

Case Report

Contribution of Electroanatomical Mapping to the Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy in a Patient with Sustained Ventricular Tachycardia

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Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a primary cardiomyopathy, characterized mainly by anatomic and functional defects of the right ventricle. In many cases its diagnosis is quite difficult in spite of the existence of defined diagnostic criteria for the disease. We describe an interesting case of a patient with sustained ventricular tachycardia derived from the right ventricular outflow tract, in whom the diagnosis of ARVC was made with the contribution of electrophysiologic study and electroanatomical mapping, as the use of all other diagnostic tests and laboratory methods had left many unanswered questions. Based on our case, but also on other studies and case reports in the literature, we conclude that electroanatomical mapping is useful for the documentation of the diagnosis of ARVC, whenever this is not clear from the use of available diagnostic tests according to the defined criteria of the disease.

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Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a primary cardiomyopathy, often familial, characterized mainly by anatomical and functional defects of the right ventricle. Microscopic findings in ARVC include replacement of diffuse areas of the right ventricular myocardium by fibrofatty tissue. The disease is manifested by life-threatening ventricular tachyarrhythmias and sudden death.¹⁻² Although there exist criteria on which to base the diagnosis of the disease,³ it is especially difficult to diagnose in some cases, because of problems concerning either the specificity of the electrocardiographic findings or the accurate representation of the structural and functional defects with the diagnostic methods available in practice.

According to recent studies, three-dimensional electroanatomical mapping

may contribute to the diagnosis of ARVC, defining pathologic areas of low potential that are compatible with fibrofatty replacement of the myocardium.^{2,4} We present here an interesting clinical case in which electroanatomical mapping with the CARTO system (Biosense-Webster) helped us in the differential diagnosis between ARVC and right ventricular outflow tachycardia (RVOT).

Case description

A 48-year-old man was referred to a district hospital because of faintness and palpitations after hard work. The patient's history included a similar episode six months before, with dizziness, diaphoresis and weakness. An electrocardiogram was performed, showing ventricular tachycardia (Figure 1) with left bundle branch block morphology

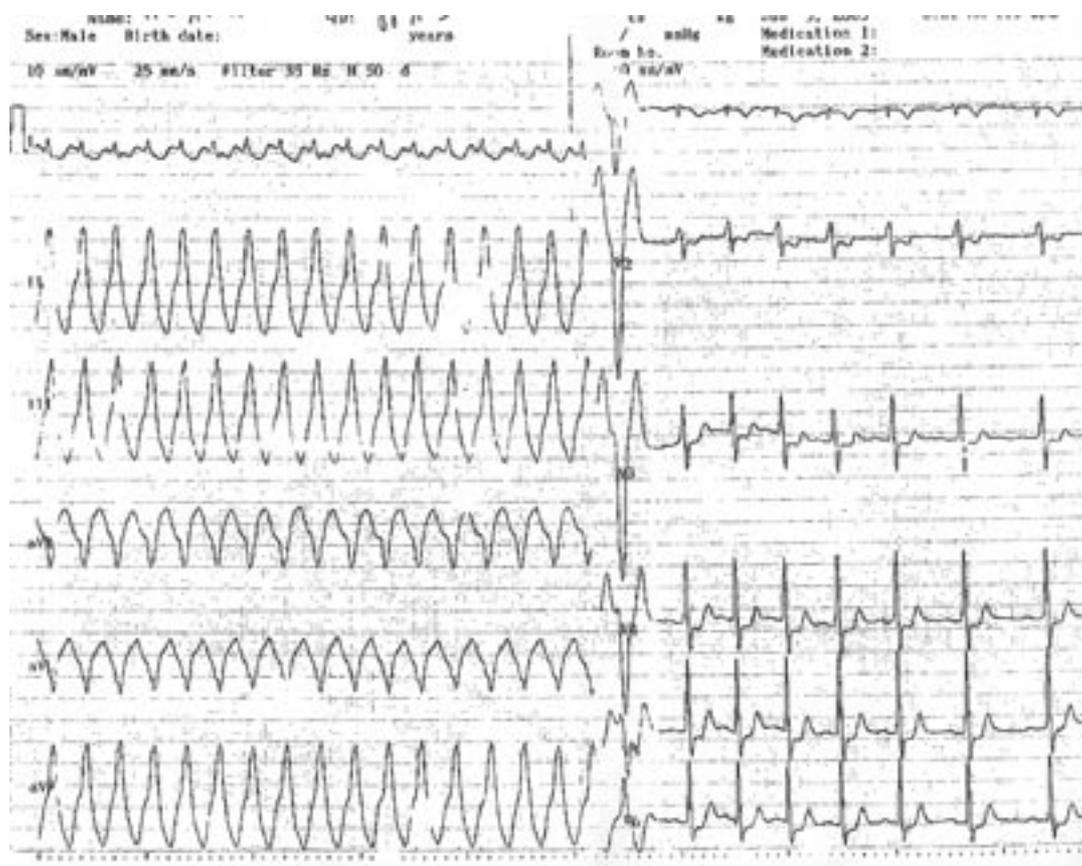


Figure 1. Emergency 12-lead electrocardiogram during the episode. A ventricular tachycardia is recorded, with left bundle branch block morphology and inferior axis, deriving from the right ventricular outflow tract. A few seconds after the beginning of the recording the tachycardia terminates spontaneously.

and inferior axis, originating from the right ventricular outflow area.

The clinical examination revealed no pathological findings. After amiodarone and β -blockers were administered a diagnostic work-up was performed for the determination of any cardiac disease. Echocardiography, coronary arteriography with left ventriculography, an exercise test and a signal averaged electrocardiogram showed no pathological findings. The 24-hour Holter recording (under treatment) was normal.

In addition, an magnetic resonance imaging (MRI) examination was performed and revealed a slight aneurysmatic protrusion in the mid portion of the right ventricular free wall, as well as dyskinetic areas of the free wall, including the outflow tract, but otherwise good systolic and diastolic function of both ventricles. After all these data had been collected it was still unclear whether the tachycardia was due to ARVC or to RVOT; the patient was referred to our hospital so that an electrophysiologic study could be performed.

The admission electrocardiogram (Figure 2) was

normal and did not show any of the known depolarization or repolarization abnormalities that are found in ARVC. The 24-hour Holter monitoring and the signal averaged electrocardiogram were also normal. Our clinic's echocardiogram reported borderline right ventricular dimensions, but with good systolic function.

After signing a consent form, the patient underwent an electrophysiologic study (EPS) and electroanatomical mapping of the right ventricle. The EPS was performed free of antiarrhythmic drugs. During programmed ventricular stimulation, the introduction of one, two and three extrastimuli, before or after the i.v administration of isoproterenol, did not provoke any ventricular arrhythmia.

Mapping

The electroanatomical mapping was performed with the CARTO system, using a 7-Fr deflectable quadri-polar mapping and ablation Navi-Star catheter (Biosense-Webster), as has already been described else-

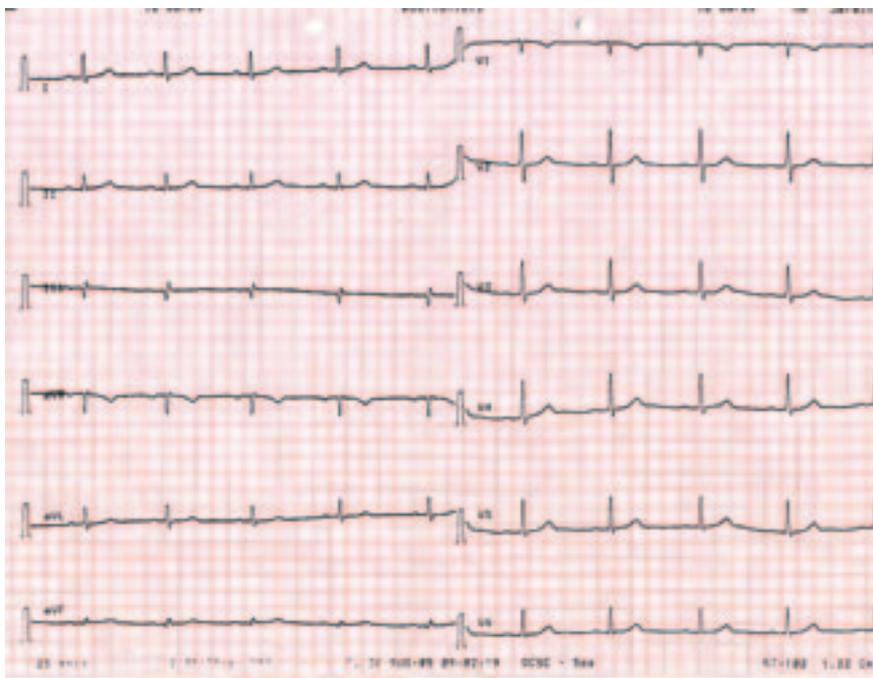


Figure 2. Recording of 12-lead electrocardiogram during admission in our hospital. There are no abnormalities of ventricular depolarization or repolarization.

where.^{2,4-8} The mapping was performed in sinus rhythm and 156 points were recorded. The results of the electroanatomical mapping are shown in Figures 3 and 4. In the right ventricle there is a clear, large, electrically inactive area extending from the right ventricular outflow tract to the subtricuspidal lateral inferior wall, as well as another, smaller one at the apex (dysplasia triangle⁹). The scar areas are delimited by an intermediate zone of voltages between 0.5 and 1.5 mV that distinguish precisely the active areas from the electrically inactive tissue. In the low voltage areas, the electrograms inscribed by the catheter's electrodes are fractionated with different waveforms (Figure 4), a finding that corresponds to a deficiency of healthy myocardial tissue.^{8,10,11}

As a result, with the aid of the electrophysiologic study and electroanatomical mapping, the presence of pathological myocardial tissue was definitively determined in certain zones of the right ventricle, thus verifying the diagnosis of ARVC. After finishing the mapping procedure, we performed an endomyocardial biopsy in the interventricular septum close to the ventricular outflow tract. The histological examination of the biopsy samples showed just the presence of areas of fibrosis among the myocardial cells. We finally implanted a cardioverter defibrillator, since the pre-syncope episodes

in combination with the recorded ventricular tachycardia placed the patient at serious risk for sudden death.

Discussion

Diagnosis of ARVC is based on established criteria, regarding mainly electrocardiographic signs combined with structural abnormalities that can be revealed by echocardiogram, ventriculography, MRI and endomyocardial biopsy. The diagnosis is documented whenever two major, one major and two minor, or four minor criteria are present.

Of particular interest is the differential diagnosis between ARVC and RVOT. The latter is a ventricular tachycardia with left bundle branch block morphology and inferior axis on the electrocardiogram, which is not combined with other cardiac diseases or obvious structural cardiac abnormalities. In our case, although morphologic abnormalities on MRI plus the recorded ventricular tachycardia gave one major and one minor criterion for the diagnosis of ARVC, these were not sufficient for a documented diagnosis. Additionally, the magnetic resonance findings are not specific for the diagnosis of ARVC, since there are studies that report similar abnormalities in patients with RVOT to those in our case.^{12,13}

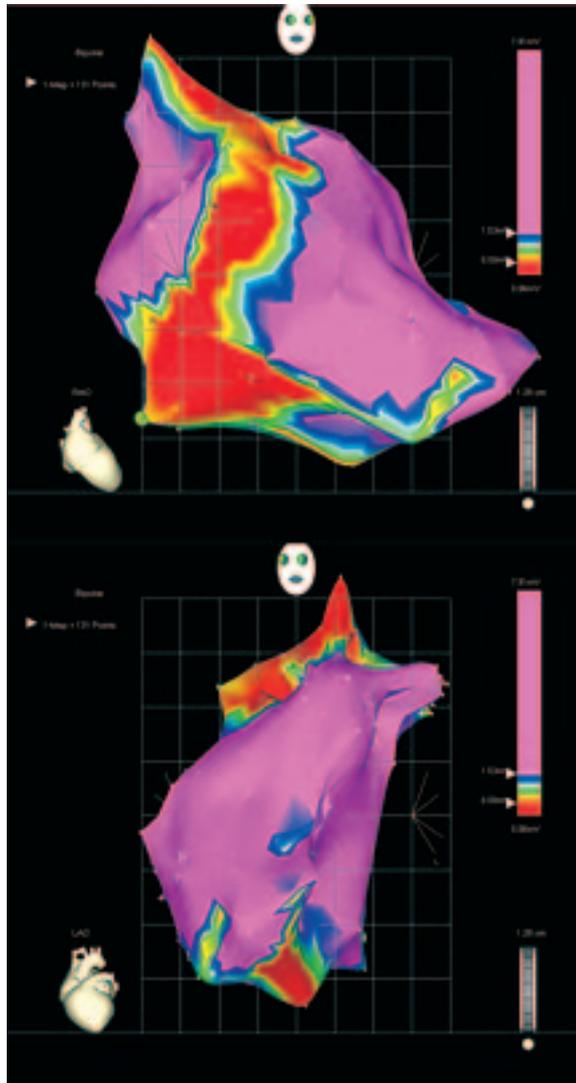


Figure 3. Electroanatomical mapping of the right ventricle with the CARTO system (30° right anterior oblique projection and 60° left anterior oblique projection). The red color shows the electrically inactive zones (scar areas) with potential <math><0.5\text{ mV}</math>, while purple represents the physiological electroanatomical areas with potential >math>>1.5\text{ mV}</math>. Intermediate colors represent the borderline areas between the physiologic and the scar areas.

However, it seems that the finding of electrically inactive areas of right ventricle, with low, fractionated potentials that are present during electroanatomical mapping, can contribute to the differential diagnosis between RVOT and ARVC. In a recent study where the CARTO system was used, Boulos et al showed that only the group of patients with ARVC presented these electrically inactive areas, while the control group and the group of patients with RVOT had no such abnormality.⁸

Moreover, the electrically inactive areas that we found in our patient are located within the known “triangle of dysplasia”,^{3,10} whose existence has already been proved in the past, giving further support to our diagnosis. In our case, the documented ventricular tachycardia originated from the right ventricular outflow tract, an area that gave abnormal findings on MRI and electroanatomical mapping, while the mid portion of the

right ventricular free wall (position of aneurysmatic protrusion according to MRI) was included in the detected zone of electroanatomical scar. Fractionated electrograms are another piece of evidence supporting the diagnosis of ARVC, since O’Donnell et al, in a comparative study of patients with ARVC or RVOT, found that these are very rarely present in patients with RVOT.¹⁴

The myocardial biopsy was performed in the interventricular septum, an area that had no abnormal findings on electroanatomical mapping. This is in accordance with the mild, non-specific, abnormal histological findings of the biopsy. The risk of cardiac tamponade forced us to avoid the right ventricular free wall during myocardial biopsy—a limitation of this invasive method for the diagnosis of ARVC.

The differential diagnosis between RVOT and ARVC is very important, because of their different

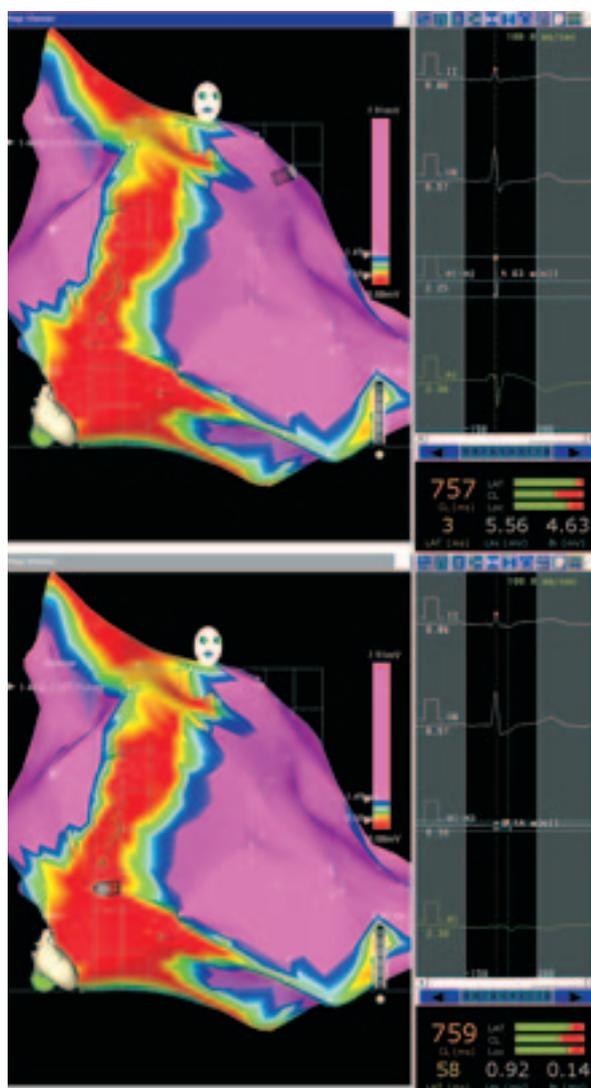


Figure 4. Electroanatomical mapping of the right ventricle with the CARTO system in 30° right anterior oblique projection. A small design indicates the position of the mapping catheter on the interventricular septum and on the free wall of the right outflow tract in the first and second image, respectively. On the right side of each image are the recordings from surface electrocardiographic leads II and V6, as well as the bipolar and monopolar endocardial recordings. In the second image fractionated electrograms in the electroanatomical scar area, with bipolar potential of just 0.14 mV, may be seen.

natural histories as well as the different therapeutic approaches to the two diseases. RVOT has a good prognosis without any particular progress of the disease with time and, in general, the theory that RVOT is an early stage of a cardiomyopathy does not appear correct.^{15,16} Whenever symptoms are especially intense, antiarrhythmic drugs can be administered or ablation of the arrhythmic focus carried out, with very good long term results.

In contrast, the natural history of ARVC is not completely known, regarding either the patients or their relatives. However, it is certain that its prognosis depends mainly on the right ventricle's electrical instability, which can provoke lethal ventricular arrhythmias at any stage of the disease.^{1,17-19}

Our patient manifested ventricular tachycardia with obvious hemodynamic intolerance, since he had

experienced pre-syncope episodes twice in the previous 6 months, which led him to the hospital where the arrhythmia was recorded. The significant contribution of electroanatomic mapping to the diagnosis of ARVC in our patient allowed the decision to implant a cardioverter defibrillator for the prevention of sudden death.

According to our case, and to other similar reports in the literature, electrophysiologic study in combination with electroanatomical mapping becomes useful for the differential diagnosis between ARVC and RVOT whenever this is not entirely clear from the use of the established diagnostic criteria of ARVC.

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