Ventricular Tachycardia Ablation in Arrhythmogenic Cardiomyopathy: A case report

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Abstract

Arrhythmogenic cardiopathy is a genetic disease that mainly affects young men and mainly involves the right ventricle. It is responsible for up to 25% of sudden deaths in children under 35 years of age [1]. To make its diagnosis, certain criteria are required, such as the characteristic electrocardiographic alterations in sinus rhythm, the presence of documented ventricular tachycardia and structural abnormalities especially in the right ventricle [2]. We present the case of a 25-year-old male patient with a confirmed diagnosis of arrhythmogenic cardiopathy who underwent ablation with a ventricular tachycardia catheter with an endocardial -epicardial approach.

Title

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Abstract

Arrhythmogenic cardiopathy is a genetic disease that mainly affects young men and mainly involves the right ventricle. It is responsible for up to 25% of sudden deaths in children under 35 years of age [1]. To make its diagnosis, certain criteria are required, such as the characteristic electrocardiographic alterations in sinus rhythm, the presence of documented ventricular tachycardia and structural abnormalities especially in the right ventricle [2].

We present the case of a 25-year-old male patient with a confirmed diagnosis of arrhythmogenic cardiopathy who underwent ablation with a ventricular tachycardia catheter with an endocardial -epicardial approach.

Keywords: arrhythmogenic cardiomyopathy, ventricular tachycardia, catheter ablation.

Introduction

A 25-year-old male patient with a history of palpitations whose sinus rhythm electrocardiogram showed the presence of negative T waves from V1 to V3, with the presence of a notch in the final part of the QRS in the DII and DIII derivatives, low voltage complexes in limb derivatives and prolonged QT interval, probably due to the use of amiodarone prescribed in another institution (Figure 1, panel A). He was admitted to the emergency room for a new episode of palpitations and it was possible to document a monomorphic tachycardia QRS wide RR regular with morphology of complete left bundle branch block (CLBBB) late transition, inferior axis and AV dissociation (Figure 1, panel B), compatible with ventricular tachycardia (VT) originated from the Right Ventricular Outflow Tract (RVOT), synchronized electrical cardioversion was performed due to hemodynamic involvement. Echocardiogram and cardiac magnetic resonance imaging showed motility alterations in both the right ventricle (RV) and the left ventricle (LV), with a slightly reduced LV ejection fraction (LVEF 49%), in addition to areas of extensive fibrosis mainly on the epicardial surface of the RV, it was initially taken to catheter ablation by endocardial approach, however, it presented an early recurrence, which is why new ablation was decided with an endocardial - epicardial approach.

Material and methods

The electro anatomical mapping system CARTO (Bio sense – Webster, Diamond Bar, California) was used, the approach to the epicardium was performed in a conventional way [3]. With a PENTARAY^{TM} NAV catheter (Bio sense – Webster, Diamond Bar, California) we proceeded to perform the endocardial electro anatomical reconstruction of the RV, as well as the epicardial surface of the entire heart (Figure 2). It should be noted that at the time of the ablation procedure the patient had complete blockage of the right branch of the bundle of His.

The bipolar endocardial voltage map (0.5-1.5 mV) of the RV showed small areas of low voltage in the tricuspid ring, in the outflow tract and in the apex; only 3.6% of the endocardial area was low voltage (<0.5mV). While most of the endocardial surface had adequate voltages, the endocardial unipolar (3.5-8.3 mV) voltage map showed many low-voltage zones anticipated wide epicardial compromise (Figure 2).

ResultsThe epicardial approach was obtained in a conventional way [3], the bipolar epicardial voltage map (0-5-1. 5 mV) revealed wide areas of low voltage, on the entire surface of the RV and in some parts of the surface of the LV, 81.3% of the entire epicardial area was low voltage. Double and fragmented potentials were identified in the RVOT (Figure 3), which is why we proceeded to apply radiofrequency with an irrigated ablation catheter THERMOCOOL ST - SF NAV bidirectional 4 mm (Bio sense – Webster, Diamond Bar, California) in this region guided by impedance drop (>10 ohms), with the objective of homogenizing the substrate in both the epicardium and the endocardium.

Once the substrate was modulated, reinduction maneuvers were made with and without isoproterenol and since VT was not induced, the procedure was considered successful. During the same hospitalization, an implantable automatic defibrillator (ICD) was placed. At 9-month follow-up, the patient has not had any recurrences.

Discussion Arrhythmogenic heart disease is a genetic disease that consists of the replacement of myocardial tissue by fibroadipose tissue [1], there are many proteins and genes involved, the most frequent being mutations of the genes that encode the proteins of desmosomes such as desmoplakin [1]. Recently in 2020 the latest diagnostic criteria were published considering that this pathology can have three variants: right dominant, left dominant and biventricular [2]. In the case presented, the patient presented alterations of contractility and fibrosis determined by cardiac magnetic resonance in both ventricles to right predominance, had typical alterations in the electrocardiogram at rest (both right and left involvement) and a VT with CLBBB morphology (Figure 1) together with the confirmed genetic mutation establish the accurate diagnosis of arrhythmogenic biventricular heart disease.

VT is common in these patients, catheter ablation is a fundamental part of management as it helps reduce recurrence, especially when the approach is endocardial and epicardial [4]. Initially epicardial embroidery was described for VT ablation in patients with chagasic heart disease [3] but has also been shown to be useful in other pathologies where epicardial involvement is greater than that of the endocardium such as arrhythmogenic heart disease [5], a recent meta-analysis showed that the endocardial and epicardial approach significantly reduces not only recurrence but all causes of mortality [5]. In our case, the patient already had a previous ablation performed with an endocardial approach only, but when presenting recurrence and considering the distribution of fibrosis described in the magnetic resonance imaging, we decided on a new endocardial and epicardial approach.

It has previously been reported that the endocardial unipolar voltage map helps predict the epicardial substrate, Hutchinson et al. by establishing those endocardial unipolar voltages below 8.27mV predict the presence of epicardial substrate [6]. In our case the endocardial bipolar voltage map showed small areas of low voltage, especially around the tricuspid ring and in the RVOT, however, the endocardial unipolar voltage map with an upper cut-off points of 8.3 mV, allowed to find extensive scar areas along the entire RV, which correlated perfectly with what was found in the standard bipolar epicardial voltage map (0.5 – 1.5 mV), where the scar was very extensive.

There are multiple ablation techniques in patients with structural heart disease [7], because the VT was not hemodynamically tolerated, we decided to apply radiofrequency in sinus rhythm, and we proceeded to perform homogenization of the substrate in the RVOT, where we found the double and fragmented potentials, after the application of radiofrequency it was not possible to induce the VT. Finally, it should be mentioned that in cases of arrhythmogenic heart disease of the RV, an ICD should be implanted regardless of the result of the ablation [1, 7]. In our case, during the same hospitalization, an ICD was implanted in the patient. At 9-month follow-up the patient has remained asymptomatic and without VT events.

Conclusion VT ablation with epicardial approach is crucial in the treatment of patients with arrhythmogenic heart disease, where fibroadipose infiltration is known to occur primarily in the epicardium. An ICD should also be implanted in these patients regardless of whether the ablation was successful or not.

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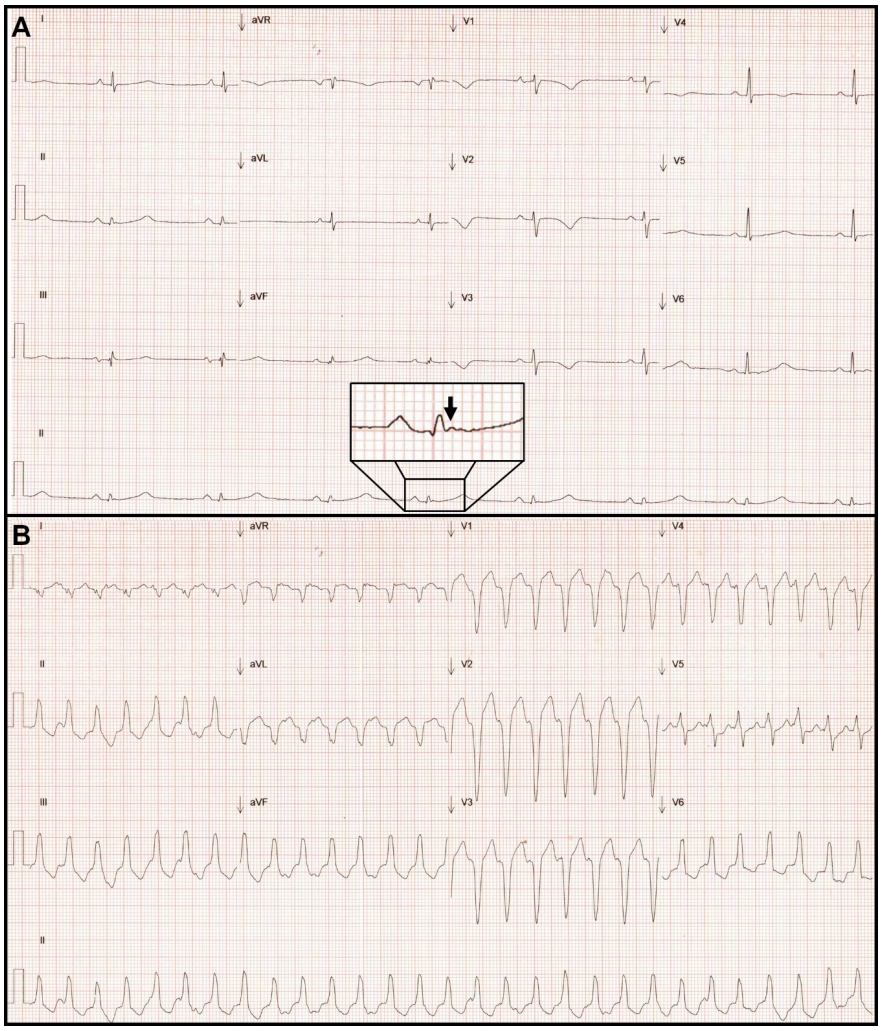

Figure 1: A. Sinus rhythm electrocardiogram, with signs of VR compromise (inverted T waves from V1 to V3, epsilon wave in the lower derivatives indicated with a black arrow) and LV (flat T wave in V4 and avL, low voltage complexes in limb derivatives), in addition to long QT interval. **B.** Ventricular tachycardia from the RV outflow tract, AV dissociation is clearly seen.

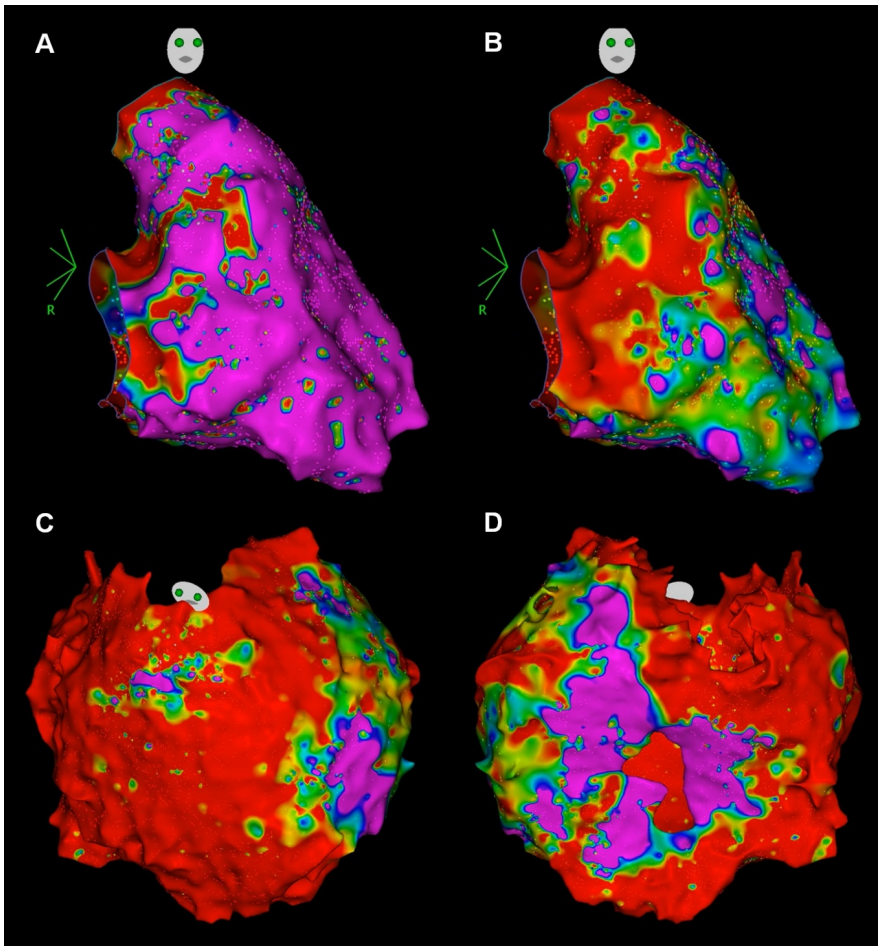

Figure 2: **A.** Bipolar endocardial voltage map (0.5 – 1.5 mV) of the RV, showing small areas of low voltage around the tricuspid ring and in the RVOT. **B.** Map of unipolar endocardial voltage (3.5 – 8.5 mV) of the RV, where a very wide area of low voltage is appreciated, which translates epicardial compromise. **C.** Previous view of the epicardial bipolar voltage map (0.5 – 1.5 mV) and **D.** Rear view of the same map, where you can see an extensive scar that involves the entire surface of the RV and part of the epicardial surface of the LV.

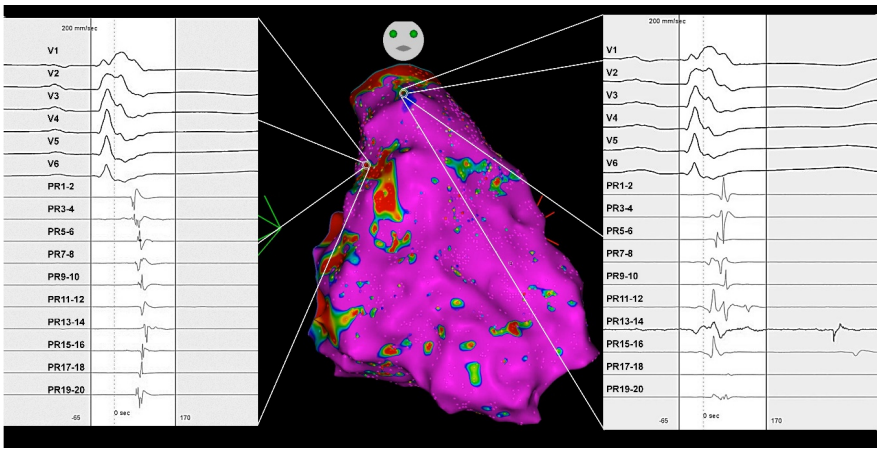

Figure 3: Recording of fragmented, double and late potentials in the RV outflow tract at a speed of 200 mm/sec. The precordial derivatives (V1 – V6) are shown in addition to the records obtained in the 10 channels of the PENTARAY ^{TM} catheter (PR 1-2 to PR 19-20).

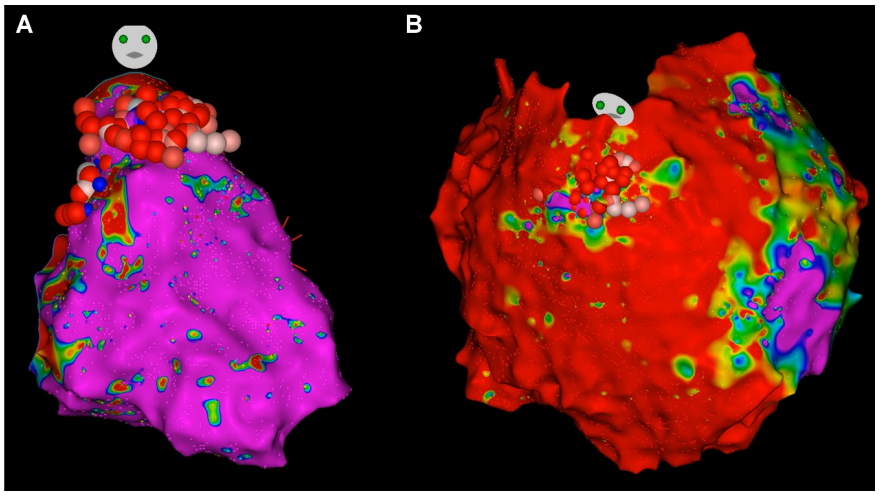

Figure 4: Ablation points on both the endocardial surface**(A)** and the epicardial surface **(B)** of the right ventricle.