Case Report

Implantable Cardioverter Defibrillator in a Patient with Eisenmenger Syndrome after Senning Repair for Transposition of the Great Arteries

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An implantation of a cardioverter-defibrillator was attempted in a 32-year-old man with atrial tachycardia, ventricular tachycardia and sinus node dysfunction. He had undergone a Senning operation and half closure of ventricular septal defect in order to correct a transposition of the great arteries. Cardiac catheterization revealed severe pulmonary hypertension and Eisenmenger syndrome. Prior knowledge of the complex cardiac anatomy obtained by magnetic resonance imaging helped in determining the suitable site for implanting the leads and planning the procedural strategy. With repletion of a large amount of saline and oral anticoagulation with warfarin, no complications related to thromboembolism occurred during a 10-month follow-up period.
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Introduction

Arrhythmias are common and generally poorly tolerated in patients with Eisenmenger syndrome and congenital heart disease.[1–3] However, implantation of an implantable cardioverter-defibrillator (ICD) is technically challenging because of the complex anatomy, atrial scar, and presence of artificial tissue. We describe here a case of atrial tachycardia (AT), ventricular tachycardia (VT) and Eisenmenger syn- drome after a Senning's operation for a transposition of the great arteries (TGA) (type II) in which an ICD was successfully implanted under the guidance of magnetic resonance imaging (MRI).

Case Report

A 32-year-old Japanese man was transferred to our hospital for treatment of AT and VT. At the age of 3, he was diagnosed with TGA and underwent

pulmonary artery banding because his pulmonary artery pressure was elevated due to an increased pulmonary blood flow through a large ventricular septal defect (VSD). Six years later, he underwent an atrial switch operation with a Senning operation and half closure of the VSD because the pulmonary vascular resistance had significantly increased to 2024 dyne·s·cm^{-5} on the cardiac catheterization.

On February 1, 2008, he had a sudden palpitation attack, and was admitted to a hospital. The diagnosis of AT with 2:1 A-V conduction was made, and a 12-lead ECG revealed a sustained wide QRS tachycardia with a right bundle branch block configuration, north-west axis, and heart rate of 134 bpm (**Figure 1A**). Intravenous treatment with verapamil and adenosine triphosphate could not terminate the AT (**Figure 1B**), and the AT was finally terminated by direct current cardioversion (**Figure 1C**). Two days after the first AT episode, the patient developed a sustained monomorphic VT with a right bundle branch block configuration, left axis

deviation, and cycle length of 280 msec (**Figure 2**), and was also terminated by direct current cardioversion. Treatment with 200 mg/day of oral amiodarone failed to control both tachycardias, but it resulted in sinus bradycardia.

He was referred to our hospital for further evaluation and treatment of the tachycardias. Despite a reduction in the amount of amiodarone to 100 mg/day, successive 12-lead ECGs after admission demonstrated a sinus bradycardia or junctional escape rhythm with a heart rate of 40 to 45 bpm. Echocardiography disclosed that the left ventricular wall motion was mildly impaired with an ejection fraction of 50%. Right heart catheterization revealed severe pulmonary arterial hypertension with a pulmonary arterial pressure of 105/50 mmHg (mean, 77 mmHg). The anatomical left ventricular and anatomical left atria pressures were 110/13 mmHg and 6/8 (6) mmHg, respectively. A left heart catheterization demonstrated an aortic pressure of 111/70 (87) mmHg. The cardiac index was 2.47
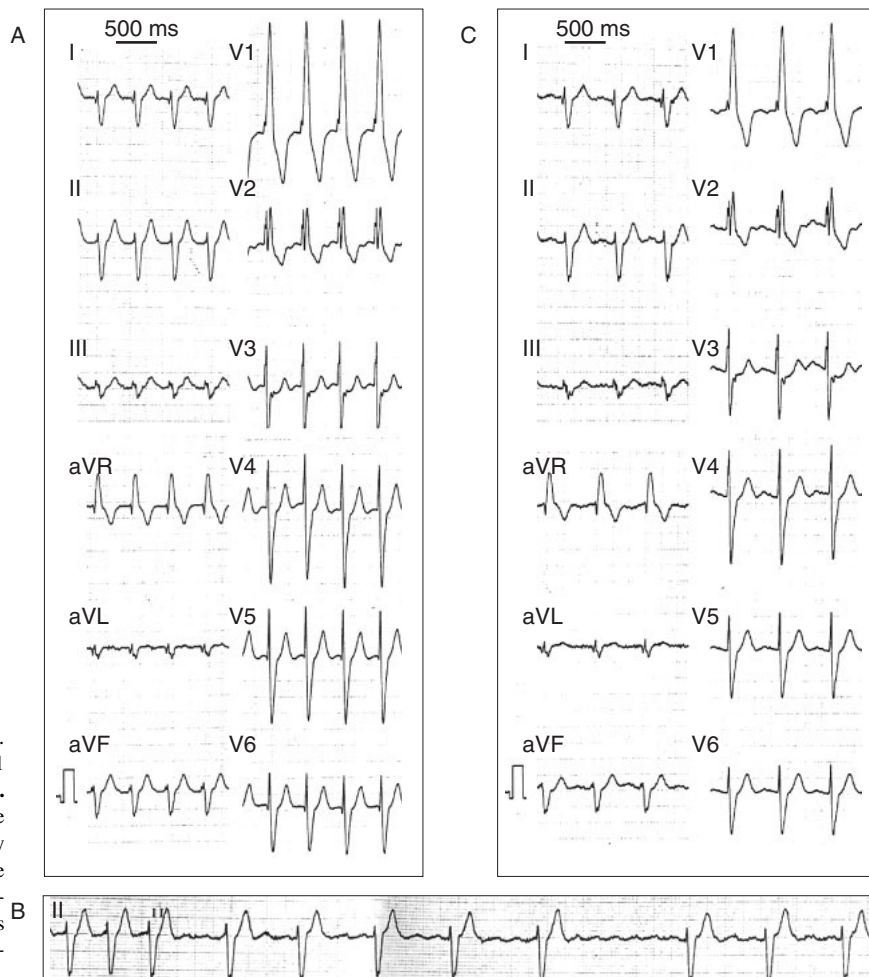

Figure 1 Twelve-lead ECGs. **A.** Atrial tachycardia with 2:1 atrioventricular conduction. **B.** Small but distinct P waves (the P-P interval was approximately 220 ms) were observed after the application of 20 mg of adenosine triphosphate. **C.** Sinus rhythm after external cardioversion.
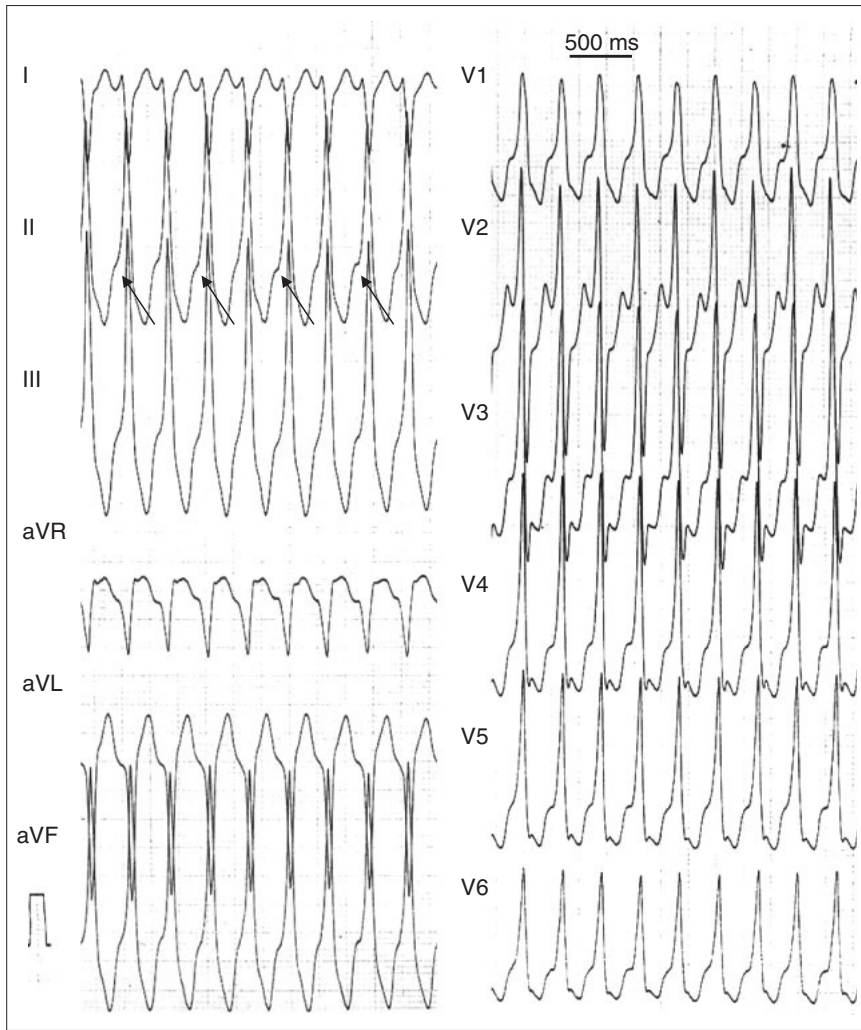
Figure 2 Twelve-lead ECG of a monomorphic sustained ventricular tachycardia.
P waves, independent of the QRS complex, were observed in lead II (arrows).

$L/min/m^2$, and the left-to-right and right-to-left (venous blood from the anatomical left ventricle to the right ventricle) shunt ratios were 0.67 and 0.18, respectively. Measurement of the arterial blood gases revealed a pH of 7.528, CO_2 tension of 28.8 mmHg, O_2 tension of 54.9 mmHg and O_2 saturation of 91.7%. Based on those results, the patient was diagnosed with Eisenmenger syndrome. Treatment with oral angiotensin converting enzyme inhibitors and bosentan was started and total doses were gradually increased.

We decided to implant an ICD device with a dual-chamber (DDD) pacing function, transvenously under local anesthesia to resolve the symptoms from his tachycardias and sinus nodal dysfunction. Considering the complex anatomy and the risk of thromboembolic complications, an electrophysiological study and catheter ablation were avoided in this case. Fortunately the AT and VT were completely suppressed with oral medications at that time. Before the operation, cardiac MRI was performed to determine the exact complex anatomy and the appropriate site for implanting the atrial pacing lead. The MRI images disclosed that the superior vena cava (SVC) was connected to the morphological left atrium (LA; **Figure 3A**), and that the roof area of the LA seemed to be the most appropriate site for the implantation of the atrial pacing lead because of the presence of an almost completely intact myocardium and the absence of any complicated structures around that area (**Figure 3A**). The morphology and appearance of the morphological left ventricle was also clearly seen in those images.

The patient underwent an implantation of a dual-chamber ICD (Virtuoso DR D164AWG, Medtronic, Minneapolis, MN, USA). Using a left-sided approach, a subclavian venous access was utilized. Angiography of the SVC was performed in multiple directions with special attention being paid to the SVC–LA junction. Under guidance with MR and
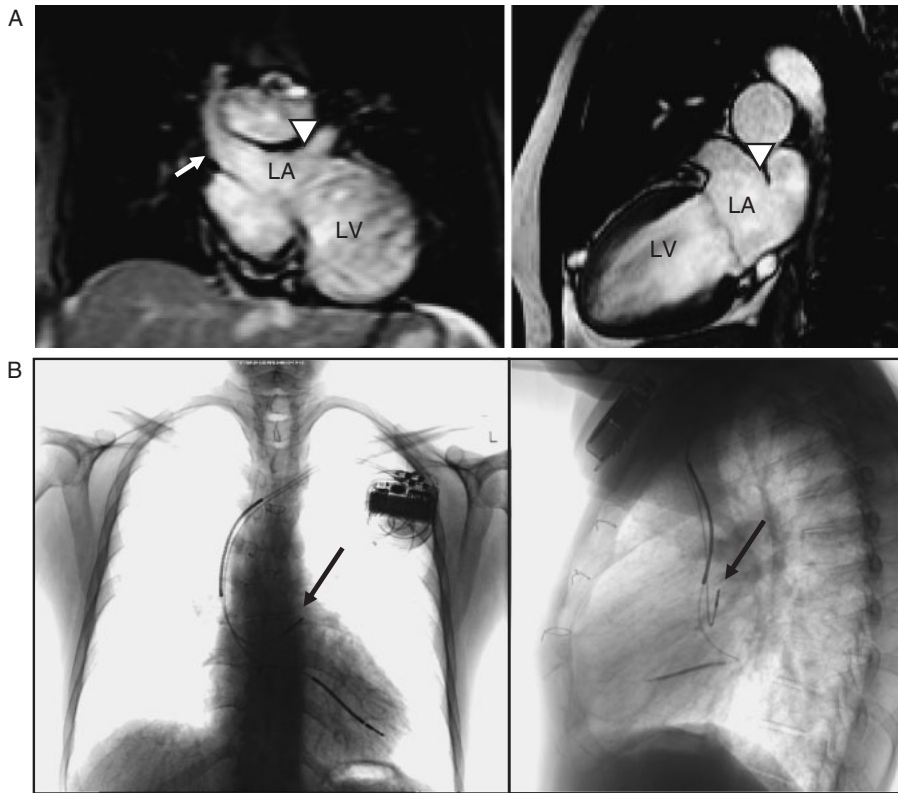
Figure 3 A. Images of balanced turbo-field echo cardiac magnetic resonance imaging (MRI) before the operation.
Coronal (left) and sagittal (right) cardiac MRI images demonstrate a superior vena cava to left atrium (LA) conduit (arrow). The arrowheads indicate the site where we planned to implant an atrial pacing lead in the LA roof.
B. Posterioranterior (left) and lateral (right) views of the chest X-ray film after the operation.
Note that the atrial lead (arrow) was implanted at the site which we determined it should be implanted using cardiac MRI guidance prior to the operation.

angiographic images and fluoroscopy, an active-fixation atrial lead was inserted into the LA, and the atrial lead (Tendril 1488T 52 cm, St. Jude Medical, St. Paul, MN, USA) was implanted at the exact site of the LA roof that we found to be an appropriate site for the implantation via the MRI (**Figure 3A**). The ICD lead (Riata ST OPTIM 7020, St. Jude Medical) was also placed in the morphological left ventricle with no difficulty (**Figure 3B**). The stability of both leads was excellent. The atrial pacing threshold was 0.9 V at a pulse width of 0.4 ms and the ventricular pacing threshold was also good at 0.5 V at a pulse width of 0.4 ms. The P wave and R wave sensing was good at 3 mV and 14 mV, respectively. There was no diaphragmatic stimulation either with atrial or ventricular pacing at high output. No far-field sensing was observed after connection to the device. Rapid ventricular fibrillation was induced by a T-wave shock and the events were successfully defibrillated with an energy of <20 J. The device was programmed as follows: (i) VT detection = 133–140 bpm for monitoring purposes only; (ii) VT detection = 141–187 bpm, therapies = anti-tachycardia pacing × 2, 20J × 1, 35J × 3 and (iii) VF detection = 188 bpm, therapies = 35J × 6. The total procedure time was 180 minutes, and there were no procedural compli-

cations. To avoid dehydration and any thromboembolic complications from the ventricular endocardial lead placement, a large amount (200 mL/h) of intravenous saline was administered during the operation. Warfarin was administered from the next day after the operation in order to keep the PT-INR (prothrombin time-internationalized ratio) at or above 2.5. A dose of 100 mg/day of aspirin was also given to the patient. The patient has done well with no recurrence of any tachycardia or potential complications including a thromboembolism during a 10-month follow-up period.

Discussion

To the best of our knowledge, this case report demonstrated for the first time that an ICD implantation was possible with no potential complications in a patient with Eisenmenger syndrome after a Senning's operation for a TGA, and that prior knowledge of the complex cardiac anatomy obtained with the MRI helped in determining the suitable site for the insertion of the pacemaker lead and in planning the procedural strategy.

Before the 1980s, most patients with TGA were treated by an atrial inflow correction using a Mustard or Senning operation. While their survival rate has

been improving, arrhythmias and late deaths are well-recognized complications.[4,5] Sudden death has been the common cause of late deaths and AT and VT have been identified as predictors of sudden death.[4,5] Sinus node dysfunction is also common in adult patients with a TGA, and sinus rhythm has been reported to be preserved in only 40% of the patients at 20 years of age.[6] An Eisenmenger physiology is a serious complication of an atrial switch repair for a TGA and occurs in approximately 7% of those who survive to adulthood.[6] Symptoms of tachycardia or a previously documented supra-ventricular tachycardia are the best predictors of sudden cardiac death,[1] and aggressive therapy with antiarrhythmic medications, pacing, or even an ICD implantation may be recommended. However, a recent study reported that neither drug therapy nor pacemakers reduce the risk of sudden death in patients with a TGA who have undergone an atrial inflow correction operation.[5] In our case, AT, VT and sinus node dysfunction were documented, and an Eisenmenger physiology was also present. Surgery and general anesthesia are well-known critical factors associated with the deterioration and death related to Eisenmenger syndrome.[2] Therefore, we think that the transvenous implantation of an ICD device with DDD pacing was indispensable for preventing any sudden death in this case.

The risk of thromboembolic complications from the ventricular endocardial lead placement was a major concern in our patient. A paradoxical embolism due to the ventricular endocardial lead placement might be a potential source for a thromboembolism, which might result in neurological complications. With repletion using a large amount of saline and oral anticoagulation with warfarin, no complications related to thromboembolisms occurred during the operation and the follow-up period in this case.

The device implantation in this case was technically challenging because of the unusual orientation of the cardiac chambers. The MRI can be used to determine the exact intracardiac anatomy and confirm the location and size of the intracardiac or extracardiac communications, especially in patients with a complex cardiac anatomy and inadequate echocardiographic windows.[7] Compared to ECG-gated computed tomography, this imaging exposes the patient to no ionizing radiation.[8] Furthermore, it can also provide information on the cardiac function and characteristics of the myocardium. In our case, prior knowledge of the cardiac anatomy from the MRI was very helpful in determining the site for the implantation of the atrial lead and in planning the procedural strategy. Therefore, we think that cardiac MRI is a very useful non-invasive method for use prior to the device implantation in patients with a complex cardiac anatomy as in this case, and a detailed assessment of the MRI images before the operation could result in a smooth, safe and successful device implantation without any complications in these patients.

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