Inappropriate defibrillator shock due to fragmented potentials derived from an extensively diseased right ventricle in a patient with arrhythmogenic right ventricular cardiomyopathy

Kentaro Goto, MD,* Shinsuke Miyazaki, MD,* Keita Handa, MD,† Kazuo Kobayashi, MD,† Masahiko Goya, MD,* Tetsuo Sasano, MD*

*From the *Department of Cardiovascular Medicine, Tokyo Medical and Dental University, Tokyo, Japan, and †Division of Cardiology, Kashiwa City Hospital, Chiba, Japan.*

Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a cardiac inherited disease characterized by the presence of right ventricle (RV) structural abnormalities and fatal arrhythmias arising from an abnormal RV, which can lead to sudden cardiac death.[1–3] Implantable cardioverter-defibrillator (ICD) therapy not only enables an effective prevention of sudden cardiac death related to lethal arrhythmias but is also associated with the risk of device-related complications, including device infections, inappropriate shocks, loss of ventricular sensing, and other lead-related complications. The importance of reducing the complications associated with ICDs in patients with inherited arrhythmias should be noted because such patients often include young adults who need long therapy periods after the implantation.

With respect to the inappropriate shocks, intelligent device programming, including long detection intervals and morphology-based discrimination algorithms, has improved the accuracy of the tachycardia discrimination and has succeeded in decreasing the rate of inappropriate shocks over the past 2 decades[4,5]; however, according to the reported studies, inappropriate shocks have an annual incidence rate of 3.7%–3.9% and an overall incidence rate of 19%–27.7% among patients with AVRC who underwent ICD implantations.[1–3,6–11] The detailed mechanisms of the inappropriate shocks in that population have not been well elucidated. Here, we report a case of inappropriate shock in a patient with ARVC, in which the QRS morphology–based discrimination algorithm did not work effectively owing to the presence of fragmented potentials derived from an extensively diseased RV.

KEY TEACHING POINTS

- Physicians need to be versed in the discrimination algorithms of implantable cardioverter-defibrillators.
- A careful evaluation of the intracardiac QRS waveforms is recommended to improve the accuracy of the QRS morphology–based tachycardia discrimination algorithm, especially in arrhythmogenic right ventricular cardiomyopathy (ARVC) patients.
- Future studies would aim to determine the accuracy of the QRS morphology–based tachycardia discrimination algorithm in patients with ARVC.

Case report

A 72-year-old man was brought to the emergency room with an acute onset of substernal chest discomfort and presyncope. Electrocardiography revealed a wide QRS regular tachycardia with a left bundle branch block configuration and inferior axis, suggesting sustained ventricular tachycardia (VT) arising from the RV outflow tract. We performed cardioversion and succeeded in restoring a sinus rhythm, seen as epsilon waves in leads V_1–V_2 and inverted T waves in leads V_1–V_4 on the 12-lead electrogram (Supplemental Figure 1). Transthoracic echocardiography and cardiac magnetic resonance imaging revealed RV enlargement, an aneurysm in the inferior RV, and late gadolinium enhancement in the mid layer of the entire RV and a part of the left ventricular septum. Based on these findings, the patient was diagnosed with ARVC.

KEYWORDS Arrhythmogenic right ventricular cardiomyopathy; Implantable cardioverter-defibrillator; QRS morphology–based analysis; Ventricular tachycardia; Inappropriate shock

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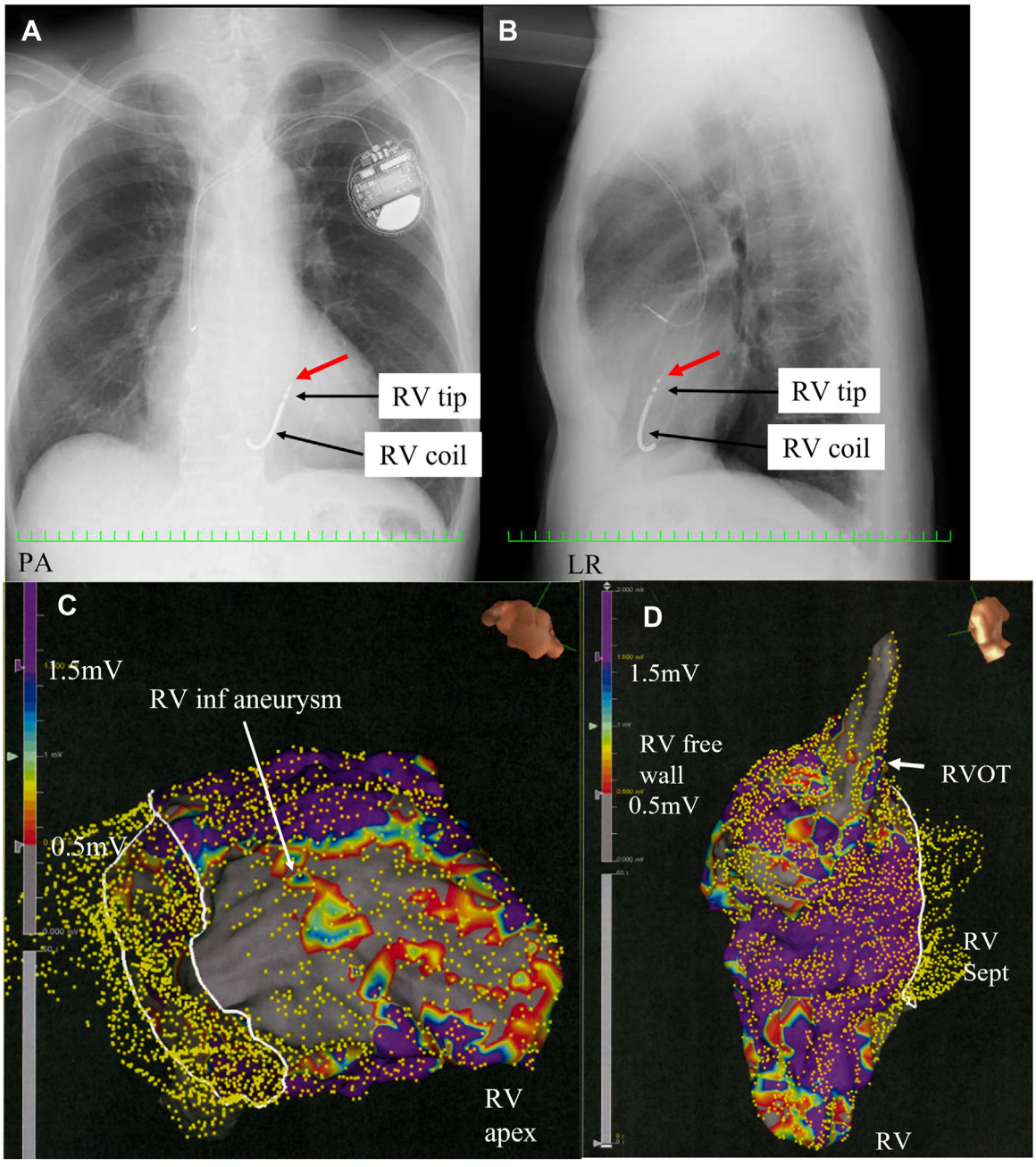

Figure 1 **A:** Chest radiograph (posterior-anterior [PA] view) after the implantable cardioverter-defibrillator (ICD) implantation. The right ventricle (RV) lead was positioned on the high RV septum (*red arrow*) because of the extensive low-voltage area at the apex and inferior wall. **B:** Chest radiograph (left-right [LR] view) after the ICD implantation. **C:** A voltage map of the RV (inferior view). An extensive low-voltage area (<1.5 mV: *gray zone*) was observed on the RV inferior wall and apex. **D:** A voltage map of the RV (superior view). An extensive low-voltage area (<1.5 mV: *gray zone*) was observed on the RV apex and RV outflow tract (RVOT). Inf = inferior; RV sept = right ventricle septum.

During an electrophysiological study, we found an extensive low-voltage area (<1.5 mV) with fragmented potentials on the inferior wall, outflow tract, and apex of the RV. The clinical VT was easily inducible and successfully eliminated by radiofrequency applications in the RV outflow tract area. Subsequently, a transvenous ICD (Cobalt XT DR DDPA2D4; Medtronic, Minneapolis, MN) was implanted. We first tried to place the RV lead around the RV apex, but the voltage was too low to confirm sensing. The RV lead (6935M Sprint Quattro MRI; Medtronic) was finally positioned on the high RV septum owing to the presence of an extensive low-voltage area in the inferior RV (Figure 1). This device categorized the tachycardia event using the "onset," which recognized acute accelerations of the V-V interval as ventricular events, and "stability," which checked the regularity of the atrial and ventricular activation cycle. Then, the other algorithms reinforced the ventricular arrhythmia detection. The PR logic (Medtronic), which discriminates tachycardias based on an atrial and ventricular activation timing categorization, and wavelet analysis

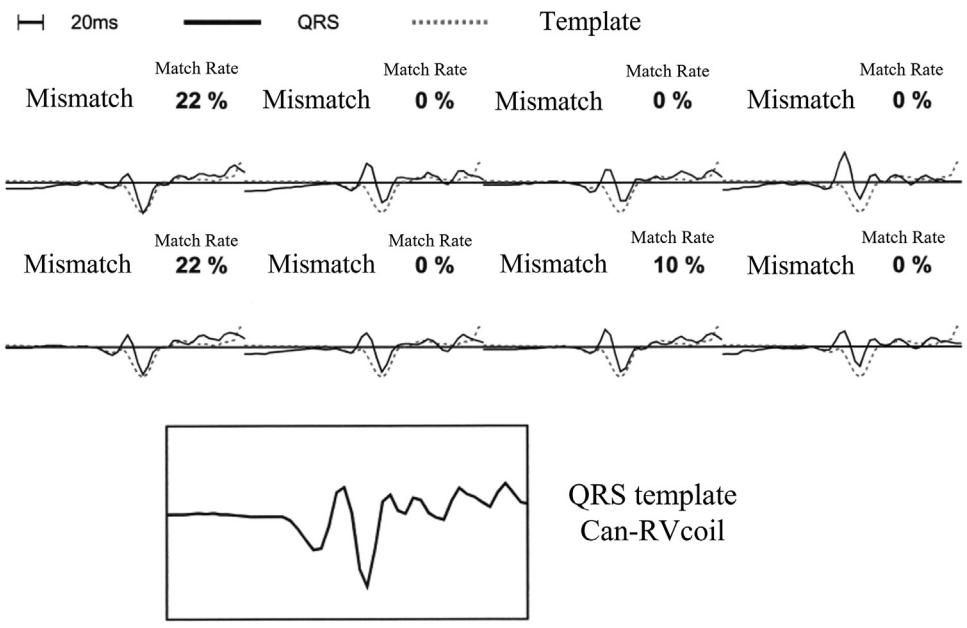

Figure 2 In the wavelet analysis, the QRS morphology match rate between the tachycardia and sinus rhythm was 0/8 (0%), all "unmatched," and the therapy was conducted. RV = right ventricle.

(Medtronic) based on the QRS morphology similarity scoring, were also used. Three zones (ventricular fibrillation zone >200 beats/min, fast VT zone 171–200 beats/min, and VT zone 150–200 beats/min) were programmed, and all were treated with intrinsic antitachycardia pacing first, followed by cardioversions.

Three months after the ICD implantation, the patient was brought to the emergency room with an inappropriate shock episode. The tachycardia episode was in the fast VT zone and was terminated by cardioversion with 40 J after ineffective intrinsic antitachycardia pacing therapy. The analysis of the intracardiac electrograms during the tachycardia revealed that (1) the preceding A-A interval determined the following V-V interval, and (2) a V-A-A-V response was observed after the antitachycardia pacing therapy, suggesting that the tachycardia was an atrial tachycardia (AT) (Supplemental Figure 2). With the discrimination algorithms, the PR logic failed to discriminate VT and AT owing to the sudden onset and 1:1 atrioventricular conduction of the tachycardia. The QRS morphology match rate of the wavelet analysis during the tachycardia was also 0/8 (0%) (Figure 2).

At this point, we noticed that the QRS waveform sensed between the can and RV coil, which was used for the wavelet analysis template, presented an abnormal and low voltage with fragmented potentials. We updated the sinus rhythm template of the wavelet analysis, but the match rate between the memorized sinus rhythm template and sinus rhythm at present was relatively low (58%–76%), even immediately after the template update (Figure 3). We changed the sensing source between the can and the RV tip, but the same phenomenon was observed. Since the abnormal QRS waveform likely reflected the electrical abnormalities of the RV, the discrimination between VT and AT seemed to be difficult with the wavelet analysis. We prescribed 100 mg/day of oral amiodarone to suppress the AT and no inappropriate shocks have occurred since then.

Discussion

ICD treatment in ARVC patients

Progressive RV structural abnormalities are a characteristic of ARVC. The efficacy of an ICD treatment in ARVC patients has been reported in many previous studies; however, it is often challenging to find an appropriate RV lead position owing to the extensive diseased RV.[1–3,6–11] The reported predictors of appropriate shocks in ARVC patients include atrial fibrillation, syncope, prior ventricular arrhythmias, male sex, and young adults, and the reported appropriate shock rate varies greatly (9.5%–10.2%/year, or a total of 24%–74%).[1–3,6–11] On the contrary, device-related complications occur in 17%–33% of ARVC patients, and inappropriate shocks occur in 3.7%–3.9%/year, with a total incidence of 19%–27.7%.[1–3,6–11] In particular, younger patients experience inappropriate shocks more frequently than the elderly,[10] and sinus tachycardia and supraventricular tachycardia are the main causes of inappropriate shocks, not oversensing owing to lead malfunctions.[9,11] Intelligent device programming with long detection intervals and other tachycardia discrimination algorithms (PR logic and wavelet analyses) have improved the accuracy of tachycardia discrimination and succeeded in decreasing the rate of inappropriate shocks even in ARVC patients. However, there are still certain cases with unavoidable inappropriate shocks caused by sinus tachycardia and atrial arrhythmias, and there are very few reports on the mechanisms of discrimination failure.

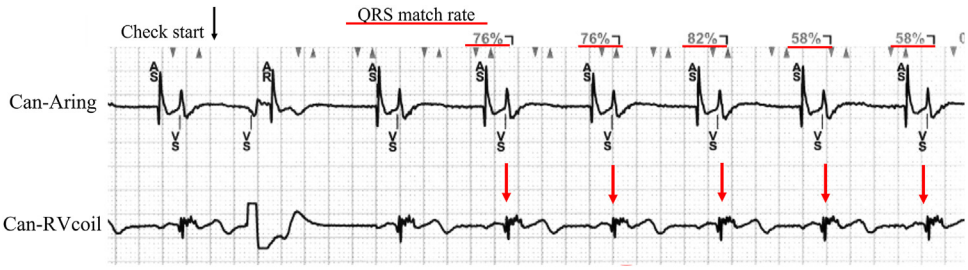

Figure 3 After the inappropriate shock episode, we updated the template of the wavelet analysis. However, the match rate between the updated sinus rhythm template and sinus rhythm at present were relatively low (58%–76%) even immediately after the sinus rhythm template update owing to the abnormal QRS waveform (*red arrows*). A ring = the ring of atrial lead; RV = right ventricle.

The discrimination algorithm of ICDs

PR logic has been reported to be an effective tool that uses the atrial and ventricular timing to discriminate sinus tachycardia and ATs from VTs.[12] To distinguish sinus tachycardia from VTs, the device evaluates 3 points: (1) a 1:1 atrioventricular conduction; (2) smaller-than-expected change in the R-R interval rate; and (3) smaller-than-expected change in the rate of the PR interval. If the tachycardia met all 3 conditions, the device recognized it as sinus tachycardia and waited for the therapy. However, it is fundamentally difficult to discriminate AT and VTs with 1:1 retrograde ventricular-atrial conduction based on the atrial/ventricular activation timing and sudden onset of the tachycardia. Therefore, an additional QRS morphology–based algorithm is necessary for the discrimination.

In the wavelet analysis, the device compared the QRS morphology between that during the tachycardia and that during the sinus rhythm, then scored the similarity. This algorithm decomposes and reconstructs the QRS morphology digitally using the Haar (square) wavelet transform. The match-percent score is based on the differences in the corresponding coefficients of the wavelet transforms.[13] If the similarity score was >70%, the QRS morphology was categorized as a "matched" wave; and if at least 3 beats of any 8 consecutive beats matched the template, the device waited for the therapy. The wavelet analysis should be effective even for ATs with 1:1 atrioventricular conduction.[5] However, in this case, the updated sinus rhythm template did not match the sinus rhythm morphology even immediately after the template update and did not work effectively. The fine fragmented QRS morphology related to severe electrical abnormalities in the RV likely caused this phenomenon during the process of the waveform decomposition and reconstruction.

To date, to the best of our knowledge, there have been no reports investigating the accuracy of QRS morphology–based algorithms in ARVC patients. A prior meta-analysis[3] revealed that the overall incidence rate of inappropriate shocks caused by atrial arrhythmias in patients with ARVC was 7.3%, which was similar to that for the other inherited arrhythmia syndromes; however, the mechanisms of the misrecognition were not analyzed. Whether the phenomenon observed in this case is unique to patients with an abnormal RV or whether patients with an abnormal left ventricle also exhibit the same phenomenon remains unresolved.

How to avoid inappropriate shocks in ARVC patients

In the present case, we positioned the RV lead on the septal wall; thus the RV coil was positioned near the RV inferior wall with an extensive low-voltage area (Figure 1). This might have resulted in the abnormal fragmented QRS morphology sensed between the can and RV coil. To the best of our knowledge, the appropriate lead location for a better discrimination has not been reported in ARVC patients.

Regarding the QRS morphology–based algorithm, we assumed that the intracardiac QRS wave morphology should be evaluated during the ICD implantation procedure before the final decision of the RV lead position. In ARVC cases, a voltage map might be helpful to decide the optimal RV lead location; however, it is still important to check the QRS waveform sensed between the can and RV coil during the implantation. In addition, given the progressive nature of ARVC, it seems to be reasonable to reevaluate the intracardiac QRS morphology during routine device check-ups, as it could change with the disease progression during the remote period.

Conclusion

Here, we reported a mechanism of an inappropriate ICD therapy in an ARVC patient. The physicians need to be versed in the discrimination algorithms of ICDs, and a careful evaluation of the intracardiac QRS waveforms is recommended to improve the accuracy of the QRS morphology–based tachycardia discrimination algorithm, especially in ARVC patients.

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Appendix

Supplementary data

Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2022.07.009.

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