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Case Report

The efficacy of pacemaker implantation for extracardiac total cavopulmonary connection in a pediatric patient with bradycardia-tachycardia syndrome

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ABSTRACT

A 9-year-old boy, diagnosed with double outlet right ventricle after birth, suffered sinus node dysfunction and non-sustained junctional tachycardia after an extracardiac total cavopulmonary connection (TCPC). Spontaneous atrial tachycardia appeared 3 years after an extracardiac TCPC. Sotalol was administered but the bradycardia was obvious. It was difficult to increase sotalol and atrial tachycardia was uncontrollable. Atrial tachycardia continued with symptoms; direct current (DC) cardioversion was frequently required. Five years after extracardiac TCPC, we implanted a pacemaker with atrial antitachycardia pacing (ATP) using epicardial leads. On day 2 post operation, wide QRS tachycardia appeared. Due to decreased blood pressure, DC cardioversion was immediately performed, but it recurred from atrial premature contraction. We judged this was atrial tachycardia with 1:1 atrioventricular conduction based on an intracardiac electrogram and it was terminated by burst atrial pacing from the pacemaker. After changing atrial pacing rate to 150 ppm, atrial tachycardia could be suppressed. Due to atrial pacing and increasing sotalol gradually, junctional tachycardia terminated spontaneously, and atrial tachycardia was not induced after pacemaker implantation. In conclusion, implantation of a pacemaker with ATP and intensification of antiarrhythmic drugs is an effective treatment strategy for pediatric patients with bradycardia-tachycardia syndrome after extracardiac TCPC.

<**Learning objective:** The treatment for bradycardia-tachycardia syndrome in children after extracardiac total cavopulmonary connection (TCPC) is challenging. The appropriate antiarrhythmic drugs for atrial tachycardia cannot be administered due to bradycardia, and it is often difficult to perform radiofrequency catheter ablation on small children. Surgical pacemaker implantation, although invasive, is the most effective treatment for bradycardia-tachycardia syndrome in small children after extracardiac TCPC.>

Introduction

The Fontan procedure has been established as a treatment to eliminate cyanosis for cyanotic heart disease [1–3]. Extracardiac total cavopulmonary connection (TCPC) procedure which is mainly performed, can result in sinus node dysfunction and radiofrequency catheter ablation is often anatomically difficult [1,4]. We report a case of bradycardia-tachycardia syndrome with extracardiac TCPC, where it has been possible to control bradycardia and atrial tachycardia via implantation of a pacemaker along with atrial antitachycardia pacing (ATP) therapy.

Case report

A 9-year-old boy developed bradycardia, junctional tachycardia, and atrial tachycardia due to sinus node dysfunction after extracardiac TCPC. A diagnosis of double outlet right ventricle with hypoplastic left ventricle and bilateral superior vena cava was made postnatally. He received staged palliation, (1) pulmonary artery banding, (2) atrial septal defect creation and bilateral bidirectional cavopulmonary connection, and (3) extracardiac TCPC using an 18 mm Gore-Tex graft. After the extracardiac TCPC, his electrocardiogram gradually showed bradycardia, junctional rhythm, and junc-

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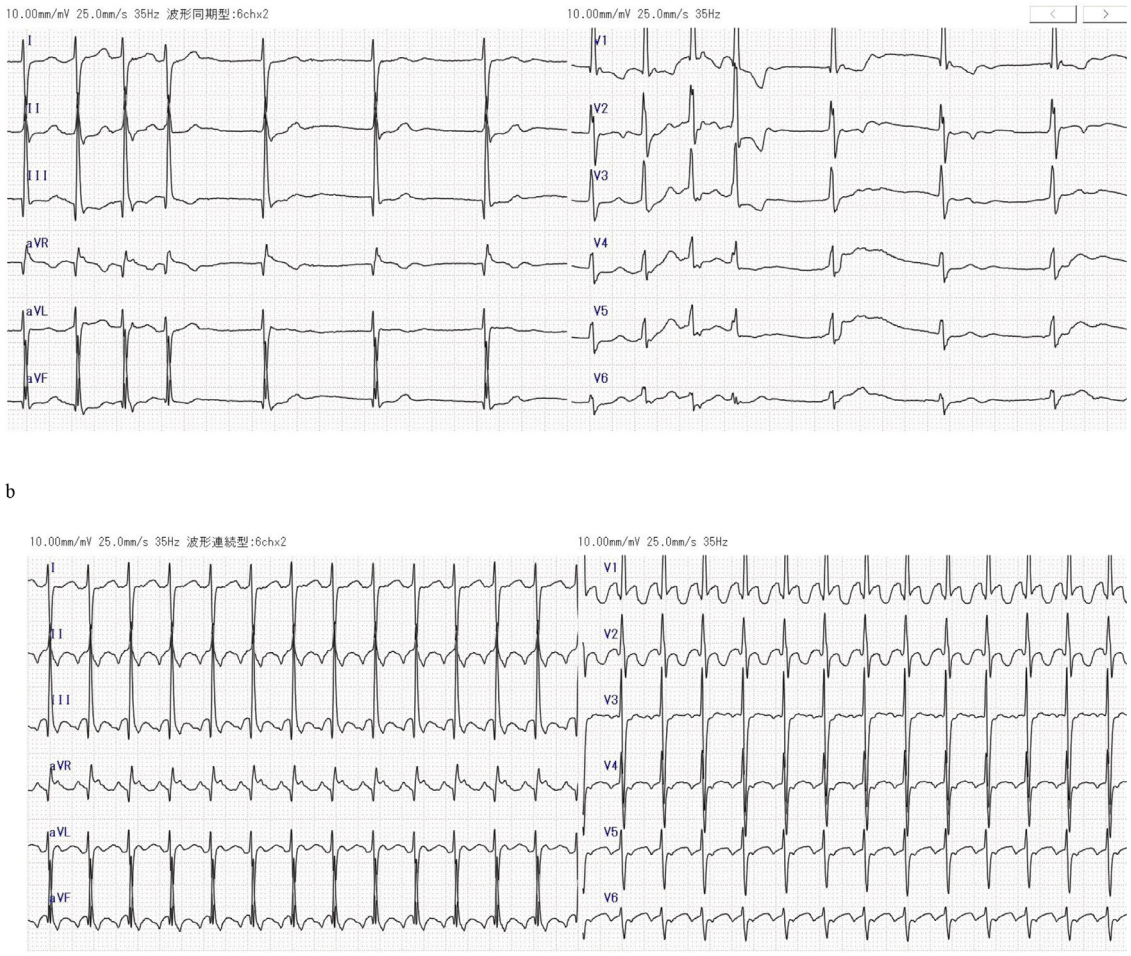

Fig. 1. 12-lead electrocardiogram of outpatient visit. (a) Short duration junctional tachycardia (150 bpm) and junctional rhythm (50–60 bpm). (b) 2:1 atrial tachycardia (170 bpm) at the emergency room.

tional tachycardia which terminated spontaneously (Fig. 1a). Spontaneous atrial tachycardia appeared 3 years after extracardiac TCPC at age 6.6 years on a Holter monitor. At 4 years after extracardiac TCPC at age 7.9 years, atrial tachycardia persisted, and he was started on oral administration of sotalol (1.5 mg/kg/day). However, bradycardia was obvious; it was difficult to increase the sotalol sufficiently, and atrial tachycardia could not be controlled. Atrial tachycardia had a ventricular rate of 154–173 beat/minutes (bpm) with 2:1 conduction (Fig. 1b). When atrial tachycardia continued for a long time, he was pale and vomiting, and direct current (DC) cardioversion was required. Cardiac catheterization showed no stenosis in the graft, and central venous pressure was 10 mmHg. The right ventricular ejection fraction (RVEF) was 62%. Although no significant pressure gradient of the pulmonary artery was revealed, angiography of left superior vena cava showed a slightly stenotic change to the pulmonary artery (Fig. 2a). It was possible to perform atrial pacing from the pulmonary artery to the left atrium (Fig. 2b), and blood pressure was stabilized rather than returning to its own rhythm (Fig. 2c, d). The Holter monitor showed a minimum heart rate of 42 bpm and many episodes of junctional tachycardia with durations of about 1–2 minutes and atrial tachycardia induced by atrial premature beats. We judged it was necessary to perform pacemaker implantation to control bradycardia and atrial tachycardia. Since atrial tachycardia requiring DC cardioversion was 2:1 conduction, a pacemaker with atrial ATP therapy was selected.

Five years after extracardiac TCPC aged 9 years, he was 113 cm and weighed 18.8 kg. We implanted a pacemaker with atrial ATP therapy (Azure XT DR, Medtronic, Minneapolis, MN, USA) using steroid-eluting epicardial leads. The ventricular lead (CapSure Epi, 4968 bipolar, 60 cm, Medtronic) was sutured to the apex of the left ventricle through a left thoracotomy, and the atrial lead (CapSure Epi, 4968, bipolar, 60 cm, Medtronic) was sutured to the right atrium through a right thoracotomy. A generator was inserted into the posterior of the rectus abdominis muscle (Fig. 3a). The atrial and ventricular lead pacing thresholds were 2.0 V at 0.4 msec and 2.0 V at 1.0 msec, respectively. On day 2 post operation, wide QRS tachycardia (273 bpm) suddenly appeared after crying (Fig. 3b). He looked pale and displayed presyncope with decreased blood pressure; the possibility of ventricular tachycardia was considered, and DC cardioversion (50 Joule) was performed after intravenous sedation. Narrow QRS tachycardia (255 bpm) recurred immediately after the premature atrial contraction (Fig. 3c). This appeared to be an atrial tachycardia of 1:1 conduction, which was confirmed by an intracardiac electrogram of the pacemaker. Burst atrial pacing (300 ppm) from the pacemaker terminated the tachy-

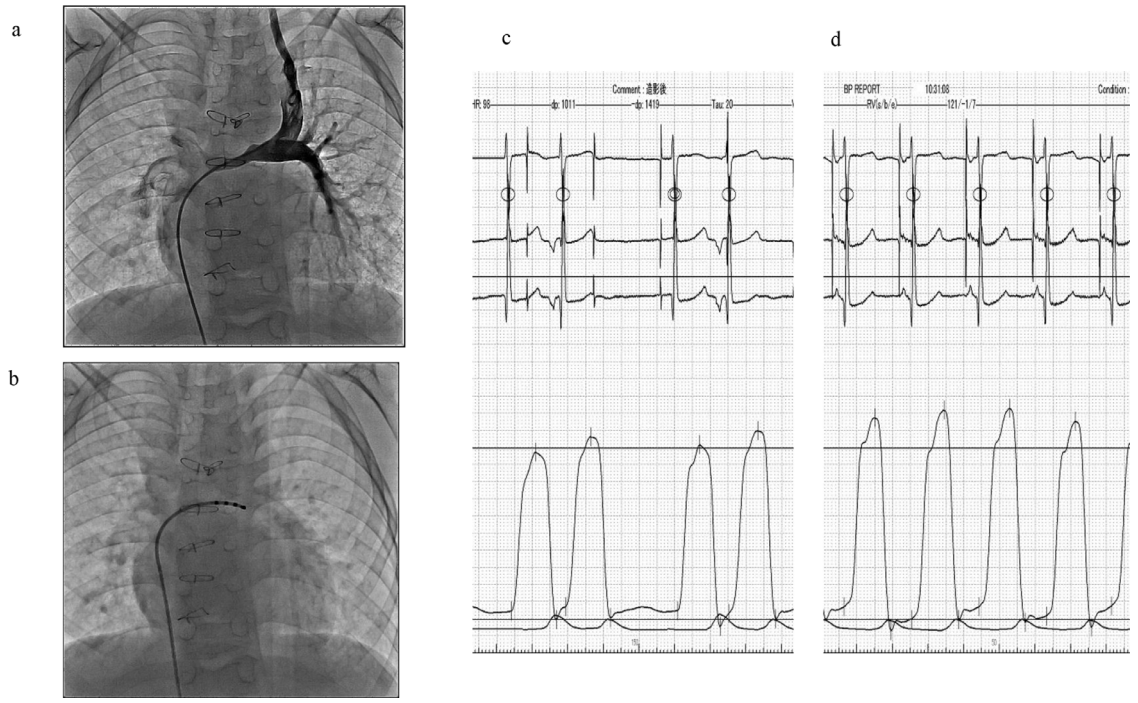

Fig. 2. Cardiac catheterization. (a) Angiography of left superior vena cava showed slightly stenotic change to pulmonary artery. (b) The catheter position of left atrium pacing from left pulmonary artery. (c) Left ventricular pressure with its own rhythm. (d) Left ventricular pressure of left atrium pacing.

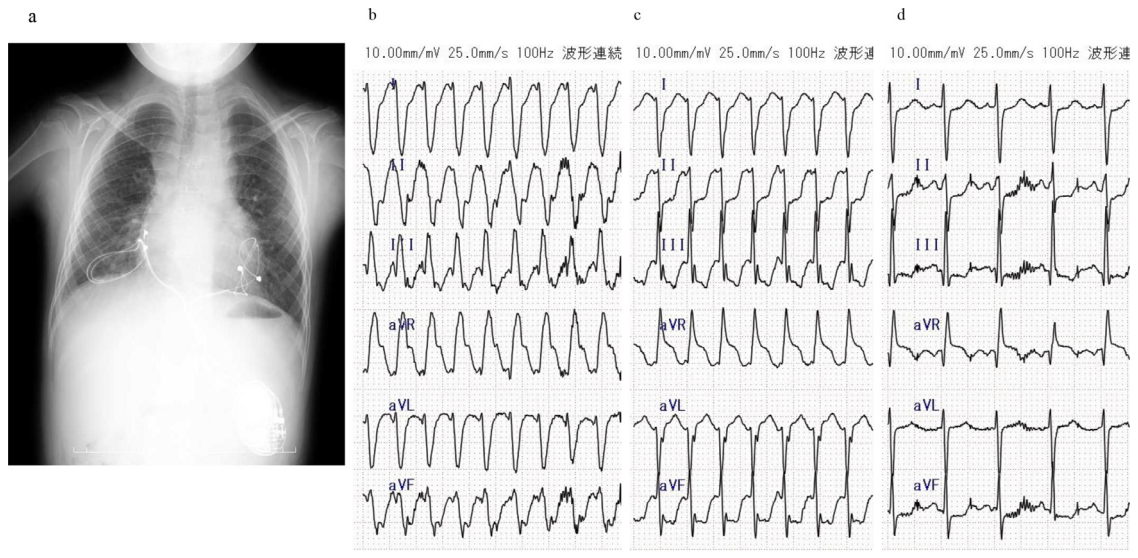

Fig. 3. Chest roentgenogram and 6-lead electrocardiogram at day 2 after pacemaker implantation. (a) Atrial, ventricular leads, and pacemaker. (b) Wide QRS tachycardia after crying. (c) Narrow QRS tachycardia after direct current cardioversion for a wide QRS tachycardia. (d) 150 pace per minutes atrial pacing after atrial burst pacing for a narrow QRS tachycardia.

cardia and relatively rapid pacing (150 ppm) was performed, resulting in suppression of atrial tachycardia (Fig. 3d). The patient was discharged without any complications on day 14 post operation. After discharge, pacing prevention algorithm and ATP therapies (using RUMP and BURST+) were enabled and the dosage of sotalol was gradually increased (up to 6 mg/kg/day). Premature atrial contractions were suppressed by atrial pacing prevention algorithm and junctional tachycardia terminated spontaneously in a short period of time. The brain natriuretic peptide (299.5 pg/ml to 11.0 pg/ml) and cardiothoracic ratio (57% to 49%) decreased with hemodynamic stability. There was no significant change in RVEF. Over 2 years of follow-up, he has needed no DC cardioversion since pacemaker implantation.

Discussion

We have herein described the case of a 9-year-old boy with extracardiac TCPC, where it has been possible to control bradycar-

dia and atrial tachycardia via implantation of a pacemaker along with atrial ATP therapy. The selection of pacemaker with atrial pacing prevention algorithm and atrial ATP therapies rather than AAI pacemaker is a better choice for small pediatric patients with bradycardia-tachycardia syndrome after extracardiac TCPC.

Pacing therapy for bradycardia with symptoms such as fatigue, dizziness, or syncope, is an effective therapy. Newcombe et al. reported a transvenous lead implantation and AAI pacing for a pediatric patient as part of an extracardiac autologous pericardial tunnel Fontan procedure [5]. In our patient, atrial pacing from the left pulmonary artery to the left atrium was possible during cardiac catheterization, and transvenous pacemaker leads could be implanted. However, we judged that stenosis of the Fontan route adversely affects hemodynamics in patients after extracardiac TCPC. Additionally, a pacemaker with atrial ATP therapy requires a ventricular lead but the transvenous approach to the ventricle for extracardiac TCPC was impossible. Therefore, surgical pacemaker implantation was chosen for this patient. Although surgical implantation is invasive, it may be an effective treatment for bradycardia and atrial tachycardia in small pediatric patients after extracardiac TCPC.

The efficacy and safety of pacing therapy with atrial ATP therapy have been reported in patients with congenital heart diseases. Efficacy rates for atrial ATP therapy for atrial tachycardia of between 43% and 72% have been reported, and this therapy is said to be effective in reducing the frequency of DC cardioversion, and there is no safety problem [6–9]. On the other hand, atrial ATP therapy is not performed for atrial tachycardias for reasons such as 1:1 atrioventricular conduction, slow atrial tachycardia under minimum setting atrial tachycardia rate, very short duration, or atrial undersensing [8]. Even in this patient, pacing therapy was not performed for 1:1 conduction junctional tachycardia lasting 1–2 minutes. Before pacemaker implantation, atrial tachycardia was induced by premature atrial contractions. Although, after pacemaker implantation, atrial tachycardia has not been induced and no atrial ATP therapies are performed, we consider it may be effective for atrial pacing prevention algorithm to suppress premature atrial contraction inducing atrial tachycardia before pacemaker implantation.

In patients with Fontan procedure, especially anatomical right ventricle, they are potentially in a state of cardiac dysfunction. Sotalol is a class III antiarrhythmic agent that is highly effective for tachyarrhythmia for congenital heart disease without affecting cardiac output [10]. In this patient, before pacemaker implantation, sotalol exacerbated bradycardia and we could not increase the dose of sotalol to control atrial tachycardia and junctional tachycardia. After pacemaker implantation, junctional tachycardia decreased with the increase in the dose of sotalol without cardiac dysfunction and was very effective for tachyarrhythmia control.

In conclusion, implantation of a pacemaker with atrial ATP therapy and intensification of antiarrhythmic drugs is an effective treatment strategy for small pediatric patients with bradycardia-tachycardia syndrome after extracardiac TCPC.

Ethics

This study was approved by our institutional Ethics Committee and written informed consent was obtained from the patient's parents.

Declaration of competing interest

The authors declare that there is no conflict of interest.

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