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CASE REPORT
Congenital heart disease

Successful transvenous implantable cardioverter-defibrillator implantation supported by preceding 3D electro-anatomical mapping for a ventricular fibrillation survivor with surgically repaired congenitally corrected transposition of the great arteries: a case report

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Background The atrial sites suitable for lead placement are limited after complex surgical atrial procedures, and lead placement can be challenging in patients with congenitally corrected transposition of the great arteries (ccTGA) after intracardiac repair.

Case summary A 34-year-old man with ccTGA, who had undergone a double-switch operation with combined Senning and Jatene operations at the age of 14 was transferred to us. He experienced faintness and suffered cardiopulmonary arrest, and electrocardiography revealed ventricular fibrillation. After conversion to sinus rhythm by urgent external defibrillation, sinus bradycardia was revealed. Electrophysiological study was done using a three-dimensional (3D) mapping system (Ensite®) to evaluate the electrical condition of atria and to decide whether atrial lead can be transvenously placed. The electrical potential of the functional right atrium was good in the lateral or posterior wall, but the threshold was high. By contrast, the roof of the functional right atrium beyond cavoatrial junction was characterized by low voltage, but in a limited region of the roof of right atrium, the threshold was satisfactory and the electrical potential was normal. Thus, 3 weeks later, we implanted a transvenous implantable cardioverter-defibrillator (ICD). We used a 3D mapping system to place the atrial lead in the limited region of the roof of the right atrium mentioned above, the threshold was 0.7 V.

Discussion Electrophysiological examination using a 3D mapping system before implantation of a dual-chamber ICD is useful because atrial sites suitable for lead placement are limited in patients.

Keywords adult congenital heart disease • congenitally corrected transposition of the great arteries • implantable cardioverter-defibrillator • ventricular fibrillation • electrophysiological study • case report

ESC Curriculum 9.7 Adult congenital heart disease • 5.10 Implantable cardioverter-defibrillators

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Learning points

- Sinus node dysfunction can be frequently encountered after extensive atrial three surgery, such as atrial switch following Senning or Mustard procedure.
- In complex congenital heart disease, electrophysiological study using a three-dimensional mapping system prior to the implantation of a rhythm device can be useful to identify the most appropriate location for atrial lead implantation.

Introduction

Patients with congenitally corrected transposition of the great arteries (ccTGA) can develop atrioventricular block related to characteristic anatomical course of conduction axis and/or to surgical closure of ventricular septal defect at the rate of 2% per year,[1] and many of those patients require pacemaker implantation.[2] On the other hand, there have been few reports of fatal ventricular arrhythmia in patients with ccTGA in the natural history of the disease.[3–6] The atrial sites suitable for lead placement are limited, and it can be anticipated that lead placement can be challenging in patients with ccTGA after double-switch operation as well as atrial switch operation for complete TGA. We report a case in which a preoperative three-dimensional (3D) mapping was useful for implantation of a dual-chamber implantable cardioverter-defibrillator (ICD) for secondary prevention due to ventricular fibrillation and sinus bradycardia in a patient with ccTGA 20 years after his double-switch operation.

Timeline

Dates	Events
Day 0	Out of hospital cardiac arrest due to ventricular fibrillation, with successful resuscitation
Day 18	Transfer to our hospital
Day 36	Electrophysiological study using a 3D system
Day 57	Implantation of a dual-chamber implantable cardioverter-defibrillator
Day 66	Discharged home

Case presentation

A 34-year-old man with ccTGA collapsed and suffered cardiopulmonary arrest. He underwent bystander cardiopulmonary resuscitation and was transported to the hospital where ventricular fibrillation was confirmed (*Figure 1*). He was immediately converted to sinus rhythm by external defibrillation. A secondary prevention ICD implantation was indicated and the patient was transferred to our tertiary hospital. Physical examination revealed a systolic murmur (3/6 on the Levine grading scale), without any other clinical manifestations of heart failure. His blood pressure was 106/62 mmHg. Electrocardiography (ECG) revealed sinus bradycardia with a heart rate of 42 b.p.m. (*Figure 2*). Twenty-four-hour Holter ECG revealed an average heart rate of 48 b.p.m., maximum heart rate of 59 b.p.m., and minimum heart rate of 38 b.p.m. Echocardiogram showed that reduced systolic function of both ventricles (left ventricular ejection fraction was 36%, right ventricular ejection fraction was 42%, and tricuspid annular plane systolic excursion was 18 mm), but there was no significant valve regurgitation or haemodynamically significant valve vitia.

He had undergone pulmonary artery banding operations twice as a child, and then underwent a double-switch operation with combined Senning and Jatene operations at the age of 14. He did not take any medications after cardiac surgery; echocardiograms were followed up about once a year until age 31 (last right ventricular ejection fraction was about 50% and relatively well preserved). Our contrast-enhanced computed tomography scan revealed that the superior vena cava and inferior vena cava were connected to the anatomical right ventricle via Senning pathway (*Figure 3*). Coronary artery angiography revealed no stenosis or anomaly of coronary arteries. We also conducted cardiac magnetic resonance imaging scan. The results revealed left ventricular ejection fraction around 30%, right ventricular ejection fraction difficult to assess, and no apparent scar. The adult congenital heart disease team decided to implant a dual-chamber ICD for ventricular fibrillation and sinus bradycardia. Implantation of the atrial lead was expected to be challenging, so it was decided to perform an electrophysiological study prior to the ICD implantation. This was also expected to reduce the implantation procedure time and thereby reduce the risk of procedure-related complications. We performed an electrophysiological study using a 3D mapping system (Ensite®) to determine the best placement site for the atrial lead.

Right femoral vein was cannulated and two sheaths were inserted. Two quadripolar electrode catheters were positioned in the right atrium and right ventricular apex, respectively. A 3D substrate map was performed in sinus rhythm. Then, we evaluated the function of sinus node and atrioventricular node by atrial pacing. The paced cycle length of atrioventricular nodal Wenckebach-type block was 620 ms. Pacing from the right atrium at a basic cycle length of 500 ms for 30-s showed that the maximum sinus node recovery time was 1728 ms. Finally, we checked electrical potential and the threshold of the right atrium. The electrical potential of the right atrium was relatively good, but the threshold was high overall [about 3 volts (V)]. The roof of the right atrium was mostly with low voltage, with the exception of a scattered limited region where the threshold was 1 V and the sensed electrical potential was normal (*Figure 4*, black arrow). Three weeks later, a dual-chamber ICD implantation was performed (Fortify Assura DR, St Jude medical; atrial lead, 2088TC-52; shock lead, 7120Q-58). We implanted a shock lead in the right ventricle and then introduced an atrial lead to the right atrium. The thresholds at the free wall side and bottom of the right atrium were high, so we placed the atrial lead at the roof of right atrium beyond cavoatrial junction using a J-shaped stylet as third attempt to implant atrial lead. The threshold there was 0.7 V. We did not perform defibrillation threshold testing. The device was programmed in DDD mode at 60–90 b.p.m. with two zones of therapy; ventricular tachycardia: ≥171 b.p.m., anti-tachycardia pacing + shock therapy [anti-tachycardia pacing × 6, 36 joule (J), 40J ×2]; ventricular fibrillation: ≥200 b.p.m., shock therapy with anti-tachycardia pacing while charging (36J, 40J × 5). *Figure 5* shows a chest X-ray 1 week after the ICD implantation. The slack did not reduce after then. We introduced bisoprolol 2.5 mg and continued after his discharge. The patient was discharged without no complications in his post-operative course.

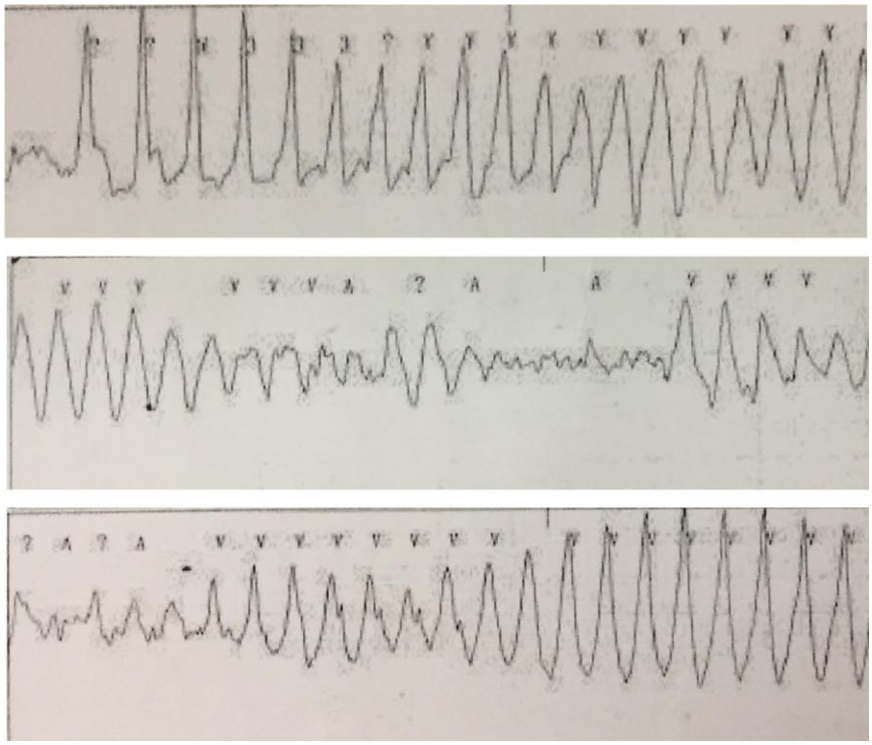


Figure 1 Monitoring electrocardiogram revealed ventricular fibrillation emergency on admission.

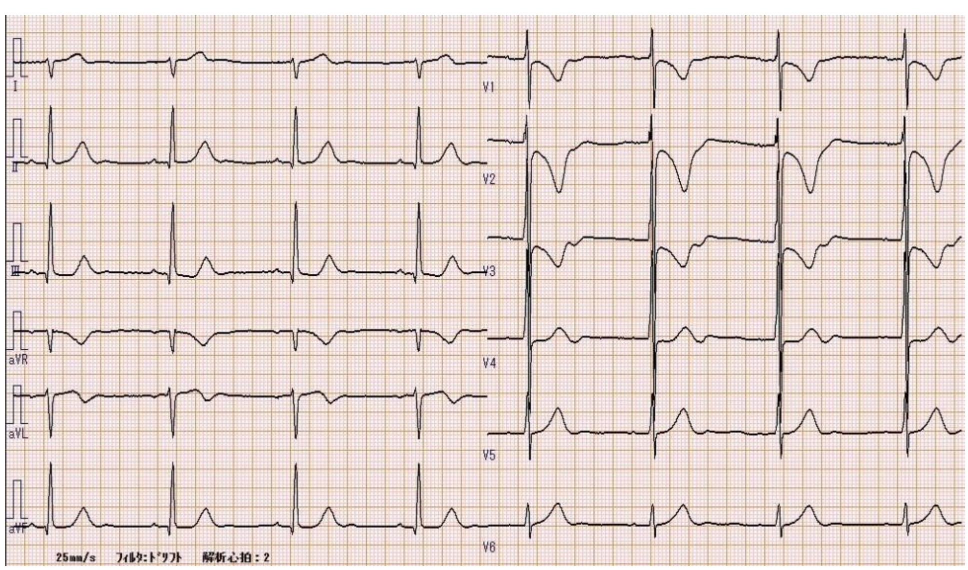


Figure 2 Electrocardiography after cardioversion revealed sinus bradycardia with low atrial voltage.

We checked the parameters of the pacemaker and leads 5 months after implantation. The amplitude of the sensed *P* wave was 5.0 mV or more and the pacing threshold was 0.5 V. The percentage of atrial pacing was 18% and that of ventricular pacing was 1%. Two years after implantation, the first appropriate therapy (anti-tachycardia pacing) was delivered for ventricular tachycardia. The patient was asymptomatic. A monomorphic ventricular tachycardia with a cycle length of 330 ms was terminated by a sequence of anti-tachycardia pacing (a burst of eight ventricular paced beats at 88% of the cycle length of ventricular tachycardia).

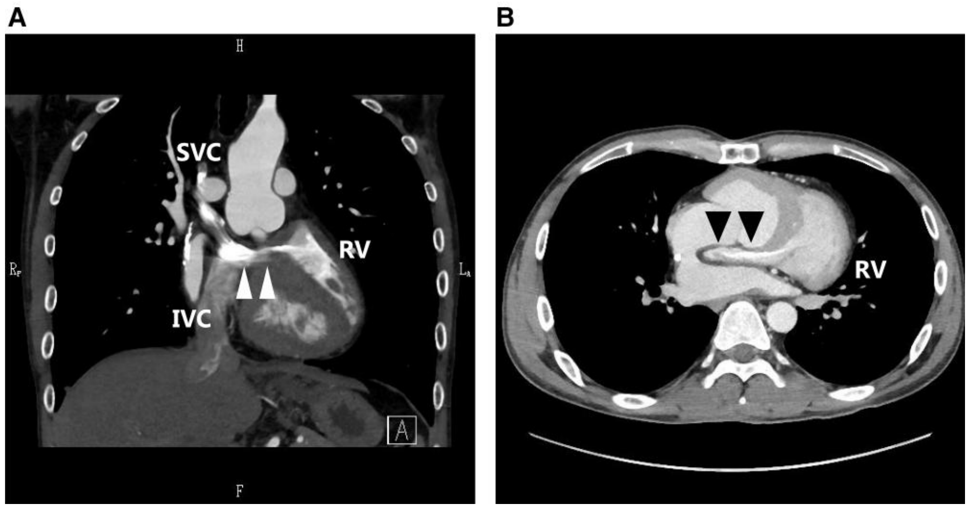


Figure 3 The superior vena cava and inferior vena cava were connected to the anatomical right ventricle on the left side via the Senning pathway. (*A*) Coronal view (white arrow: the Senning pathway); (*B*) axial view (black arrow: the Senning pathway). IVC, inferior vena cava; RV, right ventricle; SVC, superior vena cava.

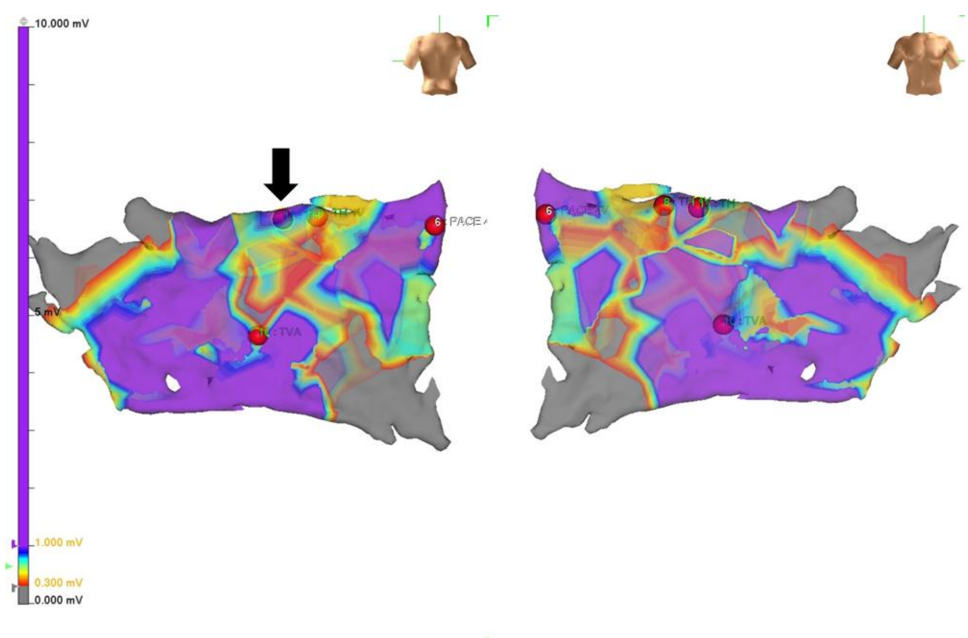


Figure 4 Electrophysiological study using 3D system (ensite®). Voltages were obtained throughout the morphological right atrium, but the areas with good pacing threshold were rather limited (round marks in the roof of superior cavoatrial channel). We implanted the atrial lead in the roof of the morphological right atrium (black arrow).

Discussion

Arrhythmia is the most common cause of sudden cardiac death in the long-term postoperative period of congenital heart disease,[7–9] and there are some reports of bradyarrhythmia in patients with ccTGA.[2,5,10] However, there are few reports of fatal ventricular arrhythmias in ccTGA.[3–5] McCombe *et al.*[4] observed 39 patients with ccTGA, and five cases suffered sudden cardiac death (per 109 patient-years); one of these patients had previously documented non-sustained ventricular tachycardia. Another retrospective report included 65

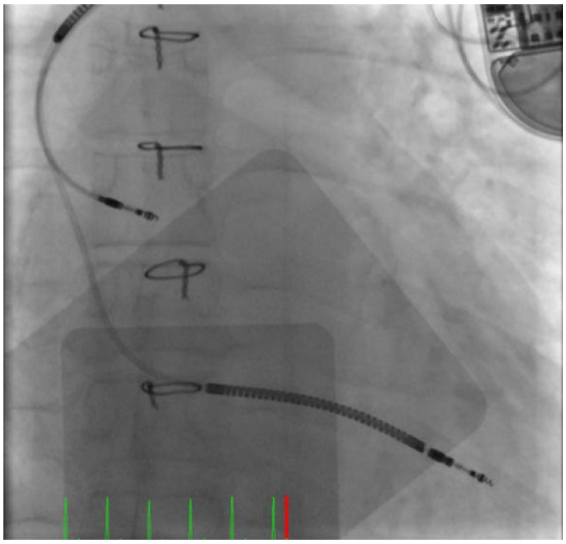


Figure 5 Fluoroscopic image: the position of the atrial and shock leads.

paediatric patients with ccTGA resulting in the death of three patients, but ventricular tachycardia was not detected during the follow-up period (median 49 months).[5]

Right ventricular failure occurs at a high rate as patients with ccTGA grow into adulthood. Double-switch surgery (Senning/Mustard + Jatene or Senning/Mustard + Rastelli) is performed to align the more muscular left ventricle with systemic rather than pulmonary circulation. In one study of patients with ccTGA, complete atrioventricular block (CAVB) was reported in 22% as a history item and 24% more experienced it after surgery; the risk of CAVB was reported to increase by 2% per year.[1] Among the various congenital heart diseases, ccTGA is the one with the most frequent occurrence of CAVB and need for pacemaker implantation.[2] In this case as well, atrial pacing by a pacemaker for sick sinus syndrome caused by Senning was required. Ventricular pacing with a single chamber might cause heart failure due to interventricular dyssynchrony. Unnecessary ventricular pacing causes a decrease in right ventricular function and increased heart failure.[11,12] The atrioventricular conduction of this patient was relatively well preserved, and he required only atrial pacing; thus, dual-chamber ICD was selected to reduce unnecessary ventricular pacing.

It was difficult to implant an atrial lead because the area of tissue suitable for implantation, i.e. a region that was healthy and had a good pacing threshold, was limited due to the prior double-switch operation. Nevertheless, we were able to obtain useful information about the best atrial site for implantation of an atrial lead by 3D mapping and were able to place the atrial lead in an appropriate site.

The reason of ventricular fibrillation was unclear in this case. There was still an episode of appropriate therapy for ventricular tachycardia after ICD implantation suggesting the presence of both ventricular arrhythmogenic substrates due to biventricular dysfunction. The reason for this biventricular dysfunction was thought as related to the characteristic of a systemic right ventricle in ccTGA and two procedures of pulmonary artery banding as a teenager. The findings of coronary angiography were normal and ventricular tachycardia due to ischaemia was unlikely. In addition to ventricular fibrillation due to bradycardia, the patient had stable ventricular tachycardia, which may indicate reentrant ventricular tachycardia with substrate associated with cardiac dysfunction.

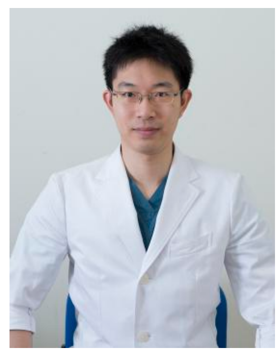
Because the patient had resuscitated ventricular fibrillation and met the indications for an ICD, a ventricular tachycardia study was not performed. The patient had a ventricular tachycardia during follow-up, thus a ventricular tachycardia study might have been warranted in this case.

When implanting dual-chamber ICDs or pacemakers in patients with ccTGA after double-switch surgery, it may be useful to perform an electrophysiological test to detect the atrial site most suitable for lead placement using a 3D map before implantation of the cardiac device.

Conclusions

We experienced a case of ventricular fibrillation and sinus bradycardia in a patient with ccTGA surgically treated 20 years ago by a double-switch operation including atrial redirection procedure. Electrophysiological examination using a 3D mapping system before implantation of a dual-chamber ICD was useful because the atrial site suitable for lead placement was limited due to his previous complex surgery in the atrium.

Lead author biography



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Supplementary material

Supplementary material is available at *European Heart Journal—Case Reports* online.

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Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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