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Case Report

Right-sided cardiac resynchronization therapy with defibrillator implantation in a patient with corrected transposition of great arteries and persistent left superior vena cava

Yuka Taguchi (MD), Katsumi Matsumoto (MD), Toshiyuki Ishikawa (MD, FJCC)*, Yutaka Ogino (MD), Hirooki Matsushita (MD), Junya Hosoda (MD)

Department of Cardiology, Yokohama City University Hospital, Yokohama, Japan

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ABSTRACT

Patients with corrected transposition of great arteries (c-TGA) are generally known to develop atrioventricular block, systemic right ventricular dysfunction, and tricuspid regurgitation over time, which are associated with tachyarrhythmia and progressive heart failure.

A 76-year-old man had been diagnosed with c-TGA. He developed a cardiopulmonary arrest while playing tennis, and an automated external defibrillator detected ventricular fibrillation (VF). Immediate cardiopulmonary resuscitation and intensive treatment were performed. He fully recovered without neurological sequelae. QRS duration was 172 ms. Echocardiography showed marked dysfunction and dyssynchrony of the systemic right ventricle (systemic right ventricular end-diastolic diameter/end-systolic diameter = 73/60 mm, systemic right ventricular ejection fraction = 34%). For secondary prevention and treatment of progressive heart failure, cardiac resynchronization therapy with defibrillator (CRT-D) implantation was recommended. Venography via the left cubital superficial vein revealed a persistent left superior vena cava (PLSVC) and giant coronary sinus that did not connect with the right superior vena cava (SVC). Because of the acute angle between the PLSVC and great cardiac vein, we selected a right-sided approach via the right SVC. We were finally able to deliver a coronary sinus lead to the lateral vein. CRT-D implantation can be achieved even in patients with c-TGA and PLSVC.

<**Learning objective:** CRT-D implantation can be achieved even in a patient with c-TGA and PLSVC.>

Introduction

Patients with congenital heart disease (CHD) who survive to adulthood constitute a growing group, due to improvements in treatment over the past few decades. This improved survival also results in long-term complications such as progressive heart failure, malignant ventricular arrhythmias, and sudden cardiac death. Ventricular dyssynchrony appears to be common, and appears early in the history of the disease. Cardiac resynchronization therapy has been reported as a potential treatment option for CHD patients [1]. However, due to the specific coronary sinus anatomy, placing a coronary sinus lead is associated with technical difficulty in this population [2].

We here present a case of cardiac resynchronization therapy with defibrillator (CRT-D) implantation in a patient with corrected transposition of great arteries (c-TGA) and a persistent left superior vena cava (PLSVC), for secondary prevention and treatment of progressive heart failure.

Case report

A 76-year-old man was diagnosed with corrected transposition of great arteries (c-TGA) 10 years previously. He first experienced an episode of syncope in 2005. His second syncopal attack was cardiopulmonary arrest while playing tennis in October 2015, and cardiopulmonary resuscitation was performed immediately. An automated external defibrillator detected ventricular defibrillation, and shocks were delivered four times. After intensive treatment, he fully recovered without neurological sequelae.

* Corresponding author at: Department of Cardiology, Yokohama City University Hospital, 3-9, Hukuura, Kanazawa-ku, Yokohama-shi, Kanagawa-ken, Japan. Fax: +81 45 701 3738.
E-mail address: tishika@yokohama-cu.ac.jp (T. Ishikawa).

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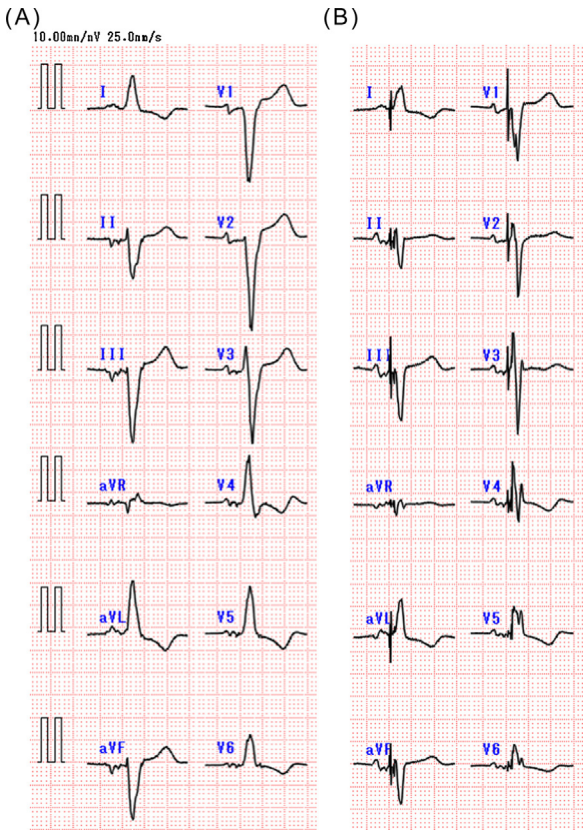

Fig. 1. 12-Lead electrocardiogram on admission showed that QRS duration was 172 ms, left bundle branch block, and QTc was 0.513 (A). After cardiac resynchronization therapy with defibrillator implantation, QRS was shortened to 137 ms (B).

Fig. 2. Venography via left cubital superficial vein showed persistent left superior vena cava that did not connect with the right superior vena cava and flowed into the giant coronary sinus.

Electrocardiography showed complete left bundle branch block, and QRS duration was 172 ms (Fig. 1).

Echocardiogram showed dysfunction and marked eyeball dyssynchrony of the systemic right ventricle (RV) with septal flash, and moderate tricuspid regurgitation. Systemic RV end-diastolic diameter and end-systolic diameter (sRVDd/Ds) were 73 and 60 mm. Ejection fraction of the systemic RV (sRVEF) was 34%. Coronary angiography showed no coronary artery stenosis. Venography via the left cubital superficial vein revealed a persistent left superior vena cava (PLSVC) that did not connect with the right superior vena cava (SVC) and flowed into the giant coronary sinus (Fig. 2).

The patient had no other cardiac anomalies such as ventricular septal defect, pulmonary stenosis, or dextrocardia. He had not undergone cardiac surgery previously.

Amiodarone administration at a dose of 200 mg per day, and medication for heart failure including carvedilol were started.

CRT-D implantation was recommended for secondary prevention and treatment of progressive heart failure.

We performed coronary artery angiography before the implantation to identify the great cardiac vein on coronary sinus phase.

Because the PLSVC made an acute angle with the great cardiac vein (Fig. 3A), we selected a right-sided approach via the right SVC for delivery of the coronary sinus lead.

CRT-D [QUADRA ASSURA 3367-40QC CRTD, St. Jude Medical (SJM), St. Paul, MN, USA] implantation was performed.

The shock lead was fixed at the systemic left ventricular apex (7122Q-58, SJM), and the atrial lead was fixed at the right atrial appendage (LPA1200M-52, SJM).

After that, we positioned the guiding sheath (CPS Direct™ Universal, right-sided, SJM) into the giant coronary sinus ostium, and inserted an inner guiding catheter (CPS Aim™ Universal, 90°, SJM) through the guiding sheath, which was used to cannulate the great cardiac vein without difficulty. The great cardiac venography was performed via inner catheter. We identified the lateral vein which was the most appropriate branch. After proceeding the wire to the branch, the coronary sinus lead (1458-86, SJM) was finally delivered to the lateral branch over the wire.

The coronary sinus lead (1458Q-86, SJM) was successfully positioned in the lateral vein through an inner guiding catheter, which was used to cannulate the great cardiac vein without difficulty (Fig. 3B). A defibrillation test was successfully performed during the operation.

After biventricular pacing, QRS duration was shortened to 137 ms (Fig. 1B). In the echocardiographic findings, sRVEF (sRVDd/Ds = 72/57 mm, sRVEF = 41%), and septal flash were improved, and moderate tricuspid regurgitation was slightly improved to mild–moderate.

During a year of follow-up, he was in New York Association 2, which was no significant change before and after implantation; however, ventricular arrhythmia has not been observed.