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SPOTLIGHT

Double-device therapy in a patient with long QT syndrome

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Congenital long QT syndrome (LQTS) is an inherited cardiac disease characterized by a prolonged QT interval and ventricular arrhythmias such as ventricular fibrillation or polymorphic ventricular tachycardia, often triggered by emotional or physical stress. β-blockers are highly effective in suppressing ventricular arrhythmias, but some LQTS patients require implantable cardioverter-defibrillator (ICD) implantation for the primary or secondary prevention of sudden cardiac death. Recently, subcutaneous implantable cardioverter defibrillator (S-ICD) has been implanted for patients with LQTS for the following reasons.[1] Anti-tachycardia pacing (ATP) is not necessary because LQTS patients typically present with ventricular fibrillation or polymorphic ventricular tachycardia, for which ATP is not effective. Given that LQTS patients requiring ICD implantation are typically young, S-ICD may be preferable to avoid venous occlusion or to reduce the possibility of defibrillator lead fracture.[2] However, there is a potential pitfall in selecting S-ICD implantation for LQTS patients. In this report, we present a case of successful bail-out from frequent appropriate S-ICD therapies by double-device therapy.

A 26-year-old man was referred to our hospital for S-ICD implantation. The patient had a history of frequent presyncope episodes when waking up to an alarm clock in the early morning. At the age of 23, he experienced syncope accompanied by palpitations in the morning, leading to hospitalization. The 12-lead ECG revealed a QTc interval of 501 ms, a notched T wave, and a heart rate lower than expected for his age, diagnosing him with congenital long QT syndrome. (Figure 1) The average heart rate on the Holter electrocardiogram before the initiation of β-blockers was 46 bpm. Repetitive lifestyle counseling, including avoiding the use of alarm clocks, was conducted; however, it was not possible to completely avoid the stress that triggers syncope. He experienced syncope even after the initiation of treatment with 0.625 mg of bisoprolol, and therefore he needed ICD implantation. Initially, the patient refused the implantation of ICD or pacing devices. Subsequently, consent was obtained only for S-ICD implantation. After admission to our hospital, he underwent S-ICD implantation. Over the next 9 months following S-ICD implantation, a total of eight ventricular arrhythmia events including six appropriate S-ICD therapies for polymorphic ventricular tachycardia or ventricular fibrillation and two episodes of spontaneous termination of polymorphic ventricular tachycardia were observed. QT prolongation and premature ventricular contractions with short-long-short sequence preceded Torsade de pointes. A premature ventricular contraction before the completion of repolarization initiated ventricular fibrillation, indicating that early afterdepolarization was likely an underlying mechanism. (Figure 2) The patient presented with sinus bradycardia, which might contribute to the occurrence of ventricular arrhythmias. To address the bradycardia issue, we implanted an atrial pacing lead in the right atrium, while retaining the S-ICD, resulting in a "double-device therapy." (Figure 3) A 12-lead ECG after the single-chamber pacemaker implantation revealed the atrial pacing and ventricular sensing rhythm at 60 beats per minute. The QTc interval was shortened from 519 to 472 ms. (Figure 4) After implanting the atrial lead, the dose of bisoprolol was increased from 0.625 to 2.5 mg. Thereafter, the patient experienced no further S-ICD therapies at all. Although the patient seemed to be LQTS type 2 based on situations and triggers of ventricular fibrillation occurring, no mutations in the KCNH2 gene were detected in the genetic test.

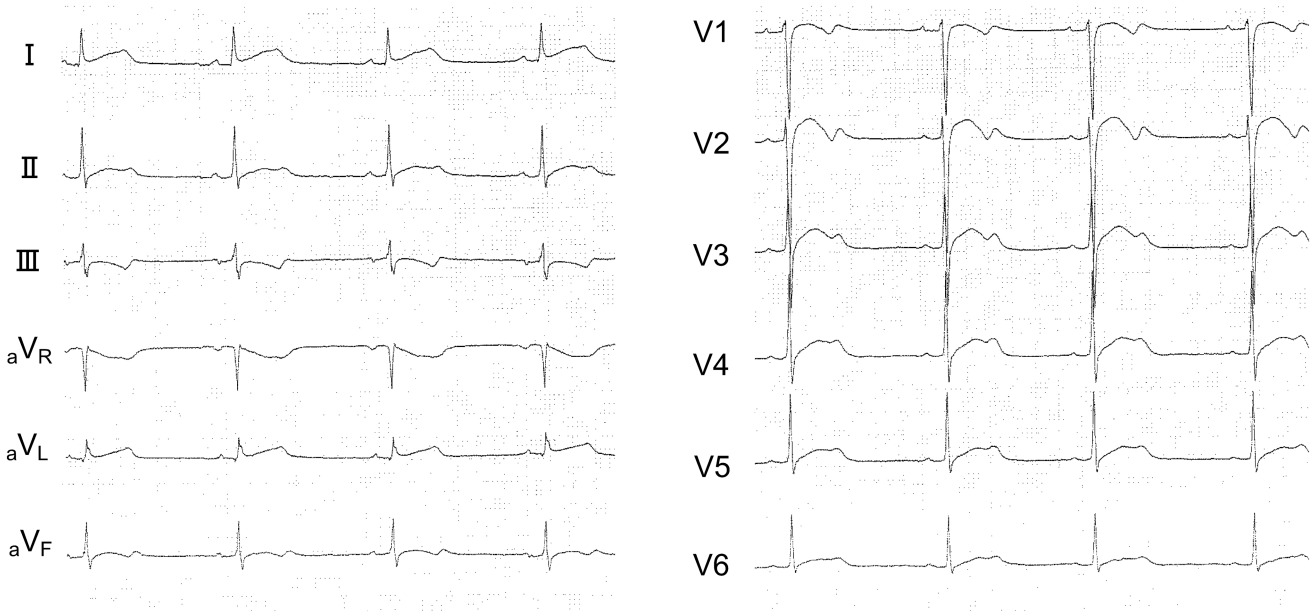

FIGURE 1 Twelve-lead electrocardiogram before the initiation of β blocker therapy. The 12-lead ECG revealed sinus rhythm of 46 beats per minute with the QTc interval of 501 ms and a notched T wave. At this point, the patient did not take β-blockers.

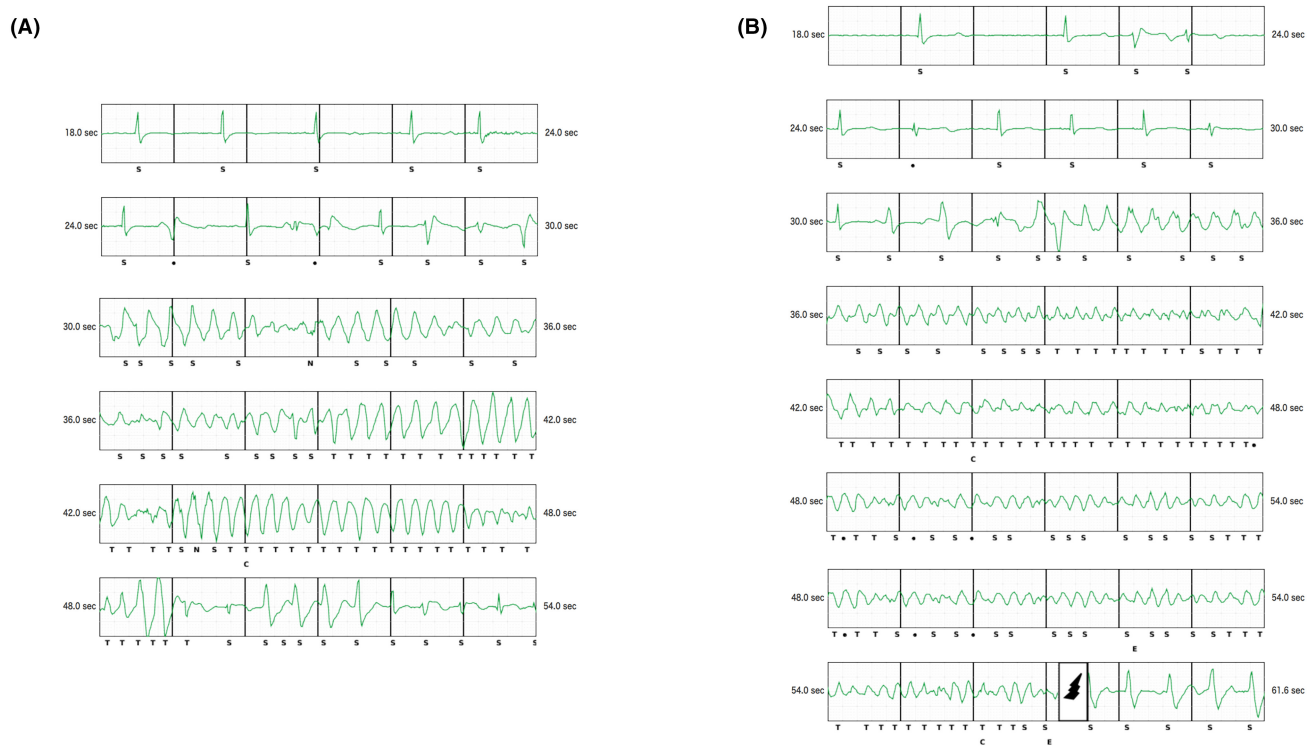

FIGURE 2 Device electrogram. (A). Torsade de pointes (TdP) was initiated by a premature ventricular contraction. Note that short-long-short sequences preceded TdP. (B). Ventricular fibrillation occurred with a premature ventricular contraction as a trigger, and the subcutaneous implantable cardioverter-defibrillator detected and terminated ventricular fibrillation. Prolonged QT interval and a premature ventricular contraction before the completion of repolarization preceded ventricular fibrillation, indicating that early afterdepolarization was likely an underlying mechanism.

In this case report, we determined two important clinical issues; (1) S-ICD implantation is not preferable for some patients with LQTS because β-blockers, a first-line treatment for LQTS, may cause bradycardia and aggravate some type of LQTS, and (2) adding an atrial lead may be effective in suppressing ventricular arrhythmias in a patient with LQTS after S-ICD implantation. The

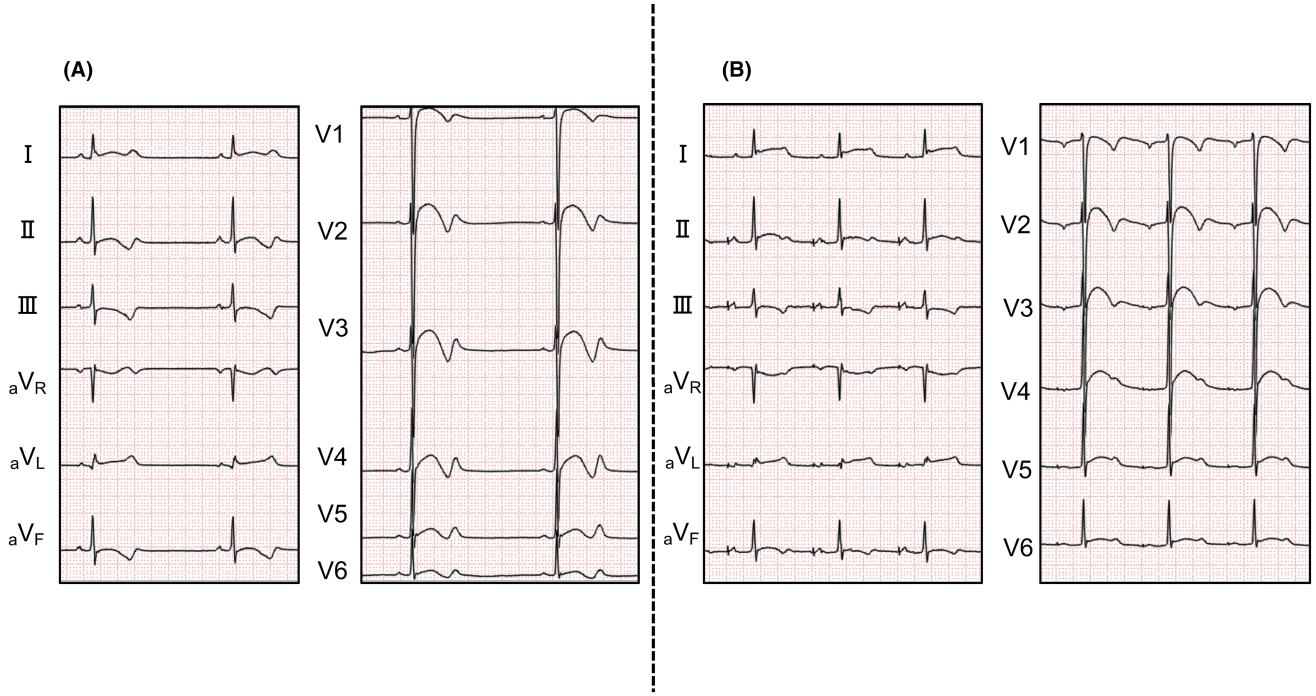

FIGURE 3 Twelve-lead electrocardiograms before and after pacemaker implantation. (A). The 12-lead electrocardiogram after S-ICD implantation shows sinus bradycardia at 36 beats per minute with a QTc interval of 519 ms. At this point, the patient was taking 0.625 mg of bisoprolol. A 12-lead electrocardiogram of this patient showed peculiar biphasic T waves in precordial leads and bradycardia-induced QT prolongation, which was associated with the initiation of Torsade de pointes. Long QT syndrome type 2 was suspected, but no mutations in the KCNH2 gene were detected in the genetic test. (B). The 12-lead electrocardiogram after single-chamber pacemaker implantation shows atrial pacing and ventricular sensing at 60 beats per minute with a QTc interval of 472 ms.

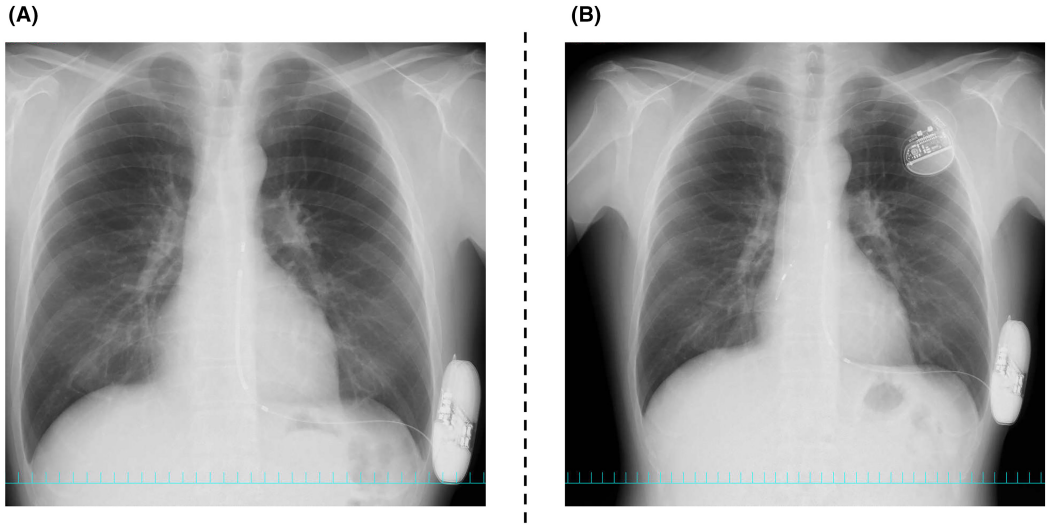

FIGURE 4 Chest x-rays before and after pacemaker implantation. (A). Before single-chamber pacemaker implantation. (B). Double-device therapy after single-chamber pacemaker implantation.

benefit of atrial pacing is shortening the QT interval and the suppression of ventricular ectopic beat triggering polymorphic ventricular tachycardia or ventricular fibrillation. Only atrial pacing might be effective for this patient, because the patient did not experience S-ICD discharge for ventricular arrhythmias after the combined therapy of atrial pacing plus β-blocker therapy. A combination of pacing therapy and β-blockers was reported to be effective for preventing ventricular arrhythmia events in LQTS.[3] The advantage of S-ICD is the avoidance of lead placement in the heart or vessels, resulting in the reduced possibility of procedure-related complications such as pocket hematoma, systemic infection, pneumothorax, lead dislodgement, or lead perforation. The S-ICD lead extraction procedure was reported to be safe, and not associated with life-threatening complications.[4] However, its

disadvantage is the inability to perform pacing therapy. Therefore, for LQTS patients, S-ICD might not be preferred due to the following reasons: Some LQTS patients have bradycardia compared with their age; the first-line treatment for LQTS involves β-blockers, which further lower heart rate; and bradycardia promotes QT prolongation and associated polymorphic VT.

There are two other treatment options to consider for this patient. One option is to initially perform transvenous ICD implantation instead of S-ICD. The other is to remove the S-ICD and implant a transvenous ICD. The advantage of these treatments is the ability to combine atrial pacing and defibrillation in a single device. However, the disadvantage is that patients with LQTS requiring ICD implantation are usually young and have a longer lifespan, which may increase the risk of future lead failure or lead extraction procedures. The implantation of an ICD lead at the right ventricle at a young age is associated with a higher risk of lead failure. Additionally, the development of connective tissue on the lead and strong adhesion to the heart and vessels make transvenous lead extraction procedures complex, leading to a higher rate of complications.[5]

Double-device therapy with S-ICD and a single-chamber pacemaker offers distinct roles to each device: The pacemaker handles atrial pacing which contributes to the suppression of ventricular arrhythmias, while the S-ICD manages detection and treatment of life-threatening arrhythmias. Double-device therapy may be a favorable treatment strategy, especially for patients requiring ICD implantation at a young age for the following reasons. First, a sufficient amount of β-blockers can be prescribed because bradycardia can be avoided by atrial pacing. Second, atrial pacing shortens the QT interval and reduces ventricular ectopic beats, preventing ventricular arrhythmias. Third, in the future, if atrial lead failure occurs and a lead extraction procedure is required, only one lead needs to be extracted. Finally, by adding one ICD lead in the right ventricle, the system can be converted to a dual-chamber ICD.

Double-device therapy for patients with LQTS combines the benefits of ICD therapy and pacing therapy. It may be a safer treatment than traditional transvenous ICD implantation especially for young patients with LQTS.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

The authors have no competing interests to disclose.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no datasets were generated or analyzed in this study.

ETHICS STATEMENT

Not applicable.

DISCLOSURE

Dr. Higuchi, Dr. Yagishita and Dr. Shoda belong to the same endowed department established by contributions from Medtronic Japan, BostonScientific, Biotronik Japan, and Abbott Medical.

PATIENT CONSENT STATEMENT

Written informed consent was obtained from the patient.

CLINICAL TRIAL REGISTRATION

Not applicable.

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