

With an advancement of imaging modalities, MRI has been useful to evaluate the end organs involvement along with cANCA, ESR, and CRP, although tissue biopsy remains the gold standard (10). In this case, the presence of cANCA along with normal eosinophil count and positive renal biopsy confirmed GPA. Cyclophosphamide therapy in GPA could lead to a DCM, but in our case, congestive cardiomyopathy was seemingly due to GPA as he was not taking any medicines (7). Endomyocardial biopsy or cardiac MRI with contrast was not performed given biopsy-positive GPA, positive inflammatory markers, and impaired renal function, but nonetheless, it could be a limitation of this case. To the extent of our knowledge, this is the fourth case with acute CHF as the initial presentation of GPA.

## Conclusion

This case reminds clinicians that acute CHF with worsening renal function could be an initial manifestation of GPA, which should be included in the differential diagnosis.

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DOI:10.14744/AnatolJCardiol.2017.7732



# Radiofrequency ablation of accessory pathways in a toddler with Ebstein's anomaly and functional single ventricle physiology

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## Introduction

The incidence of multiple accessory pathways (AP) in patients undergoing electrophysiology study (EPS) for tachycardias is higher in structural heart disease such as Ebstein's anomaly. Ablation of APs is necessary at a younger age, because tachycardia is poorly tolerated in these patients owing to compromised cardiac reserve.

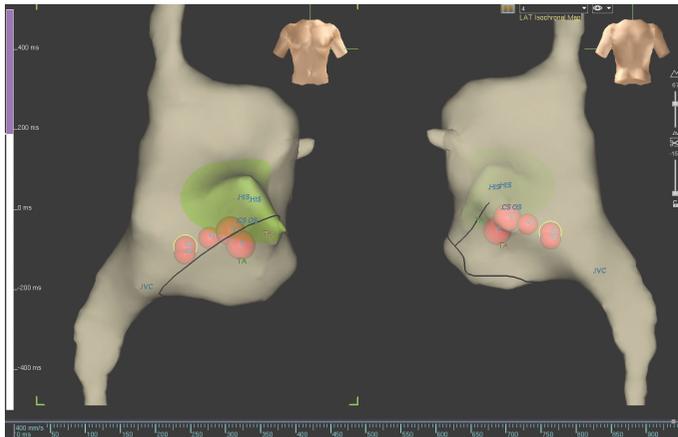
## Case Report

A 3-year-old girl, weighing 11 kg, with a diagnosis of Ebstein's anomaly was referred to our center due to recurrent supraventricular tachycardia (SVT) attacks resistance to multidrug medical therapy. She had a modified Blalock-Taussig shunt operation in the neonatal period, and thereafter suffered from recurrent SVT attacks compromising hemodynamics, requiring cardioversion. A surface electrocardiogram showed preexcitation consistent with Wolf-Parkinson-White Syndrome. An electrophysiology study with RF ablation of AP followed by hemodynamic study before bidirectional Glenn operation was planned.

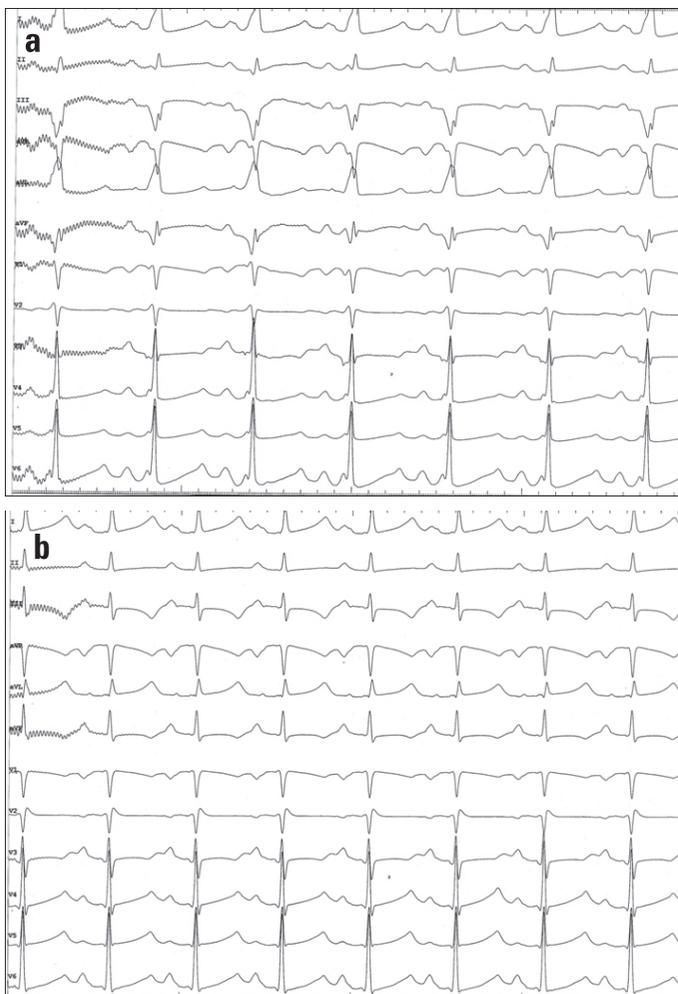
The electrophysiology study was conducted under general anesthesia. A three-dimensional mapping with the ESI system (EnSite System, St. Jude Medical, Minneapolis, MN, USA) was utilized during the procedure.

Recurrent SVT attacks induced during diagnostic catheter placement and causing hypotension and desaturation were stopped with adenosine administration. Baseline measurements were performed (AH: 82 ms, HV: 0 ms, BCL: 700 ms, PR: 115 ms, QRS: 132 ms, and QT: 450 ms).

Standard atrial stimulation protocol was carried out and orthodromic SVT with narrow QRS and tachycardia cycle length of 324 ms was induced. Because of hemodynamic compromise during SVT ESI, system mapping was done only for a short duration and the earliest VA conduction was found in right posteroseptal region of the tricuspid annulus with 63 ms (PERP: 320 ms, shortest preexcited R-R interval in AFIB: 380 ms). This region was marked via ESI system (Fig. 1) and a 5F RF ablation catheter was advanced into the right atrium positioned directly to this site. With a 50 W-50 C0 application for 2 s, AP was lost and most of the preexcitation on the 12-lead electrocardiogram was also lost



**Figure 1.** 3D electroanatomic mapping showing accessory pathways localized in posteroseptal and posterior regions of tricuspid annulus



**Figure 2.** 12-lead ECG. (a) Before ablation with preexcitation. (b) After ablation with preexcitation lost

(Fig. 2). The QRS axis in AVF and DII changed. RF was applied thrice for 60s, 60s, and 48s on the same location.

Most of the preexcitation was lost on the surface ECG, but with standard ventricular stimulation protocol, another SVT was induced and the ESI mapping during SVT revealed another

AP in the right posterior region of the tricuspid annulus. The region was marked with ESI system. A 50w-50C0 RF application for 6 s ceased the tachycardia with VA block. RF was applied four times for 60s, 30s, 30s, and 45s. VA conduction was con-santric decremental and VA block 400 ms, WLC 280 ms after the procedure.

The procedure was uncomplicated, there was no atrioventricular block, and the patient followed a normal postoperative course. After 1 week, the patient underwent a successful bidirectional Glenn operation and after 2 months of follow-up, the patient reported no further episodes of tachycardia.

## Discussion

Ebstein's anomaly, characterized by displacement of the posterior and septal leaflets of tricuspid valve downward toward the apex of the right ventricle is an uncommon congenital cardiac malformation accounting for <1% of congenital heart defects (1). Particularly, preexcitation and Wolff-Parkinson-White syndrome are more frequently associated with this anomaly (10%–29%) compared with any other congenital heart defect (2). The most frequent location of accessory pathway is the right posterior free wall (43%) (3).

Catheter ablation is a safe and effective therapy for arrhythmias seen in Ebstein's anomaly (4-6). But the anatomic challenges, in combination with the presence of multiple AP, can negatively impact ablation outcome in patients with Ebstein's anomaly (7, 8). In addition to the large right atrium, catheter stability also can be compromised by significant tricuspid insufficiency, and downward displacement of the tricuspid valve. In our case, hemodynamic compromise during tachycardia with hypotension and hypoperfusion was observed due to single ventricle physiology and multiple APs were found in EPS.

Catheter ablation of AP in infants with a 3D mapping system is a well-known safe and effective therapy, when medical therapy is ineffective (9, 10). In our case, ESI system was used to reconstruct the right and left atrial anatomy and to mark the ectopic foci for successful RF ablation.

## Conclusion

RF ablation can be used an alternative and safe method for treating patients with Ebstein's anomaly who present with AVRT, even in small children, when medical therapy is ineffective or complicated.

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DOI:10.14744/AnatolJCardiol.2017.7597



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