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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Subcutaneous ICD Implantation and Catheter Ablation

A Step-Planned Approach for Ventricular Tachycardia Management in ARVC

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ABSTRACT

Secondary prevention of sudden cardiac death in the young patient with arrhythmogenic right ventricular cardiomyopathy and hemodynamically tolerated ventricular tachycardia is still a challenging field. We present a combined approach, including subcutaneous implantable cardioverter-defibrillator (ICD) and catheter ablation, as a promising treatment to prevent both ventricular tachycardia recurrences and ICD shocks. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:185-191) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 20-year-old athlete in good health was admitted to the emergency department (ED) of another hospital for an episode of paroxysmal palpitations during physical activity. The physical examination findings were normal. The electrocardiogram (ECG) documented sinus rhythm and inverted T waves in leads V_1 to V_2 (**Figure 1**).

LEARNING OBJECTIVES

- To recognize and correctly classify arrhythmogenic cardiomyopathies by integrating different diagnostic tools.
- To emphasize the role of 3D-EAM to overcome CMR limits in RV tissue characterization.
- To consider a combined strategy including S-ICD implantation and VT ablation for managing arrhythmia recurrences in selected patients.

PAST MEDICAL HISTORY

The past medical history was unremarkable. In particular, the patient had no familial history of sudden cardiac death or syncopal episodes.

DIFFERENTIAL DIAGNOSIS

Effort-related palpitations can result from supraventricular or ventricular tachyarrhythmias occurring in patients with or without structural heart disease.

INVESTIGATIONS

The echocardiographic examination results were normal. Cardiac magnetic resonance (CMR) revealed a

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ABBREVIATIONS AND ACRONYMS

ARVC = arrhythmogenic right ventricular cardiomyopathy

ATP = antitachycardia pacing

CA = catheter ablation

CMR = cardiac magnetic resonance

ECG = electrocardiogram

ED = emergency department

EP = electrophysiological

ICD = implantable cardioverter-defibrillator

LBBB = left bundle branch block

LP = late potential

LV = left ventricle

NSVT = nonsustained ventricular tachycardia

PVS = programmed ventricular stimulation

RV = right ventricular

S-ICD = subcutaneous implantable cardioverter-defibrillator

VT = ventricular tachycardia

3D-EAM = 3-dimensional electroanatomical map

subepicardial region of late enhancement at the middle posterolateral aspect of the left ventricle (LV); other findings were unremarkable. Results of a stress test were negative for ST-segment T-wave changes and arrhythmias.

A diagnosis of healed myocarditis[1] was hypothesized, and the patient underwent implantation of a loop recorder (Reveal Linq, Medtronic) to correlate symptoms with possible cardiac rhythm disturbances. The first electronic control of the device revealed multiple episodes of nonsustained ventricular tachycardia (NSVT), as well as a single episode of symptomatic (palpitations) sustained monomorphic ventricular tachycardia (VT) lasting 51 seconds (cycle length 280 milliseconds).

MANAGEMENT

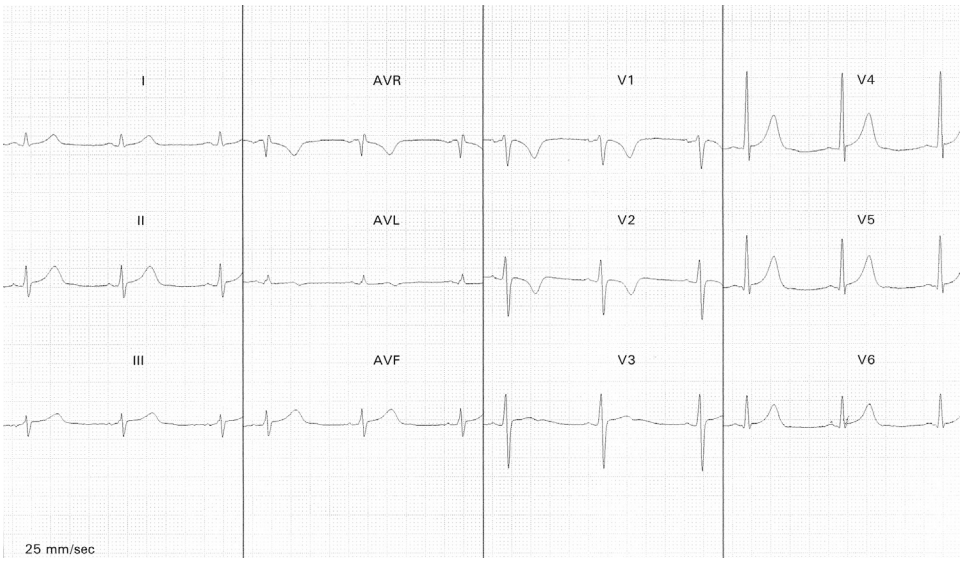
Nadolol was ineffective in VT prevention. Therefore, the patient was referred to our hospital (Humanitas Mater Domini Clinical Institute, Castellanza, Italy) for electrophysiological (EP) evaluation and possible catheter ablation (CA).

During the EP study, multiple monomorphic and polymorphic NSVTs with fast cycle length (250-280 ms) were induced with programmed ventricular stimulation (PVS). Notably, all induced VTs had a left bundle branch block (LBBB) configuration (**Figure 2**), consistent with a possible origin from the right ventricle.

Hence, on the basis of this finding and the ECG repolarization abnormalities, a borderline diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC) was hypothesized (1 major and 1 minor criteria),[2] and an endocardial 3-dimensional electroanatomical map (3D-EAM, CARTO-3 System, Biosense Webster) of the right ventricle was performed. The bipolar map revealed a small (19.6 cm^2) abnormal voltage area (≤1.5 mV) localized at the anterior aspect of the outflow tract (**Figure 3A**). Herein, only far-field late potentials (LPs) were recorded (**Figures 3B and 3C**). The unipolar map confirmed a wider (53.2 cm^2) abnormal voltage area (≤5.5 mV) extending from the anterior outflow tract to the superior anterolateral aspect of the right ventricle, consistent with an epicardial scar (**Figure 3D**).

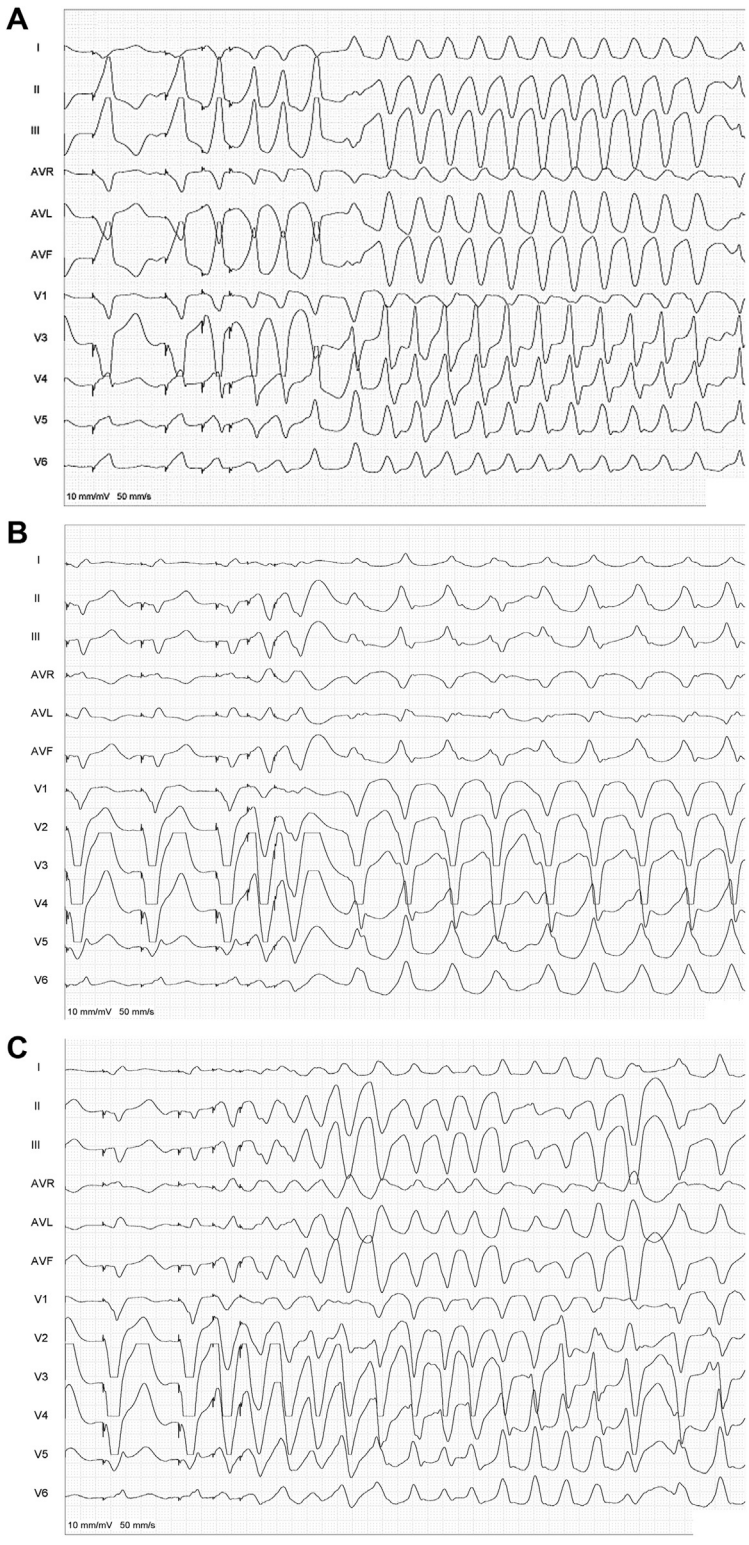
CMR repeated 1 year after the first scan documented mild right ventricular (RV) dilation with akinesia of the basal inflow tract (third minor criterion for ARVC).[2] The subepicardial band of late enhancement at the posterolateral aspect of the LV was confirmed. Given the very likely diagnosis of ARVC, and after having carefully balanced risks and benefits, a subcutaneous implantable cardioverter-defibrillator

FIGURE 1 Baseline 12-Lead Electrocardiogram



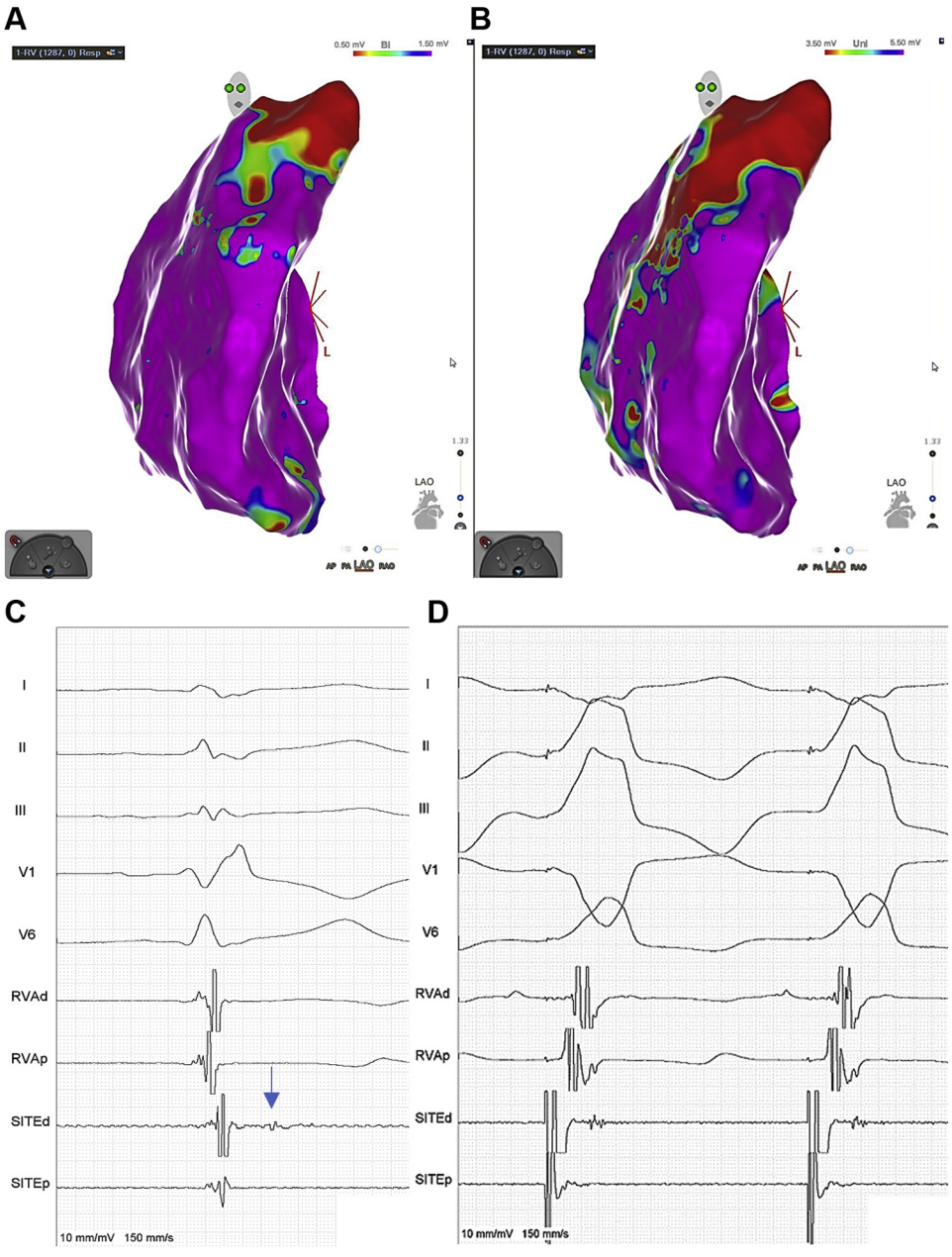
The tracings show inverted T waves in leads V_1 to V_2.

FIGURE 2 **Results of Programmed Ventricular Stimulation**



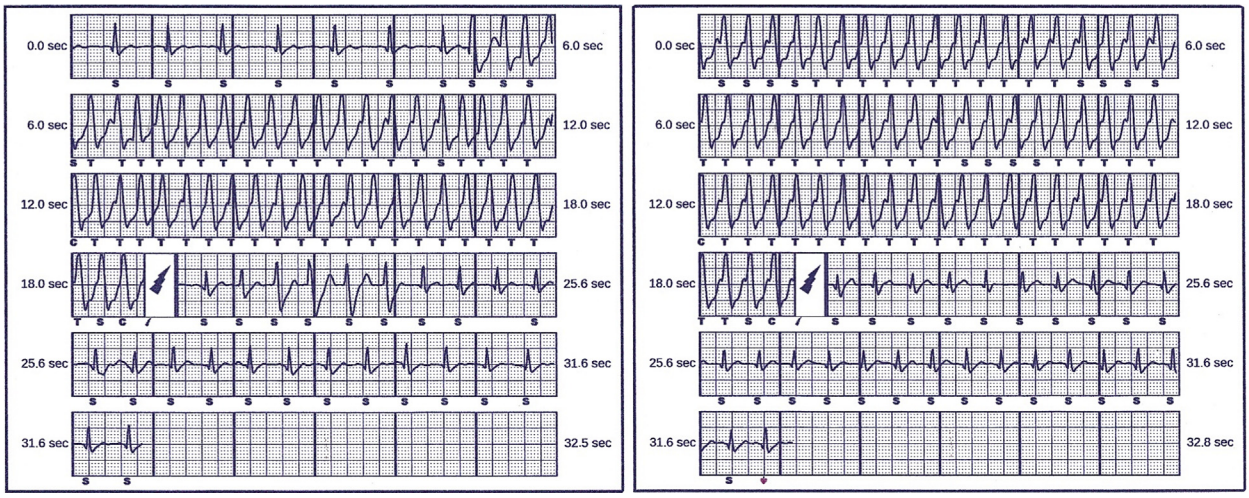
Multiple induced **(A and B)** monomorphic or **(C)** polymorphic nonsustained ventricular tachycardias with left bundle branch block configuration and different axis deviation.

FIGURE 3 Endocardial Electroanatomical Map and Intracardiac Recordings



(A to D) Electroanatomical mapping and recording. **(A)** A right ventricular bipolar endocardial electroanatomical map (EAM) shows a region (surface 19.6 cm^2) of abnormal voltages consistent with dense scar (≤0.5 mV) **(red)** and a border zone (0.5-1.5 mV) **(yellow to blue)** located at the anterior aspect of the outflow tract. Within this area, **(C)** far-field late potentials were recorded **(blue arrows),** and **(D)** ventricular pacing resulted in local ventricular capture (short-stimulus QRS interval) without late potential capture, thus confirming their remote origin (from epicardium). **(B)** The right ventricular unipolar endocardial substrate map shows a more extensive epicardial scar (53.2 cm^2). The right bundle branch block pattern in **C** was mechanically induced. LAO = left anterior oblique; RVA = right ventricular apex; SITE = mapping catheter.

FIGURE 4 Monomorphic Sustained Ventricular Tachycardia Successfully Treated by Subcutaneous Implantable Cardioverter-Defibrillator Shock



(S-ICD) was inserted according to current guidelines.[3] Conversely, ablation was not deemed indicated at that time in view of the very likely need for an epicardial approach with its associated complications. Nonetheless, it should have been considered after the S-ICD interventions.

One week after discharge, the patient was readmitted to the ED because of an arrhythmic storm. An electronic control of the device documented 10 episodes of monomorphic sustained VT, 6 of which were treated by successful ICD shocks (**Figure 4**). Administration of intravenous amiodarone during monitoring in the intensive care unit was effective in preventing early VT recurrences.

Given the young age of the patient, the limited efficacy of antiarrhythmic drugs, and the high success rate of ablation[4] in preventing recurrent VT in ARVC, an ablative strategy was pursued. An epicardial approach was directly attempted because of the absence of endocardial targets, as shown by the 3D-EAM findings.

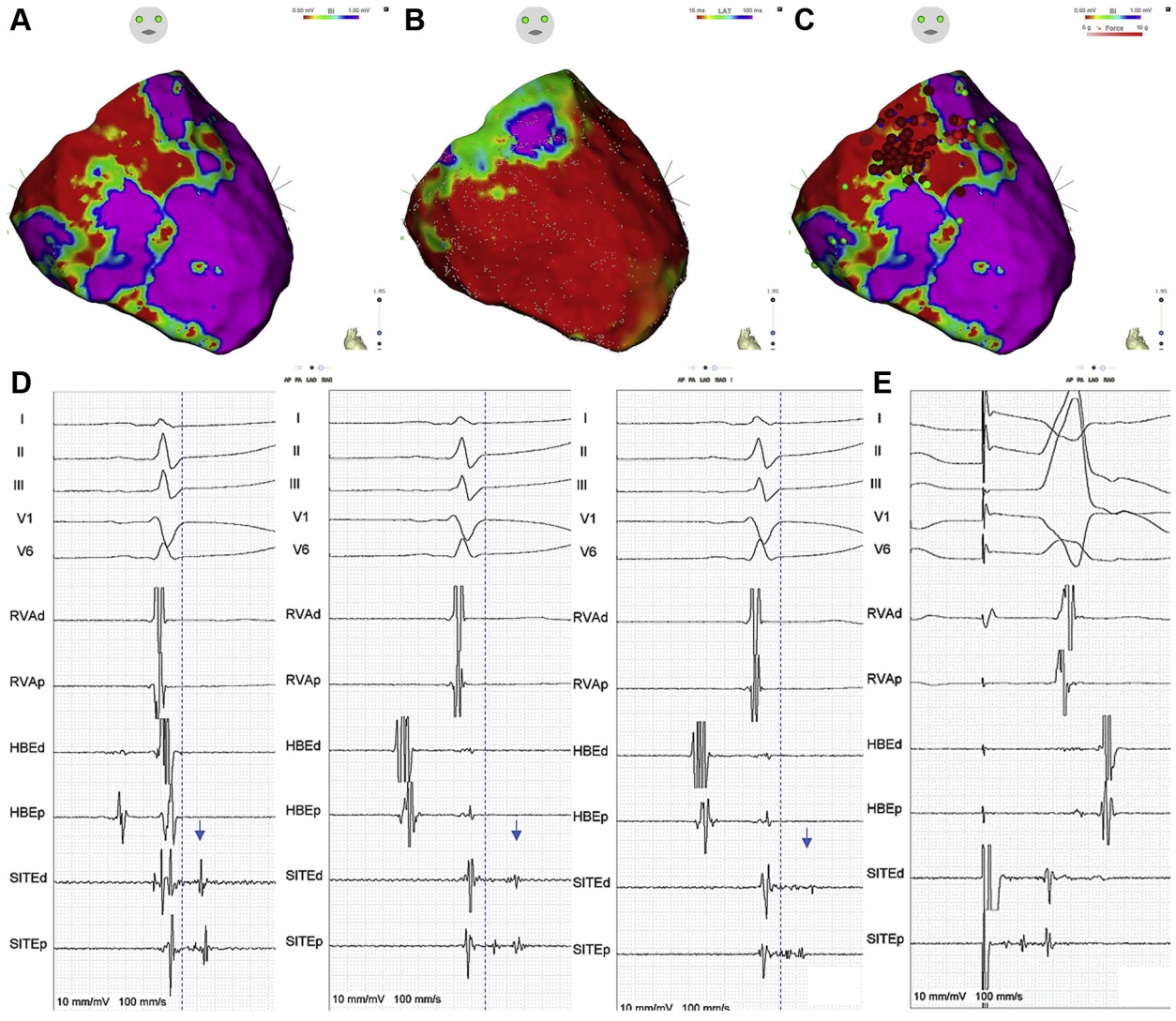
An open irrigated catheter with contact force (Thermocool Smart Touch, Biosense Webster) was used for epicardial substrate mapping and ablation. A large (51 cm^2) epicardial area of abnormal voltage electrograms (≤1 mV, just opposed to the endocardial region of abnormal unipolar voltage recordings) was detected (**Figure 5A**). Multiple low-voltage and long-duration LPs (**Figures 5B and 5D**) were recorded within the scar region. Herein, ventricular pacing resulted in local LP capture with a long-stimulus QRS interval, possible expression of slow conduction from isles of excitable cardiomyocytes to the healthy ventricular myocardium through fibrofatty tissue (**Figure 5E**). Abolition of all LPs was successfully achieved (**Figure 5C**) at the ablation sites. At the end of the procedure, no ventricular arrhythmia was inducible by PVS.

DISCUSSION

The diagnosis of ARVC is often challenging, and it requires integration of information gathered from different diagnostic tools. As in the described case, ECG depolarization or repolarization abnormalities and VTs with LBBB configuration mostly represent the first signs leading to clinical suspicion of ARVC. Given the small thickness, CMR has a limitation in RV wall tissue characterization.[5] Instead, 3D-EAM could have a prognostic value in predicting the arrhythmic risk in ARVC, and it provided helpful insight to formulate the correct diagnosis in the present case.[5] Conversely, the subepicardial band of late enhancement at the posterolateral aspect of the LV was diagnostically confounding, and it could have represented either biventricular involvement of the arrhythmogenic cardiomyopathy or the result of healed myocarditis.

Regarding treatment, patients with scar-related re-entrant VT mostly benefit from antitachycardia pacing (ATP), which has been demonstrated to be effective and safe compared with ICD shocks and to improve quality of life in different clinical settings.[6,7] In patients with ARVC, appropriate ICD therapies (including ATP interventions) are frequent, but complication rates are very high (about 20%,

FIGURE 5 Epicardial Substrate Map



(A) The bipolar map shows an extensive abnormal voltage area (<1 mV) **(red to blue)** mirroring in location that detected at the endocardial electroanatomical map (Figure 3D). **(B)** The right ventricular activation map during sinus rhythm demonstrates a centripetal gradient of delayed activation **(red to purple)** within the abnormal voltage area. **(C)** Ablation sites shown by **red dots. (D)** Examples of single or multicomponent late potentials **(arrows)** with different activation delays recorded during sinus rhythm within the abnormal substrate. The **vertical lines** indicate the end of the QRS complex. **(E)** Electrical stimulation from this area results in direct late potential capture with a prolonged-stimulus QRS interval (see also text). HBE = His bundle electrogram; RVA = right ventricular apex; SITE = ablation catheter.

including infections and lead malfunctions).[8] Despite the lack of ATP capability, the S-ICD may provide a valid alternative to overcome some limitations of the transvenous systems. Indeed, the S-ICD has been effectively inserted as secondary prevention, as demonstrated by real-life registries and randomized clinical trials.[9,10] Furthermore, endocardial-epicardial CA seems to be very effective in reducing ICD interventions in patients with ARVC even in the long term, with approximately 70% freedom from VT recurrences.[4] Therefore, CA represents a promising solution to the lack of ATP in patients ARVC who would otherwise be eligible for an S-ICD. Conversely, CA is associated with possible major complications, especially when the epicardial approach is used;[11] thus, it should be reserved for tertiary centers. Notably, our patient underwent S-ICD insertion with the aim of eventually treating recurrent VTs with ablation. So

far, this type of step-planned, combined approach for management of scar-related arrhythmias in ARVC has only rarely been described and should be considered more frequently in selected patients.

FOLLOW-UP

The patient remained free from ICD interventions in the next 24 months. Furthermore, genetic analyses revealed 3 different heterozygous variants of *LMNA*, *PKP2*, and *DSG2*. Although the *LMNA* defect is very likely coincidental, the *PKP2* and *DSG2* mutations are definitely associated with ARVC and support possible involvement of the LV in the cardiomyopathy.

CONCLUSIONS

Clinical management of young patients with established or suspected arrhythmogenic cardiomyopathies is often challenging and always requires an in-depth balance of the risks and benefits among different therapeutic options. The combined strategy including S-ICD insertion followed by CA of eventually occurring VTs seems to be an effective therapeutic option, and it could avoid the complications related to the transvenous systems in the long term.

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KEY WORDS arrhythmogenic right ventricular cardiomyopathy, subcutaneous ICD, ventricular tachycardia ablation