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CASE REPORT

INTERMEDIATE

CLINICAL CASE: SURGERY AND INTERVENTIONS

Successful Ablation of Sustained Monomorphic Ventricular Tachycardia in a Patient With Mitral Annular Disjunction

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ABSTRACT

Mitral annular disjunction (MAD) with or without mitral valve prolapse is associated with sudden death. Observed arrhythmias are usually ventricular ectopic beats originating from the papillary muscles. We describe a successful ablation of sustained monomorphic ventricular tachycardia from an epicardial focus in a patient with MAD. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:895-901) © 2022 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/).

A 53-year-old man presented with unprovoked syncope and multiple shocks from his implantable cardioverter-defibrillator (ICD). The result of physical examination was unremarkable.

MEDICAL HISTORY

He had been admitted to another hospital 4 months earlier with syncope. A diagnostic coronary angiogram was normal. An echocardiogram showed normal ejection fraction without mitral valve prolapse (MVP). Cardiac magnetic resonance (**Figure 1**) demonstrated mitral annular disjunction (MAD) at the posterobasal area of the left ventricle (LV) with abnormal wall motion but no late gadolinium enhancement (LGE).

Ventricular tachycardia (VT) was documented during that admission and an ICD was implanted. He underwent electrophysiological study with endocardial mapping, but ablation was not performed because of the lack of clear ablation targets. In particular, there was no endocardial scar or obvious focus during VT.

He presented at our institution 4 months later with recurrent ICD shocks.

LEARNING OBJECTIVES

- To recognize mitral annular disjunction with or without mitral valve prolapse as a cause of ventricular tachycardia and sudden cardiac death.
- To understand the mechanisms by which mitral annular disjunction can cause ventricular tachycardia.
- To review the role of catheter ablation in the management of these cases.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included VT storm, with the VT focus originating from the area of MAD.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

ICD = implantable cardioverter-defibrillator

LGE = late gadolinium enhancement

LV = left ventricle

MAD = mitral annular disjunction

MDI = maximum deflection index

MVP = mitral valve prolapse

SCD = sudden cardiac death

VA = ventricular arrhythmias

VEB = ventricular ectopic beat

VF = ventricular fibrillation

VT = ventricular tachycardia

INVESTIGATIONS

Interrogation of the ICD revealed that he had received 17 appropriate shocks after failed antitachycardia pacing from the device.

A 12-lead electrocardiogram (ECG) documented ventricular ectopic beats (VEBs) originating from the posterobasal area of the LV, which corresponded to the area of MAD seen on cardiac magnetic resonance (Figure 2). The morphology of the VEBs suggested an epicardial origin (Figure 3).

MANAGEMENT

An electrophysiological study with a view toward ablation was undertaken because of the patient's presentation with VT storm.

Epicardial access was obtained by use of an 18-gauge Tuohy needle from an anterior approach, and a short Agilis deflectable sheath was positioned in the pericardial space. Retrograde aortic access and transseptal access to the LV were also obtained.

Mapping was first performed over the epicardium by use of the CARTO electroanatomical mapping system with a Pentaray multielectrode catheter. There were low-voltage areas at the posterolateral base caused by epicardial fat, but no clear area of fractionation was seen. Likewise, no scar or abnormal Purkinje potentials were recorded endocardially. Pacemapping of the clinical VEB was performed from the endocardial LV posterobasal region with a morphology match of 84% (Figure 4).

Ablation was performed at this location by use of a 4.0-mm tip irrigated catheter. Radiofrequency energy at 40 W was delivered in a unipolar fashion with temperature and impedance monitoring, with an irrigation rate of 17 mL/min. After ablation at this location (of pacemap match), incessant monomorphic VT appeared, which was morphologically very similar to the clinical VEB (Figure 5).

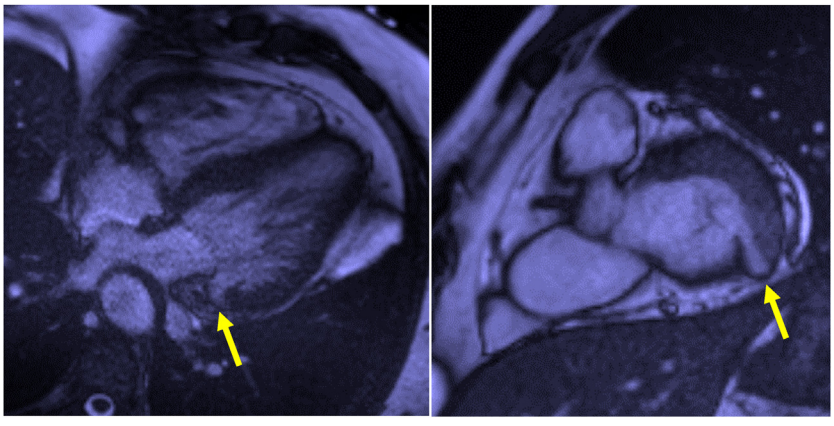
Mapping was performed during VT both endocardially and epicardially. The ventricular activation was noted to be earlier on the epicardium, where a small-fractionated signal during VT preceded the QRS by 30 ms (Figure 6). Entrainment was not possible because of the absence of capture with epicardial pacing. Ablation at this location (where the signal was recorded) at 40 W terminated the VT (Figure 7). Before ablation, coronary angiography was performed to ensure adequate distance between the ablation catheter and the coronary arteries.

DISCUSSION

MVP has been associated with sudden cardiac death (SCD).[1-5] Whereas the mechanism is not completely clear, it is thought to be strongly related to the presence of MAD.[1,6,7] MAD is defined as mitral annular detachment from the basal LV myocardium with an abnormal systolic excursion of the leaflet hinge point into the left atrium.[6,8,9]

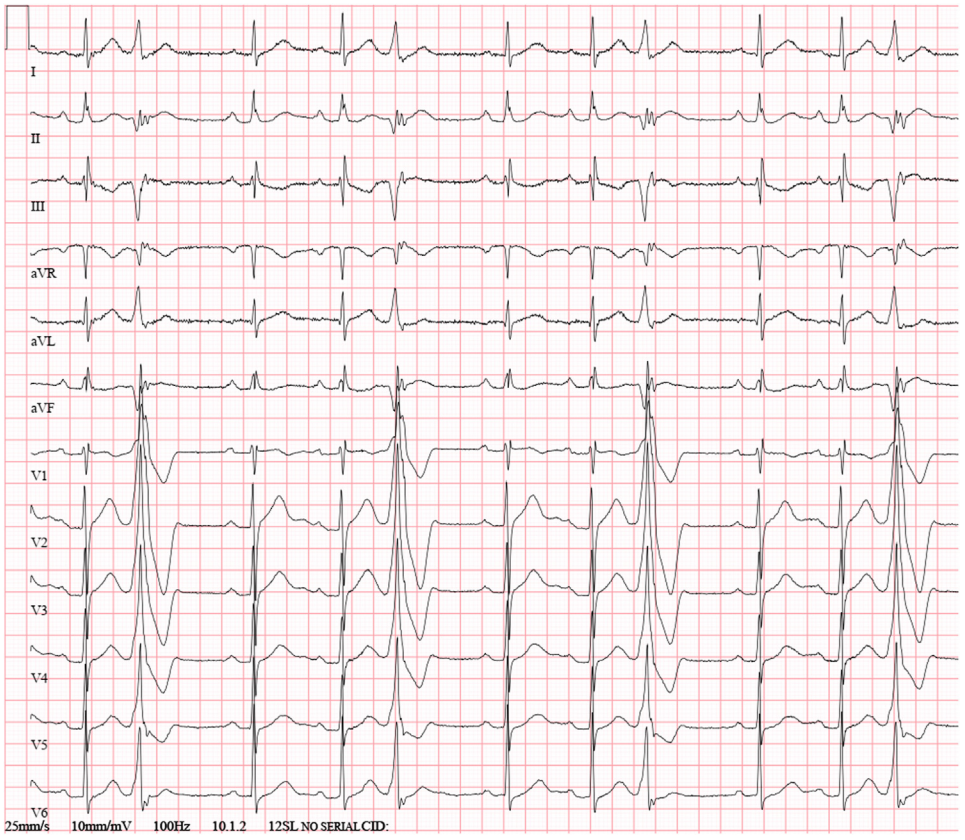
In a large study on MVP, the odds ratio of severe ventricular arrhythmias (VA), defined as VT ≥180 beats/min and/or VT/VF, was 6.97 for those with MAD.[1]

FIGURE 1 Cardiac Magnetic Resonance Image



Arrows indicate the area of mitral annular disjunction.

FIGURE 2 **Electrocardiogram of VEB Originating from the Posterobasal LV**



The ventricular ectopic beat (VEB) has a right bundle branch block morphology indicating its origin from the left ventricle (LV). All the QRS complexes for the VEB are positive in the precordial leads, consistent with a basal origin. Lead III is negative, and lead II is almost equiphasic. Thus, the VEB origin is posterior, with a direction of activation perpendicular to the vector of lead II.

Although first described in the context of MVP, where it occurs in one-third of patients,[10] MAD is observed also in the absence of MVP.[6,11,12] In the one-fifth of patients with MAD who do not have concomitant MVP,[11] the risk of SCD appears to be higher.[11] In that study, 34% of patients with MAD had VA, the occurrence of which correlated with longer MAD distance and the presence of papillary muscle fibrosis.[11]

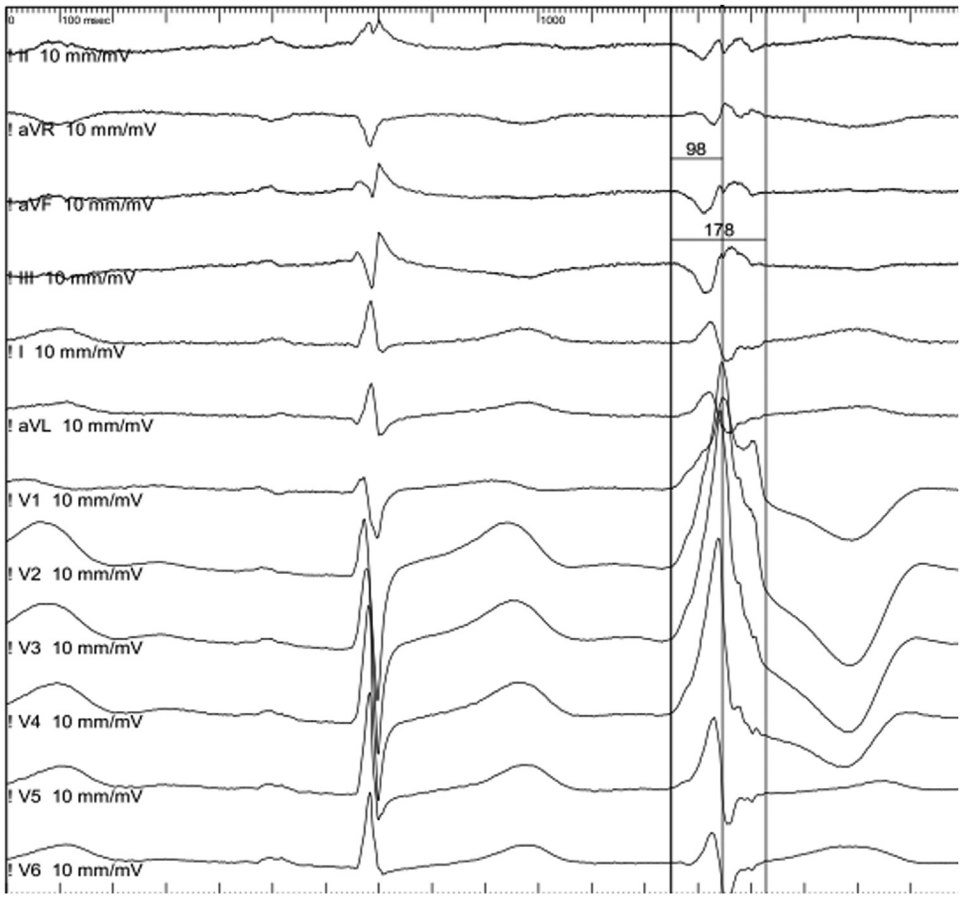
Catheter ablation in the setting of MAD has involved mainly VEBs in isolation or as triggers for ventricular fibrillation (VF).

Our case is unique because it is the first description, to our knowledge, of a clinical sustained monomorphic VT originating from the area of MAD and its successful elimination with catheter ablation. The likely mechanism of this VT was abnormal automaticity caused by diseased Purkinje fiber endings resulting from abnormal wall stress and ensuing fibrosis.

It is hypothesized that the abnormal motion due to MAD causes abnormal tension on the papillary muscles and the LV basal inferolateral myocardium,[13] which then leads to fibrosis that becomes the substrate for VA.[5] On cardiac magnetic resonance, late gadolinium enhancement (LGE), if present, was usually seen in the papillary muscles and was independently associated with VA.[14] However, many patients with VA did not exhibit LGE on cardiac magnetic resonance.[15] In these cases, myocardial fibrosis may have been too diffuse to be seen as LGE but may be suggested instead by reduced post-contrast T1 times on cardiac magnetic resonance,[16] and this finding has been associated with the occurrence of VA.

A trigger, usually a VEB from the papillary muscle or fascicle,[17,18] then initiates VA, resulting in cardiac arrest. Abnormal stretch of the papillary muscles, apart from causing fibrosis, lengthens the refractory period[19] and predisposes to ectopy. Purkinje fibers

FIGURE 3 MDI of 0.55 Suggesting an Epicardial Origin for the VEB



Maximum deflection index (MDI) of 0.55 suggesting an epicardial origin for the ventricular ectopic beat (VEB). The MDI is derived from the electrocardiogram, where MDI = TMD/QRSd. TMD = time from start of QRS to maximum deflection in any precordial lead; QRSd = total QRS duration. A MDI ratio of ≥0.55 suggests an epicardial origin.

are known to arborize into the papillary muscles and mitral annular area. In our case, the VT focus could have exited instead on the epicardial surface.

In a study of patients with MVP who underwent ablation for VF, the most common source of VEB trigger was from the Purkinje network near the papillary muscles and left fascicles.[20] Ablation resulted in VF storm in 1 patient, suggesting automaticity as the mechanism.[20]

FOLLOW-UP

The patient did not have further episodes of ventricular tachycardia after 3 months following the ablation.

CONCLUSIONS

Mitral annular disjunction can lead to ventricular arrhythmias, some of which are automatic and can be successfully eliminated with catheter ablation.

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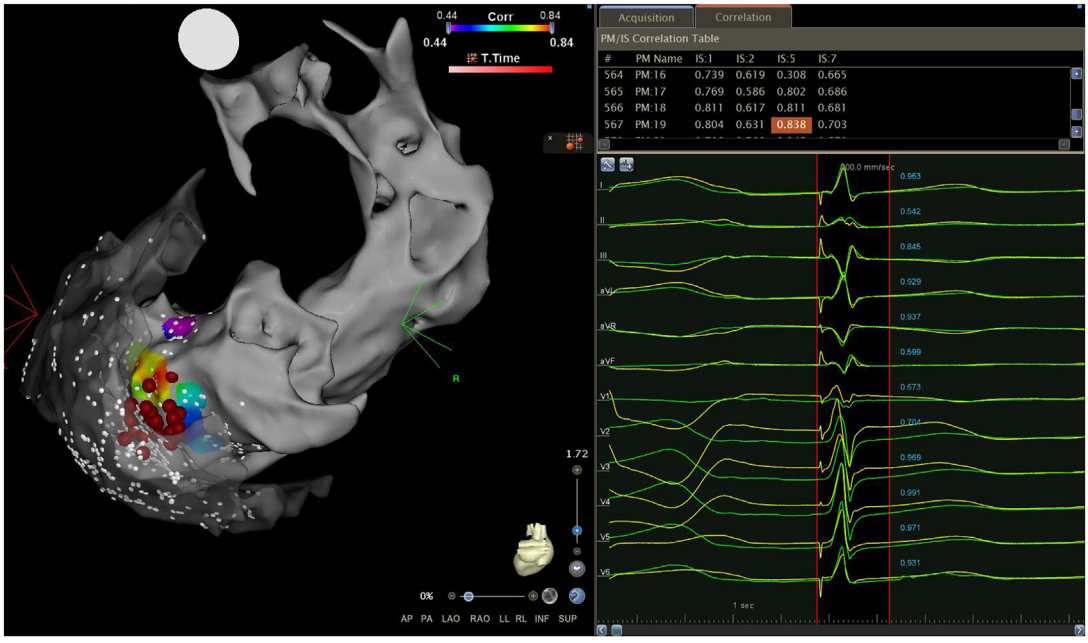
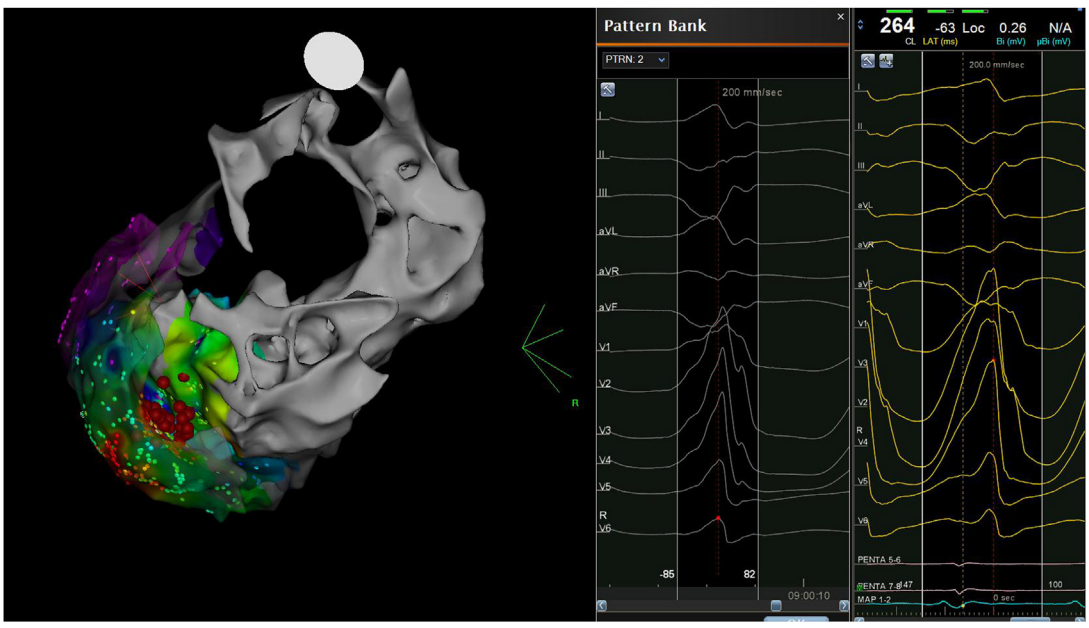
FIGURE 4 **Pacemap Morphology Match for VEB**



Color bar denotes degree of match with native ventricular ectopic beat (VEB).

FIGURE 5 **Electrocardiographic Morphology for VEB Versus VT**



(Left) Ventricular ectopic beat (VEB); **(right)** ventricular tachycardia (VT); 3D map showing location of the endocardial ablation spot for the VEB directly opposite the successful epicardial ablation location for the VT. **Color code** reflects activation timing during VT.

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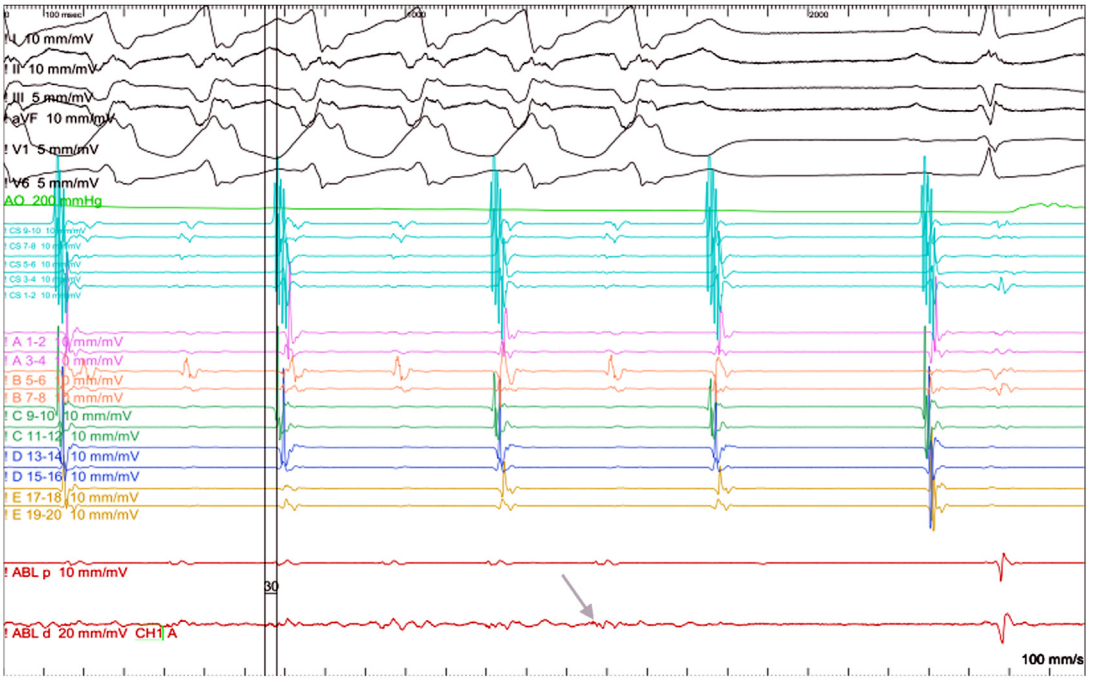
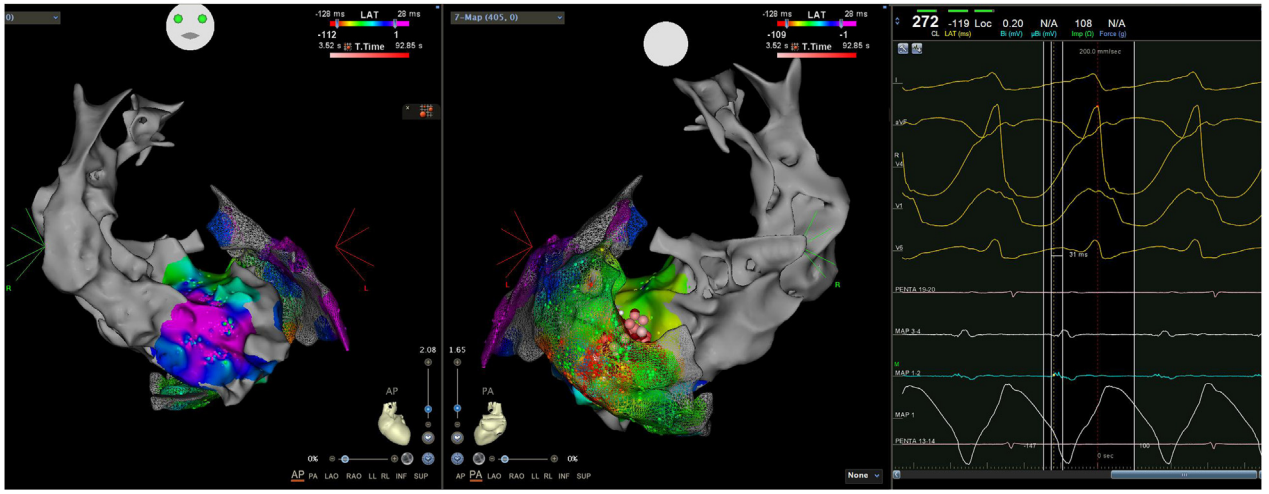
FIGURE 6 Intracardiac Electrograms During VT



Arrow shows low-amplitude sharp signal preceding QRS by 30 ms. VT = ventricular tachycardia.

FIGURE 7 Electroanatomical Map and Electrograms at Successful Ablation Site



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KEY WORDS **mitral annular disjunction, ventricular tachycardia**