

# Pacemaker implantation via femoral vein and successful arrhythmia management in an elderly patient with Fontan circulation: a case report

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## Background

The frequency of arrhythmias increases after the Fontan operation over time; atrial tachycardia (AT) and sinus node dysfunction (SND) are frequently observed.

## Case summary

Our patient was 63-year-old woman who underwent a lateral tunnel Fontan operation for double outlet right ventricle at age 36. She experienced paroxysmal AT for 1 year, and antiarrhythmic medication was not feasible due to symptomatic SND. Computed tomography revealed a 45 mm-sized thrombus in the high right atrium (RA). The patient had three coexisting conditions: paroxysmal AT, symptomatic SND, and the right atrial thrombus, for which total cavopulmonary connection conversion and epicardial pacemaker implantation (PMI) would have been effective; however, given her age and comorbidities, surgical treatment was considered high risk. Catheter ablation was avoided because of the right atrial thrombus. Finally, a transvenous pacemaker was implanted via the right femoral vein to avoid the right atrial thrombus and severe venous tortuosity from the left subclavian vein to the RA. After PMI, the patient was prescribed amiodarone and bisoprolol for AT suppression. Atrial tachycardia occurred once in the third month after discharge. We increased the dose of amiodarone, and she has been tachycardia-free.

## Discussion

Transvenous PMI must be considered in cases where open thoracic surgery or catheter ablation cannot be performed. This is the first report of transvenous PMI via the right femoral vein and successful AT and SND management in an elderly Fontan patient.

## Keywords

Transvenous pacemaker • Fontan operation • Sinus node dysfunction • Atrial tachycardia • Case report

## ESC Curriculum

9.7 Adult congenital heart disease • 5.9 Pacemakers

## Learning points

- The femoral approach may be useful for transvenous pacemaker implantation in Fontan patients who with atrial tachycardia (AT) and sinus node dysfunction (SND).
- Herein, AT and SND were successfully managed with a transvenous pacemaker and antiarrhythmic medications in an elderly Fontan patient.

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## Introduction

Since the first Fontan operation reported in 1971, several modifications have improved clinical outcomes.<sup>1,2</sup> Approximately 40% of patients with complex congenital heart disease (CHD) reach 60 years of age.<sup>3</sup> The prevalence of arrhythmias caused by structural remodeling is increasing with the life expectancy of such patients.<sup>4</sup>

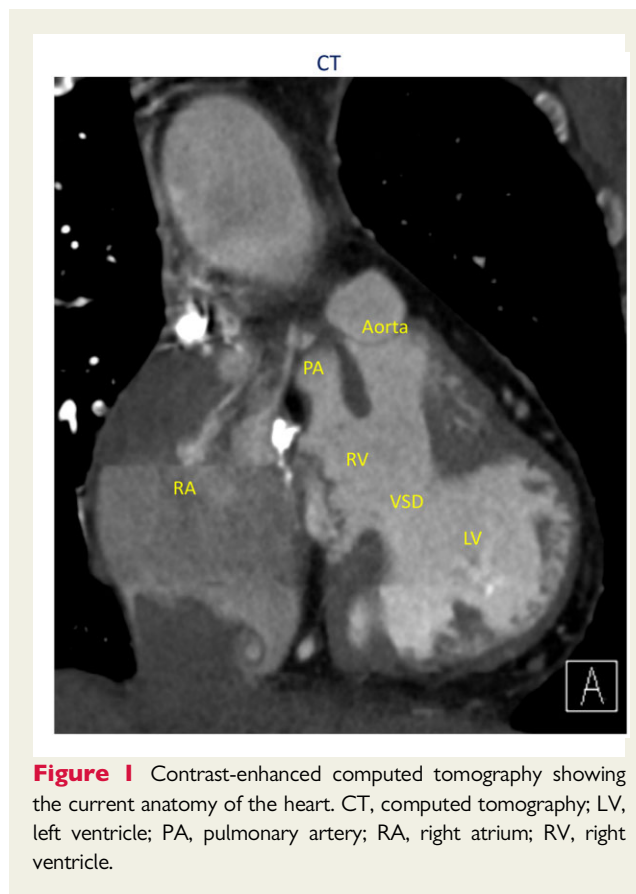
Arrhythmia prevalence after the Fontan operation is ~50%, 20 years postoperatively.<sup>5</sup> Atrial tachycardia (AT) and sinus node dysfunction (SND) are commonly observed in Fontan patients; both conditions can coexist. The procedure for pacemaker implantation (PMI), transvenous or epicardial, in such patients remains controversial<sup>6</sup>; transvenous PMI is challenging due to the anatomical complexity.<sup>7</sup> Herein, we report the case of an elderly Fontan patient successfully treated with PMI via the right common femoral vein (CFV) and antiarrhythmic drugs for AT and SND.

## Timeline

| Age             | Event  |
|-----------------|--|
| 7 months of age | This patient presented with cyanosis at 7 months of age and was suspected to have congenital heart disease.                                |
| 6 years of age  | She began to complain of shortness of breath on exertion.  |
| Unknown         | The diagnosis of double outlet of right ventricle was made.  |
| 10 years of age | She underwent a Glenn operation.   |
| 36 years of age | She underwent a lateral tunnel Fontan operation.   |
| 62 years of age | Atrial tachycardia (AT) and sinus node dysfunction (SND) were detected.  |
| 63 years of age | She was referred to our hospital.  |
| After admission | A transvenous pacemaker was implanted via the right common femoral vein. She was prescribed antiarrhythmic medications for AT suppression. |
| After discharge | Atrial tachycardia and SND were successfully managed with a transvenous pacemaker and antiarrhythmic medications.                          |

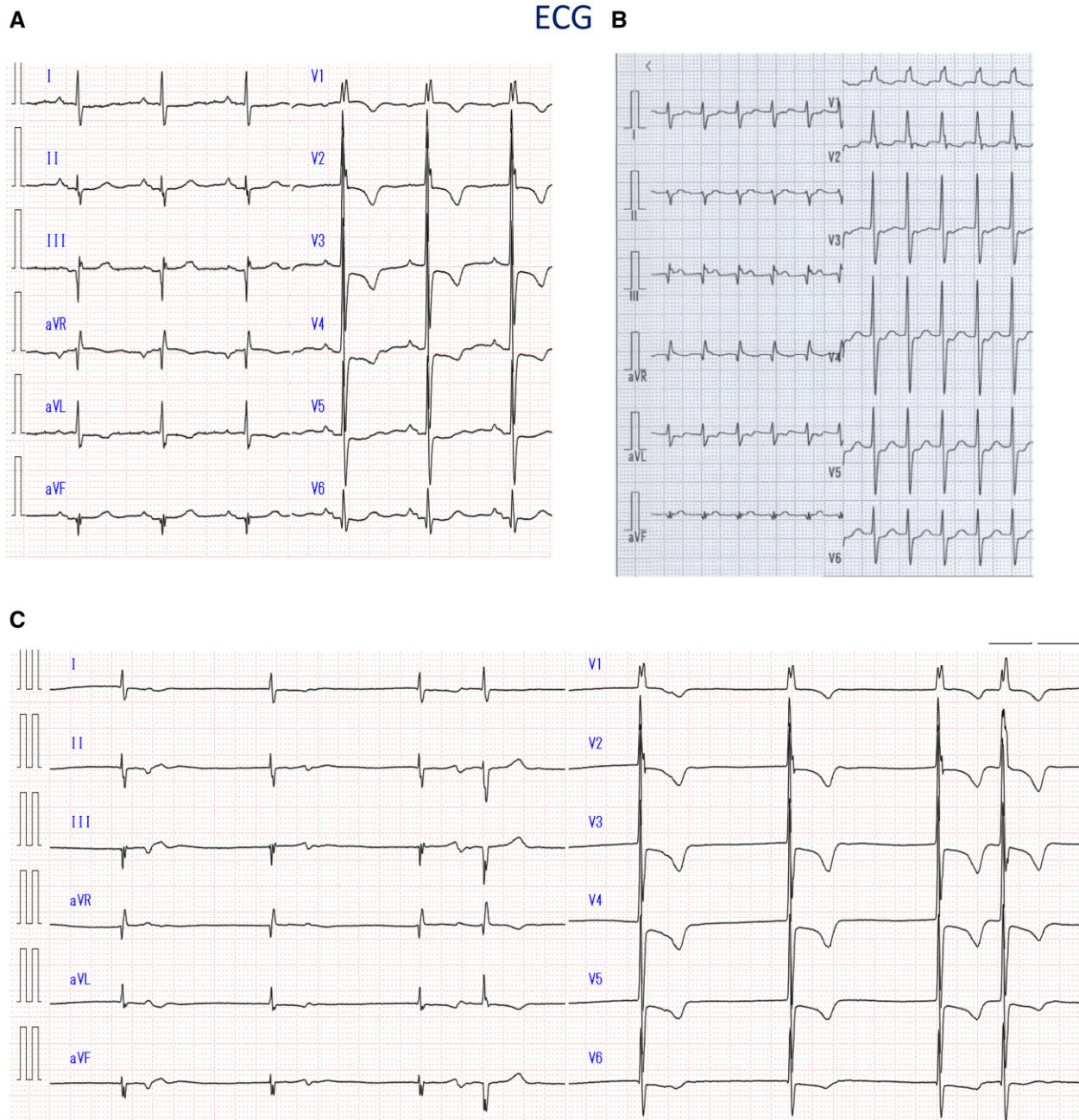
## Case presentation

Our patient was a 63-year-old woman who presented with cyanosis at 7 months of age and was suspected to have CHD. At age 6, she complained of shortness of breath and decreased physical activity and was diagnosed with double outlet right ventricle with remote ventricular septal defect and pulmonary stenosis (*Figure 1*); at age 10, she underwent a Glenn operation. At ages 30, 36, and 43, the patient



**Figure 1** Contrast-enhanced computed tomography showing the current anatomy of the heart. CT, computed tomography; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle.

experienced shortness of breath and other symptoms, underwent a lateral tunnel Fontan operation and was lost to follow-up, respectively. She visited a referring physician at age 62 because of worsening palpitations; a 12-lead electrocardiogram detected AT (*Figure 2B*). She was prescribed 100 mg of amiodarone, which was subsequently reduced to 50 mg because of the appearance of symptomatic SND; however, this dose was not effective for the AT (*Figure 2C*). The patient required frequent cardioversion, and she was referred to our hospital for catheter ablation (CA). She had a pulse rate of 46 beats per minute, blood pressure of 118/69 mmHg, and oxygen saturation of 89% on room air, with no leg oedema or heart murmur. She was taking azosemide (60 mg) and warfarin (1.5 mg). She had New York Heart Association class III heart failure and chronic kidney disease with an estimated glomerular filtration rate of 49 mL/min/1.73 m<sup>2</sup> (normal range: 57–102 mL/min/1.73 m<sup>2</sup>). Blood tests showed hyperbilirubinaemia with a total bilirubin of 1.9 mg/dL (0.4–1.5 mg/dL). Computed tomography showed a small amount of ascites. Liver ultrasonography revealed congestion and moderate to severe liver fibrosis, leading to a diagnosis of cirrhosis. Echocardiography showed that the systemic ventricle was the left ventricle and its contraction was slightly reduced, with an end-diastolic area of 34.9 cm<sup>2</sup> (10–24 cm<sup>2</sup>), end-systolic area of 23.2 cm<sup>2</sup> (3–15 cm<sup>2</sup>), and fractional area change of 34% (35–56%).<sup>8</sup> Magnetic resonance imaging revealed a left ventricular end-diastolic volume, end-systolic volume, and ejection fraction of 69 mL (42–100 mL), 30 mL (16–52 mL), and 57% (42–68%), respectively.<sup>8</sup> Computed tomography revealed a 45 mm-sized

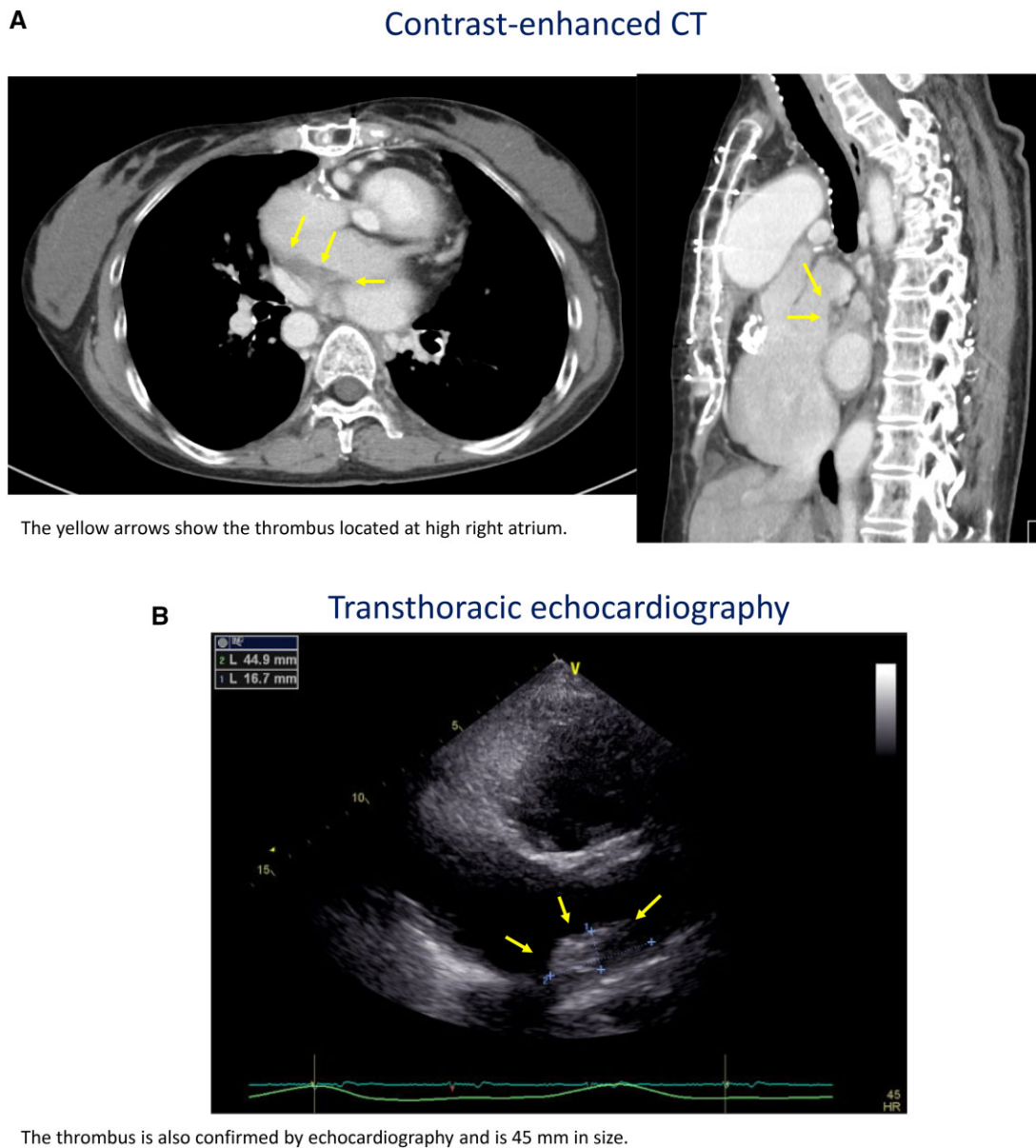


**Figure 2** (A) Electrocardiogram during sinus rhythm with a pulse rate of 70 b.p.m. (B) The electrocardiogram during atrial tachycardia with a pulse rate of 160 b.p.m. (C) The electrocardiogram during sinus arrest with escape beats.

thrombus located in the high right atrium (RA) (Figure 3A), confirmed by echocardiography (Figure 3B). Pulmonary capillary wedge pressure, right atrial pressure, and systemic ventricle end-diastolic pressure were 14, 12, and 11 mmHg, respectively.

The patient had three coexisting conditions: paroxysmal AT, symptomatic SND, and the right atrial thrombus, for which total cavopulmonary connection (TCPC) conversion with an extracardiac

conduit and epicardial PMI would have been effective. However, she had no family support for life after discharge. Given the age, liver cirrhosis, renal impairment, and lack of family support, TCPC conversion was considered a high-risk option. A multidisciplinary team of cardiologists, surgeons, anaesthetists, and paediatricians selected transvenous PMI and antiarrhythmic drugs for treatment because it was less invasive. The patient also preferred a less invasive treatment.

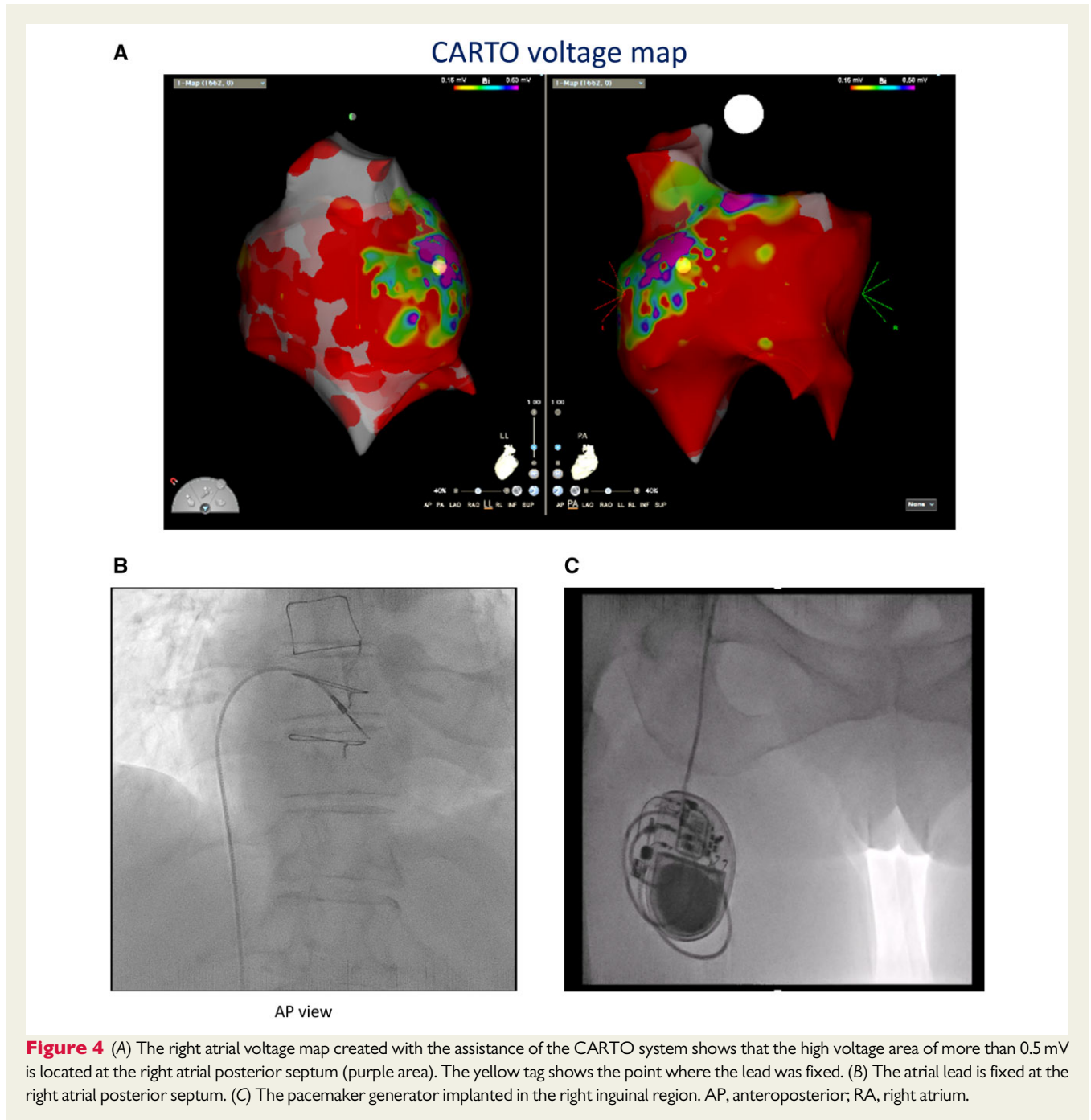


**Figure 3** Contrast-enhanced computed tomography (A) and echocardiography (B) showing a 45 mm-sized thrombus (yellow arrows) at the high right atrium.

The pectoral approach was avoided because of the high right atrial thrombus and severe tortuosity from the left subclavian vein to the RA. A transvenous pacemaker was implanted via the right CFV.

Venous access was obtained using the left CFV. Right atrial voltage mapping was performed using a multi-electrode mapping catheter (DECANAV, Biosense Webster, Inc., Diamond Bar, CA, USA) with a three-dimensional mapping system (CARTO, Biosense Webster, Inc., Diamond Bar, CA, USA) (Figure 4A). While most of the RA had low voltage, suitable pacing tissue was found in the posterior septum, where the voltage remained relatively high. A peel-away sheath for the pacing lead was inserted into the right CFV. An

85-cm active fixation atrial lead (Medtronic, Minneapolis, MN, USA) was implanted and connected to a Medtronic Azure W2SR01 generator (Figure 4B and C). Subsequently, the patient was prescribed amiodarone and bisoprolol for AT suppression. Lead dislodgement or haematoma was not observed. She was discharged on warfarin with a target international normalised ratio of 2.0–3.0. At the most recent visit (12 months from PMI), chest radiography revealed a stable lead position with normal lead function. Atrial tachycardia occurred once in the third month after discharge. We increased the dose of amiodarone from 100 to 200 mg and she has been tachycardia-free. The dose of bisoprolol was gradually



increased to 3.75 mg. The amiodarone dose has been carefully adjusted with regular monitoring of the patient's liver function, amiodarone blood levels, and KL-6 levels.

## Discussion

In Fontan patients, optimal arrhythmia treatment requires a multidisciplinary approach to determine the appropriate treatment.<sup>4</sup> In this case, while TCPC conversion and epicardial PMI

could have improved the AT, SND, and right atrial thrombus, the patient's age, liver cirrhosis, renal impairment, and lack of family support made surgical treatment high risk. A transvenous pacemaker was implanted via the right CFV, avoiding the right atrial thrombus, tortuous venous course, and repeat sternotomy.

To the best of our knowledge, this is the first report of successful transvenous PMI via the right CFV in an elderly Fontan patient with successful AT management. This case also serves as an additional example of transvenous PMI in an elderly Fontan patient who had both AT and SND.

Arrhythmia is the most frequent complication after the Fontan operation that increases over time.<sup>5</sup> In a previous report analysing 599 Fontan patients, 13% required PMI, with SND being the most common indication.<sup>9</sup>

Herein, CA was avoided because of a thrombus in the Fontan circulation. Moreover, CA in patients with complex CHD may not completely control AT; it has lower success and higher recurrence rates.<sup>10</sup>

Epicardial pacemakers may be used in Fontan patients; however, the epicardial approach requires thoracotomy or sternotomy, resulting in longer hospital stays and perioperative complications. Transvenous pacemakers are superior to epicardial pacemakers in terms of lead thresholds and lead longevity.<sup>6,11,12</sup>

A predominant concern is thromboembolic events from the transvenous leads. Herein, the patient was prescribed warfarin with a target international normalised ratio of 2.0–3.0 to prevent thrombosis. Segar et al.<sup>12</sup> reported that eight Fontan patients with transvenous pacemakers did not experience thrombosis. A larger trial comparing the safety and efficacy of transvenous and epicardial pacemakers is required in the Fontan population.

Atrial anti-tachycardia pacing (ATP) from implantable cardiac devices effectively suppresses AT in patients with CHD.<sup>13</sup> In this case, we used a single chamber atrial pacemaker, which cannot automatically deliver ATP. Manual ATP will be considered if the AT recurs. However, CA may be the only effective treatment option for frequent AT recurrence, with close attention paid to the thrombus.

The femoral approach is useful for transvenous PMI in Fontan patients with limited vein access. Atrial tachycardia and SND were successfully managed with a transvenous pacemaker and antiarrhythmic medications.

## Lead author biography



Tomofumi Mizuno, MD, graduated from Faculty of Medicine Okayama University, Japan in 2012. He completed 2 years of Japanese post-graduate residency programme at Okayama University Hospital in Japan. Then he has worked as a fellow in cardiology at Fukuyama City Hospital. Currently, he is working as a clinical fellow at Department of Cardiovascular Medicine Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences, Japan. He is a Fellow of the Japanese Society of Internal Medicine.

## 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 this case 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.

**Conflict of interest:** N.N. and H.M. are affiliated with the department endowed by Japan Medtronic Inc.

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