

Case Report

Fontan Conversion with Novel Direct Ablation After Childbirth: Report of a Case

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Abstract

A 38-year-old woman underwent atriopulmonary Fontan surgery at age 18 years and subsequently successfully delivered a girl by cesarean section at age 34. Her condition later deteriorated due to atrial tachyarrhythmia and progressed to New York Heart Association (NYHA) class IV heart failure. Her treatment, at age 36, comprised total cavopulmonary connection conversion, direct right atrial ablation with bipolar radiofrequency devices, the creation of an atrial septal defect, and placement of a dual-chamber permanent pacemaker. Three years after the conversion, her condition has improved to NYHA class I.

Key words Adult · Congenital heart disease · Fontan · Arrhythmia therapy · Reoperation

Introduction

Pregnancy and childbirth are considerable challenges for patients who have undergone a Fontan operation. Indeed, there have been only a few reports of successful pregnancies and deliveries in those who have undergone such a procedure.^{1–4} In addition, Fontan conversion is sometimes necessary in adults who begin to show symptoms of failing Fontan circulation.^{5,6}

We herein describe a patient who underwent a Fontan conversion 2 years after she successfully delivered a healthy female infant. To the best of our knowledge, this is the first case report of total cavopulmonary connection (TCPC) conversion with arrhythmia surgery in a

patient who had delivered a baby after an atriopulmonary (AP) Fontan procedure.

Case Report

At age 18 years, the patient underwent AP Fontan surgery in our department under the diagnoses of corrected transposition of the great arteries with a hypoplastic left ventricle, large ventricular septal defect, and pulmonary stenosis. Although she reported a couple of brief episodes of palpitation after the AP Fontan procedure, her heart maintained a normal sinus rhythm. Sixteen years later, she became pregnant. She developed atrial tachycardia (AT) at 3 months' gestation, which was successfully treated with antiarrhythmic agents. She successfully delivered a healthy female infant by cesarean section in the obstetrics department of our hospital. However, her AT recurred at 10 days postpartum and continued to recur thereafter. She began complaining of palpitations, orthopnea, and shortness of breath. In addition, her serum level of brain natriuretic peptide (BNP) increased to 728.9 pg/ml. Her condition deteriorated due to repeated AT, and ultimately progressed to New York Heart Association (NYHA) functional class IV heart failure.

Echocardiography revealed a smoke-like echo in the giant right atrium (RA). Contrast-enhanced computed tomography (CT) showed marked accumulation of contrast medium in the bottom of the giant RA (Fig. 1), and three-dimensional CT showed a markedly enlarged RA, superior vena cava (SVC), and inferior vena cava (IVC) (Fig. 2). Cardiac catheterization revealed marked central venous stagnation and a marked pressure elevation of 18–22 mmHg throughout the SVC, IVC, giant RA, and pulmonary arteries.

Electroanatomical mapping (CARTO XP; Biosense Webster, Diamond Bar, CA, USA) of right atrial activation was performed. Incisional re-entrant AT and several

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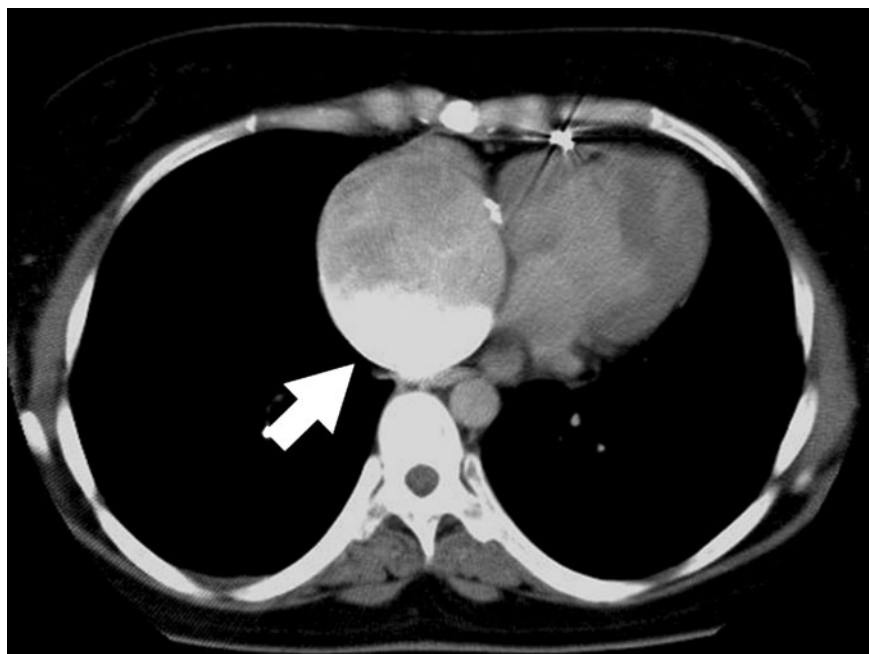


Fig. 1. Preoperative computed tomography shows marked accumulation of contrast medium (*arrow*) in the dorsal portion of a giant right atrium

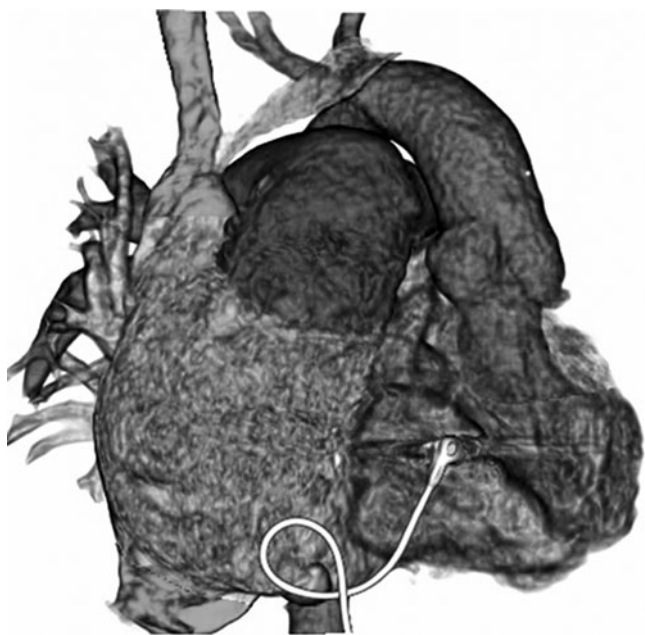


Fig. 2. Preoperative three-dimensional computed tomography

areas of focal AT were observed. Isopotential mapping differentiated the incisional scars from areas of degeneration. Catheter ablation was repeatedly attempted when the patient was aged 35 years; however, recurrent AT persisted because the radiofrequency energy was insufficient owing to blood congestion. The patient was thus admitted for TCPC conversion with arrhythmia surgery at age 36. After establishing cardiopulmonary

bypass (CPB) and performing a bidirectional Glenn anastomosis, the enlarged and thickly fibrous RA wall was extensively resected. After division of the IVC, direct ablation was performed using bipolar radiofrequency (RF) devices (Isolator Transpolar Clamp, Isolator Transpolar Pen; AtriCure, West Chester, OH, USA).⁷ In principle, ablation lines were drawn according to the modified right-sided Maze procedure described by Mavroudis et al. (Fig. 3).⁵ A pen-type bipolar RF device was used mainly for the atrial septum and in the region of the atrioventricular annulus. A bipolar RF clamp was extensively applied to the isthmus block portion and to the atrial wall near the excision lines. Extracardiac TCPC was performed by using a 24-mm diameter vascular graft (Gore-Tex Vascular Grafts; Gore & Associates, Phoenix, AZ, USA). Aortic cross-clamping was limited to the creation of an atrial septal defect,⁶ the duration of which was 17 min. After weaning from CPB, DDD-pacing leads (Myopore lead; Enpath Medical, Minneapolis, MN, USA) and a generator (Sole DR; ELA Medical SAS, Le Plessis Robinson, France) were implanted.

Postoperatively, the central venous pressure decreased to 10 mmHg. One episode of AT was observed, at 2 days after surgery. However, this resolved after the intravenous administration of an ultrashort-acting β -blocker (landiolol hydrochloride [Onoact]; Ono Pharmaceutical, Osaka, Japan) during her stay in the intensive care unit. Three-dimensional CT confirmed a satisfactory cardiac configuration (Fig. 4).

Three years after the conversion, there has been no obvious recurrence of AT. The serum BNP level also

decreased to within the normal range (the most recent level was 17.1 pg/ml). The patient is now in NYHA functional class I, and is fully able to perform housework and care for her child. Normal sinus rhythm has

also been maintained since the surgery, although a DDD pacemaker rhythm has occasionally been observed on Holter monitoring. The patient has had no complaints.

Discussion

Patients who have undergone a Fontan procedure and desire to deliver a child face significant challenges. In addition, TCPC conversion is often required in adults who have undergone an AP Fontan procedure. However, TCPC conversion after successful delivery has rarely been described, because patients with Fontan circulation have major difficulties during pregnancy and delivery^{1,2} and TCPC conversion is a somewhat complicated procedure that is associated with substantial perioperative mortality and morbidity.⁸ This is the first report of a patient with Fontan circulation undergoing TCPC conversion with arrhythmia surgery after successful delivery.

The patient underwent all of the procedures (AP Fontan, cesarean section, catheter ablation, and TCPC conversion) at our hospital over a 20-year period. The success of these procedures absolutely depended on close and meticulous collaboration among the obstetricians, pediatric and adult cardiologists, and cardiovascular surgeons. The rate of first-trimester spontaneous miscarriages in the Fontan population is approximately 33%, which is double the rate in healthy females.¹⁻⁴ In the present case, the TCPC conversion was performed after childbirth. However, the patient's pregnancy and delivery triggered a breakdown in the AP Fontan circu-

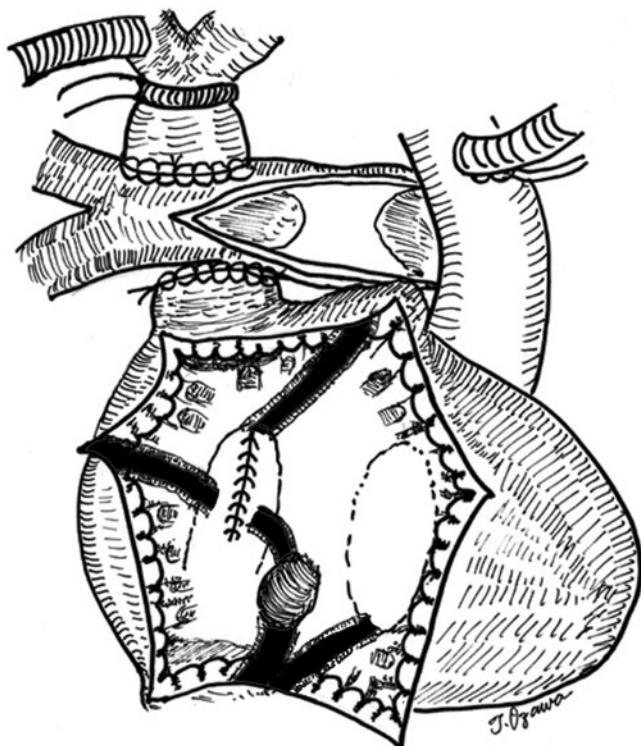


Fig. 3. Direct right atrial ablation lines are indicated by *thick black lines*

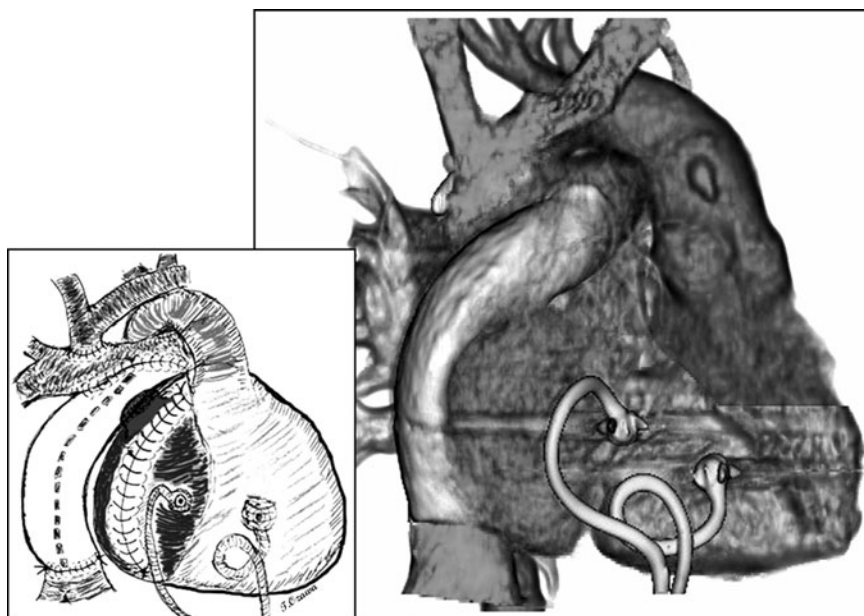


Fig. 4. Postoperative three-dimensional computed tomography alongside the operative schema, showing the configuration of the extracardiac total cavopulmonary connection

lation, as indicated by chronic AT and obvious deterioration in the NYHA functional class after pregnancy. Low cardiac output accompanying the Fontan circulation results in low blood flow to the uterus and placenta, which could result in various problems such as non-reassuring fetal status and intolerance of labor.⁹ Therefore if a failure in AP Fontan circulation appears likely, TCPC conversion should be performed before pregnancy.

During our operation, direct atrial ablation was performed in addition to TCPC conversion. It is questionable whether the use of the conversion procedure for hemodynamic restoration from classic Fontan¹⁰ alone can improve atrial tachyarrhythmia. Takahashi et al.⁸ demonstrated that concomitant arrhythmia surgery reduced subsequent arrhythmia events. Our patient has had no obvious episode of tachyarrhythmia for 3 years postoperatively, which suggests that arrhythmia surgery, including direct ablation and placement of a dual-chamber pacemaker, should be performed concomitantly with TCPC conversion, as it increases the safety of postoperative care and improves long-term outcomes.^{8,11}

With regard to the arrhythmia procedure, both clamp and pen-type bipolar devices were used for various ablation lines, according to the cardiac wall pattern in this case. During the past decade a number of surgical treatments for arrhythmia have been investigated, although the main target of these treatments has been atrial fibrillation (AF). A common concern in arrhythmia surgery has been to replace the cut-and-sew technique with linear lines of ablation by using a variety of energy sources, including unipolar and bipolar RF ablation, lasers, microwaves,¹² and cryoablation. Although all of these techniques have had some success, cryoablation is the standard procedure for arrhythmia surgery in cases of Fontan conversion.^{5,6} However, a comparative study of cryoablation and RF ablation in the Maze procedure showed that RF ablation was simpler and as effective.¹³ Moreover, many experimental studies have indicated that bipolar RF ablation results in the most consistent transmural lesions.^{14–17} In a failing Fontan case with a very thick atrial wall, the creation of a transmural lesion is critical for successfully treating the chronic AT.

The method used to create lesions differs greatly in the cut-and-sew and ablation procedures,¹⁸ and an incision always creates a conduction block across the suture line. However, although the bipolar RF ablation device allows the surgeon to isolate targeted tissue and create transmural lesions,^{14–17} it is not absolutely perfect because all the lesions produced a clinical conduction block.¹⁸ Indeed, one study showed that gaps smaller than 1 mm can allow propagation of AF.¹⁹ On the other hand, Lall et al.¹⁸ demonstrated that a cut-and-sew group (Cox-Maze III procedure) had significantly

longer operation and cross-clamp times than did groups treated with bipolar RF or cryoablation (Cox-Maze IV procedure), and that surgical outcomes for Cox-Maze IV patients were similar to those for Cox-Maze III patients at 1 year of follow-up. Therefore, we believe that our choice of bipolar RF ablation with TCPC conversion is consistent with their outcomes,^{13,17,18} and that proper application of a clamp-type device in patients with a thickened RA wall after an AP Fontan procedure should ensure continuous transmural lesions.⁷ In addition, novel bipolar RF devices that need no ground pad are useful, as they enable rapid ablation and shorter operation and CPB times.

In conclusion, we experienced a very rare case of TCPC conversion with arrhythmia surgery for a female patient who delivered a healthy child after undergoing an AP Fontan procedure, and the new bipolar ablation devices were useful and effective for treating her recurrent AT with failing Fontan circulation.

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Conflict of Interest Statement. The authors have no conflict of interest to declare.

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