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RHYTHM DISORDERS AND ELECTROPHYSIOLOGY

CASE REPORT: CLINICAL CASE

Life-Saving Cooperative Function of a Subcutaneous Cardioverter-Defibrillator and a Pacemaker in Dextrocardia With Heart Failure

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ABSTRACT

A 47-year-old man with congenitally corrected transposition of the great arteries, dextro-cardia, heart failure, and pacemaker-dependency presented with nonsustained ventricular tachycardia. He underwent a right-sided transmuscular cardioverter-defibrillator insertion after appropriate testing. One year later, life-saving antiarrhythmic therapy was applied by the subcutaneous defibrillator while the appropriate pacemaker functioning supported heart rhythm. (JACC Case Rep. 2024;29:102714) © 2024 The Authors. 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/).

HISTORY OF PRESENTATION

A 47-year-old man with history of congenitally corrected transposition of the great arteries (cCTGA), dextrocardia with situs inversus (Figure 1), and a chronically implanted dual-chamber pacemaker because of an intermittent 2:1 atrioventricular block presented for a follow-up to the pacemaker clinic of our hospital. He was clinically stable. The pacemaker interrogation revealed an episode of nonsustained ventricular tachycardia, while the patient was pacemaker-dependent. The arrhythmia was monomorphic, lasted 10 seconds, and had a mean ventricular rate of 185 beats/min; it was associated with a presyncopal episode. A recent echocardiographic examination showed a dilated systemic ventricle with a reduced ejection fraction of 40%. Insertion of a cardioverter-defibrillator was deemed appropriate.

TAKE-HOME MESSAGES

- A rare case of a pacemaker-dependent patient is presented, with congenital heart disease, dextrocardia, heart failure, and ventricular tachycardia successfully treated by a subcutaneous defibrillator.
- Testing for potentially hazardous interactions between implanted electronic devices is of extreme importance, especially in pacemaker-dependent patients with unique heart disease characteristics.

PAST MEDICAL HISTORY

The patient's heart condition had been diagnosed 4 years earlier. At that time, a 2:1 atrioventricular block necessitated a dual-chamber pacemaker insertion. The systemic ventricle was already dilated, with an ejection fraction of 40%. Medical therapy was initiated, and he was asymptomatic; however, 6 months

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ABBREVIATIONS AND ACRONYMS

cCTGA = congenitally corrected transposition of the great arteries

CRT-D = cardiac resynchronization therapy with pacemaker-defibrillator

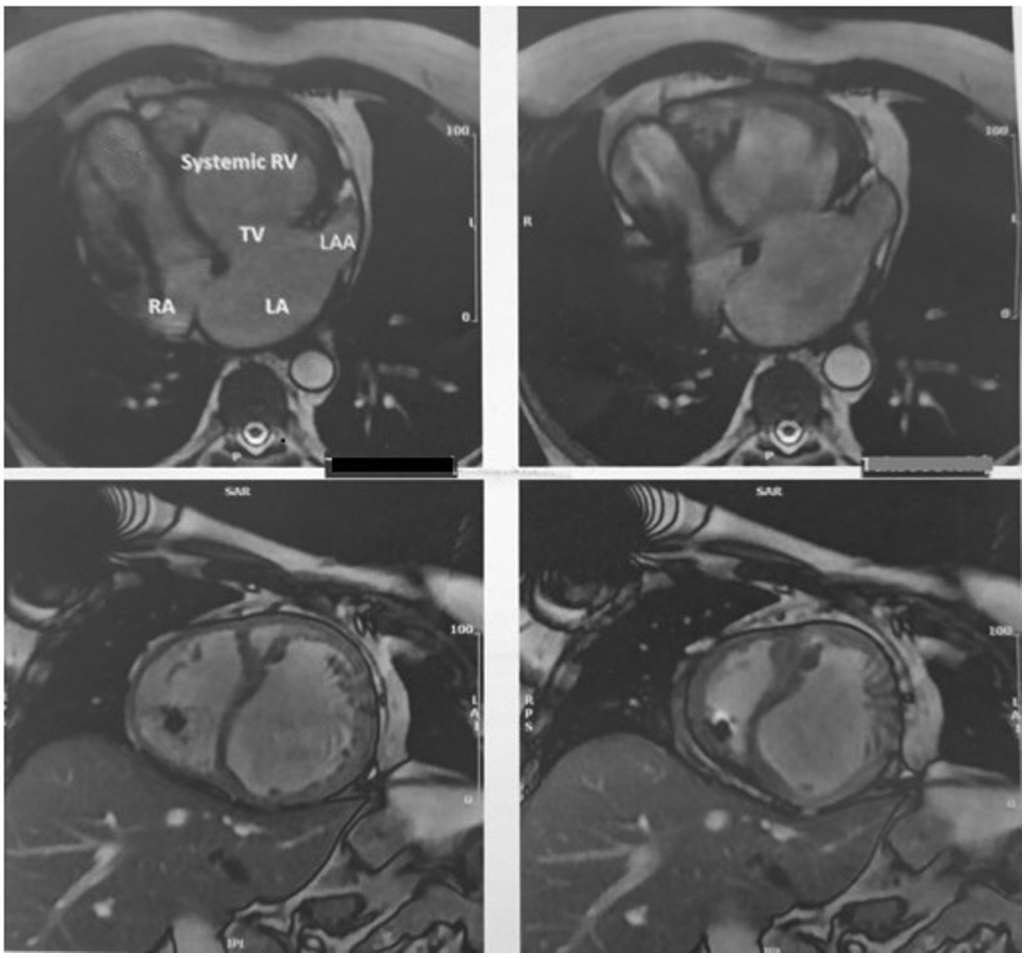
S-ICD = subcutaneous implantable cardioverter-defibrillator

later, persistent atrial fibrillation ensued, and the pacemaker was programmed to VVIR mode.

DIFFERENTIAL DIAGNOSIS

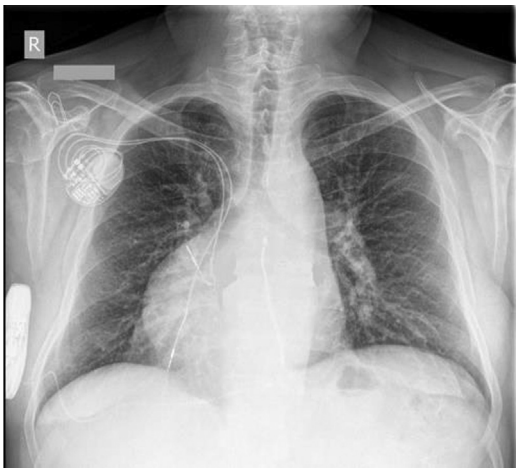
A question raised was whether we might better implant a subcutaneous implantable cardioverter-defibrillator (S-ICD) or upgrade to cardiac resynchronization therapy with a pacemaker-defibrillator (CRT-D). S-ICDs were clinically available, and our impression was that an S-ICD would encompass the whole heart better than a transvenous one, whereas the pacemaker was already in place and functioning well in VVIR mode. Few initial reports in the literature already supported the possibility of right-sided ICDs and the appropriate functioning of S-ICDs together with permanent pacemakers, especially in VVIR mode,[1,2] albeit in left-sided hearts. Another possible option would be to

FIGURE 1 Cardiac Magnetic Resonance Image Conducted Just Before Pacemaker Implantation



Note that the apex of the heart is pointing to the right, and this is a mirror image of a congenitally corrected transposition of the great arteries heart to the right side of the thorax. The right ventricle in an anterior-upper position is connected to the left atrium, not the right atrium, in the context of the congenitally corrected transposition of the great arteries.

FIGURE 2 Right-Sided Subcutaneous Implantable Cardioverter-Defibrillator



A right-sided subcutaneous implantable cardioverter-defibrillator was implanted intermuscularly, with the defibrillator lead on the left. The pre-existing transvenous pacemaker is also right-sided.

upgrade to a transvenous CRT-D system; however, this could be deferred for a later stage of the disease, because no overt functional or echocardiographic deterioration had been observed after pacing. A previously conducted coronary angiography with levophase imaging had not demonstrated any sizable coronary sinus branches, a finding reducing the possibility of a successful CRT-D implantation. Therefore, a right-sided S-ICD was considered preferable. The patient's' body mass index was >25 kg/m^2, potentially being a cause of increased defibrillation resistance.

INVESTIGATIONS

The patient was screened for right-sided S-ICD suitability, with the lead on the left side of the sternum, to encompass the whole heart as much as possible. Because he was pacemaker-dependent, the 100% paced electrocardiogram was tested in the supine and upright position at various gains and pacing outputs (up to 5 V/1 ms, in bipolar and unipolar configurations). The screening templates were positioned at a mirror-like right thoracic position, and 2 of 3 surface electrocardiographic leads (sensing vectors) were considered acceptable, by using Boston Scientific screening templates. Evaluation of QRS morphologies and pacing stimuli at various clinical voltage parameters was performed. The tracings were evaluated by 2 independent reviewers.

MANAGEMENT

A right-sided S-ICD was implanted intermuscularly (model Emblem A219, Boston Scientific) (**Figure 2**). Deep sedation was used, with an anesthesiologist on call. The defibrillation impedance was 130 Ω, and we proceeded to acute testing of the system efficacy by inducing ventricular fibrillation and assessing defibrillation success and prompt pacemaker functioning immediately during and after tachyarrhythmia termination. Testing was also conducted by inducing ventricular fibrillation while the pulse generator was programmed to pace asynchronously at high-output settings (5 V/1 ms)[3] to ensure appropriate detection of ventricular fibrillation by the S-ICD in case the pacemaker undersenses the arrhythmia and is not inhibited by it. The second defibrillation impedance was 140 Ω.

DISCUSSION

Overall experience with simultaneous use of the S-ICD and a permanent pacemaker is limited,[2] especially in very rare cases, such as the case presented here. The prevalence of cCTGA is 1 in 33,000 live births, whereas mesocardia and dextrocardia occur in a minority (15%-20%) of cCTGA cases.[4,5]

Right-sided lead positions have been reported to increase S-ICD eligibility in certain types of congenital heart disease, such as tetralogy of Fallot,[1] whereas the generator remains on the left. For patients with dextrocardia in the setting of cCTGA and heart failure, to our knowledge, no previous experience has been reported.

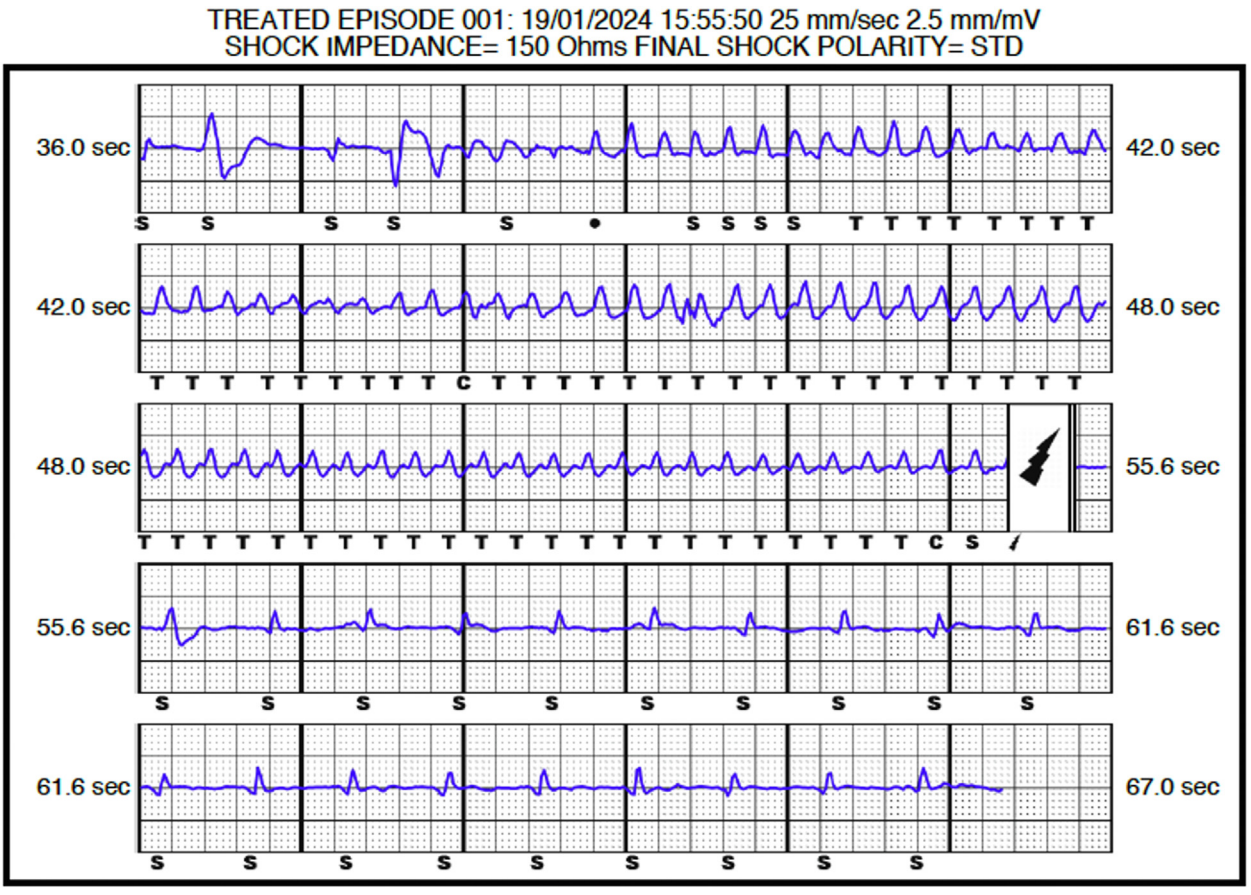
A similar case as ours, although totally right-sided (ie, both lead and can), S-ICD implantation has been reported[6] in a nonpaced patient with dextrocardia. However, the need for pacing complicates the situation, because a potential adverse interaction between the 2 devices might well be life-threatening. In addition, we opted for a left-sided defibrillator lead to encompass the heart as much as possible, although an increased defibrillator resistance might constitute a problem.

Recently, a series of 11 patients was presented,[7] all with complex congenital heart disease, an S-ICD, and a previously implanted pacemaker. In 2 patients, the S-ICD lead position was on the right of the sternum and the S-ICD can position on the left; no patient had dextrocardia.

FOLLOW-UP

One year after S-ICD implantation, the patient reported sudden loss of consciousness and was

FIGURE 3 Electrocardiogram of a Rapid Ventricular Tachyarrhythmia



A rapid ventricular tachyarrhythmia was promptly treated by the defibrillator, whereas pacemaker activity resumed evenly after high-voltage therapy. After successful defibrillation (third electrocardiographic strip, end), the sensed ventricular rhythm (S) is pacemaker-mediated (fourth and fifth electrocardiographic strip). The pacemaker was programmed in VVIR mode, 60-110 beats/min, pacing output 3/0.4 V/ms, bipolar sense 4 mV.

examined in our pacemaker clinic. A rapid ventricular tachyarrhythmia was observed and promptly treated by the defibrillator, and pacemaker activity resumed evenly after high-voltage therapy (Figure 3). The shock impedance was 150 Ω and time to treatment 10 seconds. A pacemaker interrogation showed that this event had not been sensed by the pacemaker.

CONCLUSIONS

In unique situations such as the present case, combined use of an S-ICD and a permanent pacemaker seems to be a relatively safe option, despite the fact that arrhythmia undersensing by the pacemaker may be a possibility. The S-ICD was implanted in a patient with dextrocardia and heart failure due to cCTGA, and this was safe, technically simple, and eventually life-saving.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS **congenitally corrected transposition of the great arteries, dextrocardia, pacemaker, subcutaneous defibrillator**