

# Finding the right pathway is the key to success

Adam J. Graham, MRCP,<sup>\*</sup> Jonathan M. Behar, PhD, MRCP,<sup>\*</sup> Simon Sporton, MD, FRCP,<sup>\*</sup> Anthony Chow, MD, MRCP,<sup>\*</sup> Mehul Dhinoja, MD, MRCP, FHRS,<sup>\*</sup> Pier D. Lambiase, PhD, FRCP, FHRS<sup>\*†</sup>

From the <sup>\*</sup>Barts Heart Centre, Barts Health NHS Trust, London, United Kingdom, and <sup>†</sup>Institute of Cardiovascular Science, University College London, London, United Kingdom.

## Introduction

We present a case involving an initially unsuccessful ablation attempt of a right ventricular outflow tract (RVOT) ventricular tachycardia (VT) and the subsequent management that provided a definitive diagnosis and effective treatment. VT with left bundle branch block morphology and an inferior axis is often caused by focal tachycardia arising from the RVOT. The differential for this arrhythmia includes a macro-reentrant VT. This can be due to arrhythmogenic right ventricular cardiomyopathy (ARVC),<sup>1</sup> which is diagnosed using a combination of clinical cues and electrical and noninvasive imaging to meet the Task Force Criteria.<sup>2</sup> This case demonstrates the difficulties with diagnosis in the early sub-clinical phase of ARVC and illustrates the pathophysiology of reentrant VT.

## Case report

A 24-year-old woman was referred to a tertiary cardiac center with recurrent, hemodynamically tolerated broad complex tachycardia, consistent with RVOT-VT. She had been hospitalized on 10 occasions for symptomatic palpitations in the preceding 3 months, having undergone several direct current cardioversions. There was no family history of premature sudden cardiac death.

Prior investigations included a normal echocardiogram and 12-lead electrocardiogram (ECG). Cardiac magnetic resonance imaging (CMR) was normal with the exception of minor irregularities in the free wall of the right ventricle (RV). A 24-hour Holter recording revealed a high burden of ventricular ectopy (VE), which correlated with patient symptoms. A trial of medical therapy was undertaken, initially with bisoprolol, then concomitantly with diltiazem and subsequently sotalol monotherapy.

Previous elective catheter ablation of the VEs and RVOT VT was attempted on 2 occasions by identifying the site of

## KEY TEACHING POINTS

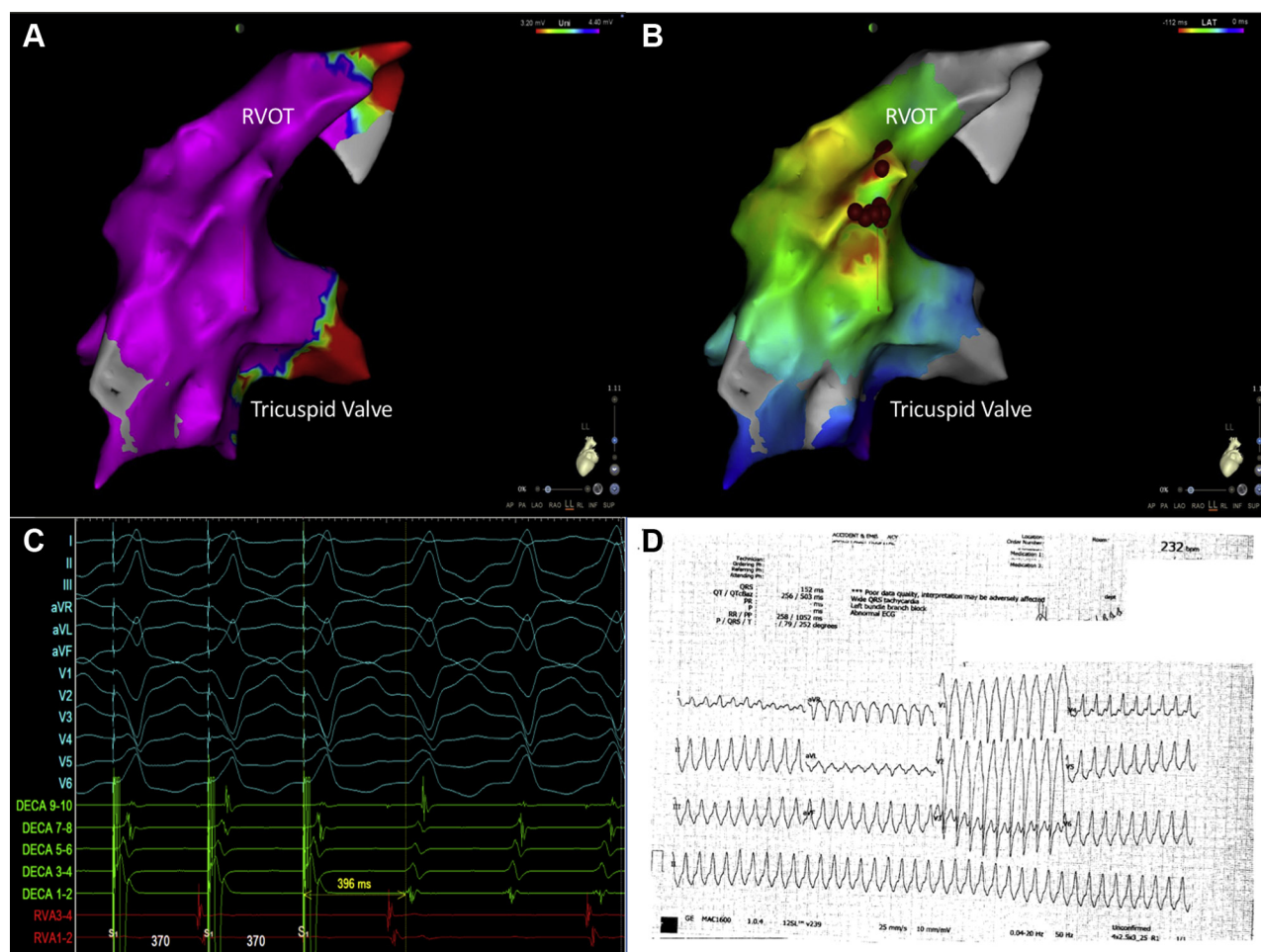
- A high index of suspicion for arrhythmogenic right ventricular cardiomyopathy is necessary even if Task Force Criteria are not initially met, and one should recognize that right ventricle repolarization abnormalities are not always fixed during the disease course.
- Extensive epicardial electrophysiological abnormalities can be present in the absence of endocardial correlates and pathology seen noninvasively with cardiac magnetic resonance imaging and echocardiography.
- Reentrant circuits can be fully delineated with detailed multipolar mapping.
- A failed endocardial approach should warrant early consideration of epicardial access.

earliest activation using 3-dimensional (3D) mapping (CARTO v3, Johnson & Johnson, New Brunswick, NJ). For both cases, the origin was mapped to the RVOT and ablation during VT terminated the tachycardia. On the second occasion programmed electrical stimulation was performed at the end of the case, but the clinical VT was still inducible. Of note, neither endocardial scar nor a diastolic pathway was seen during either study. The patient had further episodes of tachycardia and was referred to our center.

Given the frequency of her tachycardia, she was admitted from clinic and treatment with flecainide was initiated. Her 12-lead ECGs showed T-wave inversion (TWI) in V<sub>1</sub>–V<sub>3</sub>, which contrasted with normal T wave on prior ECGs. An exercise ECG demonstrated VT with similar morphology to the previous clinical events, with a slightly slower rate. The repolarization changes seen on the 12-lead ECG and the resistance of the arrhythmia to endocardial ablation prompted a reevaluation of the diagnosis. An electrophysiology study was scheduled with a high index of suspicion for an arrhythmogenic cardiomyopathy, given the borderline

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**Address reprint requests and correspondence:** Dr Pier D. Lambiase, Cardiology Department, Barts Heart Centre, Barts Health NHS Trust, West Smithfield, London, EC1A 7BE, UK. E-mail address: [p.lambiase@ucl.ac.uk](mailto:p.lambiase@ucl.ac.uk).



**Figure 1** **A:** Left lateral unipolar voltage map of right ventricle endocardium. **B:** Left lateral view of the endocardial activation map during ventricular tachycardia (VT). Ablation lesions are displayed on the right ventricular outflow tract septum with earliest activation in red. Note the diffuse endocardial early activation indicative of an epicardial-to-endocardial breakthrough. **C:** Intracardiac electrograms with surface electrocardiogram (ECG) showing broad complex tachycardia with left bundle branch block (LBBB) morphology with an inferior axis. **D:** A 12-lead surface ECG of VT on admission showing the same LBBB and inferior axis as during the case. RVOT = right ventricular outflow tract.

diagnosis comprised of 1 major (12-lead ECG TWI) and 1 minor criteria (VT with inferior axis).

VT was easily inducible at the start of the procedure with a 600-ms drive and S2 at 320 ms from the RV apex. The VT was consistent with the clinical VT previously demonstrated (Figure 1D). The case was performed with 3D mapping (CARTO v3). The RV endocardium was mapped using a decapolar catheter with the site of earliest activation in the septal portion of the RVOT, with no diastolic electrograms seen (Figure 1). Entrainment was performed with a postpacing interval of 26 ms (Figure 1C). Ablation in this area terminated VT after 20 seconds of radiofrequency (RF) energy delivery. Further consolidative adjacent lesions were delivered. Reinduction of broad complex tachycardia was possible using a 600-ms drive with S2 at 320 ms and S3 at 300 ms.

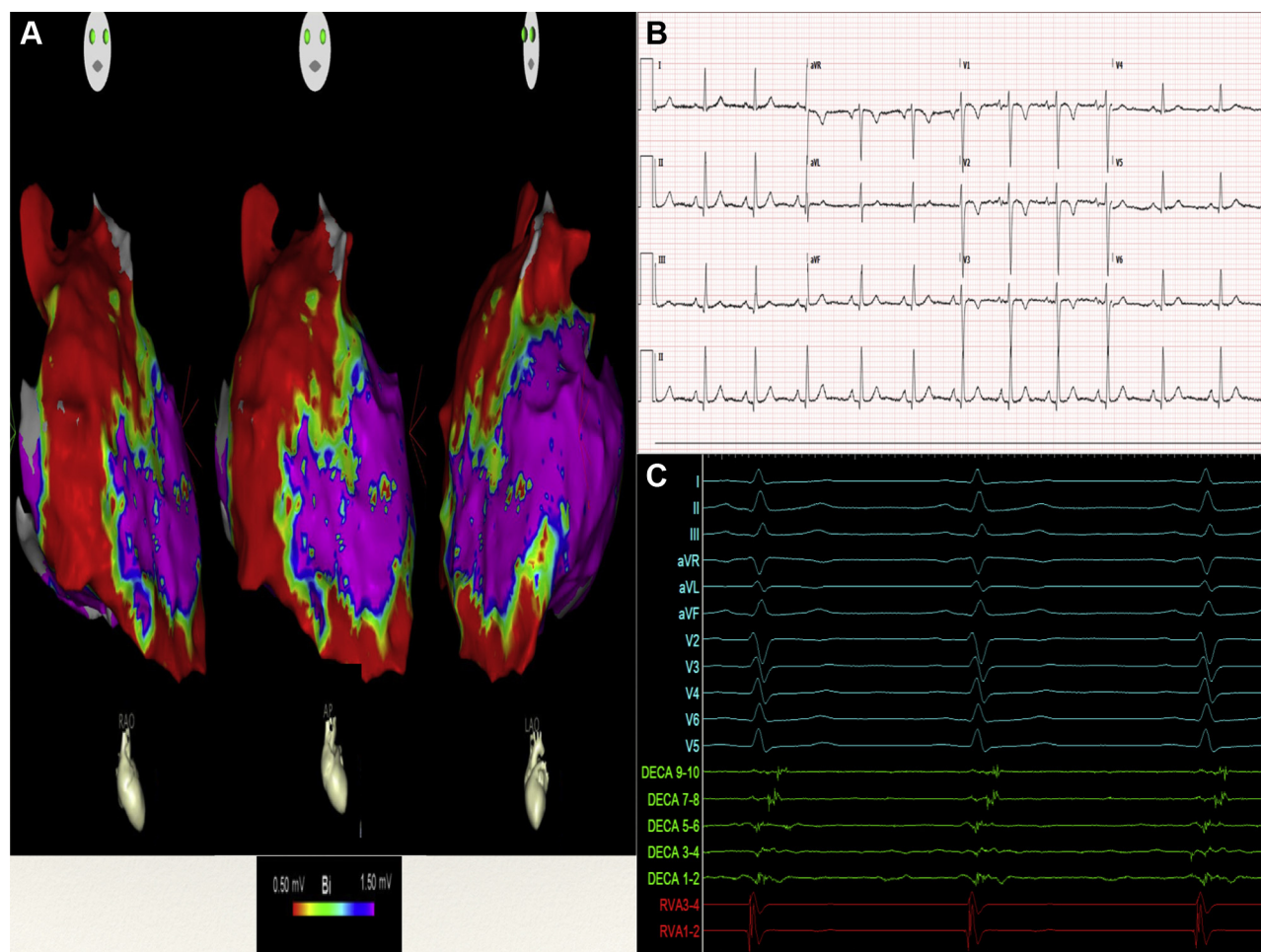
A decision was made to perform epicardial access and mapping; this was achieved via a subxiphisternal approach. VT spontaneously terminated during epicardial access. Figure 2A shows a sinus rhythm voltage map of the epicardial surface created using the decapolar catheter and

the fractionated electrograms that are often seen in ARVC cases (Figure 2C).<sup>3</sup> Figure 2B shows the 12-lead ECG during sinus rhythm with T-wave inversion in leads V<sub>1</sub>–V<sub>3</sub>. VT was reinduced with the aforementioned induction protocol and the entire diastolic pathway mapped with the decapolar catheter, as illustrated in Figure 3B. Figure 3A shows the activation map during VT.

Entrainment was attempted in the diastolic pathway; however, myocardial capture was not possible despite maximum output. Ablation in this region resulted in immediate termination of this tachycardia (Figure 3C). Further lesions were applied to this area and no further VT was inducible despite a full Wellens protocol. The patient was discharged on bisoprolol and has reported no further symptoms on follow-up in clinic 3 months post ablation.

## Discussion

This case illustrates key principles for management of VT in ARVC. The reentrant mechanism of scar-related VT, along



**Figure 2** A: Epicardial bipolar voltage map created during sinus rhythm (SR), with bipolar voltage setting at 0.5 mV to 1.5 mV. Maps are displayed in right anterior oblique (RAO), anterior posterior (AP), and left anterior oblique (LAO) from left to right. B: Resting 12-lead electrocardiogram showing T-wave inversion in V<sub>1</sub>–V<sub>3</sub>. C: Late and fractionated potentials seen recorded epicardially from the decapolar catheter during SR.

with the anatomic and functional substrate that supports it, has been well described.<sup>4</sup> However, it is rare to see this substrate clearly demonstrated in the intact human heart—especially given that the majority of VT induced during catheter ablation is unstable<sup>5</sup>—although with advances in electroanatomic mapping systems and the tools utilized, it is hoped that it will become more commonplace.<sup>6</sup>

The alignment of the decapolar catheter allowed us to anatomically demarcate the entire isthmus from its entrance, represented by the local electrograms on electrode poles 9–10 of the decapolar catheter (Figure 3B), to the exit site seen later in diastole on poles 3–4. The signal on 1–2 represents the far-field signal from global ventricular activation, with potentially a small near-field component at the onset of the far-field electrogram. Unfortunately, entrainment was not possible owing to lack of capture of the tissue underlying the catheter. This lack of capture is a common problem encountered when attempting to pace from scarred areas<sup>7</sup> and areas of epicardial fat.<sup>8</sup>

Further evidence that this site represents an essential component of the VT circuit is provided by the termination with epicardial ablation and the subsequent nonindu-

cibility of the tachycardia. Interestingly, termination also occurred during endocardial ablation. This could be on account of the RV wall being thin, leading to delivery of RF energy endocardially causing edema epicardially, or owing to endocardial components of the circuit being affected.<sup>8</sup>

Endo + epicardial vs endocardial-only VT ablation for ARVC has been shown to improve outcomes.<sup>9</sup> The diagnostic uncertainty that exists in cases such as this can delay attempts at potentially curative epicardial ablation and the implantation of an implantable cardioverter-defibrillator. The repolarization abnormalities detected on her 12-lead ECG on presentation at our center was a key factor in highlighting an ARVC diagnosis. TWI in the RV precordial leads has been shown to accurately differentiate ARVC from RVOT VT in the normal heart.<sup>10</sup> The fact that TWI was absent in the index ECGs made the diagnosis more difficult. Repolarization abnormalities can be transient and change over time in ARVC; hence multiple ECGs over time should be reviewed in suspected cases.<sup>11</sup> Additionally, in cases of diagnostic uncertainty an endomyocardial biopsy can be considered as a tool to confirm the diagnosis.<sup>12</sup>





**Figure 3** A: Anteroposterior view of activation map of ventricular tachycardia (VT) superimposed onto full epicardial geometry for orientation. Early and late activation are displayed in red and blue, respectively. The area of early activation shows the diastolic activation from the isthmus of the VT circuit. B: Surface electrocardiogram leads and epicardial bipolar electrograms recorded from a decapolar catheter on the isthmus during VT. Entrance to the isthmus can be seen on poles 9–10, with diastolic electrograms seen on subsequent poles following the pathway to the exit on 1–2. RVA catheter recordings are from the right ventricular apex. C: Termination within a few seconds of ablation on the area marked with Visitags on A.

In conclusion, this case demonstrates the distinct components of the epicardial VT circuit present in a patient with previously undiagnosed ARVC and clearly illustrates the important role of an epicardial ablation approach to define the substrate.

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