

# Failure of a cardiac resynchronization therapy-defibrillator sensing lead to correctly detect a fatal arrhythmia because of a newly emerged intraventricular conduction abnormality induced by cardiac sarcoidosis

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## ABSTRACT

Here, we report the case of a 59-year-old man with cardiac sarcoidosis (CS) who experienced the failure of a cardiac resynchronization therapy-defibrillator (CRT-D) because the sensing lead failed to operate correctly. The sensing lead, which was inserted into the right ventricular outflow tract (RVOT), failed because of a newly emerged intraventricular conduction abnormality (IVCA) resulting from the progression of the CS). For determining the cause of the failure, an electrophysiological study was conducted with catheters placed to the right ventricle apex (RVA) and RVOT. The IVCA was not seen during rapid pacing at 180 bpm but was observed during rapid pacing at over 190 bpm; however, this phenomenon may also develop as a result of the restitution property of conduction velocity due to the relative ventricular refractory period, even in the absence of CS. A voltage map recorded by the CARTO mapping system revealed a markedly low voltage area between the RVA and RVOT. Therefore, we assumed the heart rate for the ventricular tachyarrhythmia was underestimated by the CRT-D sensing lead and had thereby led to its operation failure. An appropriate intervention was accomplished by inserting an additional sensing lead to the RVA, and the same phenomenon has not occurred to date. To our knowledge, no similar report has previously been published, and thus makes this case an extremely rare and didactic case.

**Key words:** cardiac sarcoidosis, CRT-D, under-count, ventricular tachyarrhythmia.

## INTRODUCTION

The cardiac resynchronization therapy-defibrillator (CRT-D) and implantable cardioverter defibrillator (ICD) for ventricular tachyarrhythmia (VT) and ventricular fibrillation (VF) have tremendously contributed to a better prognosis.<sup>1</sup> However, CRT-Ds and ICDs occasionally fail to respond to life-threatening arrhythmias in clinical situations. Of importance, the inadequate detection and treatment of the causes for the sensing failure can result in catastrophic consequences, including death. Hence, a full awareness of the causes and proper treatment of the

CRT-D operation failure is vital.

We report here a case in which the progression of an underlying cardiac condition caused a new intra-cardiac electrical barrier and resulted in the failure of the sensing lead. To our knowledge, no similar report has been published, thus making this case an extremely rare and didactic case.

## CASE REPORT

A 59-year-old Japanese man had a history of admission for heart failure occurring twice in the period of a year. Coronary angiography revealed a normal coronary artery, while cardiac ultrasound showed the thinning of the basal anteroseptal wall accompanied by diffuse left ventricular hypokinesia. His electrocardiogram showed a complete right bundle branch block with a first-degree

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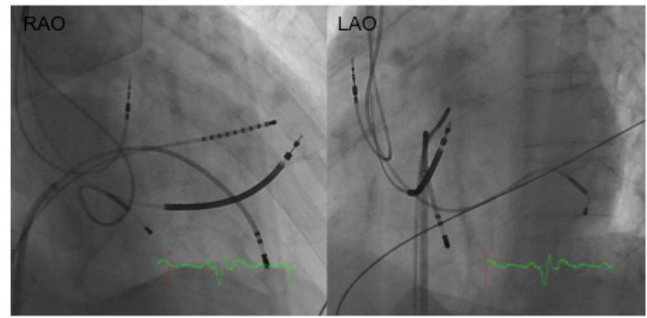
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atrioventricular block. However, it progressed to complete atrioventricular block during follow-up. Combining the progressive atrioventricular block and the characteristic findings on cardiac ultrasound, we diagnosed the patient with cardiac sarcoidosis (CS). Although a temporary pacemaker was implanted via the jugular vein, a ventricular tachyarrhythmia (VT) storm occurred the next day. Therefore, a CRT-D was implanted in the patient after his hemodynamics and VT was stabilized. In the present patient, the progression from first-degree atrioventricular block to complete atrioventricular block was very rapid, and required only 3 months. Therefore, we decided to implant a CRT-D instead of an ICD due to the high risk of atrioventricular block progression from complete right bundle block to complete atrioventricular block in the near future. A shock test just after the CRT-D implantation was normal. After the operation, heart failure and VT were absent, and the patient was treated with a  $\beta$ -blocker, angiotensin receptor blocker, and amiodarone.

After 5 months, the patient visited our hospital emergency room (ER) because he noticed an alarm was sounding from the CRT-D battery. Since he was alert and no sound was audible, he was instructed to wait in the waiting room. However, he was moved into the ER promptly thereafter because he was found unconscious after only a few minutes. The ER staff immediately started to monitor him and observed VF. Therefore, CPR and cardioversion was initiated because the CRT-D did not apply a shock to him despite the VF. After cardioversion, he recovered to a sinus rhythm and was admitted to our hospital for further evaluation.

An electrophysiological study was conducted because a careful review of the CRT-D suggested the sensing lead was under-sensing. A 7-Fr 4-mm-tip ablation catheter (Navistar R-curve, Johnson and Johnson, Japan) and a 6-Fr 10-pole electrode catheter (Snake, Japan) were inserted from the right femoral vein and positioned in the right ventricle apex (RVA) and right ventricular outflow tract (RVOT), respectively. (Figure 1). VT at 180 bpm introduced by a T-wave shock was only recognized as a 90-bpm VT by the sensing lead. Meanwhile, the R wave amplitude detected by the sensing lead was sufficient. This led to the assumption that a rate-dependent functional block could be occurring in the left ventricle. To verify this assumption, we used rapid pacing from the RVA using the CARTO system. Rapid pacing at 180 bpm from the RVA was conducted to the RVOT catheter in a 1:1 manner. However, a pacing frequency over 190 bpm caused a Wenckebach-like IVCA, and the RVOT catheter recorded in a 2:1–3:2 conduction manner (Figure 2). During VT induced by rapid pacing,



**Figure 1: RAO and LAO view of the catheter position during the electrophysiological study.**

LAO: left anterior oblique RAO: right anterior oblique

under-counting was evident (Figure 3). According to the voltage map obtained through the CARTO system, the RVA to the inferior wall showed a normal voltage, but the lower RVOT, where the sensing lead was located, showed a markedly low voltage (Figure 4).

Therefore, we concluded that the progression of CS had resulted in the development of a new intraventricular conduction abnormality (IVCA) and caused the CRT-D sensing lead to under-sense the tachyarrhythmia at over 190 bpm. Later, the patient received an additional lead implantation at the base of the RVA, and the CRT-D was confirmed as operating properly. We initially attempted to place the RV lead at the low interventricular septum or the apex; however, we were unable to identify a site with sufficient electric potential and threshold for sensing and pacing despite a thorough investigation, and therefore placed the RV lead at the RVOT. In addition, because we opted for CRT-D therapy for the treatment of complete atrioventricular block, we believed that it would be more effective to place the lead at the RVOT than at the apex. During 24 months of follow-up, VT was occasionally seen, but shock failure was not observed.

## DISCUSSION

Sarcoidosis is a systemic disease of an unknown etiology and is characterized by the formation of noncaseating granulomas in various tissues.<sup>2</sup> Its incidence in the Japanese population is 1.01 per 100,000 individuals (males, 0.73; females, 1.28), and its peak incidence occurs at 20–34 years of age for male patients and 50–60 years of age for female patients. Importantly, a cardiac involvement rate of 23.0% was found in an investigation of 1027 sarcoidosis Japanese patients.<sup>3</sup> Moreover, Watanabe et al reported that after 3 months of follow up, 33% of patients with an ICD implantation for CS had an R wave amplitude of smaller than 5.0 mV.<sup>4</sup> In light of this previous finding, the current report suggests that CS is progressive in a

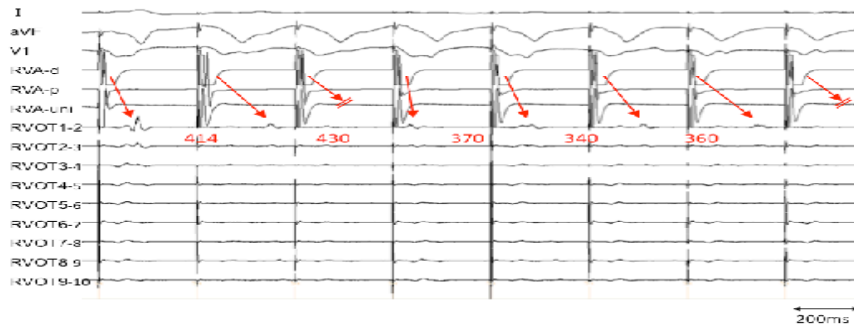


Figure 2: When rapid pacing at 200 bpm was conducted from the RVA, each stimulation did not conduct in the same manner as that conducted during pacing at 180 bpm. The time from the RVA stimulation to conduction to the RVOT gradually elongated until there was no conduction.

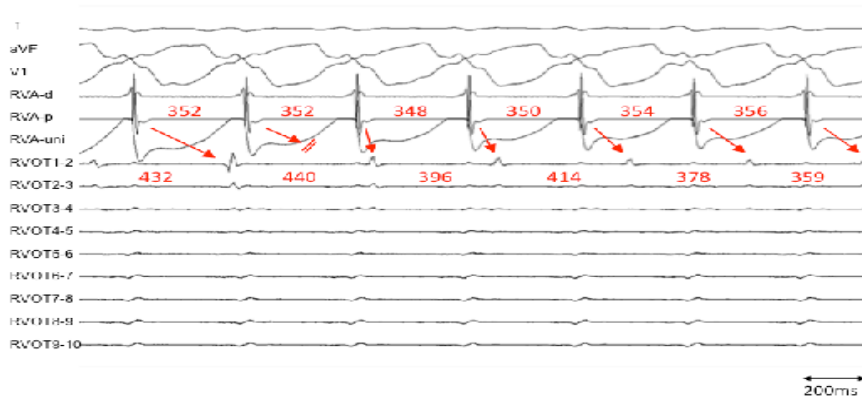


Figure 3: Electrode placed at RVOT was under-counting the actual heart rates for VT. Electrode placed at the RVA was responding in a 1:1 manner with the surface electrocardiogram.

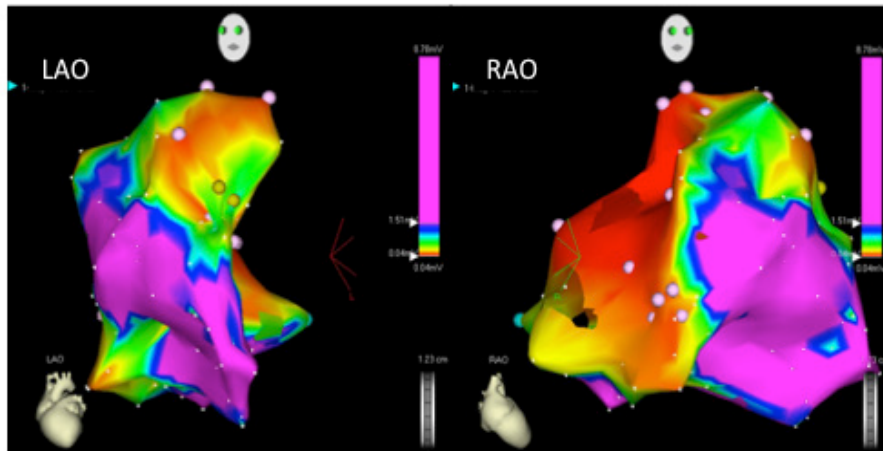


Figure 4: RAO and LAO view of voltage map obtained by the CARTO map system. The red and orange areas represent low voltage areas. The site where the electrode placed at the upper RV and RVOT were surrounded by low voltage areas.

considerable portion of cases, and that this progression could be responsible for the low-voltage myocardium in the present case.

The clinical manifestation of CS varies. Patients present with complete atrioventricular block, bundle block, VT, and congestive heart failure at rates of 23–30%, 12–32%, 23–30% and 25–65% respectively.<sup>5</sup> The echocardiography characteristics for CS range widely and include diffuse local wall motion abnormalities, dilated cardiomyopathy, contraction or relaxation of the left ventricle, and thinness or hypertrophy of the interventricular septum. The diagnostic criteria for CS includes 2 of the following 4 requirements: 1) high degree atrioventricular block, 2) thinness of the basal septum of the left ventricle, and 3), abnormal enhancement of the heart as shown by Gallium 67 citrate scintigraphy, and 4) left ventricular dysfunction, with an ejection fraction of less than 60%.<sup>6</sup> The present case was diagnosed as CS because criteria 1, 2, and 4 were fulfilled.

Treatment for CS with chronic heart failure includes proven medicines, such as angiotensin receptor blockers,  $\beta$ -blockers, and aldosterone antagonists. Steroids are also a potential option for CS treatment. Although no randomized clinical trials have examined the effectiveness of corticosteroid on CS, several reports have shown the effectiveness of corticosteroid (prednisolone 30 mg/day) tapered to 5–15 mg/day for 6–12 months. This administration schedule improved cardiac function and lowered the frequency of arrhythmia.<sup>7-8</sup> In the present case, the sensing lead was placed at a lower RVOT because the sensing threshold and R wave amplitude were sufficient to sense and pace. However, the progression of the CS caused fibrosis of the myocardium surrounding the sensing lead and worked as an electrical barrier, which led to a functional block. This patient was prescribed 5 mg/day of prednisolone, but it was considered to be ineffective in the present case because of the newly emerged fibrosis of the myocardium.

To our knowledge, this report is the first one describing CRT-D shock failure as a result of under-counting because of newly emerged IVCA, despite possessing ample amplitude. Because treatment through medication can be difficult, an additional lead should be a potential

treatment option. However, because data regarding the patient's CARTO map prior to the addition of the extra lead were not available, further study is required to confirm our theory. Moreover, this phenomenon may also have resulted from the intrinsic electrophysiological properties, independent of CS. We reached this conclusion because, in the present case, 2 consecutive shocks were successfully applied for ventricular tachycardia at a rate of >190 bpm when the CRT-D was first implanted. Therefore, we believe that this episode involving an unsuccessful under-sense event can be attributed to the progression of the underlying disease.

In conclusion, in a case of CRT-D operation failure in correctly detecting VF/VT, the possibility of a newly emerged IVCA resulting from the progression of an underlying disease should be included in the differential diagnosis list.

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## CONFLICTS OF INTEREST

The authors have no disclosures.

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