Arrhythmogenic right ventricular dysplasia presenting as right ventricular outflow tract tachycardia

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Abstract A case of a 51-year old male is presented. A left bundle branch block inferior axis tachycardia was manifest. At electrophysiological study this tachycardia was inducible and was ablated in the septal right ventricular outflow tract (RVOT). Two other tachycardias were identified both with right bundle branch block (RBBB) morphology raising the suspicion of diffuse pathology. Arrhythmogenic right ventricular dysplasia (ARVD) was confirmed by right ventricular angiography and magnetic resonance imaging (MRI). An implantable cardioverter defibrillator (ICD) was implanted and an appropriate shock was later delivered.

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case report

A 51-year old Caucasian male was admitted to the intensive care unit with palpitations and dyspnoea. The surface ECG showed sustained ventricular tachycardia (VT) displaying the typical morphology of RVOT tachycardia with an inferior axis and left bundle branch block morphology (Fig. 1A). Coronary artery disease was ruled out by coronary angiography and echocardiography showed normal left ventricular function without valvular disease. There was no history of syncope or sudden cardiac death in the family. The patient experienced a similar, self-terminating episode accompanied by near fainting one year earlier. The ECG during sinus rhythm displayed no overt abnormalities. The presumed clinical idiopathic RVOT tachycardia was reproducibly inducible by programmed ventricular stimulation (S₁ 350 ms, S₂ 260 ms, S₃ 260 ms) without isoproterenol indicating a reentry tachycardia. This arrhythmia was successfully treated with catheter-based radio-frequency ablation from the septal, subpulmonary
During post-ablation programmed ventricular stimulation without isoproteronol, a second sustained monomorphic VT originating from the left apical, posteroseptal region (extreme right axis, RBBB morphology) was induced (Fig. 1B). Finally, a third sustained left-sided VT showing no characteristics of an idiopathic VT was provoked by programmed ventricular stimulation (Fig. 1C). Subsequently, a right ventricular angiogram was performed and revealed an aneurysm of the subpulmonary wall, characteristic of ARVD (Fig. 2). The diagnosis was confirmed by regional wall movement abnormalities and wall thinning on MRI. Therefore, the patient received an implantable defibrillator (ICD) that delivered an appropriate shock for polymorphic VT four months later.

Discussion

Idiopathic ventricular tachycardias (IVT) occur in patients without structural heart disease and are responsive to ablative and medical therapy [1]. RVOT is the most common type of IVT and can also occur in ARVD, a condition that is not readily identified by standard diagnostic measures [2]. It is critical to identify patients with ARVD since this condition has been implicated in up to 20% of sudden cardiac deaths in patients less than 35 years old and may require more aggressive treatment such as the

![Figure 1](image1.png)

**Figure 1** (A) Ablated clinical ventricular tachycardia (cycle length 290 ms) displaying the typical features of RVOT tachycardia (inferior axis, left bundle branch block morphology), (B) induced left ventricular tachycardia (cycle length 350 ms) with superior axis and right bundle branch block morphology, (C) second induced left ventricular tachycardia (cycle length 260 ms).

![Figure 2](image2.png)

**Figure 2** Right ventricular angiogram with typical subpulmonary aneurysm (arrows) indicative of ARVD.
implantation of an ICD [3]. During the electrophysiological study (EPS), induction of a reentrant VT with critically timed extrastimuli, the presence of fragmented intracardiac electrograms, and the induction of more than one VT should prompt high suspicion for the existence of ARVD [4,5]. In the present case, the second and third induced VT (Fig. 1B and C) displayed a right bundle branch block morphology, indicating a left ventricular origin. Since left ventricular involvement can be shown histologically in up to 76% of patients with ARVD, induction of VT with a right bundle branch block morphology is not uncommon in this condition [6]. MRI is an important contributor to the diagnosis of ARVD if right ventricular wall movement abnormalities can be demonstrated [7]. However, MRI has been implicated in overdiagnosis of ARVD since free-wall thinning and increased intramyocardial fat are also found in healthy subjects [8].

In patients presenting with a VT displaying the characteristic ECG-morphologies of RVOT tachycardia, underlying ARVD should be suspected if the VT shows characteristics of a reentrant ventricular tachycardia and if more than one VT can be induced. In this situation, a right ventricular angiogram can readily be performed in order to confirm the diagnosis of ARVD.

References