

Bicuspid aortic valve supporting supra-ventricular “substrate” for multiple ventricular tachycardias



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Introduction

The great arteries are an important source of arrhythmias, including premature ventricular contractions, ventricular tachycardia (VT), atrial tachycardia, and accessory pathways.¹ The source of ventricular arrhythmias is thought to be muscular fibers extending from the outflow tracts to a variable extent above the valves.² Ventricular arrhythmias from the aortic cusps exhibit complex patterns of activation breakout attributed to preferential conduction along complex insulated myocardial fibers; complexity in regional anatomy also makes mapping and ablation challenging.^{3,4} We present a unique case of bicuspid aortic valve harboring the necessary “substrate” for multiple reentrant VTs above and below the aortic valve in a patient with ischemic cardiomyopathy whose VTs were unrelated to underlying infarct substrate.

Case Report

A 61-year-old man with prior inferior wall myocardial infarction, coronary bypass grafting, and ischemic cardiomyopathy (ejection fraction 40%) was referred for catheter ablation of sustained monomorphic VT after he experienced recurrent defibrillator shocks refractory to sotalol and amiodarone. He had undergone repair of a dilated ascending aorta at the time of his bypass grafting; bicuspid aortic valve was also present but without dysfunction, and hence not replaced at the time of surgery. Preprocedural stress imaging

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showed an inferior myocardial infarct without ischemia; transthoracic echocardiography showed absence of bicuspid valvular stenosis or regurgitation, and coronary angiography showed patent grafts.

In the electrophysiology laboratory, 2 VTs were repeatedly inducible with programmed ventricular stimulation and with catheter manipulation within the fused cusps (Figure 1A). VT1 was right bundle morphology in lead V₁, inferior axis, with a cycle length of 400 ms, and was consistent with the spontaneous clinical VT. VT2 had a QR morphology in lead V₁, right inferior axis, with a cycle length of 465 ms, and was not seen spontaneously. The left ventricle and aortic root were mapped retrogradely using an irrigated catheter (Thermocool SurroundFlow; Biosense Webster, Diamond Bar, CA) guided by intracardiac echocardiography integrated into the CARTO mapping system (Biosense Webster). The left and right coronary cusps were fused (Figure 1B, Movie 1, available online). A preponderance of late potentials were noted in sinus rhythm within the fused left-right cusp, especially at the commissure, where pace mapping produced near-perfect matches for both VT1 and VT2 with long stimulus to QRS delays (>80 ms) (Figure 1C, D). Coronary angiography showed absence of coronary anomalies and proposed site of ablation was >1 cm from the left main ostium (Figure 1E). During induction of VT1, a long-duration fractionated electrogram was noted within the common cusp, with unipolar signals showing a QS pattern and a sharp downward deflection (Figure 1F). Radiofrequency ablation at this site terminated VT1 and rendered both VTs noninducible (Figure 1G). Additional ablation lesions were delivered in the adjacent region within the sinuses of the fused cusps at sites that captured with pacing (10 mA, 2 ms), rendering the region electrically inexcitable with pacing and abolishing the late potentials within the commissure. The subvalvular region was also mapped, showing a small periaortic low-voltage region consistent with scar; however, pace maps at these sites did not resemble either VT1 or VT2. Some empiric ablation was performed at sites that capture with long stimulus to QRS delays. However, as the best pace maps and late potentials were seen in the supra-ventricular region, ablation was focused in this area. Repeat Doppler echocardiography of the aortic

KEY TEACHING POINTS

- The aortic cusps or sinuses are an important source of ventricular arrhythmias. The origin is attributed to muscular fibers extending from the outflow tracts to a variable extent above the valves.
- This case report shows that fused aortic commissures in a bicuspid aortic valve were also capable of providing the necessary to form the “supravalvular substrate” for multiple reentrant sustained monomorphic ventricular tachycardias, akin to that reported to occur in otherwise normal trileaflet valves. The mechanism is also likely mediated by muscular extensions from the left ventricular outflow tracts above the valve.
- Additionally, such ventricular tachycardias in this patient with ischemic cardiomyopathy were unrelated to inferior wall infarct substrate.

valve showed no evidence of regurgitation. The patient had recurrence of VT1 1 month later, which was successfully ablated in the periaortic left ventricular outflow tract immediately below the site of ablation in the cusps in the prior procedure. At this site, attempts at entrainment of VT prior to ablation demonstrated termination without global capture (third beat, as shown by arrow in [Figure 1H](#); green dot representing this site shown in [Figure 1I](#)). The patient remained free of VT at 1-year follow-up.

Discussion

This case illustrates, firstly, that the fused aortic commissures in bicuspid aortic valves are also capable of providing the necessary “supravalvular substrate” for multiple reentrant sustained monomorphic VTs, akin to that reported to occur in otherwise normal trileaflet valves. The mechanism is likely muscular extension above the valve cusps from the left ventricular outflow tract. Second, patients with ischemic cardiomyopathy may develop ventricular arrhythmia because of substrate unrelated to the prior infarction.

The aortic cusps are well recognized as a source of premature ventricular contractions and VT.^{1,5} Sleeves of myocardium commonly extend for a variable length above the semilunar valves to the great arteries.³ Extension is more common in the right (54% in 1 autopsy study) than the left cusp (24%).² Lack of inhibitive electrotonic interactions from surrounding and remote cardiomyocytes owing to scarcity of muscle bundles above the semilunar valves may promote ventricular ectopy.^{6,7} Intervening regions of fibrosis, for example, in bicuspid valves may further inhibit this electrotonic interaction. Prior reports of VT arising from within the cusps and immediately below the valve suggest

the presence of substrate capable of reentry in and around the aortic commissures.^{4,8}

This is the first reported case of supravalvular “substrate” supported by the presence of a bicuspid aortic valve with fused left and right commissures, likely mediated by muscular extensions from the left ventricular outflow tract above the fused commissure. We found a preponderance of late potentials within the common cusp, where pacing captured with long stimulus to QRS delays provided evidence for slow conduction and produced near-identical pace map matches to the induced VT. VTs were reliably inducible and terminated with programmed stimulation; and ablation terminated >1 VT at this site. Moreover, low-amplitude fractionated electrograms were present during induced VT with ablation at these sites terminating one VT, and rendering them both noninducible.⁹ In addition, termination without global capture during the second procedure suggested that the region immediately below the valve in the outflow tract was also a critical area of the likely reentry circuit, as has been previously shown in patients with prior coronary disease.¹⁰ Although limited empiric ablation was performed in the subvalvular region in the first procedure, it is unclear if extensive ablation through the low-voltage periaortic region would have avoided a subsequent procedure by eliminating all potential exits of the VTs. It is plausible that generous supravalvular myocardial extension from fusion of the right and left cusps where myocardial extension is the most prominent, when combined with the enhanced fibrosis commonly seen in fused cusps, promoted the development of substrate for reentry in this patient. Pacing at 1 site in the fused commissures replicated 2 different pace map morphologies of VT. This is consistent with the finding that multiple exits of VT are possibly attributed to preferential conduction along myocardial fibers traveling to surrounding structures, such as the right ventricular outflow tract, epicardial left ventricular summit, great cardiac vein, and infravalvular left ventricular outflow tract, which often makes pace mapping unreliable in the supravalvular left ventricular outflow tract.¹¹

Lastly, the presence of outflow tract arrhythmia in this patient also illustrates that not all ventricular arrhythmias may arise from anticipated regions of substrate provided by prior infarction.¹² In 1 study of patients with coronary artery disease and VT, regions of abnormal voltage giving rise to VT had a basal perivalvular distribution or were epicardially remote from anticipated infarction sites, suggesting the presence of concurrent nonischemic substrate for VT.¹²

In conclusion, this case provides electrophysiologic data and anatomic imaging to illustrate the presence of substrate above and below the valve for multiple VTs in a unique patient with a fused bicuspid valve, and highlights the importance of recognizing that ventricular arrhythmias may arise from regions unrelated to anticipated substrate of prior infarction in patients with ischemic cardiomyopathy.

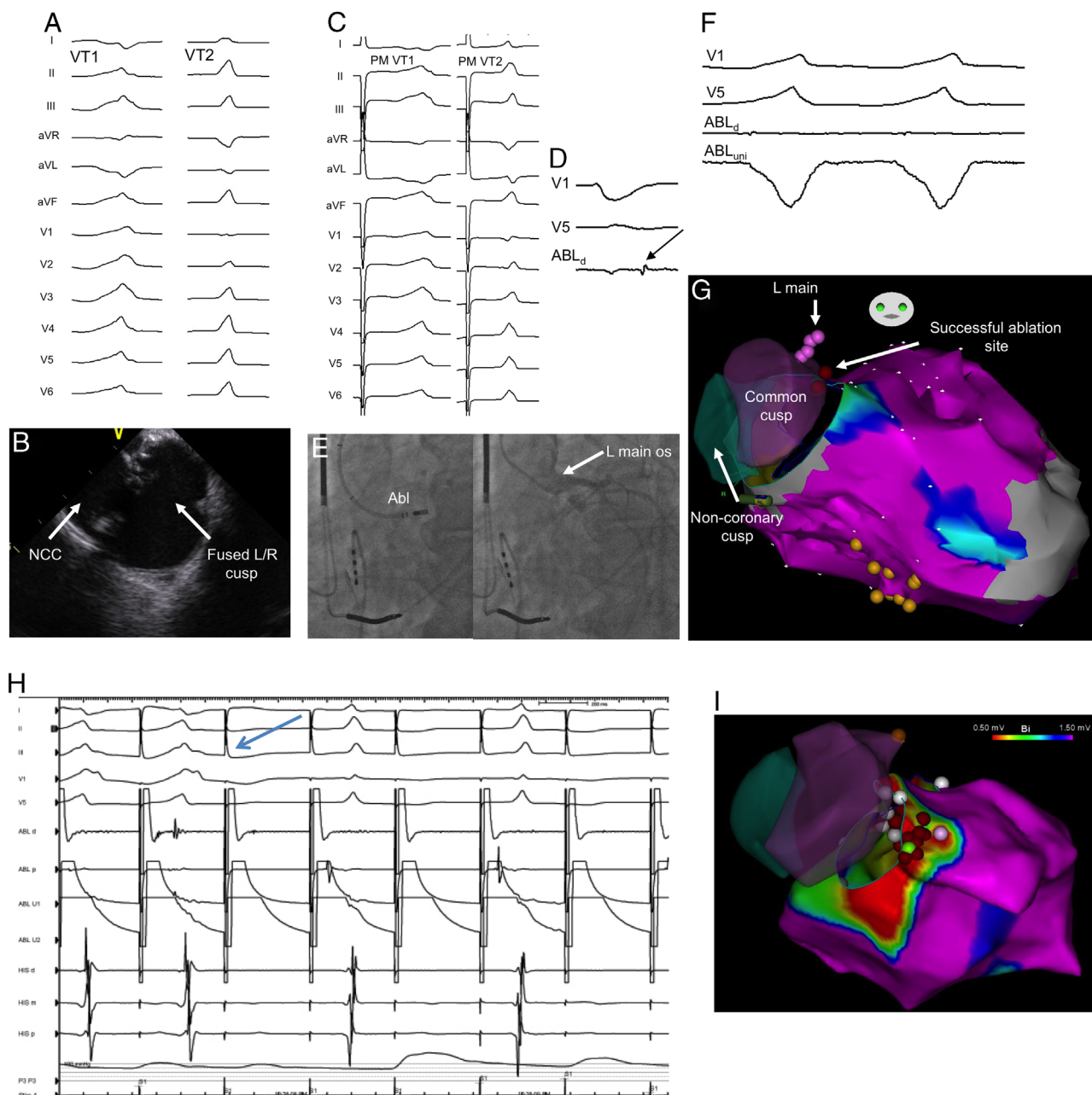


Figure 1 Two ventricular tachycardias (VTs) were repeatedly inducible with programmed ventricular stimulation and with catheter manipulation within the fused cusps. A: Induced VT1 and VT2. B: Fused left and right commissures on intracardiac echocardiography. C: Excellent pace matches for both VTs, with long stimulus to QRS delay at sites with late potentials (D) in the left-right fused bicuspid commissure > 1 cm below the left main coronary ostium (E), where long-duration fractionated signal with a unipolar QS was seen during VT 1 (F, G). VT recurred 1 month later and entrainment attempt at the periaortic left ventricular outflow tract below the site of previous ablation showed termination without global capture (arrow, H; site in green in I). Despite a prior inferior wall infarction, there was periaortic low voltage with minimal inferior wall scar, more consistent with a nonischemic distribution of scar.

**Appendix
Supplementary data**

Supplementary data associated with this article can be found in the online version at <http://dx.doi.org/10.1016/j.hrcre.2016.09.006>.

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