

# The mid-term surgical results of Fontan conversion with antiarrhythmia surgery

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## Abstract

**OBJECTIVES:** We investigated the mid-term surgical results of Fontan conversion with antiarrhythmias surgery and permanent pacemaker implantation, which were complications of a previous Fontan operation.

**METHODS:** From January 1996 to November 2011, we performed Fontan conversion in 31 of 260 Fontan cases (M:F = 17:14, mean age = 19.0 years). The Fontan conversion from atriopulmonary connection (APC) to extracardiac conduit (ECC) was performed in 20 patients, APC to lateral tunnel in 5 and lateral tunnel to ECC in 6. The clinical outcomes and improvements of arrhythmias were analysed. The types of arrhythmias included atrial flutter in 21 patients, atrial fibrillation with flutter in 4 patients and junctional tachycardia in 3 patients.

**RESULTS:** Twenty-six patients (83.9%) required antiarrhythmia surgery (isthmus cryoablation = 9, right-sided maze = 13, bilateral maze = 4). In addition, 23 patients (74.2%) received a permanent pacemaker. The New York Heart Association functional class (NYHA Fc) was statistically improved after the surgery during the 6.5-year median follow-up duration (preoperative NYHA Fc = 1.77, postoperative NYHA Fc = 1.13,  $n = 15$ ,  $P = 0.001$ ). There were 4 late mortalities. Actuarial 5-year survival was  $90.0 \pm 5.5\%$ . And freedom from arrhythmia was  $91.8 \pm 5.5\%$ , at 5 years. Normal sinus rhythm was maintained in 12 patients (38.7%), pacing-dependent rhythm in 10 patients (32.3%) and intermittent pacing-dependent rhythm in 4 patients (12.9%).

**CONCLUSIONS:** Fontan conversion with antiarrhythmia surgery and permanent pacemaker implantation is safe and improves the clinical outcome and arrhythmias.

**Keywords:** Fontan conversion • Antiarrhythmia surgery • Pacemaker implantation

## INTRODUCTION

Since the first reported Fontan operation [1], surgical modifications have been incorporated due to complications after the Fontan operation with increasing duration of follow-up. The surgery has considerable potential morbidities, including thromboembolisms, atrial arrhythmias, venous pathway obstruction and mortality in atriopulmonary artery connection (APC) [2]. Total cavopulmonary artery connection (TCPC) with intracardiac baffling, which showed good early results, was introduced in 1988 [3]; however, late arrhythmias were still problematic. Thus, TCPC using extracardiac conduit (ECC) was introduced to minimize the right atrial load.

Fontan conversion with antiarrhythmia surgery and permanent pacemaker implantation is one of the options for treating these complications. There are several studies that have reported clinical

improvement, including exercise tolerance, New York Heart Association functional class (NYHA Fc) improvement and rhythm consideration after Fontan conversion with antiarrhythmia surgery [4–6]. However, whether there is improvement in ventricular function and protein-losing enteropathy (PLE) is unclear. Recently, a study from our centre reported that Fontan conversion with concomitant antiarrhythmias surgery and permanent pacemaker implantation was safe, improved the NYHA Fc and resulted in a low incidence of arrhythmia recurrence [7]. However, there was limited evaluation of surgical results after Fontan conversion with arrhythmia surgery and permanent pacemaker implantation due to the small number of patients and the short-term follow-up. Thus, we re-evaluated the surgical results of Fontan conversion with antiarrhythmia surgery in this study.

## MATERIALS AND METHODS

The study protocol was approved by the Institutional Review Board of the University of Seoul National University Hospital (No. H-1303-004-468). The procedures were performed in accordance with institutional guidelines for the protection of patient confidentiality. The requirement for patient consent was waived due to the retrospective nature of the study. We retrospectively reviewed the 31 patients who had undergone Fontan conversion out of 260 Fontan cases from January 1996 to November 2011.

### Surgical strategy

We performed the Fontan conversion from APC to lateral tunnel connection in the early series. However, we changed the surgical conversion strategy from APC or lateral tunnel connection to ECC Fontan conversion in 2002. We have described the surgical techniques in detail in a previous study [7].

We standardized the antiarrhythmia surgery by performing isthmus ablations for atrial reentry tachycardia, modified right-sided maze procedures for atrial flutter, and bilateral maze procedures for atrial fibrillation. The right-sided maze procedure was performed in four distinct locations between the posterior rim of the atrial septal defect (ASD) and the posterior edge or resected atrial wall, between the superior rim of the ASD and resected right atrial appendage, between the inferior vena cava (IVC) and the rim of the coronary sinus and between the IVC and the annulus of the right-sided atrioventricular valve. The left-sided maze procedure was performed in the following three regions: encircling the pulmonary veins, between the lower pulmonary veins and the mitral valve annulus and at the orifice of the left atrial appendage. We used a 10-mm linear probe [CCS-200, Cooper Surgical, Shelton, CT, USA] for 90–120 s based on the tissue thickness at  $-70^{\circ}\text{C}$ .

We tried to implant the permanent epicardial pacemaker simultaneously during the Fontan conversion. We tried to find the implantation site of the atrial lead around the sinoatrial node and of the ventricular lead at the ventricular apex. In addition, we used the bipolar steroid-eluting type lead in almost all cases. However, when implantation was difficult, we used the screw-type pacemaker lead at the ventricular site. The pacemaker implantation mode was dependent upon the atrioventricular node conduction status, and programming of the pacemaker was individualized to the patient.

### Patient follow-up

During the follow-up, patients were monitored using the functional classification of the NYHA, echocardiography, electrocardiography, continuous 24-h electrocardiographic monitoring, exercise pulmonary function test (PET) and pacemaker analysis. Early mortality was defined as death related to the operation within 30 days.

### Clinical outcomes

The median follow-up duration was 6.5 years (range 0.1–15.6 years). The median age at initial Fontan operation was 3.6 years (range: 1.3–15.8 years). The median age at Fontan conversion was 18.8

years (range 6.6–18.8 years). The mean conversion duration from initial Fontan operation was  $14.4 \pm 5.3$  years. The median duration of arrhythmias from detection to operation was 30.0 months (range 0.5–210.0 months,  $n = 17$ ). The mean cardiopulmonary bypass time was  $250.4 \pm 76.3$  min. The median extubation time was 15.0 h (range 2.0–474.0 h). In addition, the median time in the intensive care unit was 3.8 days (range 0.9–44.6 days). The median hospital stay was 19.5 days (range 9.0–61.0 days). The median length of chest tube drainage was 10.0 days (range 3.0–34.0 days). The mean graft size in ECC Fontan conversion cases was  $23.0 \text{ mm} \pm 1.2 \text{ mm}$  ( $n = 26$ ). Clinical outcomes are described in Table 1.

Preoperative symptoms included dyspnoea on exertion in 22 (71.0%) patients, palpitations in 8 (25.8%), fatigue in 1 (3.2%) and chest discomfort in 1 (3.2%). The most common indication for operation in the 33 cases was arrhythmias in 26 patients (78.8%), including atrial flutter in 21 (63.6%) patients, sinus node dysfunction in 6 (18.2%) and atrial fibrillation in 3 (9.1%), atrioventricular valve regurgitation (AVVR) in 11 (35.5%) patients and thrombus in 10 patients (32.3%) (Table 2). Twenty-six patients (83.9%) required the arrhythmia surgery (isthmus cryoablation = 9, right-sided maze = 13 and bilateral maze = 4) (Fig. 1). In addition, 23 patients (74.2%) received permanent pacemaker implantation (DDDR mode = 15, DDD = 6, and AAIR = 2) (Medtronic, Inc., Minneapolis, MN, USA, in 21 patients and St. Jude Medical, Sylmar, CA, USA, in 2 patients).

Tricuspid atresia was seen in 9 patients (29.0%) and was the most common diagnosis, followed by heterotaxy syndrome in 7 (22.6%) and double-outlet right ventricle (DORV) in 5 patients (16.1%). The right ventricle (RV) dominant ventricle in 17 patients (54.8%) was most commonly observed. The initial Fontan operations were atriopulmonary connection (APC) in 25 (80.6%) patients and lateral tunnel Fontan operation in 6 patients (19.4%). We performed the Fontan conversion from APC to lateral tunnel in 5 patients (16.1%), to ECC in 20 (64.5%) and from lateral tunnel to ECC in 6 (19.4%). Fontan fenestration during the first operation and Fontan conversion were performed in 2 patients (APC = 1, lateral tunnel = 1) and in 3 patients (lateral tunnel = 1, ECC = 2), respectively. The clinical characteristics of the patients are given in Table 2. Additional procedures were performed in 22 patients (71.0%) during the Fontan conversion. RA reduction plasty ( $n = 14$ , 45.1%) and thrombectomy ( $n = 8$ , 25.8%) were the most common additional procedures (Table 3).

### Statistical analysis

All data were expressed as means  $\pm$  standard deviations or as medians (ranges). Comparisons between continuous variables were performed using Student's *t*-tests. The analysis was performed with Fisher's exact test for categorical variables. Survival was estimated using the Kaplan–Meier method. A log-rank test was used for comparisons between factors. A *P*-value of  $<0.05$  was considered statistically significant. These analyses were performed using the SPSS (SPSS version 19.0, SPSS, Inc., Chicago, IL, USA) statistical package.

## RESULTS

### Surgical results

There was no early mortality. A total of 4 mortalities (1 in-hospital death and 3 late mortalities) occurred during the 6.5-year median

**Table 1:** Clinical characteristics of the patients

Variables	Values	Percent (%)
Age (year, mean ± SD)	14.4 ± 5.3	
Male/female ratio	17:14	
Body weight (kg, mean ± SD)	52.7 ± 12.9	
Diagnosis (n)	9	29.0
TA	7	22.6
Heterotaxy	5	16.1
DORV	5	16.1
UVH (RV type)	4	12.9
ccTGA	1	3.2
PA with IVS		
Ventricle type (n)		
RV dominant	17	54.8
LV dominant	11	35.5
Both	3	9.7
Initial Fontan type		
APC	25	80.6
Lateral tunnel	6	19.4
CPB time (mean ± SD, min)	250 ± 76.3	
Graft size (mean ± SD, mm)	23.0 ± 1.2 (n = 26)	
Extubation time (median, h)	15 (2.0–474.0)	
ICU stay (median, days)	3.8 (0.9–44.6)	
Hospital stay (median, days)	19.5 (9.0–61.0)	

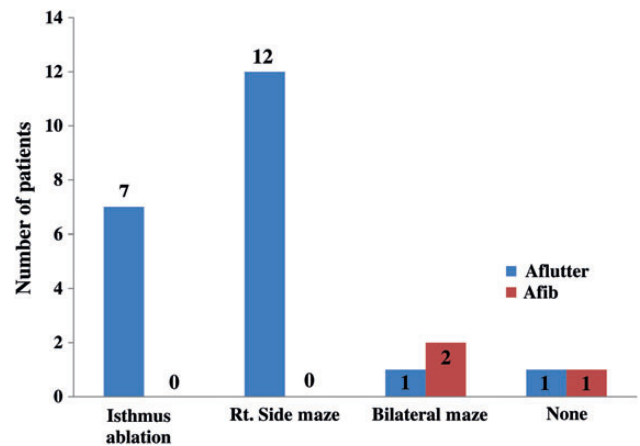
APC: atriopulmonary connection; ccTGA: congenitally corrected transposition of aorta; CPB: cardiopulmonary bypass; DORV: double-outlet right ventricle; ICU: intensive care unit; LV: left ventricle; MA: mitral atresia; PA with IVS: pulmonary atresia with intact ventricular septum; RV: right ventricle; SD: standard deviation; TA: tricuspid atresia; UVH: univentricular heart.

**Table 2:** Indications for operation

Indications for operation	n	Percent (%)
Atrial arrhythmia	33	
Atrial flutter	21	63.6
Aflutter + Afibrillation	3	9.1
SA node dysfunction	6	18.2
Junctional tachycardia	3	9.1
AVVR (>moderate)	11	35.5
Thrombus	10	32.3
Ventricular dysfunction	9	29.0
LC	5	16.1
RPV compression	3	9.7
Fontan pathway narrowing	3	9.7
Residual R-L shunt	3	9.7
PLE	3	9.7

Aflutter: atrial flutter; Afibrillation: atrial fibrillation; SA: sinoatrial; AVVR: atrioventricular valve regurgitation; RPV: right pulmonary vein; LC: liver cirrhosis; PLE: protein-losing enteropathy; R-L: right to left.

follow-up period (range 0.1–15.6 years). Actuarial 5-year survival was 90.0 ± 5.5%. One patient who had unbalanced AVSD and RV type functional single-ventricle underwent the ECC Fontan conversion at 23.2 years of age. He had preoperative AVVR, ventricular dysfunction and PLE. He died 35 days after the surgery due to Fontan failure. The second patient, who had DORV and preoperative PLE, underwent the ECC Fontan conversion at 18.1 years of age. He died 1.7 years after the operation due to Fontan failure. The third patient, who had heterotaxy syndrome, preoperative



**Figure 1:** Antiarrhythmia surgery in Fontan conversion according to arrhythmia type. We mainly performed isthmus ablation and right-sided maze procedures in atrial flutter and the bilateral maze procedure in atrial fibrillation. Aflutter: atrial flutter; Afib: atrial fibrillation; Rt: right.

**Table 3:** Additional procedures with Fontan conversion

Additional procedures	n (%)
RA reduction plasty	14 (45.1)
Thrombectomy	8 (25.8)
AV valve repair	8 (25.8)
AV valve replacement	6 (19.4)
PA angioplasty	5 (16.1)
Fenestration	3 (9.7)
Total	44

AV: atrioventricular; RA: right atrium.

AVVR and ventricular dysfunction, underwent the ECC Fontan conversion. He died 5 months later due to ventricular dysfunction. The last patient, who had tricuspid atresia, underwent the lateral tunnel type Fontan conversion and expired 12 years later due to an unknown cause. Twelve postoperative complications occurred in 9 patients (29.0%, 95% confidence interval (CI): 15.9%, 46.8%). Postoperative bleeding (n = 3) was the most common complication, followed by diaphragm palsy, prolonged pleural effusion, postoperative mediastinitis and transient seizures (n = 2).

Freedom from arrhythmia was 91.8 ± 5.5% at 5 years (Fig. 2). After Fontan conversion, normal sinus rhythm was maintained in 12 patients (38.7%, 95% CI: 23.7%, 56.2%), pacing-dependent rhythm in 10 patients (32.3%, 95% CI: 15.5%, 50.0%) (atrial pacing-dependent = 3, atrial and ventricular pacing-dependent = 6 and ventricular pacing-dependent = 1) and intermittent pacing-dependent rhythm in 4 patients (12.9%, 95% CI: 4.5%, 29.5%) (atrial pacing-dependent = 1 and ventricular pacing-dependent = 3). The median follow-up duration was 2.1 years (range 0.0–13.9 years) (Fig. 3). We used the antiarrhythmic medication (sotalol) in 8 patients at the latest follow-up. Among 8 patients, pacing-dependent rhythm was shown in 3 patients, normal sinus rhythm in 2 patients, atrial fibrillation in 2 patients and atrial flutter in 1 patient.

NYHA Fc improvement was statistically significant after Fontan conversion during the 5-year median follow-up (range 2.5–14.4 years) (P = 0.001) (mean preoperative Fc = 1.77, postoperative NYHA Fc = 1.13, n = 15, P < 0.001) (Fig. 4). Postoperative maximal oxygen consumption (MVO2) was 25.6 ± 4.9 (normal range

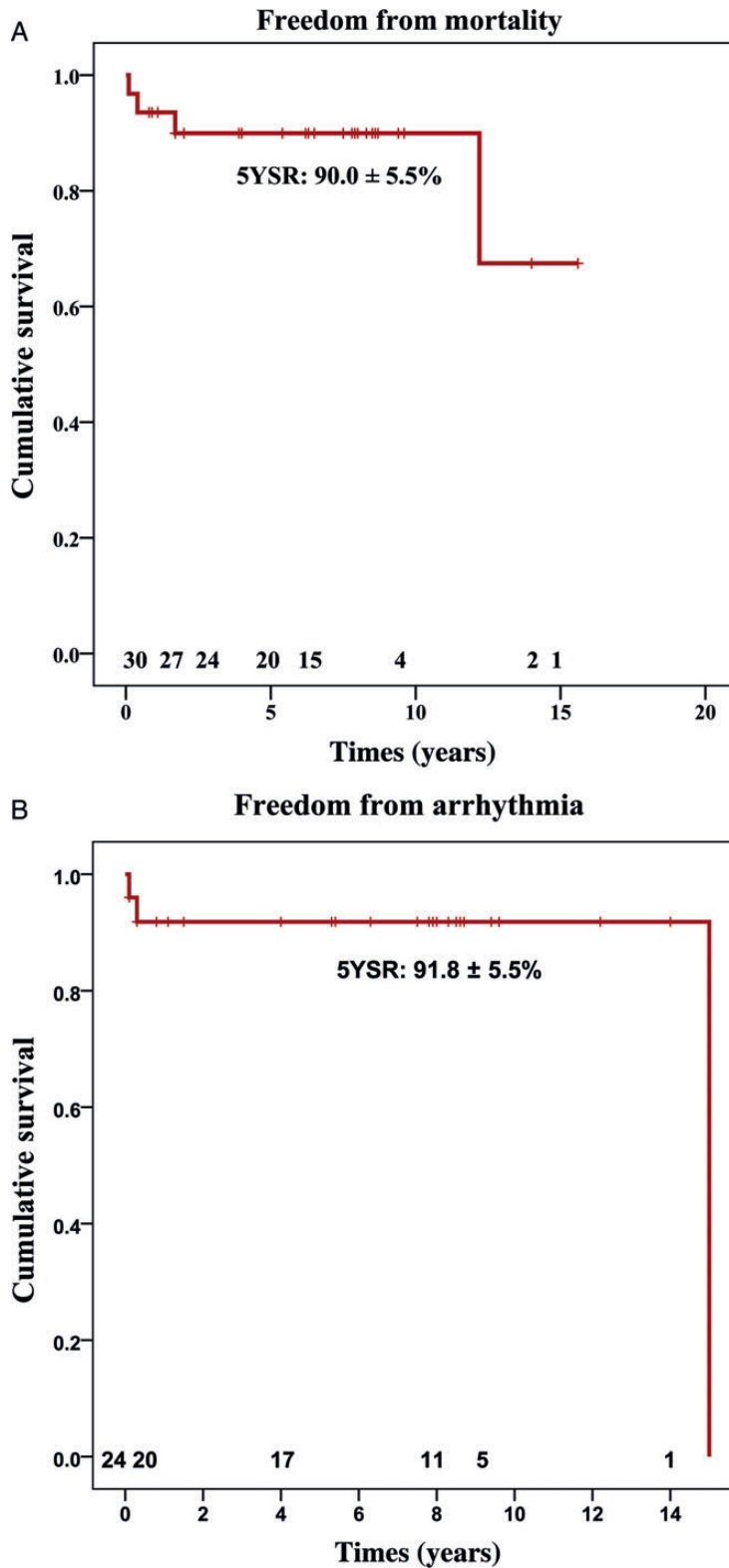
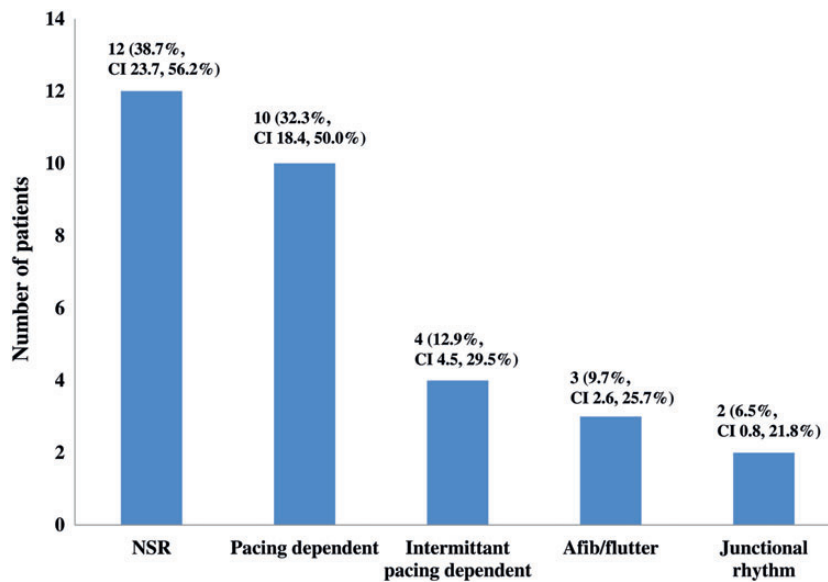


Figure 2: Freedom from mortality (A) and arrhythmia (B). YSR: year survival rate.

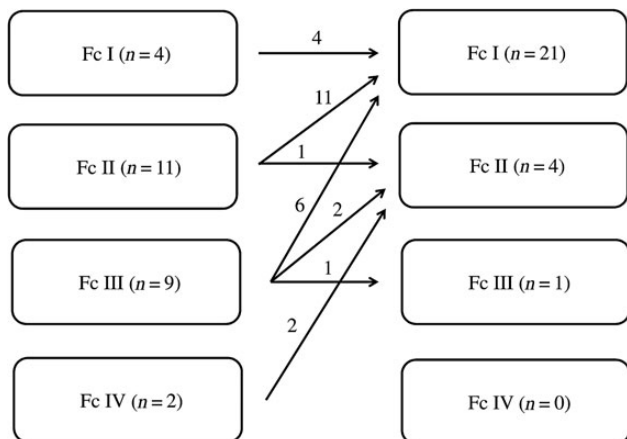
>30 ml/kg/min,  $n = 20$ ), and the minute ventilation/production of  $\text{CO}_2$  ( $\text{VE}/\text{VCO}_2$ ) slope during the exercise PFT was  $36.4 \pm 5.6$  ( $n = 20$ ) (normal range 26–29).

In a small number of patients ( $n = 9$ ), we evaluated the changes in ventricular function after Fontan conversion using

echocardiography. The median follow-up duration was 2.4 years (range 0.2–8.6 years). Although ventricular ejection fraction ( $39.2 \pm 8.3$  to  $45.0 \pm 3.6$ ,  $P = 0.246$ ), Tei index ( $0.6 \pm 0.1$ – $0.5 \pm 0.0$ ,  $P = 0.871$ ) and  $\text{dP}/\text{dt}$  ( $784.0 \pm 102.1$ – $810.2 \pm 77.1$ ,  $P = 0.956$ ) were improved after Fontan conversion, the improvements were not



**Figure 3:** Rhythm analysis after Fontan conversion. Twenty-six patients (83.9%) showed normal sinus or pacing-dependent regular rhythms. Afib: atrial fibrillation; Aflutter: atrial flutter; CI: 95% confidence interval; NSR: normal sinus rhythm.



**Figure 4:** NYHA functional class improvement ( $n = 26$ ). There was a statistically significant improvement in the New York Heart Association functional class (NYHA Fc) ( $P < 0.001$ ).

statistically significant. There were 3 patients with PLE. Two patients expired after Fontan conversion at 35 days and 1.7 days, and 1 patient showed no improvement in PLE during the 8.6-year follow-up.

**DISCUSSION**

In our series of 31 patients with failed Fontan circulation and arrhythmia, we accomplished the Fontan conversion including antiarrhythmia surgery with low operative mortality. The benefit of arrhythmia surgery with the Fontan conversion surgery was confirmed. In addition, NYHA Fc was improved, although we did not compare the functional improvement using the exercise pulmonary test.

Several studies have reported that almost half of the patients undergoing the Fontan procedure developed atrial arrhythmias in the AP Fontan connection, usually in association with significant

haemodynamic abnormalities in at least 50% of the patients in the long-term follow-up [2, 8]. Thus, the Fontan conversion with antiarrhythmia surgery was introduced, and several studies have reported the results for arrhythmia control [5, 9-11]. Here, we identified the effectiveness of arrhythmia surgery. Fontan conversion without antiarrhythmia surgery had a high recurrence rate of arrhythmias, and the poor haemodynamics has been addressed in several reports [12-14]. Mavroudis *et al.* [11] identified the anatomical circuits responsible for atrial arrhythmias using preoperative and intraoperative mapping. The isthmus was found to be confined on at least one aspect by a surgical repair site, resulting in the tachycardia mechanism [15]. Thus, we included isthmus ablation in all patients with antiarrhythmia surgery. In addition, we tried to perform the right-sided maze procedure in patients with atrial flutter and added the left-sided maze procedure in patients with atrial fibrillation. This strategy resulted in good arrhythmia-free rates during the postoperative follow-up. Although there is a study reporting that a right-sided maze procedure has an effect in atrial fibrillation [16], we thought that the right-sided maze procedure for atrial fibrillation had to be limited to selected cases.

We observed sinus node dysfunction in 6 patients (18.2%). Sinus node dysfunction may result from right atrial enlargement or from sinus node injury during the multistage Fontan operation [17]. In addition, sinus node dysfunction associated with bradycardia may predispose the patient to atrial flutter or fibrillation by promoting more variable refractory periods of the atrial muscles and may induce the re-entry mechanism [17]. Thus, we planned to implant an atrial pacemaker to prevent the bradycardia episodes, which may induce supraventricular or ventricular tachycardia [18]. In this study, we identified 14 (45.1%) patients who had continuous or intermittent pacing-dependent rhythms during the follow-up. Long duration of atrial fibrillation or flutter were risk factors for decreased sinus rhythm conversion rates, and the duration of atrial fibrillation or flutter to the Fontan conversion was relatively long and varied from 0.5 to 210 months in this study [19]. Thus, we emphasize the importance of permanent pacemaker implantation to maintain atrioventricular synchrony.



Giardini *et al.* [20] reported that Fontan conversion was associated with improvement of cardiopulmonary function and heart failure symptoms. However, they did not prove improvement in ventricular ejection fraction after Fontan conversion. We also found improvement in heart failure symptoms. However, we found that the absolute values of exercise PFT were in the sub-normal range after Fontan conversion at the 5.0-year median follow-up, but we did not compare the preoperative and postoperative PFT. In addition, we did not find a statistically significant improvement in ventricular function after Fontan conversion due to the small number of evaluations of ventricular function and the lack of an accurate definition of ventricular function in single-ventricle physiology. Although cardiac transplantation has been a surgical option for patients in whom the Fontan procedure has failed, the surgical results have to be taken into consideration due to the relatively high mortality rate [21, 22]. Thus, we expect that Fontan conversion may be the primary treatment of choice in selected patients after a failed Fontan procedure.

There are controversies about the PLE improvement after Fontan conversion [9, 10]. In this study, we observed that all 3 PLE patients had a high morbidity and mortality, similar to several previous reports [23, 24]. Two patients died after Fontan conversion, and 1 patient showed no improvement in PLE. Thus, further studies are needed to determine whether there are PLE improvements after Fontan conversion.

## CONCLUSIONS

Fontan conversion with antiarrhythmia surgery and permanent pacemaker implantation is a safe and reliable procedure. Fontan conversion is associated with low operative mortality and improvement in NYHA functional class, and has a low incidence of recurrent arrhythmias.

## Limitations of the study

There are several limitations to this study. This study was performed at a single centre and is limited by its retrospective nature. In addition, we recently checked the exercise PFT in Fontan conversion patients. Thus, we did not match the preoperative and postoperative pulmonary function in Fontan conversion patients. We tried to determine the improvement in ventricular function after Fontan conversion. However, it is difficult to draw any conclusions due to the small number of patients in this study and the lack of absolute criteria for ventricular function in single-ventricle physiology on echocardiography.

**Conflict of interest:** none declared.

## REFERENCES

- Fontan F, Mounicot FB, Baudet E, Simonneau J, Gordo J, Gouffrant JM. [Correction] of tricuspid atresia. 2 cases 'corrected' using a new surgical technic. *Ann Chir Thorac Cardiovasc* 1971;10:39-47.
- Cromme-Dijkhuis AH, Hess J, Hahlen K, Henkens CM, Bink-Boelkens MT, Eygelaar AA *et al.* Specific sequelae after Fontan operation at mid- and long-term follow-up. Arrhythmia, liver dysfunction, and coagulation disorders. *J Thorac Cardiovasc Surg* 1993;106:1126-32.
- de Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. Experimental studies and early clinical experience. *J Thorac Cardiovasc Surg* 1988;96:682-95.
- Backer CL, Deal BJ, Mavroudis C, Franklin WH, Stewart RD. Conversion of the failed Fontan circulation. *Cardiol Young* 2006;16(Suppl 1):85-91.
- Mavroudis C, Backer CL, Deal BJ, Stewart RD, Franklin WH, Tsao S *et al.* Evolving anatomic and electrophysiological considerations associated with Fontan conversion. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2007;136-45.
- Mavroudis C, Deal BJ, Backer CL. The beneficial effects of total cavopulmonary conversion and arrhythmia surgery for the failed Fontan. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2002;5:12-24.
- Kim WH, Lim HG, Lee JR, Rho JR, Bae EJ, Noh CI *et al.* Fontan conversion with arrhythmia surgery. *Eur J Cardiothorac Surg* 2005;27:250-7.
- Driscoll DJ, Offord KP, Feldt RH, Schaff HV, Puga FJ, Danielson GK. Five- to fifteen-year follow-up after Fontan operation. *Circulation* 1992;85:469-96.
- Sheikh AM, Tang AT, Roman K, Baig K, Mehta R, Morgan J *et al.* The failing Fontan circulation: successful conversion of atriopulmonary connections. *J Thorac Cardiovasc Surg* 2004;128:60-6.
- Weinstein S, Cua C, Chan D, Davis JT. Outcome of symptomatic patients undergoing extracardiac Fontan conversion and cryoablation. *J Thorac Cardiovasc Surg* 2003;126:529-36.
- Mavroudis C, Backer CL, Deal BJ, Johnsrude C, Strasburger J. Total cavopulmonary conversion and maze procedure for patients with failure of the Fontan operation. *J Thorac Cardiovasc Surg* 2001;122:863-71.
- Balaji S, Johnson TB, Sade RM, Case CL, Gillette PC. Management of atrial flutter after the Fontan procedure. *J Am Coll Cardiol* 1994;23:1209-15.
- Marcelletti CF, Hanley FL, Mavroudis C, McElhinney DB, Abella RF, Marianeschi SM *et al.* Revision of previous Fontan connections to total extracardiac cavopulmonary anastomosis: a multicentre experience. *J Thorac Cardiovasc Surg* 2000;119:340-6.
- van Son JA, Mohr FW, Harnschi J, Schneider P, Hess H, Haas GS. Conversion of atriopulmonary or lateral atrial tunnel cavopulmonary anastomosis to extracardiac conduit Fontan modification. *Eur J Cardiothorac Surg* 1999;15:150-7; discussion 57-8.
- Kalman JM, VanHare GF, Olgin JE, Saxon LA, Stark SI, Lesh MD. Ablation of 'incisional' reentrant atrial tachycardia complicating surgery for congenital heart disease. Use of entrainment to define a critical isthmus of conduction. *Circulation* 1996;93:502-12.
- Theodoro DA, Danielson GK, Porter CJ, Warnes CA. Right-sided maze procedure for right atrial arrhythmias in congenital heart disease. *Ann Thorac Surg* 1998;65:149-53; discussion 53-4.
- Manning PB, Mayer JE Jr, Wernovsky G, Fishberger SB, Walsh EP. Staged operation to Fontan increases the incidence of sinoatrial node dysfunction. *J Thorac Cardiovasc Surg* 1996;111:833-9; discussion 39-40.
- Silka MJ, Manwill JR, Kron J, McAnulty JH. Bradycardia-mediated tachyarrhythmias in congenital heart disease and responses to chronic pacing at physiological rates. *Am J Cardiol* 1990;65:488-93.
- Chen MC, Chang JP, Chen YL. Surgical treatment of atrial fibrillation with concomitant mitral valve disease: an Asian review. *Chang Gung med J* 2008;31:538-45.
- Giardini A, Pace Napoleone C, Specchia S, Dondi A, Formigari R, Oppido G *et al.* Conversion of atriopulmonary Fontan to extracardiac total cavopulmonary connection improves cardiopulmonary function. *Int J Cardiol* 2006;113:341-4.
- Jayakumar KA, Addonizio LJ, Kichuk-Chrisant MR, Galantowicz ME, Lamour JM, Quaegebeur JM *et al.* Cardiac transplantation after the Fontan or Glenn procedure. *J Am Coll Cardiol* 2004;44:2065-72.
- Michielon G, Parisi F, Squitieri C, Carotti A, Gagliardi G, Pasquini L *et al.* Orthotopic heart transplantation for congenital heart disease: an alternative for high-risk Fontan candidates? *Circulation* 2003;108(Suppl 1):II140-9.
- Mertens L, Hagler DJ, Sauer U, Somerville J, Gewillig M. Protein-losing enteropathy after the Fontan operation: an international multicentre study. PLE study group. *J Thorac Cardiovasc Surg* 1998;115:1063-73.
- Rogers LS, Glatz AC, Ravishankar C, Spray TL, Nicolson SC, Rychik J *et al.* 18 Years of the Fontan operation at a single institution: results from 771 consecutive patients. *J Am Coll Cardiol* 2012;60:1018-25.