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RHYTHM DISORDERS

CLINICAL CASE

Subcutaneous Implantable Cardioverter-Defibrillator Implantation in a Hereditary Dystonia Patient With Bilateral Deep Brain Stimulation

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ABSTRACT

The effect of noise from the deep brain stimulator (DBS) or neurologic symptoms of hereditary dystonia on the subcutaneous implantable cardioverter-defibrillator (S-ICD) remains unknown. Therefore, herein, we present the first case of S-ICD implantation in a patient with hereditary dystonia who underwent DBS. (JACC Case Rep. 2025;30:103108) © 2025 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 57-year-old man experienced cardiac arrest, and the electrocardiogram (ECG) revealed ventricular fibrillation (VF). The patient was transferred to another hospital after resuming the heartbeat after one defibrillation (no-flow time: 11 minutes). After examination, including emergency coronary angiography, he was diagnosed with VF induced by a coronary artery spasm and was referred to our hospital. He was treated with hypothermic therapy, and no neurologic sequelae of cardiopulmonary arrest were observed. However, these physical and psychological stressors exacerbated the neurologic symptoms of hereditary dystonia, including involuntary muscle movements or tremors, making remaining at rest for long periods difficult. By expanding the degree of rest through rehabilitation, neurologic symptoms decreased with a reduction in stress.

TAKE-HOME MESSAGES

- This case highlights the importance of the precautions needed for cardiac implantable electronic devices in patients with hereditary dystonia and DBS.
- An S-ICD was not affected by noise from DBS and involuntary movements in this case. It is important to understand the characteristics of DBS and S-ICD, as well as the patient's condition at preoperative screening, during surgery, and at follow-up.

PAST MEDICAL HISTORY

History of hereditary dystonia with a *DYT1-TOR1A* mutation was revealed by genetic testing. The dystonic symptoms first appeared as involuntary movements. The symptoms extended from the right lower

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ABBREVIATIONS AND ACRONYMS

DBS = deep brain stimulation

DFT = defibrillation threshold test

ECG = electrocardiogram

ICD = implantable cardioverter-defibrillator

SCD = sudden cardiac death

S-ICD = subcutaneous implantable cardioverter-defibrillator

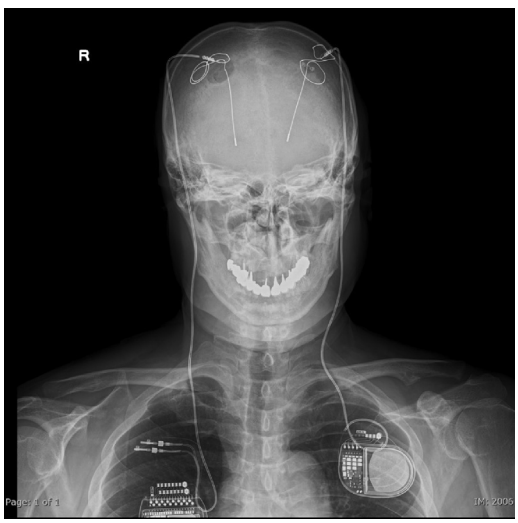
VF = ventricular fibrillation

limb to the left lower limb and trunk. By 14 years of age, the patient was using crutches, and by 20 years of age he was in a wheelchair. At 39 years of age, he underwent deep brain stimulation (DBS) at another hospital and was able to walk on his own. DBS was bilateral, and both generators were implanted in the upper thoracic region (left generator, Activa SC; right generator, Activa RC, Medtronic). The left generator was bipolar and nonrechargeable (frequency, 130 Hz; amplitude, 3.0 V [voltage mode]; and pulse width, 330 μs), and the right generator was unipolar and rechargeable (frequency, 125 Hz; amplitude, 3.3 mA [current mode]; and pulse width, 60 μs) as these settings were appropriate for this patient. Each lead was connected to the globus pallidus in the cranium from the generator through the subcutis (Figure 1).

DIFFERENTIAL DIAGNOSIS

The patient had a current smoking history and experienced cardiac arrest in the early morning of admission. The rhythm strip initially showed VF (Figure 2A). The 12-lead ECG after defibrillation revealed ST-segment elevation at V_2-V_4 and reciprocal changes in the contralateral leads with DBS noise (Figure 2B). In his emergency coronary angiography, the improvement of ST-segment elevation in the ECG was observed, and there was no more than 50% stenosis or no plaque disruption on optical coherence tomography in the coronary arteries, which was determined to be the most likely cause of coronary artery spasm. A coronary artery spasm provocation test was not performed because of the history of cardiac arrest caused by VF, and it was deemed unsafe.

FIGURE 1 Preprocedural Head and Upper Chest Radiograph



Generators for bilateral deep brain stimulation are implanted into the upper chest. Each lead is connected to the globus pallidus in the cranium, using a generator through the subcutis.

INVESTIGATIONS

Transthoracic echocardiography revealed no wall-motion abnormalities, valvular disease, or heart failure. Screening for subcutaneous implantable cardioverter-defibrillators (S-ICDs) (Boston Scientific) on the primary and secondary vectors was performed in the supine, standing, and sitting positions, although concerns existed regarding noise from DBS and noise from involuntary movements (Figure 3).

MANAGEMENT

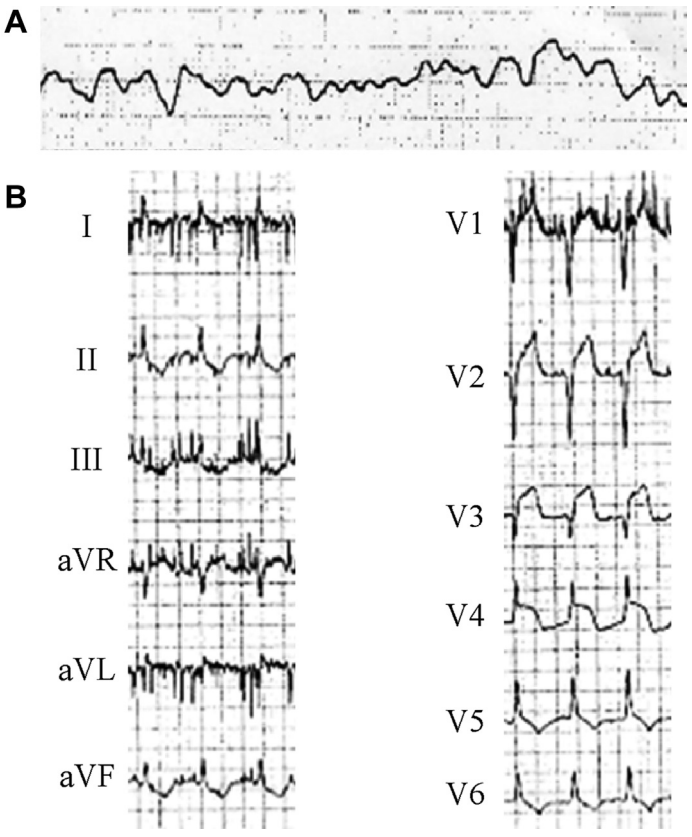
After VF resuscitation, implantable cardioverter-defibrillator (ICD) therapy was selected for secondary prevention of sudden cardiac death (SCD) in addition to medical therapy using coronary vasodilators.[1,2] The wearable cardioverter-defibrillator was considered but could not be used because of a lack of patient cooperation caused by worsening dystonic symptoms. Instead, he was treated in a hospital in which defibrillation could be performed until ICD implantation. ICD options include transvenous ICD and S-ICD. S-ICD was selected in this case after considering the noise caused by DBS and involuntary movements, positional constraints of moving from a dual to a triple generator, absence of pacing, and possible future infections and lead problems. The patient was administered 10 mg diazepam as a preoperative anesthetic to prevent anxiety-induced worsening of involuntary movements during surgery. In addition to 1% lidocaine 200 mg as a local anesthetic, the surgery was performed under sedation with dexmedetomidine hydrochloride (loading dose: 6 μg/kg/h infused for 10 minutes; maintenance dose: 0.2-0.7 μg/kg/h). Respiratory management was provided by noninvasive ventilation, and DBS was continued during surgery. The S-ICD was implanted

20 cm away from the DBS generator. Diathermy was performed using an indifferent electrode in the leg because of the need for a large surgical wound for S-ICD implantation. Defibrillation threshold test (DFT) was performed at the end of the procedure, and the induced VF was terminated with a single defibrillation of 65 J and an impedance of 64 Ω without any problems with the DBS generators. **Figure 4** presents the chest radiograph obtained after S-ICD implantation. No oversensing of DBS or involuntary movements was observed (**Figure 5**). To prevent the coronary artery spasm recurrence, the patient stopped smoking and was administered calcium channel blockers and coronary vasodilators.

DISCUSSION

S-ICD is increasingly widely used for both primary and secondary SCD prevention because of ventricular arrhythmias.[3] DBS treatment is used worldwide, mainly in diseases associated with tremor, including Parkinson disease. DBS is also used to treat hereditary dystonia, particularly in patients with *DYT1* mutations. Patients with hereditary dystonia experience greater involuntary movements than those with Parkinson disease, and the application of S-ICD in these patients has not been characterized extensively.[4] Two studies have reported the application of S-ICD in patients with Parkinson disease; however, no previous studies have reported the application of S-ICD in patients with hereditary dystonia.[5,6] S-ICD was selected for this case based on the following 5 perspectives (**Table 1**): The noise from DBS and involuntary movements could pass ECG screening for S-ICDs; no history of bradycardia or other conditions needing permanent pacing; avoiding the upper pectoral area in which the DBS generators are implanted; reducing the risk of systemic infection; and avoiding lead damage caused by involuntary movements. The S-ICD allows frequencies of 3 to 40 Hz to pass and eliminates the DBS frequencies above 60 Hz. Hereditary dystonia requires more extensive stimulation than Parkinson syndrome, with larger pulse widths and higher amplitude settings than Parkinson syndrome. Even if the frequency is within the filter range, large pulse widths and high amplitude can be a problem, making preoperative ECG screening and S-ICD checks at the time of surgery important. In addition to pulse width and amplitude, the positions of the DBS and S-ICD, vector configuration, and distance of the S-ICD system are also important for S-ICD sensing; therefore, careful preoperative ECG screening and checking during surgery are also important. During the operation, the anesthetic dosage was adjusted carefully, as advised by the neurosurgeon, to reduce pain and anxiety.[7] A distance of at least 20 cm between the 2 generators is recommended, and was used as a reference for positioning in this case.[8]

FIGURE 2 Electrocardiograms at the Cardiac Event

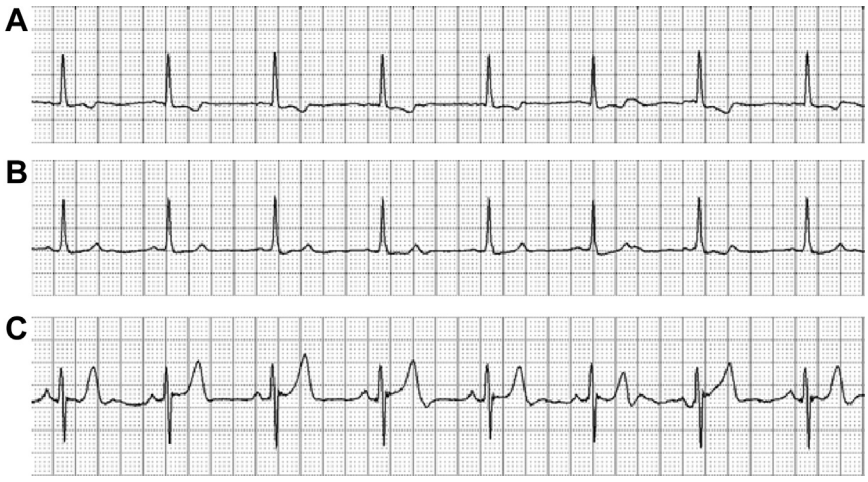


(A) The initial waveform indicates ventricular fibrillation. (B) The 12-lead electrocardiogram after defibrillation reveals ST-segment elevation at V_2-V_4 and reciprocal changes in the contralateral leads with deep brain stimulation noise.

FOLLOW-UP

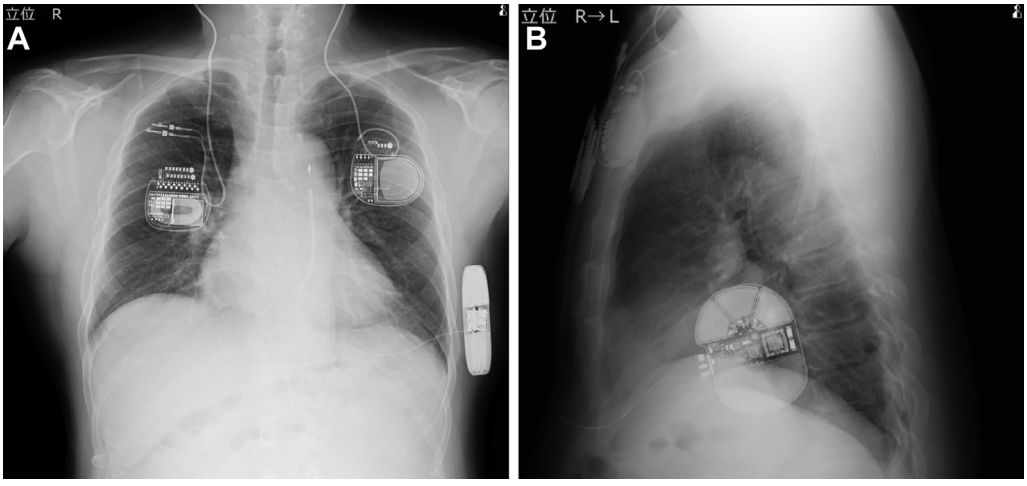
No evidence of inappropriate shocks of the S-ICD was reported at the 12-month follow-up. One DBS battery replacement operation was performed during the

FIGURE 3 Preoperative Electrocardiogram Screening



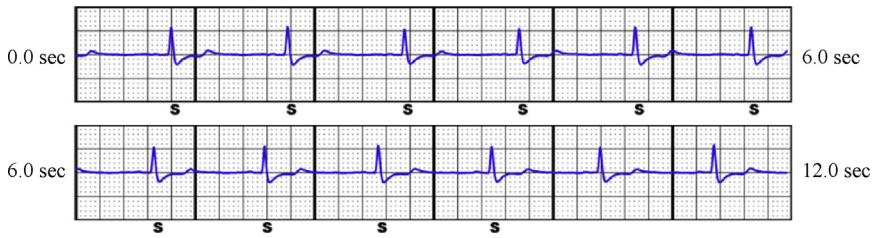
Preoperative electrocardiographic screening for subcutaneous implantable cardioverter-defibrillators. (A) Primary passed. (B) Secondary passed. (C) Alternative not passed.

FIGURE 4 Postprocedural Chest Radiograph



The S-ICD generator and a lead are implanted. There is sufficient distance between the S-ICD generator and left deep brain stimulation generator. (A) Posteroanterior projection chest radiograph. (B) Right-lateral projection chest radiograph. S-ICD = subcutaneous implantable cardioverter-defibrillator.

FIGURE 5 A Subcutaneous Implantable Cardioverter-Defibrillator Electrogram Recording Postimplantation



Subcutaneous implantable cardioverter-defibrillator electrogram recordings reveal appropriate sensing in the primary vector after implantation.

follow-up period without any device infection signs. Future ICD therapy has been reported to affect DBS generators instead of no effects in testing during operation.[9] We have confirmed appropriate S-ICD shock for induced VF at the time of surgery; however, no studies have reported the appropriate shock needed for clinical ventricular arrhythmias in patients with DBS. The possibility of device damage caused by the shock should be considered and it is important to defibrillate with minimal energy and to check DBS generators after the shock. In this case, 2 defibrillations both at the VF (automated external defibrillator with biphasic 150 J) and at the DFT (S-ICD with biphasic 65 J) did not affect DBS. It is also important to check the S-ICD to ensure that it is within the filter range when the settings of DBS are changed. Regular follow-ups will be needed to control hereditary dystonia, avoid worsening involuntary movements; control coronary artery spasm; prevent ICD activation; and manage devices, including future battery replacement.

TABLE 1 Assessment for Cardiac Implantable Electronic Device Selection in Patients With DBS

The noise from deep brain stimulation (bipolar or unipolar) and involuntary movements
Does the patient's condition necessitate permanent pacing or not?
The positional relation between DBS generators and new generator
Increasing risk of infection caused by multiple devices
The possibility of lead damage by involuntary movements

CONCLUSIONS

We performed S-ICD implantation in a patient with hereditary dystonia who underwent DBS. Selecting the type of ICD device and performing the surgery was enabled by careful assessment of DBS and the disease. Twelve months after the operation, the patient was followed-up without any adverse events, including inappropriate ICD therapy.

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KEY WORDS **deep brain stimulation, hereditary dystonia, subcutaneous implantable cardioverter-defibrillator, sudden cardiac death, ventricular fibrillation**