Usefulness of lead repositioning to improve subcutaneous electrocardiogram sensing in patients with arrhythmogenic right ventricular cardiomyopathy with subcutaneous implantable cardioverter-defibrillator

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Introduction

The EMBLEM entirely subcutaneous implantable cardioverter-defibrillator (S-ICD) system (Boston Scientific, Marlborough, MA) was introduced as a new alternative to the conventional transvenous ICD and has been expected to reduce device-related complications, especially in young patients who require long-term lead placement.1,2 Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a well-known hereditary disease recognized as a cause of sudden cardiac death (SCD) in young adults.3 However, the usefulness of S-ICD in patients with ARVC has not been established because of the low QRS amplitude of subcutaneous electrocardiogram (S-ECG) followed by the high incidence of inappropriate shock (IAS) delivery owing to oversensing.4,5 Here we report 2 cases of ARVC in which S-ECG had sensing problems, which were resolved by repositioning the S-ICD leads.

Case report

Case 1

This case involved a 17-year-old male patient who had no family history of SCD and had not shown ECG abnormalities. After exercise at a gymnasium, the patient suddenly developed ventricular fibrillation (VF) and lost consciousness. The patient was resuscitated by an automated external defibrillator and transferred to our hospital immediately. As a result of the examination performed after hospitalization, a definite diagnosis of ARVC was made based on the task force criteria.6 However, sustained ventricular tachycardia (VT) or VF could not be induced through programmed ventricular stimulation in an electrophysiological study, and late gadolinium enhancement was not observed in cardiac magnetic resonance imaging. Therefore, we determined that this patient was experiencing an early phase of ARVC, and he underwent S-ICD implantation as a secondary prevention for VF. The device pocket was formed in the intermuscular space between the serratus anterior and latissimus dorsi, and the S-ICD lead was placed on the left parasternal area according to the result of the preimplant screening using the 3-incision technique. A defibrillation test was performed during the procedure, and VF was induced and then terminated by a single 65-J shock. The time to shock therapy was 12.4 seconds, and the postshock impedance of the S-ICD lead was 46 ohms.

KEY TEACHING POINTS

- Temporal change of QRS amplitude in subcutaneous electrocardiogram (S-ECG) can occur in patients with arrhythmogenic right ventricular cardiomyopathy (ARVC).
- Repositioning of subcutaneous implantable cardioverter-defibrillator lead is useful for achieving better S-ECG in patients with ARVC.
- Exercise test after lead repositioning is necessary for prevention of oversensing and achieving optimal sensing.

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Thirteen months after the S-ICD implantation, the patient experienced a first IAS delivery when he was wiping his hair with a towel after bathing. The S-ICD was set to use the primary sensing vector at the time of IAS, and the Smart Pass (SP) algorithm turned off automatically owing to attenuated QRS amplitude. S-ECG showed myopotential interference (MPI) that led to IAS delivery owing to oversensing (Figure 1A). The authors evaluated the MPI in the same situation as IAS delivery and changed the sensing vector from the primary sensing vector to the alternate sensing vector to avoid MPI. However, 6 months after changing of the sensing vector, the patient experienced a second IAS delivery when he was operating his smartphone in the left lateral decubitus position. The S-ICD was set to use the alternate sensing vector, and the SP algorithm turned off automatically owing to attenuated QRS amplitude (Figure 1B). Temporal changes in the QRS amplitude of S-ECG caused by disease progression were observed in all sensing vectors, especially in the alternate sensing vector. Although the secondary sensing vector also had low QRS amplitude, the authors utilized the secondary sensing vector because it was the only sensing vector that did not cause IAS. Unfortunately, the patient experienced a third IAS delivery owing to MPI under the SP algorithm when he was lying down and operating his smartphone. Therefore, the authors performed an automated screening test (AST) on surface ECG again with a different electrode positioning. First, the authors evaluated the right-side parasternal tunneling, but the QRS amplitude of S-ECG was low, and screening failed for all vectors. Then, the authors performed an AST by moving the distal electrode approximately 1 intercostal space downward, and the proximal electrode was moved to the left, so that left ventricular ECG was more reflected in S-ECG. As a result, the QRS amplitude of S-ECG increased in 2 out of 3 sensing vectors and passed the AST. Therefore, we performed S-ICD lead extraction and repositioning under fluoroscopy, so that the lead was in the same position as previously described (Figure 2). After repositioning of the S-ICD lead, the QRS amplitude of S-ECG increased and 2 (secondary and alternate sensing vectors) out of 3 vectors were suitable for S-ECG sensing. The authors turned the SP algorithm on just after the S-ICD implantation procedure and performed MPI induction test about a week after implantation to determine the optimal sensing vector without MPI. In the MPI induction test that reproduced physical activities at the time of IAS, slight MPI was still observed in all sensing vectors, but most MPIs were identified as noise events. Because the secondary sensing vector had the highest QRS/T ratio among the 3 vectors and was less affected by postural changes, the authors selected this vector as the optimal sensing vector, and there was no IAS for 12 months thereafter.

Case 2

This case involved a 19-year-old man who was referred to the authors’ hospital for detailed examination for an abnormal ECG at medical examination. The patient had no family history of SCD but had a history of unexplained syncope. As a result of the examination performed after hospitalization, the diagnosis of ARVC was made based on the modified task force criteria for ARVC. Cardiac magnetic resonance imaging demonstrated late gadolinium
enhancement in the entire enlarged right ventricle, and fatty degeneration and edematous changes were found on the posterolateral wall of the base of the tricuspid valve. Furthermore, sustained VT/VF was induced by programmed ventricular stimulation in an electrophysiological study. Therefore, we decided to implant an S-ICD to primarily prevent SCD, adhering to the class IIa indication of ICD implantation based on the 2018 JCS/JHRS Guideline for Non-Pharmacotherapy of Cardiac Arrhythmias.8 However, all preimplant screening of S-ECG at normal electrode position failed. Then, the authors performed an AST by moving the distal electrode approximately 1 intercostal space downward, and the proximal electrode was moved to the left, so that left ventricular ECG was more reflected in S-ECG. As a result, in S-ECG, the QRS/T ratio improved and passed screening in all sensing vectors. Therefore, we performed S-ICD implantation with modified left-side parasternal tunneling. During the implantation procedure, the distal electrode was placed on the middle of the sternum to reduce the risk of MPI, and the proximal electrode was placed to the left of the normal position as mentioned above (Figure 3A). A defibrillation test was performed during the procedure, and VF was induced and then terminated by a single 65-J shock. After S-ICD implantation, the QRS amplitude of S-ECG increased in all sensing vectors and passed preimplant screening. We performed a novel tube exercise test9 and treadmill exercise test to determine the optimal sensing vector that does not cause MPI. In the results of both exercise tests, no significant MPI and T-wave oversensing were detected when the alternate sensing vector was used (Figure 3B). Therefore, the authors selected the alternate sensing vector, which was recommended by the S-ICD automatically as the optimal sensing vector, and there was no IAS delivery for 5 months thereafter.

**Discussion**

In this report, we demonstrated the usefulness of S-ICD lead repositioning in improving S-ECG sensing in young adult patients with ARVC. It was also possible to reduce the risk of IAS delivery by selecting individually optimized sensing vectors according to the results of an exercise test under modified S-ECG sensing.

It has been suggested that fatal events occurring before overt structural myocardial changes may be caused by a primarily electrical mechanism in patients with ARVC, as a consequence of the crosstalk of genetically defective desmosomal proteins with the voltage-gated sodium channel complex, leading to reduced sodium currents and arrhythogenic mechanisms similar to those in Brugada syndrome.10 It has also been known that the phenotype of fatal arrhythmias in patients with ARVC is age-dependent. Older patients with advanced disease more often experience reentrant VT around a myocardial fibrofatty scar, whereas in young patients it is common to experience sudden onsets of VF reflecting acute electrical instability in the early phases of the disease. Furthermore, S-ICD is also recommended to minimize the incidence of long-term lead-related complications. Therefore, S-ICD is expected to prevent SCD, especially in young patients with ARVC.

However, the precise clinical role of S-ICD in patients with ARVC remains to be defined. One reason is the beneficial effect of antitachycardia pacing that is expected to be highly effective in terminating scar-related reentrant...
VT, which is often observed in patients with ARVC. Another reason is the possibility of morphological changes in S-ECG leading to cardiac oversensing and IAS delivery. An Italian multicenter registry, which enrolled 44 young patients who underwent S-ICD implantation (mean age of 37 years; mean left ventricular ejection fraction of 53%; primary prevention of 59%), demonstrated a high incidence of IAS delivery. Among the enrolled patients, 6 (14%) experienced 8 IAS deliveries, consisting of 4 cardiac oversensing and 4 noncardiac oversensing. In addition, a transatlantic experience, which enrolled 29 young patients with ARVC who underwent S-ICD implantation (mean age of 34 years, mean LVEF of 56%, primary prevention of 59%), has demonstrated a high incidence of IAS. Among the enrolled patients, 6 (21%) experienced 39 IAS deliveries owing to oversensing. By these evidence, S-ICD implantation in patients with ARVC has still been limited.

The usefulness of lead and device repositioning in preventing IAS deliveries has been well known. The most important benefit of lead repositioning is improved sensing. The sensing of S-ICD is largely dependent on the QRS/T ratio of S-ECG, which is vulnerable to changes caused by physical activities or the progression of underlying heart diseases. The decrease in the QRS amplitude is more likely...

Figure 3  A: Chest radiograph (left) and 3-dimensional computed tomographic (right) images after subcutaneous implantable cardioverter-defibrillator (S-ICD) implantation. B: Comparison of changes in subcutaneous electrocardiogram (S-ECG) during the treadmill exercise test. During the treadmill exercise test, myopotential interference (MPI) and/or T-wave oversensing were observed when the primary and secondary sensing vectors were used. On the other hand, no significant MPI and T-wave oversensing were detected when the alternate sensing vector was used.
to lead to oversensing such as T-wave or myopotential oversensing. In patients with ARVC, the decrease in the QRS amplitude owing to degeneration of the right ventricular myocardium progresses over time. Therefore, it is suggested that more reflection on left ventricular ECGs is useful in accurately improving S-ECG sensing. In other words, it is important to move the proximal electrode to the left to create a sensing vector different from the conventional sensing vector that more closely reflects the left ventricular ECG. In this report, we demonstrated the usefulness of electrode repositioning in improving S-ECG sensing in young patients with ARVC. However, a concern in the method used in our cases was that repositioned electrodes, especially the proximal electrode that was directly above or near the pectoralis major muscle, would be more vulnerable to MPI. Certainly, even after repositioning, slight MPI was observed in some sensing vectors in the first case, but it was possible to avoid MPI by selecting the optimal sensing vector based on the results of an exercise test. In the second case, placing the distal electrode on the sternum reduced the risk of MPI, as shown in the results of the exercise test (Figure 3B). Furthermore, the higher QRS amplitude achieved by the modified lead positioning enabled the continued use of the SP algorithm and reduced the risk of T-wave oversensing. Thus, S-ICD implantation in patients with ARVC with modified lead positioning may provide many benefits.

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