Transvenous shock-only implantable cardioverter defibrillator for tricuspid valve atresia after an atrio-pulmonary Fontan surgery

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Abstract

A 42-year-old woman with tricuspid atresia who underwent a Fontan surgery **(**atrio-

pulmonary connection) was admitted to our hospital due to symptomatic ventricular tachycardia. A defibrillation lead was implanted in a distal site of a coronary vein since there was no usual entry to the ventricle. Ventricular pacing was impossible due to the high threshold, however, good sensing was obtained. Three years later, she felt palpitations and a subsequent shock therapy while climbing stairs. The cardioverter data showed that an appropriate cardioversion therapy successfully converted ventricular tachycardia to normal rhythm.

Keywords: Fontan surgery, atrio-plumonary connection, ventricular tachycardia, implantable cardioverter defibrillator, coronary venous lead

1. Introduction

Ventricular tachycardia (VT) rarely occurs in patients after a Fontan surgery, however, it can be fatal.[1] Placement of an implantable cardioverter defibrillator (ICD) lead in the ventricle is not possible because there is no entry in the presence of a Fontan with an atrio-pulmonary connection (APC). Some case reports have previously documented the placement of an ICD lead in coronary veins (CV) after tricuspid valve surgery.[2] To the best of our knowledge, we for the first time report a case of a transvenous ICD implantation with a defibrillation lead into the CV for tricuspid atresia after a Fontan surgery, and demonstrated a successful therapy with a shock-only ICD in such a difficult situation.

2. Case report

A 42-year-old woman with tricuspid atresia had undergone an APC-Fontan surgery at the age of 6 years old. The patient presented with frequent attacks of AT (Figure 1A) and sick sinus syndrome (SSS) (Figure 1B) from 26 years old. She had an episode of palpitations with faintness and dyspnea at 42 years old and the 12-lead electrocardiogram revealed a sustained VT with a cycle length of 270 ms (Figure 1C). We considered that she needed permanent atrial pacing for SSS with an atrial anti-tachycardia pacing (ATP) feature for AT, and an ICD for VT. Three-dimensional computed tomography showed a huge right atrium (RA) and dilated coronary sinus ostium (Figure 2A, 2B). Cardiac catheterization revealed a high central venous pressure of 13 mmHg, normal coronary arteries without any stenosis, and multiple small CV branches to the coronary sinus. We decided to implant an ICD system with transvenous atrial and ventricular leads instead of surgically-placed epicardial leads to reduce the risk of the procedure.

We inserted a FINELINE II Sterox EZ lead (Boston Scientific, Marlborough, MA,

USA) and anchored it to the inferior septum of the RA with an atrial pacing threshold of 2.0 V at 0.4 msec (Figures 2C, 2D). Selective CV angiography revealed a large ostium and small CV posterior branch with a defect of the great CV. A Durata™ ICD lead (Abbott, St. Paul, MN, USA) was advanced into the distal portion of the CV branch (Figures 2C, 2D). Although acceptable ventricular sensing with an amplitude of 5.8 mV and successful defibrillation with 10 joules were achieved, ventricular pacing was impossible at a pacing output of 10 V. Then, we implanted an Evera® XT ICD DR (Medtronic, Minneapolis, USA), resulting in full atrial pacing with the automatic atrial ATP programmed for an AT of more than 150 bpm and a single therapy zone for ventricular tachyarrhythmias of more than 180 bpm without any ventricular ATP programming. Anticoagulation therapy with warfarin was started to prevent any thrombotic complications after the ICD implantation.

Three years later when she climbed stairs, she had palpitations and faintness and felt a subsequent shock from the cardioversion. The ICD telemetry data showed an appropriate cardioversion therapy for VT (Figure 3). The atrial pacing was stable, and the AT attacks were successfully terminated by the automatic atrial ATP over four years after the ICD therapy.

3．Discussion

To the best of our knowledge, this is the first report demonstrating a successful transvenous ICD implantation with a CV lead in a patient with tricuspid atresia who underwent an APC-Fontan surgery and had an episode of an appropriate ICD therapy. The epicardial ICD implantation technique has been used and Cannon et al. reported the surgical placement of an ICD coil directly in the pericardial sac with no pacing or sensing capabilities.[4] In the epicardial lead placement, a repeat thoracotomy can be associated with significant risks and failures. Furthermore, our patient was subjected not only to VT but also to SSS and

AT, which required permanent atrial pacing and atrial ATP. The endocardial ICD system with automatic atrial ATP was considered better for our patient in this difficult situation even if the ventricular pacing capability could not be achieved.

A technique of an ICD lead implantation into the middle cardiac vein has been described.[2] In our patient, the anatomical difficulty of the CV prevented the ideal implantation of an ICD lead for ventricular pacing and defibrillation. However, the shock-only ICD could successfully treat the VT with an appropriate shock, and additionally, the SSS and AT with atrial pacing.

The long-term safety of a defibrillating coil lead in the CV is unknown. CV thromboses may be problematic following invasive right heart procedures such as a central venous catheter placement, CV cannulation, and catheter ablation in Fontan patients.[5] Anticoagulation with warfarin can be reasonable for preventing CV thromboses and other thromboembolic complications.

In conclusion, our case report showed that the implantation of a transvenous atrial lead and CV lead was feasible, and that a shock-only ICD functioned successfully to treat VT in a patient with a difficult anatomy.

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Figure Legends

Figure 1A: Twelve-lead ECG recorded during atrial tachycardia.

Figure 1B: Twelve-lead ECG recorded during bradycardia due to sinus node dysfunction.

Figure 1C: Twelve-lead ECG recorded during ventricular tachycardia.

Figure 2A: Three-dimensional computed tomographic anteroposterior view of the cardiac chambers and aorta (Ao).

Figure 2B: Three-dimensional computed tomographic anteroposterior view of the dilated right atrium (RA), dilated coronary sinus (CS), and small posterior coronary vein.

Figures 2C: Chest radiograph in the anteroposterior view after successful atrial and ventricular pacing through the coronary vein. The white arrow shows the atrial lead. The black arrow shows the implantable cardioverter defibrillator (ICD) lead.

Figure 2D: Three-dimensional computed tomographic anteroposterior view after the device implantation. The white arrow shows the atrial lead. The black arrow shows the ICD lead.

PA pulmonary artery, SVC superior vena cava, IVC inferior vena cava, LV left ventricle

Figure 3: Intracardiac electrogram during ventricular tachycardia (cycle length 320 ms) using a device programmer. The red circle shows an appropriate cardioversion therapy with 36 J.

A atrium, V ventricle