CASE REPORT

A novel treatment strategy for therapy refractory ventricular arrhythmias in the setting of arrhythmogenic right ventricular dysplasia

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Arrhythmogenic right ventricular dysplasia (ARVD) is a major cause of ventricular tachycardia and cardiac arrest in young adults. The ideal management of this genetic disorder is individual. The treatment options are antiarrhythmic drug therapy, transcatheter radiofrequency catheter ablation, implantable cardioverter defibrillator therapy, and surgical treatment [Kies P, Bootsma M, Bax J, Schalij MJ, van der Wall EE. Arrhythmogenic right ventricular dysplasia/cardiomyopathy: screening, diagnosis, and treatment. Heart Rhythm 2006;3:225–34; Verma A, Kilicaslan F, Schweikert RA et al. Short- and long-term success of substrate-based mapping and ablation of ventricular tachycardia in arrhythmogenic right ventricular dysplasia. Circulation 2005;111:3209–16]. In the following, we describe a unique case of a young patient, presenting with therapy refractory ventricular arrhythmias in the setting of ARVD, who following failed catheter ablations, has been successfully treated with beating heart cryoablation.

KEYWORDS
Arrhythmogenic right ventricular dysplasia; Beating heart surgery; Cryoablation; Radiofrequency catheter ablation; Sternotomy

Clinical summary

An 18-year-old athlete was admitted to our centre for treatment of symptomatic recurrent ventricular arrhythmias in the setting of arrhythmogenic right ventricular dysplasia (ARVD). The diagnosis of ARVD was based on electrocardiogram (ECG) (Figure 1A and B), transoesophageal echocardiography (TEE), magnetic resonance imaging (MRI) findings, and the history of recurrent ventricular tachycardia (VT) attacks with left bundle branch block (LBBB) morphology (two minor and one major criteria). The TEE and MRI scan revealed a slightly dilated right ventricle (RV) and impaired contraction in the free wall of the RV. The anatomo-histopathological examination at the time of surgery confirmed the definitive diagnosis of ARVD (ventricular muscle was replaced by fatty and fibrous tissue). The patient’s medical history consisted of recurrent exercise-induced monomorphic VTs requiring DC cardioversion at cycle lengths ranging from 350 to 400 ms, without a history of syncope. Physical examination during admission was normal. Antiarrhythmic therapy was initiated, however the VT recurred. Subsequently, an electro-physiological study (EPS) was performed. The programmed ventricular stimulation with a basic cycle lengths of 500 and 600 ms and a single extra systole induced sustained monomorphic VT with LBBB and inferior axis morphology at cycle lengths ranging from 350 to 400 ms. Endocardial activation and entrainment mapping was performed during tachycardia. Radiofrequency (RF) catheter ablation was performed at the best target sites. However, post-ablation the programmed stimulation demonstrated still inducible monomorphic VTs with slightly different multiple morphologies, but always with LBBB and inferior axis at cycle lengths ranging from 350 to 400 ms. The patient was started on a beta-blocker and amiodarone treatment but had further clinical episodes of VT during exercise at cycle lengths ranging from 430 to 500 ms. He had another RF and cryothermy catheter ablation at 3 months intervals. During these procedures, CARTO mapping in sinus rhythm was performed to characterize the arrhythmia substrate, and activation mapping during the inducible tachycardias. Linear lesions were performed in an attempt to encircle the scar low-potential area containing also the earliest activation sites involving the RV outflow tract (RVOT) and anterior...
basal free wall. However, in spite of extensive ablations, several VT morphologies remained inducible with one extra systole at a basic cycle length of 500 and 600 ms. Subsequently, in spite of a combination of amiodarone and sotalol therapy, the patient presented with clinical recur-
rences of the same monomorphic VTs with cycle length of 460–550 ms. Given the incessant and drug resistant nature of the arrhythmia and the repeatedly failed ablations, a novel surgical treatment alternative other than disarticula-
tion was proposed. In order to perform a detailed mapping and to detect the possible epicardial origin of the arrhythmogenic focus, a beating heart epicardial mapping with cryo-
ablation was planned.

Under general anaesthesia, a median sternotomy was per-
formed. A TEE examination confirmed no evident thrombus in the cardiac chambers. There was a normal atrio-
ventricular (AV) and ventriculo-atrial concordance. On gross inspection, there was a dysplastic and aneurysmatic RV wall extending to the RVOT, and having a paradoxical systolic motion (expansion during systole instead of contrac-
tion). We performed a detailed activation mapping during sinus rhythm and tachycardia using a duodecapolar 
mapping band (with 5 mm interelectrode distance). For the ablation we used standard cryoablation probes (Frigitronics; CooperVision Inc., Lake Forest, CA). The mapping of the epicardial surface of the RV in sinus rhythm revealed delayed, fragmented, low amplitude local activation, suggesting important arrhythmogenic zones in the mid-right AV groove, RVOT, pulmonary annulus, and apical RV. VT at a cycle length of 400 ms was inducible with one extra systole at a basic cycle length of 500 ms and well tolerated. During the induced tachycardia, very early fragmented activation was detected in the anterior base of the RVOT. During point ablation of this target (Figure 1C) the tachycardia terminated. This target site was within an approximately 2 cm distance from the best endocardial target sites at the previous endocardial attempts. Subsequently, further linear lesions were created in this zone and between the anterior infundibulum, RV apex, and inferior or diaphragmatic aspect of the RV (Figure 2), the so-called ‘triangle of dysplasia’, using cryothermy (−60°C for 2 min) in order to encircle and isolate the RVOT and RV areas with abnormal local potentials (Figure 3). Following the ablations, the ventricular stimu-
lation in spite of aggressive stimulation protocols at three basic cycle lengths (430, 500, and 600 ms) with three extra systoles in the presence of isoproterenol infusion failed to induce any ventricular arrhythmias. The temporary epicardial pacemaker leads were placed and the chest was closed in standard fashion.
An EPS after one week, in the absence of any anti-arrhythmic drug treatment and in spite of aggressive stimulation protocols with three basic cycle lengths (430, 500, and 600 ms) and three extra systoles showed no inducible ventricular arrhythmias. After an uneventful recovery, the patient was discharged without medication on the eighth post-operative day. The patient remained symptom-free for a 6-month period following the intervention. A control transthoracic echocardiography after 9 months revealed the same findings as reported preoperatively; a slightly dilated RV and an impaired contraction in the free wall of the RV.

Discussion

The arrhythmogenic RV dysplasia is an important cause of sudden death, often exertional, in young individuals and athletes. The two major causes of death are severe ventricular arrhythmias and progressive heart failure, with a similar expected rate of approximately 1% a year. The characteristic pathological finding is the fibro-fatty replacement of the RV myocardium. One of the major goals when treating ARVD is to prevent sudden cardiac death. Available therapies include antiarrhythmic medications, defibrillator implantation, RF ablation, and other surgical interventions. Arrhythmias can be controlled with drugs such as sotalol or amiodarone, or medications that block the stimulating effects of adrenaline in the heart such as β-blockers. Some high-risk patients who have episodes of ventricular fibrillation or VT require implantable cardioverter defibrillator (ICD) therapy. RF ablation should be reserved for patients who have recurrent, therapy refractory recurrent ventricular arrhythmias in spite of antiarrhythmic therapy and/or recurrent frequent ICD shocks.

In our case, failure of the medical therapy and several catheter ablations (RF and cryotherapy), the frequency and the symptomatic character of the arrhythmia and the goal to improve the quality of life, were the major reasons leading to the decision to proceed with surgical intervention. We felt that in our young patient, the eventual success of a surgical procedure could postpone heart transplantation, which always remains a final option (especially for patients with bilateral ventricular involvement which was not present in this case). Similarly, ICD therapy would likely not have been helpful given the slow nature of the VT (<120/min). Post-operatively, we decided to postpone the ICD implant, given the absence of involvement of the left ventricle, of any spontaneous or induced ventricular arrhythmias, episodes of syncope or family history of sudden death. Disarticulation of the RV was not considered owing to the invasive nature and high post-operative complication rate of this type of complex surgery. A thoracoscopic technique instead of sternotomy, and transthoracic epicardial catheter ablation could have been an option in this young patient, however, some zones could likely not have been accessible for mapping and ablation through these approaches.

In our case, beating heart cryoablation through sternotomy was a feasible and effective treatment strategy. We believe that this technique can be recommended as an alternative treatment for disarticulation surgery in patients with therapy-refractory recurrent ventricular arrhythmias, following failed catheter ablations in the setting of ARVD.

References