A CASE FOR EDUCATION

Ventricular tachycardia catheter ablation in arrhythmogenic right ventricular cardiomyopathy

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Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterized by progressive fibro-fatty replacement of the myocardium. These islands of surviving myocardial tissue create the substrate for recurrent sustained ventricular tachycardia (VT), which constitutes the major clinical issue. A significant proportion of patients have a clinical course characterized by ventricular arrhythmias, with important changes in their quality of life and long-term prognosis. Anti-arrhythmic drugs may not result in a very efficient control of the relapses and, often, are poorly tolerated. Catheter ablation has been demonstrated to be effective at reducing VT episodes and implantable cardiac defibrillator (ICD) therapies in ARVC patients. Herein we describe our interventional approach to VT ablation in the setting of ARVC.

Procedural settings

Before the procedure

ICD therapies are deactivated at the beginning of the procedure. When the 12-lead electrocardiogram (ECG) of the target VT is not available, noninvasive programmed stimulation is performed the day before or at the time of the procedure to provide a pattern of the inducible morphologies. In the case of stable electrical reference required by the chosen mapping system, a catheter is advanced into the coronary sinus, generally through the right jugular vein because of easy engagement and good stability achieved. The procedure is performed under general anesthesia because epicardial access is standard in patients with ARVC.

The electrophysiological study

The programmed stimulation is performed at multiple drive cycles (600 msec drive and 400 msec) up to 4 extrastimuli, at baseline and under isoproterenol administration. The 12-lead ECG morphology of the induced VT is kept as reference through the entire clinical case. Overdrive pacing or external DC shock may be required to interrupt the arrhythmia.

Approaching the case

Following the reconstruction of the 3-dimensional (3D) geometry of the endocardial and epicardial surfaces with the ablation catheter, high-density mapping is performed to provide (1) bipolar/unipolar voltage and (2) local activation time maps. A precise reconstruction of the geometry of the chamber of interest is achieved with the ablation catheter because of the lower distance between the distal tip and the magnet. Furthermore, an overall better handling and maneuverability allows a more precise reconstruction of the 3D geometry.

Current technology supports the use of the multispline and multielectrode catheter, which allows the creation, during sinus rhythm, of high-density maps with definition of the electrical substrate characterization. Local activation maps are generated to provide a propagation pattern of sinus rhythm, leading to identification of areas with abnormal (and/or late) activity and expression of slow conduction. Steerable sheaths are often required to provide support during endocardial mapping and ablation. These must always be highlighted on the map with colored tags.

Electrical imaging of the ventricular tachycardia

VT induction is generally attempted when the multielectrode catheter is place into or adjacent to these areas with conduction delay to maximize the possibilities of recording diastolic activity in case of short-lived arrhythmia or arrhythmias that are hemodynamically not tolerated. In case of tolerated VTs, following precise electrical imaging of the diastolic pathway, the ablation catheter is advanced at sites showing entry/isthmus activity in an attempt to prove VT interruption by radiofrequency (RF). After VT interruption, RF delivery is maintained during sinus rhythm with the endpoint of

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achieving abolition of near-field electrical activity at sites where diastolic activity had been imaged during VT.

If the VT is not tolerated hemodynamically, the arrhythmia is promptly terminated by overdrive pacing or external DC shock. In this case RF delivery is then performed during sinus rhythm, aiming at the same endpoint as described above. If the target is epicardial, following completion of the epicardial ablation, careful evaluation of electrical activity at endocardial adjacent sites is undertaken to abolish any residual activity located endocardially. If the VT procedure began endocardially, the same process is applied epicardially.

Following the achievement of substrate modification endpoints during sinus rhythm (elimination of late potentials [LPs] and/or near-field electrical activity at site where diastolic activity had been imaged during VT), programmed electrical stimulation is performed under the same protocol used at baseline, also following isoproterenol administration titrated to achievement of sinus rate of 120 to 140 beats per minute.

**Power setting**
Up to 50 watts may be used with standard RF irrigated tip ablation catheter. The rate of impedance drop is used to decide when to stop RF delivery. There should be a careful evaluation of local impedance: a drop higher than 20 ohms should lead to RF interruption. Contact force monitoring can provide an additional feature to improve RF ablation. Coronary angiography is not routinely performed except in those situations where simultaneous atrial and ventricular electrograms (EGMs) are recorded, meaning the catheter is sitting in the atrioventricular groove, or when epicardial ablation is performed at the acute margin or very basally at the presumed location of the interventricular posterior artery. Phrenic nerve, unlike left-sided VT ablation, testing is not an issue in this type of procedure because the nerve runs posterior to the atrioventricular groove over the right atrium.

**Case report**
We describe the case of a 21-year-old man with a history of cardiac arrest followed by prompt cardiac resuscitation. ARVC was diagnosed by cardiac magnetic resonance. He received a dual-chamber ICD and was discharged with beta blockers titrated to the maximal tolerated dose. At 1 year follow-up a monomorphic VT (left bundle branch block morphology with inferior axis, cycle length [CL] 310 msec) was documented. VT catheter ablation was indicated. Noninvasive programmed stimulation was undertaken and induced a VT with CL of 310 msec, inferior axis, left bundle branch block morphology, and transition in lead V5 (Figure 1).

**Epicardial access**
As per protocol epicardial puncture was performed to achieve access to the epicardial surface. The access must be always performed under general anesthesia; this approach is preferred owing to a lower complication rate and better patient management in case of severe complications, such as perforation of the right ventricle (RV) wall.

An Agilis EPI (Abbott Medical, Saint Paul, MN) is then used to deliver the ablation catheter to create the 3D map; following this, a bipolar voltage map is generated by a multiplespline mapping catheter (PentaRay catheter, Biosense Webster, Diamond Bar, CA) (Figure 2A), revealing a large area of scar extending from the right ventricular outflow tract to the lateral side of the tricuspid annulus and to the basal/inferior aspect of the RV. LPs have been recorded (Figure 3) over a limited area on the area shown in a second map (Figure 2B) within an activation map (Supplemental Video).
Inducing ventricular tachycardia

As per our common practice, before VT induction, the multispline mapping catheter is advanced over the area where LPs have been identified (Figure 4).

The clinical VT was induced and the mapping catheter was quickly moved over the epicardial surface to cover and record the full diastolic pathway of the VT (as described in Figure 5). The figure shows that the entire CL of the VT has been recorded epicardially, above the area of LP before identification.

Since the VT was hemodynamically tolerated, the mapping catheter was rapidly exchanged with the ablation catheter (SmartTouch SF, Biosense Webster). As shown in Figure 6, EGMs mapped between the proximal and distal tip of the ablation catheter covered 76% of the VT CL. Ten seconds of RF ablation (50 watts, $35\degree C$) terminated the VT (Figure 7).

Remap

A critical endpoint of substrate modification of VT by catheter ablation is the proven abolition of near-field electrical activity at the remap during sinus rhythm. The first remap (Figure 8B) of the epicardial surface after first pulses of RF delivered at the isthmus site of the VT circuit showed an incomplete abolition of LPs when compared to the local activation performed at the beginning of the procedure (Figure 8A). Further RF was delivered at that site (12 minutes, 50 watts, $35\degree C$) and a second remap (Figure 8C) proved electrical silence with complete abolition of abnormal activity.

Endocardial map revealed absence of late activity and no further RF was delivered (Figure 9).

Induction

Acute success of the procedure could be therefore claimed when several attempts failed to reinduce any VT (Figure 10).

LP areas and VT circuits: Not always coincident

We describe a second case of a 19-year-old subject with AVRC diagnosed after several syncopal episodes with ECG abnormalities in V1–V3 (T-wave inversion) and cardiac magnetic resonance scan positive for major criteria. ICD was implanted following the occurrence of sustained VT. Ten months later, the patient received multiple shock therapies leading to indication for catheter ablation. Epicardial mapping documented a large area of low voltage along the basal, mid-ventricular anterior wall of the RV, extended to the outflow tract and to the basal, inferior aspect of the RV. LPs were mapped on the anterior surface of the right outflow tract (Figure 11). Interestingly, when VT was induced, the entire circuit of the tachycardia was mapped (Figure 12) on a different site compared to where EGM abnormalities during sinus rhythm were previously identified (Figure 13).

The entire diastolic pathway could be tracked and the area displayed on the electroanatomic map. Ablation was performed at the documented isthmus site of the VT circuit with documented interruption. In this case one can observe that the area of abnormal propagation during sinus rhythm, identified by the highlighted yellow line, is unrelated to the area where diastolic activity was imaged.
Follow-up
At 12 months both cases described showed no recurrences of ventricular arrhythmias.

Discussion
Through these 2 clinical cases we describe our approach to catheter ablation in the setting of ARVC based on the following workflow: (1) substrate mapping during sinus rhythm to define reproducible areas of interest; (2) VT induction to rapidly identify the areas of the reentrant circuit (entry, isthmus, and exit); this constitutes a significant step forward from a pure sinus rhythm–based substrate modification, since it provides a direct link between sinus rhythm abnormalities and mid-diastolic activity during VT; (3) beyond the achievement of noninducibility we routinely strive to reach an original endpoint of electrical silence at the region where abnormalities have been mapped during VT.

ARVC frequently requires an epicardial approach. The disease is the result of progressive fibro-fatty infiltration of the myocardium proceeding from epicardium to endocardium layers. The modification of the electrical diseased substrate together with the identification of the clinical VT have provided higher long-term success rates (approximately 60%–80%). Endocardial bipolar EGM can fail to detect diseased areas in about 30% of ARVC patients and endocardial-only ablation has been shown to provide a modest freedom from VT recurrence. The finding of unipolar low-voltage recordings at the endocardium site, in the absence of bipolar EGM abnormalities, has been correlated to changes in the epicardial substrate.

A priority choice in every ARVC case is to go with an epicardial approach as the first-line approach. In keeping with this and with the literature, the first-line epicardial approach is standard at our institution. Once the epicardial access is gained, high-density sinus rhythm substrate...
mapping is performed to allow the identification of propagation abnormalities. Significant time and effort must be dedicated during the procedure to identify areas of low voltage and, more relevantly, abnormal potentials. The identification of areas of abnormal late activity is important because this will be the starting position for the VT activation map.

Arrhythmia intolerance and/or termination are reasons to develop a time-efficient strategy of VT mapping. The placement of a multielectrode mapping catheter at the location of maximal abnormal potentials is a starting point, even if there are some exceptions (and these are not rare), such as the second clinical case hereby presented; an

Figure 5  The entire diastolic pathway (A: entrance-isthmus; B: exit) is shown on the splines of the PentaRay catheter (Biosense Webster, Diamond Bar, CA).

Figure 6  Epicardial recordings at the proximal and distal tip of the ablation catheter. Ablation catheter is most likely to be at the isthmus/exit site of the ventricular tachycardia circuit.
ablation strategy based only on the abnormal substrate mapped during sinus rhythm would have failed to abolish the VT reentrant mechanism.

The chance to map even untolerated VTs is increased thanks to the advent of multielectrode mapping tool catheters. The multielectrode catheter is swept from its original position where LPs are recorded to adjacent areas to encompass the entire circuit. Using this mapping modality, the vast majority of short-lasting VT or untolerated VT can be mapped to an extent that provides the full disclosure of relevant areas involved in the reentry circuit. RF ablation can be subsequently delivered in cases of hemodynamically tolerated VT, aiming at the interruption. In cases of nontolerated VT, RF is delivered during sinus rhythm at the areas where the VT isthmus has been identified. Prevention of VT inducibility at the termination of the procedure is accepted as the optimal endpoint of VT induction. However, one must keep in mind that achievement of VT noninducibility status may occur as a consequence of the unreliable and probabilistic test nature of the electrophysiological study.

Figure 7 Documentation of ventricular tachycardia interruption during radiofrequency delivery (10 mm/s speed).

Figure 8 Epicardial maps with representation of the progressive abolition of late potentials (LPs) achieved during the case. A: The site of LPs before radiofrequency (RF) catheter ablation. B: Residual LPs after catheter ablation performed at the isthmus site, resulting in ventricular tachycardia (VT) termination. C: A re-map is then performed, which documents almost complete abolition of LPs. The window of interest is set to show signal from 40 to 80 msec (reference: QRS positive peak). Electrograms (EGMs) located inside the QRS are displayed with a red color (40–50 msec) while EGMs arriving after QRS end are progressively displayed as yellow, green, blue, and (latest) violet color. LAT = local activation time.
In fact, VT may be transiently noninducible for a variety of confounding reasons, which include altered autonomic tone, residual effect of anesthesia, and the inflammation and the transient tissue edema consequent to RF ablation, which may lead to noninducible VT and to the erroneous presumption that VT has been treated successfully.

**Two sides of the same coin or complementary information?**

The proof that areas of abnormal substrate previously identified as linked to the VT circuit are no longer operational during sinus rhythm (near-field electrical silence) provides an important endpoint. Similarly, in our prior study we have demonstrated that the strongest predictor of VT recurrence is the persistence of LPs. Therefore the achievement of complete electrical silence, at the site where the ventricular tachycardia has been electrically imaged, is mandatory and correlates with a better outcome.

**Conclusion**

The vast majority of VT in the setting of ARVC requires an epicardial approach; therefore general anesthesia should always be considered to allow epicardial puncture as the first strategy. Through these clinical cases we have shown our procedure workflow based on the identification of substrate abnormalities during sinus rhythm, linking this to the diastolic activity during VT. The activation
Figure 11  High-resolution bipolar voltage of the epicardial map (A) with late activity (B).

Figure 12  Electrogram recordings at the entrance (A) and isthmus-exit site (B).
map should always be attempted starting the mapping from areas of LPs recorded during sinus rhythm. A complete electrical imaging of the diastolic pathway with multielectrode catheters should always be amenable because precise identification of the isthmus site can be made. The aim of catheter ablation should always be to acutely interrupt the ventricular arrhythmia. Subsequently, a step forward is to prove the complete abolition of the near-field (late) potentials at the areas linked with the diastolic activity. LP areas may predict where the isthmus site could be but, not rarely, also be distant from the VT circuit.

Appendix
Supplementary data
Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2019.07.009.

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