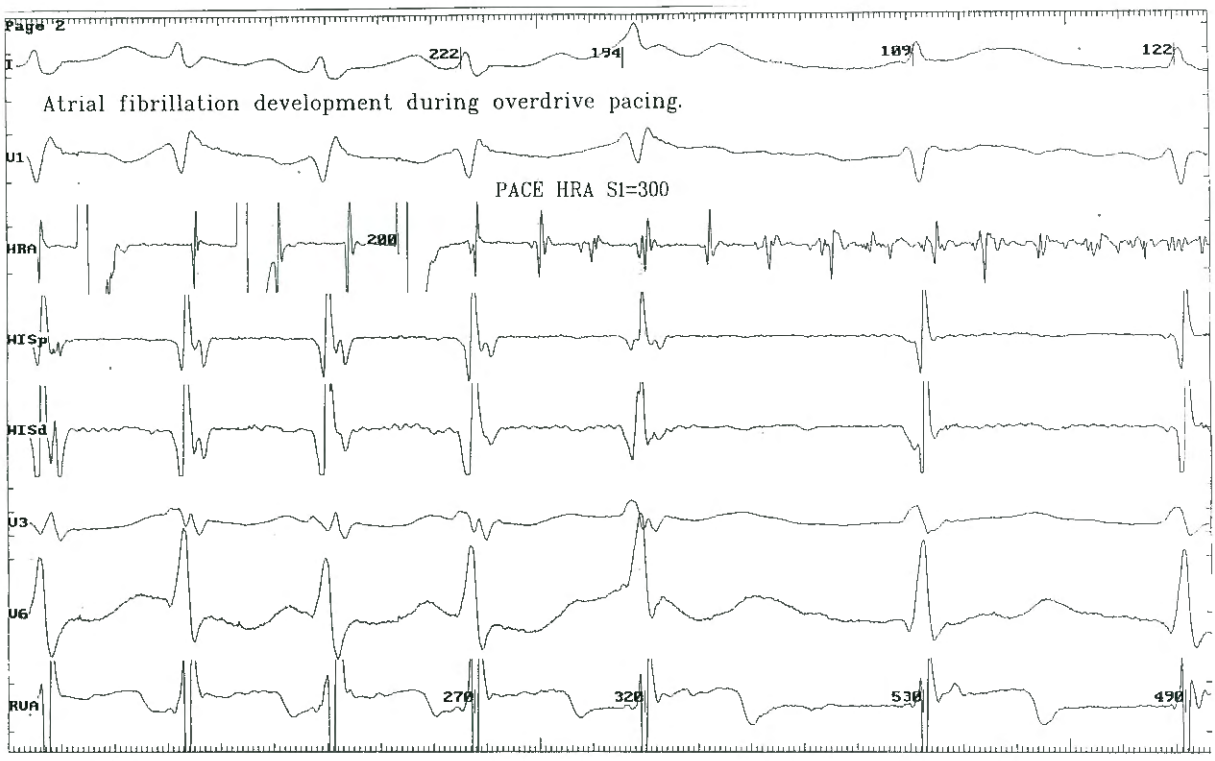

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Figure 2: *Atrial fibrillation development during overdrive pacing of orthodromic atrioventricular reentrant tachycardia. Scale: 100 mm/sec; 1 mV*

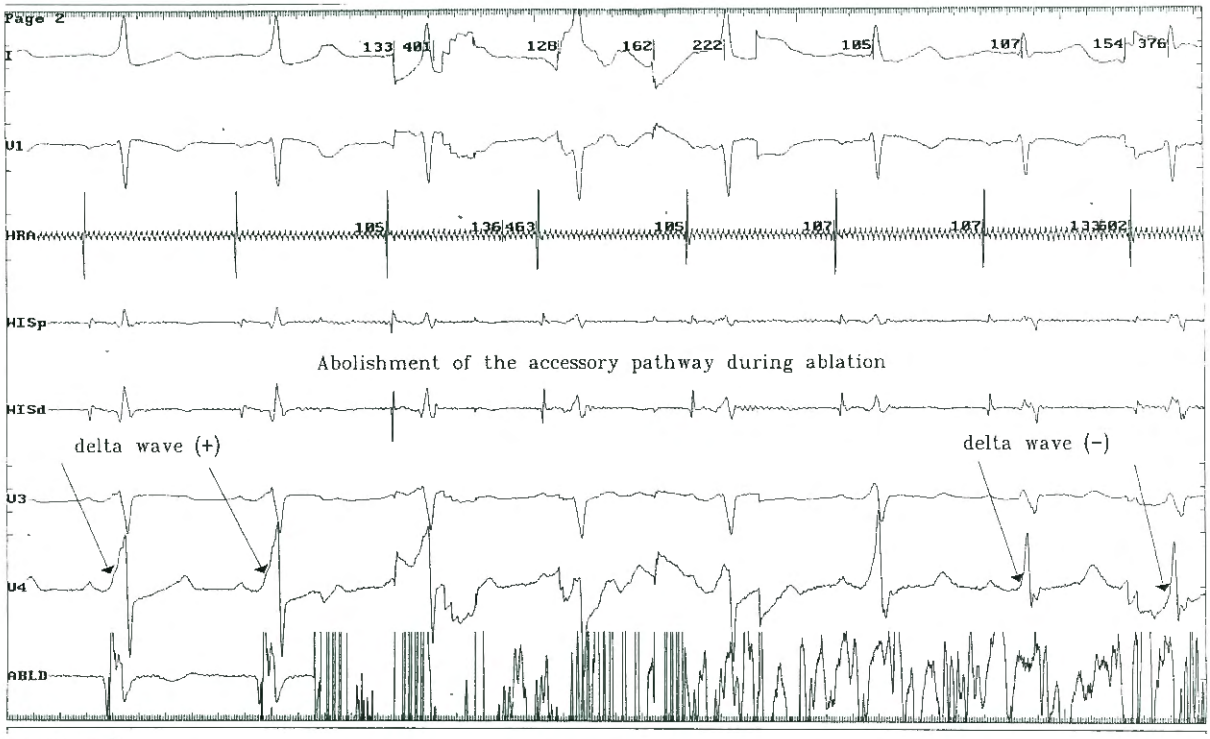

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Figure 3: *Abolishment of accessory pathway during ablation. Scale: 50 mm/sec; 1 mV*

DISCUSSION

Patients with EA are known to have a high potential for developing tachyarrhythmias. Most of these tachycardias are based on accessory pathways located along the anomalous atrioventricular valve, found in up to 30% of this patient group[2]. Next to this main representative for congenital arrhythmogenic substrates, various types of acquired tachycardia were found in patients with EA, such as atrial ectopic tachycardia, atrial flutter, atrial reentrant tachycardia, atrial fibrillation and ventricular arrhythmias. The clinical manifestations of this anomaly are quite variable, depending upon the spectrum of pathology and the presence of associated malformations. It is well documented that a considerable proportion of these patients are able to survive into adult life[2].

There are 2 standard techniques for the definitive treatment of supraventricular tachycardia in this population;[1] radiofrequency catheter ablation (RFA) in electrophysiology laboratory as in our case, and[2] intraoperative cryoablation or direct surgical division of the accessory pathways. Results are, in general, good with an acute RFA success rate of 81% for all ablation targets, although recurrence rate is high. Cryoablation and surgical division with intraoperative mapping have been used with excellent results, although both approaches are time consuming[3,4].

In general, RFA in patients with congenital heart disease and arrhythmia is both safe and effective and may be the preferred approach to the treatment in some patients. Such therapy carries the potential for a definitive treatment and should be taken early into consideration, especially those who are refractory to medication. In patients who are to undergo surgical correction or palliation, preoperative radiofrequency ablation of the tachycardia substrate or the incorporation the ablation procedure into the intracardiac surgical repair is effective and may be preferred to operative accessory pathway division. However, longer procedure and radiation exposure times were necessary to achieve a high success rate in patients with congenital heart disease. In the case of RF catheter ablation for patient with EA, close attention is indispensable in order to accomplish it safely and successfully, because of the anatomical and functional difference peculiar to EA[3]. The location of the atrioventricular node was displaced from the usual position to postero-inferior area of Koch's triangle in 1/5 cases[5]. In patients with EA and reentrant atrioventricular tachycardias, factors likely to account for failure of RF catheter ablation include an accessory pathways located along the atrialized right ventricle and the abnormal morphology of endocardial activation potentials generated in this region[6]. In patients with EA, accessory pathways are generally right-sided. The ablation of the accessory pathway in the situation with Ebstein's anomaly can be quite challenging. The presence of significant tricuspid regurgitation and the downward displacement of the tricuspid valve leaflets may make stable catheter position difficult on the right AV groove. However, the single most difficult factor in such patients is the difficulty in achieving an adequate temperature at the catheter tip despite maximum voltage. This is most likely due to the large right atrium and atrialized right ventricle. Regarding the tendency to atrial fibrillation and multiple accessory pathways in patients with EA, one must be prepared to try a variety of catheter approaches, both from the inferior vena cava as well as from the internal jugular or right subclavian vein, and one might consider the use of long venous sheaths to allow for better catheter stability. A 4 French mapping catheter introduced directly into the right coronary artery around the AV groove may help the fluoroscopic identification of the right AV groove, and allows precise mapping of signals at the AV groove. The use of temperature monitoring and/or temperature control is mandatory in patients with Ebstein's anomaly to differentiate between lack of success due to incorrect catheter position versus inadequate temperature.

Operation for WPW syndrome may become indicated for RF ablation failure, when additional procedures are required[7]. Surgical therapy is a conservative alternative to a lifetime of medical therapy in young, otherwise healthy patients with the WPW syndrome. Repairing EA without correction of associated arrhythmia may result in sudden death. Catheter or surgical ablation is indicated for various symptomatic tachyarrhythmias in EA[8]. Detailed preoperative electrophysiological evaluation in patients with EA is mandatory. Aggressive surgical intervention of the associated arrhythmias in addition to anatomic correction can reduce the sudden death in EA[8]. The combined approach of tricuspid valve repair and surgical ablation of accessory atrioventricular connections has been proved to be safe and effective[9]. It should be

kept in mind, that the absence of manifestations of RBBB (usual in patients with EA without preexcitation) in the presence of EA diagnosed by hemodynamic study or echocardiography should let us think of the coexistence of the preexcitation. Thus preoperative EPS should be concerned before surgery in order to identify the accessory pathway .

In conclusion, if any major indication for operation was absent, for instance patient becomes symptomatic either because of paradoxical embolism or because of worsening of the tricuspid regurgitation, radiofrequency ablation of WPW in patients with EA should be performed. If this is unsuccessful or there is operative indication with coexistence of the WPW syndrome, surgical interruption of accessory pathway or surgical radiofrequency ablation can be used safely and effectively and seems to be useful with the guidance of preoperative EPS.

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