A case report of pre-implantation feasibility test for combining leadless pacemaker and subcutaneous implantable cardioverter-defibrillator in adult congenital heart disease

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Abstract

New cardiac implantable electronic devices (CIEDs), such as leadless pacemakers and subcutaneous implantable cardioverter defibrillators (S-ICDs), are being used in patients with adult congenital heart disease. The selection of CIEDs often requires careful consideration due to technical challenges posed by a unique heart structure. A 27-year-old man following a surgical tetralogy of Fallot (TOF) repair developed non-sustained ventricular tachycardia, sick sinus syndrome, and complete atrioventricular block. He had a history of recurrent bacteremia. We discussed about the use of a combination of leadless pacemaker and S-ICD as a non-transvenous CIED after considering the infection risk and decided to select the appropriate CIED after a pre-implantation test. Ventricular tachycardia was not induced in the electrophysiological study. Although he did not need an ICD at that point, patients after TOF repair are at a high risk for ventricular tachycardia later in life. We measured the local pacing threshold and R-wave amplitude and performed an S-ICD screening for paced-QRS. Finally, we implanted a leadless pacemaker safely with the option to add an S-ICD if needed. A pre-implantation test could help future decisions regarding combinations of leadless pacemakers with S-ICDs in patients with adult congenital heart disease.

Learning Objectives

The appropriate cardiac implantable electronic devices (CIED) selection in patients with adult congenital heart disease often requires careful consideration. The pre-implantation feasibility test for combining a leadless pacemaker (LP) and a subcutaneous implantable cardioverter defibrillator aided decision-making in CIED selection and safe LP implantation procedure in the unique heart structure.

Introduction

Cardiac implantable electronic device (CIED) selection for adult congenital heart disease (ACHD) often requires careful consideration. There are technical challenges due to the unique heart structure following a repair surgery. Herein, we present a case of non-sustained ventricular tachycardia (NSVT) with bradyarrhythmia after a history of recurrent bacteremia following tetralogy of Fallot (TOF) repair. After a pre-implantation feasibility test, a leadless pacemaker (LP) was chosen, anticipating a future combination with a subcutaneous implantable cardioverter-defibrillator (S-ICD). The pre-implantation test aided decision-making in CIED selection and LP implantation procedure.

Case report

A 27-year-old, post-TOF repair, man underwent pulmonary artery valve replacement three years earlier for prosthetic valve endocarditis caused by *Staphylococcus aureus*. Postoperatively, the patient developed asymptomatic persistent sick sinus syndrome (SSS) and complete atrioventricular block (CAVB) with ventricular escape rhythm with a rate of 35–50 bpm (Fig. 1A) without symptoms of heart failure. At the age of 25, *Staphylococcus aureus* bacteremia recurred. Regardless of a thorough examination, no infection foci were identified. Antibiotic treatment was administered for six months. Although he had no further recurrences of bacteremia, he remained at risk for experiencing bacteremia and pacemaker infection. We refrained from recommending pacemaker implantation. At a routine visit, he exhibited

symptomatic NSVT with a long coupling interval (1220 ms) and an irregular RR interval (530–752 ms; Fig. 1B). No significant heart failure symptoms were noted. The serum N-terminal pro b-type natriuretic peptide (NT-proBNP) level was mildly elevated (188 pg/mL). Transthoracic echocardiogram revealed left ventricular ejection fraction and right ventricular (RV) fractional area change of 62% and 45%, respectively. Mild pulmonary valve regurgitation and mild RV outflow tract obstruction were observed. Cardiac computed tomography revealed pectoris excavatum, a clockwise-rotated heart, an interventricular septum (IVS) protruding toward the RV (Fig. 1C), a small RA, and a dilated RV chamber (Fig. 1D).

We recommended CIED therapy; he met a relative indication for pacemaker implantation because of the bradyarrhythmia. We assessed implantable cardioverter-defibrillator (ICD) indications for post-TOF repair NSVT. Given the recurrent bacteremia and surgical invasiveness, we opted against transvenous and epicardial CIED and explored the feasibility of a combination of LP and S-ICD via cardiac catheterization.

Regarding the electrophysiological study (EPS), we used an 8.5-Fr deflectable sheath (Leftee^{TM}, Japan Lifeline, Tokyo) and a deflectable electrode catheter (EPstar Snake^{TM}, Japan Lifeline) to attain close contact against the IVS (Fig. 2A). Catheter manipulation proved unusual due to a unique anatomy. We performed ventricular tachycardia (VT) induction with burst stimuli (cycle length: up to 240 ms) and single/double extra stimuli (up to S2S3 240 ms) from the RV apex and outflow tract with and without isoproterenol. VT was not induced. After right ventriculography (Fig. 2B), we measured the local pacing thresholds and R-wave amplitude in the IVS (Fig. 2C). No. 3 and No. 8 (Fig. 2B) were within the acceptable range of pacing conditions (Fig. 2C). For potential VT development, we performed S-ICD screening under RV

pacing. Because we anticipated RV pacing-dependent after LP implantation, we had to test T-wave oversensing, and QRS undersensing due to low paced QRS amplitude. The paced-QRS at No. 3 and 8 passed the test in the alternative vector (Fig. 2C). No. 8 placed at the longitudinal mid portion of the high IVS was considered the best LP implantation site because of its lowest pacing threshold.

Since VT was not inducible, we selected an LP (Micra^{TM} AV, Medtronic, Minneapolis, USA) without an S-ICD. We implanted the LP near site No. 8, referring to the right anterior oblique (RAO) 0° and left anterior oblique (LAO) 90° projections (Fig. 2D). The pacing parameters were: threshold, 0.38 V at 0.24 ms pulse width; R-wave amplitude, 8.1 mV; and impedance, 620 ohm. LP implantation was completed (skin to skin: 34 minutes, Fig. 3A). Due to SSS, the pacing mode was set to VVIR 70 ppm (Fig. 3B). The QRS duration decreased from 156 to 140 ms and LP-paced QRS morphology passed S-ICD screening at alternative vector in both supine and sitting positions (Fig. 3C). After discharge, he remained stable without VT events. The RV chamber size and NT-proBNP levels normalized and the LP parameters remained unchanged.

Discussion

The patient following TOF repair experienced NSVT with bradyarrhythmia. Because of a history of recurrent bacteremia, LP implantation was performed, anticipating a future combination with an S-ICD. The pre-implantation feasibility test aided decision-making during CIED selection and enabled safe implantation.

CIED selections

First, we discussed whether CIEDs were needed for his treatment. Although he met the relative indication criteria of a pacemaker, CIEDs implantation had been deferred because of high risk for infection before this admission. The mechanism of NSVT was considered automaticity mediated by bradycardia, which could be suppressed by overdrive pacing [1] and mildly elevated NT-pro-BNP might indicate bradycardia caused cardiac stress. Taken together, we recommended CIEDs therapy. Transvenous CIEDs were avoided due to the history of recurrent bacteremia. [2] Epicardial CIEDs might be inappropriate due to their invasiveness after open-chest surgeries, posing an infection risk. The LP posed a considerably lower risk of infection [3] and could be safely implanted after TOF repair. [4] Despite the presence of SSS and CAVB, we selected VVI-LP because he had normal LV function without significant heart failure symptoms. The IVS near the right bundle branch was considered the optimal implantation site. A retrievable LP, AVEIR^{TM} (Abbott, Abbott Park, IL), might have been desirable rather than Micra^{TM}, because the patient was young and susceptible to infection. However, the first domestic launch of AVEIR^{TM} was three months before the procedure and we had no experience at that time. Furthermore, the procedure was considered unusual due to the patient's cardiac deformity. Therefore, we selected Micra^{TM}. We also discussed post-TOF repair ICD indications. The risk factors for sudden cardiac death following TOF repair include LV systolic or diastolic dysfunction, NSVT, QRS duration ≥180 ms, extensive RV fibrosis, and inducible sustained VT. According to the Japanese guidelines, patients with more than three risk factors are indicated for ICDs. [5] Our patient had only one risk factor (NSVT) and did not meet the ICD indications at that time. Although ICD was unnecessary at the time, the VT risk could increase when patients with TOF become middle-aged. [6] Close observation is warranted.

Pre-implantation feasibility test

The pre-implantation feasibility test aimed to assess appropriate CIED selection, the LP implantation strategy, and the detection ability of S-ICD for LP-paced QRS. [7] This test had three advantages. First, we grew accustomed to catheter manipulation in the unique morphology. Second, we could confirm the optimal fluoroscopic projection angles. In this case, with an extremely clockwise rotated heart, an RAO 0° and an LAO 90° were appropriate. Third, we could identify the target implantation site in advance. The results of EPS and S-ICD screening greatly differed by site, possibly due to local scarring and the artificial patch. This information might minimize the number of LP deployments and reduce complications. The real LP-paced QRS also passed the S-ICD screening, suggesting reliability of the feasibility test results. However, we should recognize three concerns. First, later disease progression can alter the paced QRS morphology, leading to S-ICD detection failure. Second, LP might provide pacing during ventricular tachyarrhythmia due to undersensing, and the pacing artifacts might reset the sensing threshold, causing S-ICD detection delay. This could be tested only by VF induction during real implantation. Third, the current S-ICD and LP are provided by different manufacturers, and their combination is off-label. The results from the MODULAR-ATP clinical trial on the efficacy and safety of intercommunicating LP and S-ICD by the same manufacturer might help enhance feasibility. [8]

In conclusion, the pre-implantation feasibility test was useful for CIED selection and securing LP implantation in a patient with ACHD.

Declaration of competing interest

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Consent

The authors confirm that written consent for the submission and publication of this case report, including images and associated text, was obtained from the patient in line with the Committee on Publication Ethics guidelines.

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Figure legends

Fig. 1: Examination at admission. (A) Twelve-lead electrocardiogram (ECG) at a routine visit. (B) Twelve-lead ECG shows non-sustained ventricular tachycardia (NSVT). The coupling interval and RR intervals during NSVT. (C) Cardiac computed tomography. Horizontal section. (D) Cardiac computed tomography. Volume-rendered image.

Fig. 2: Pre-implantation feasibility test. (A) Electrophysiological test (EPS) using deflectable sheath and electrode catheter. (B) Right ventriculography. The number shows the pacing site. (C) Result of EPS. (D) Fluoroscopic image during leadless pacemaker implantation.

Fig. 3: After leadless pacemaker (LP) implantation. (A) Chest radiography. (B) Twelve-lead electrocardiogram. (C) Result of the subcutaneous implantable cardioverter-defibrillator screening after LP implantation.