CASE PRESENTATION

Long Term Follow-Up in a Patient with Arrhythmogenic Right Ventricular Dysplasia

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ABSTRACT

This case report describes eight years of follow-up in a young adult with arrhythmogenic right ventricular dysplasia (ARVD). He presented with exertional palpitations, symmetric T wave inversions and possible epsilon waves in the right precordial leads on electrocardiogram (EKG), raising suspicion for ARVD. Transthoracic echocardiography revealed a dilated and excessively trabeculated right ventricle (RV), and cardiac magnetic resonance imaging showed fatty infiltration of the RV myocardium. These findings established the diagnosis of ARVD, and given his palpitations, a defibrillator was implanted. Over the next years, he had several episodes of ventricular tachycardia requiring therapy from his device, despite escalating medical therapy. He therefore underwent radiofrequency catheter ablation for the VT, which successfully controlled the VT.

Keywords: arrhythmogenic right ventricular dysplasia, ventricular arrhythmia, intracardiac defibrillator, radiofrequency catheter ablation.

REZUMAT

Descriem cazul unui pacient tanăr cu displazie aritmogenă de ventricul drept (DAVD) și urmărirea acestuia pe o perioadă de 8 ani. Diagnosticul prezumptiv a fost pus pe baza simptomatologiei clinice (palpitații la efort fizic), modificărilor electrocardiografice (unde T negative, simetrice și unde epsilon în derivațiile precordiale drepte). Ecocardiografia transtoracică a evidențiat un ventricul drept cu trabeculații excesive, iar rezonanța magnetică nucleară a relevat prezența infiltrării grăsoase a miocardului ventricular drept. După stabilirea diagnosticului de certitudine, pacientului i s-a implantat un defibrilator cardiac. Pe parcursul următorilor ani, sub tratament antiaritmic, pacientul a prezentat mai multe episoade de tahicardie ventriculară, care au fost terminate prin șocuri electrice interne. Ablația prin cateter cu curent de radiofrecvență a constituit atitudinea terapeutică de succes la acest pacient.

Cuvinte cheie: displazie aritmogenă de ventricul drept, aritmie ventriculară, defibrilator intracardiac, ablație prin cateter cu radiofrecvență.

INTRODUCTION

Arrhythmogenic right ventricular dysplasia (ARVD) is an inherited condition affecting predominantly the right ventricle (RV) with fatty or fibro-fatty substitution of myocytes, leading to segmental or global dilation. The fibrofatty replacement leads to ventricular arrhythmia and risk of sudden death in its early stages, and to right ventricular or biventricular failure in its later stages[1].

The disease starts usually in the subepicardium, expands to the subendocardium of the RV, and only rarely advances to the left ventricle (LV). The clinical manifestation of ARVD is unpredictable, and is probably connected with the chronological evolution of the myocardial injury. Accordingly, ARVD patients are sorted into 4 clinicopathological stages (phases): hidden phase (silent), arrhythmic phase, global dysfunction of the RV, and biventricular pump failure. Usually at the age of 30 to 40 years, ARVD patients report of exercise-induced palpitations because ventricular arrhythmias (VA) that arise in the RV[2]. The majority of patients with ARVD usually remain in the second stage of the illness. At this stage, the disease is defined by segmental or global RV myocardial lesions with remaining normal myocardium[3]. Cardiac mortality in ARVD patients is caused by heart failure and sudden cardiac death (SCD). Those who have experienced ventricular tachycardia (VT), have an elevated SCD risk[4,5].

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CASE REPORT

A 35-year-old patient with existing cardiovascular risk factors (overweight, dyslipidemia, former smoker) and Hashimoto's thyroiditis, with no personal history of cardiovascular disease and no family history of SCD, was referred to our hospital in February 2012 after a syncopal episode, that occured at rest and was preceded by dizziness and rapid palpitations. He reported dyspnea at medium exertion over a few months.

The ECG (Figure 1) showed sinus rhythm with frequent ventricular premature complexes (VPC) of left bundle branch block (LBBB) pattern, indicating an inferior RV origin; T wave inversions in the inferior and precordial leads, S waves in all precordial leads, and possible epsilon waves in leads DII, DIII, and aVF were noticed. Late ventricular potentials were also present (Figure 2).

ARVD was suspected at transthoracic echocardiography (TTE) assessment (Figure 3). Impaired, dilated, and excessive trabeculated RV with dilatation of the RV outflow tract were TTE`s main findings.

Holter electrocardiography monitoring for 24 hours detected isolated supraventricular and ventricular premature complexes (episodes of ventricular bigeminy, couplets and triplets). Repeated resting ECG captured VPC and one episode of non-sustained ventricular tachycardia with a LBBB pattern. Coronary angiography showed normal coronary arteries.

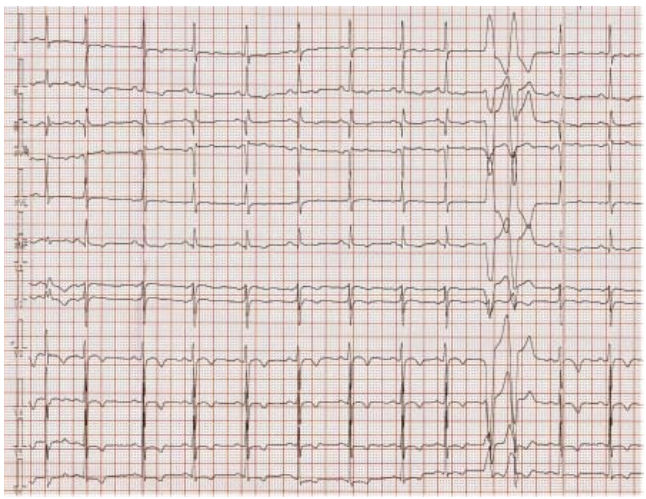

Figure 1. 12-leads ECG. Sinus ryhthm with repetitive ventricular premature beats having a left ventricular branch block pattern, of inferior right ventricular origin; T waves inversions and S waves and in all precordial leads; possible epsilon waves in leads DII, DIII, aVF.

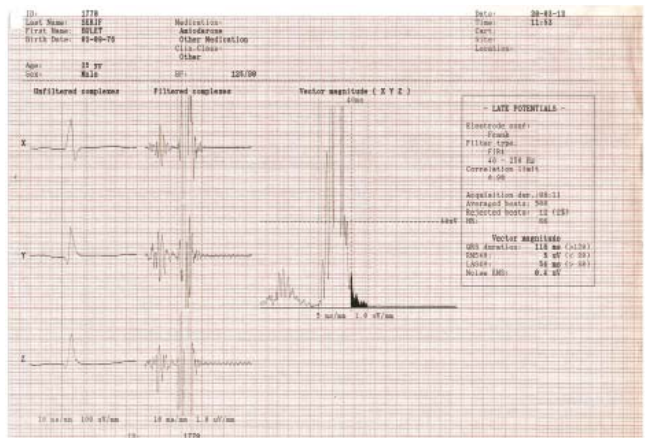

Figure 2. Ventricular late potentials: QRS duration = 116 msec, RMS40 = 6uV (<20); LAS40 = 54ms (>38), noise RMS = 0.3uV.

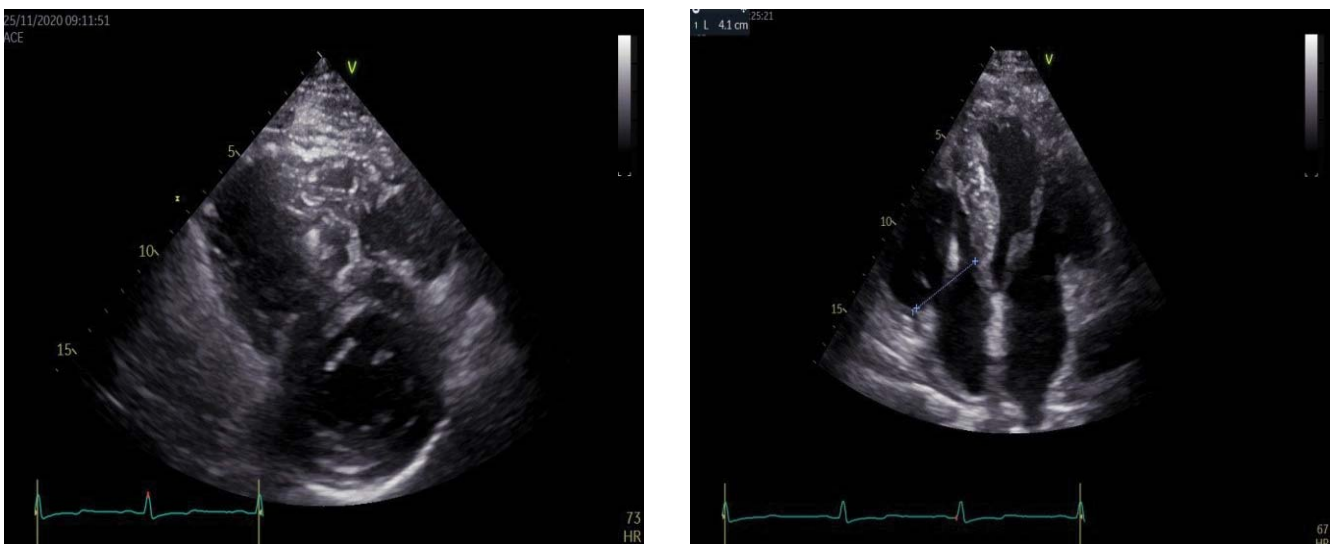

Figure 3. Transthoracic echocardiography
Left: parasternal short-axis view at mitral valve level showing right ventricular (RV) dilation with excessive trabeculations. Right: apical four-chamber view showing increased RV diameter at the base and the intracardiac defibrillator lead positioned in the RV apex.

We referred the patient to cardiac magnetic resonance imaging (MRI – Figure 4), which in the following years after the diagnosis of ARVD, magnetic resonance imaging was not performed again. Right and left ventricular function was assessed by transthoracic echocardiography. RV function remained the same, did not alter progressively during the follow-up. The LV had normal wall motion and function (LV ejection fraction =70%), but presented focal areas of late gadolinium enhancement in the basal segment of the lateral-inferior wall.

One year later the patient was hospitalized for several appropriate therapies delivered by the ICD, including anti-tachycardia pacing (ATP) initiated and shocks. The ECG at admission showed sustained ventricular tachycardia originating from the basal RV free wall, with a rate of 162 beats per minute (bpm) (Figure 5).

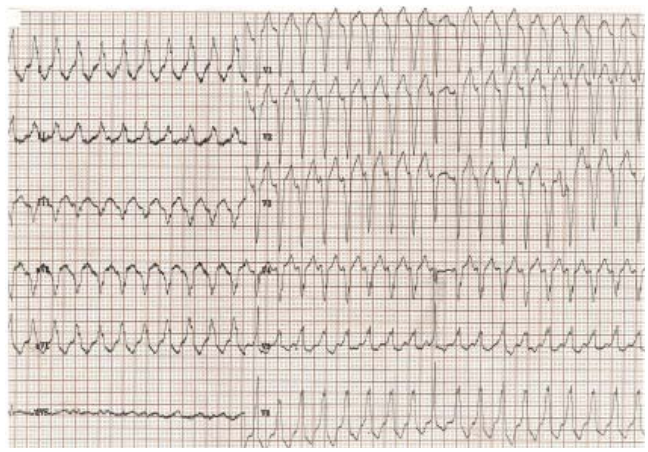

Figure 5. Sustained ventricular tachycardia originating in the right ventricle, with an appearance of left bundle branch block, heart rate of 162 beats per minute. Notice fusion QRS complex as patonomonic ECG sign of VT.

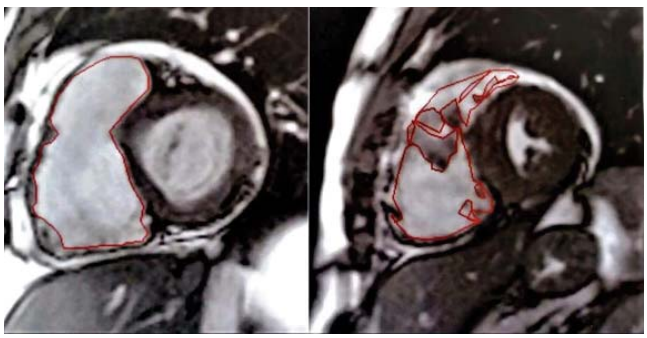

Figure 4. MRI performed before implantation demonstrated localised bulging of the RV, excessive trabeculations and increased RV end-diastolic and end-systolic volumes. The patient underwent implantation of a St. Jude Medical single-chamber ICD, programmed in VVI mode with a base frequency of 40 bpm. Antiarrhythmic therapy with amiodarone and beta-blocker was initiated, but the patient missed the follow-ups, did not adhere to the prescribed medication, and did not respect the recommendation of limited physical activity.

After the reinitiation of amiodarone, an electrophysiologic study was performed but VT was not induced (January 2014), so conventional VT ablation was not performed.

He was followed closely clinically, and after one year, only amiodarone was discontinued, the therapy with beta-blocker was continued. The patient played football as a hobby. We suggested him to avoid vigurous and competitive physical activity and to reduce exercise duration and dose per week.

Aerobic exercises and endurance training should be avoided by this category of patients.

Other restrictions included: smoking, alcohol and energy drinks.

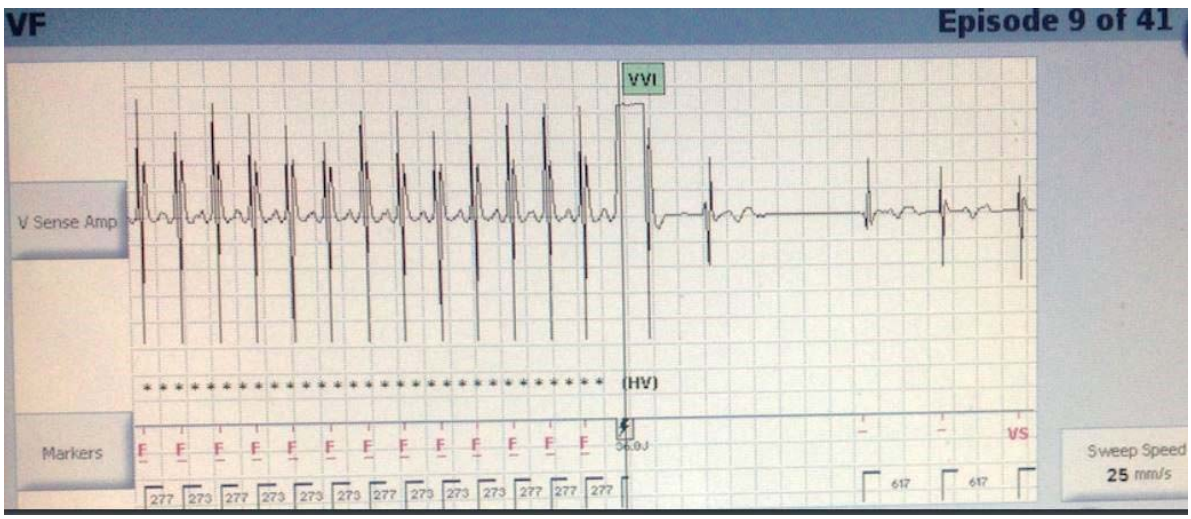

Figure 6. ICD interrogation. Intracardiac electrogram showing VF detection and internal cardioversion in this patient. F = ventricular fibrillation window; VS=ventricular sensed beat.

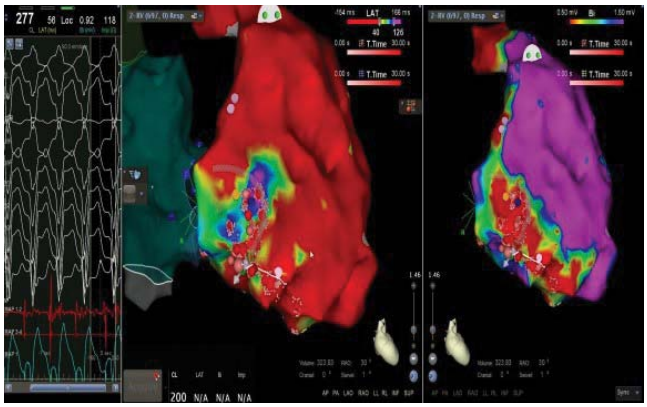

Figure 7. Three-dimensional mapping system (Carto-3), right lateral view. The scar is located in the lateral free RV wall. The red dots indicate serial ablations applied inside the scar region according to defined targets. ECG 12 derivation represents the VT of inferior axis LBBB like, showing origin inside the scar. Scar definition was done by voltage map (unipolar middle image; bipolar right image).

In May 2019, seven years after the implant, the ICD was changed due to End of Life (EOL) indication (Medtronic Protecta XT). One year later he was re-admitted for recurrent VT episodes with appropriate therapy delivered by the ICD (Figure 6; 4 episodes in one week, failed ATP, the VT was terminated by internal shock delivery). Radiofrequency (RF) catheter ablation was performed using a three-dimensional (3D) mapping system (Carto-3).

The 3D electroanatomic CARTO map was achieved in sinus rhythm and substrate ablation was performed (voltage and activation map was done). „Normal" endocardial regions were described as those with an amplitude in bipolar electrogram of 1.5 mV, „scars" were defined as areas having an amplitude of 0.5 mV, and "abnormal" region was defined as having a bipolar electrogram amplitude of 0.5 to 1.5 mV. Substrate ablation target besides the voltage criteria, included critical sites (delayed, split and fragmented electrograms)[6]. During cathether manipulation VT was induced (Figure 7), but activation map was not achievable due to unstable hemodynamics of the patient (electrical cardioversion was performed immediately after syncope occurred).

The endpoint of each application was signal attenuation / complete elimination of these potentials. Programmed RV stimulation was performed after the completion of ablation, up to 4 extrastimuli at 2 different RV sites were used. The procedure was successful as no VT was inducible.

DISCUSSION

ARVD is a progressive disorder that often leads to RV failure in the long term. Adapted treatment strategies in ARVD plan to suppress or effectively end the recurrent VA, and to prevent SCD by antiarrhythmic pharmacotherapy, catheter ablation, or an ICD[7]. In our patient, incidental EKG findings after episodes of palpitations led to the discovery of a potentially life-threatening disease and to subsequent ICD implantation and antiarrhythmic therapy. The patient experienced after implantation many episodes of recurrent VTs and ICD shocks, initially with successful antiarrhythmic therapy; we can also speculate that drastically reduction in exercise led to a period of no arrhythmia. The special medical requirement of avoiding excessive physical activity was efficient, followed by clinical improvement. Thus the patient understood the risk of aggravation of his condition in case of continuing sustained physical activity. There are limited data published regarding the amount of exercise that may worsen the outcome. Yet, the evolution led finally to RF catheter ablation indication which was successfully performed. This case illustrates successful follow-up and survival in a life-threatening disease as ARVD. Close follow-up and complex attitude permitted an almost normal life course in a young patient.

However, due to progressive nature of ARVD recurrence rates are not insignificant, and ablation does not eliminate the need for ICD placement[8,9]. There are few data and case reports in long term follow-up of ARVD patients.

Compliance with ethics requirements:

The authors declare no conflict of interest regarding this article. The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national law. Informed consent was obtained from all the patients included in the study.

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