

# Surgical Therapy of Arrhythmias in Single-Ventricle Patients Undergoing Fontan or Fontan Conversion

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**ABSTRACT** *Background:* Arrhythmia is detrimental to Fontan hemodynamics. Clinical outcomes among Fontan patients who underwent antiarrhythmic treatment were retrospectively reviewed. *Methods:* From January 1996 to January 2007, 182 patients underwent a Fontan procedure, including Fontan conversion. Thirty-nine of the 182 patients showed various arrhythmias pre- or post-Fontan operations, and were treated surgically including Fontan conversion (18 patients) or medically. The authors analyzed the outcomes of arrhythmia treatments retrospectively. *Results:* Thirty-nine patients (21.4%) showed various arrhythmias, such as atrial flutter, atrial fibrillation, junctional rhythm, sinus node dysfunction, or brady tachyarrhythmia pre- or post-Fontan procedure. Follow-up duration was  $13.1 \pm 8.7$  years (11 months to 325 months). Atrial flutter and fibrillation only developed in 17 patients who received atriopulmonary connection Fontan, and who were treated by Fontan conversion with concomitant procedures such as Cox-maze procedure (two patients), right-side maze and pacemaker implantation (five patients), right atrial isthmus ablation (four patients), right atrial isthmus cryoablation and pacemaker implantation (five patients), and only pacemaker implantation (one patient). The 21 patients who showed arrhythmia at the time of the Fontan procedure underwent the following procedures concomitantly: right atrial isthmus cryoablation with pacemaker implantation (one patient), right atrial isthmus cryoablation (one patient), or pacemaker implantation (nine patients). The remaining 10 patients, who showed junctional rhythm, sinus bradycardia, or intermittent ectopic beats, were managed medically. There were two late mortalities due to protein-losing enteropathy. As a result, 33 patients (89.2%) maintained atrioventricular synchrony, 19 in sinus rhythm and 14 supported by a DDD-type pacemaker. The remaining four patients (10.8%) showed persistent junctional rhythm with a stable hemodynamic status. *Conclusions:* The various arrhythmias in Fontan patients were well controlled by aggressive surgical management. doi: 10.1111/j.1540-8191.2009.00914.x (*J Card Surg* 2009;24:738-741)

It is well known that arrhythmia is detrimental to Fontan hemodynamics, and that it affects long-term morbidity and mortality, and thus, many surgeons and physicians try to manage arrhythmia aggressively in Fontan patients or Fontan candidates. The aim of this study was to assess the clinical statuses of Fontan patients with arrhythmias that had been treated medically or surgically.

## METHODS

Between January 1996 and January 2007, 182 patients underwent a Fontan-type operation at the Seoul National University Children's Hospital. We performed lateral tunnel Fontan in 67 patients, extracardiac conduit Fontan in 97, and Fontan conversion in 18 patients. The overall follow-up duration for these 182 patients was  $13.1 \pm 8.7$  years (11 to 325 months). Thirty-nine (21.4%) of the 182 patients showed various arrhythmias, such as atrial flutter, atrial fibrillation, sinus node dysfunction, brady- or tachyarrhythmia, or junctional rhythm. Diagnoses included univentricular heart (right ventricle type in 15 and left ventricle type in one patient), tricuspid atresia (nine patients), heterotaxy syndrome (right isomerism in two, and left isomerism in two patients), criss-cross heart (two patients), transposition of great arteries with hypoplastic left ventricle (two patients), double outlet right ventricle with

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**TABLE 1**  
**Numbers of Patients Who Underwent Different Treatment Strategies by Study Group**

	Fontan Conversion ± Concomitant Procedure (18)	Fontan ± concomitant Procedure (11)	Medical Treatment (10)
Concomitant procedure	Cox-maze (2)	RA isthmus ablation + pacemaker implantation (1)	
	RA maze (5)	RA isthmus ablation (1)	
Permanent pacemaker	RA isthmus ablation (9)	Pacemaker implantation only (9)	
Mortality	11	10	
AV synchrony/ living patients	1	1	7/10
	17/17	9/10	

RA = right atrium; AV = atrioventricle.

tricuspid valve straddling, and others (four patients). We retrospectively reviewed the clinical outcomes and functional statuses of the 39 arrhythmia patients after antiarrhythmic treatments. The Seoul National University Hospital Institutional Review Board (IRB) reviewed and approved this study and individual consent was waived (IRB No.: H-0808-032-253).

### RESULTS

We divided the 39 arrhythmia patients into three groups by treatment strategy as follows: Fontan conversion operation with or without a concomitant antiarrhythmic procedure for previous Fontan patients (group 1, 18 patients), Fontan operation with or without a concomitant antiarrhythmic procedure for Fontan candidate patients (group 2, 11 patients), and the medically treated group (group 3, 10 patients) (Table 1).

In group 1, which contained 18 patients, we performed a Fontan conversion procedure. There were 14 atriopulmonary Fontan and four lateral tunnel Fontan patients before Fontan conversion. In this study, atrial flutter and fibrillation were only found in this group. The mean interval between the previous Fontan operation and Fontan conversion was  $13.0 \pm 4.0$  years. We performed concomitant arrhythmia surgery, such as Cox-maze (two patients), right-side maze (five patients), right atrial isthmus ablation (nine patients) in 16 patients, and 11 permanent pacemaker implantations. In three of these 16 patients, surgical antiarrhythmic procedures were performed prophylactically, because their electrocardiogram (ECG) findings raised suspicion of sinus node dysfunction, sinus rhythm with a short PR interval, and intermittent bradycardia, respectively. Excepting the one patient who expired, all living patients in group 1 had an atrioventricular synchronic status. In group 2, the Fontan procedure with or without a concomitant surgical arrhythmic treatment group, there were 11 patients. Of these, two patients, who had shown arrhythmia before the Fontan procedure, underwent concomitant arrhythmia surgery, namely, right atrial isthmus cryoablation with or without permanent pacemaker implantation. The remaining nine patients underwent the Fontan procedure with pacemaker implantation. As a result, excluding a pa-

tient who expired and a patient with junctional rhythm, nine patients maintained an atrioventricular synchronic status at last follow-up. We have managed the patients who showed the arrhythmia such as junctional rhythm in groups 1 and 2 after surgical procedure with antiarrhythmic agents, digoxin or amiodarone. In group 3, which contained 10 patients, arrhythmias were managed medically after the Fontan procedure. These patients showed various arrhythmias, such as junctional rhythm, sinus bradycardia, and intermittent ectopic beats. Seven of these 10 patients showed normal sinus rhythm conversion during medical treatment, and the three remaining patients, though not converted to sinus rhythm, showed a stable hemodynamic status at last follow-up visits.

We encountered four isomerism patients (two left and two right) in this study. The two left isomerism patients showed bradycardia and sinoatrial node dysfunction with atrial flutter, respectively, and the two right isomerism patients also showed arrhythmias, for example, sinus node dysfunction and complete atrioventricular block. One of the right isomerism patients expired due to protein-losing enteropathy. In fact, both of the two late mortalities were related to protein-losing enteropathy; one of these patients suffered from liver cirrhosis, Child class C.

Finally, 33 of the 37 living patients (89.2%) maintained atrioventricle synchrony. Fifteen patients showed a normal sinus rhythm, and 14 patients were supported by a permanent pacemaker in an atrioventricular synchronic status. Another four of these 33 patients showed sinus bradycardia. All sinus rhythm patients had New York Heart Association (NYHA) functional class I or II. Four nonsinus rhythm patients (four of 37) were in junctional rhythm status, but their hemodynamic statuses were tolerable.

### DISCUSSION

The maintenance of a normal sinus rhythm is a precondition of a stable hemodynamic status in all heart disease patients. In terms of biventricular physiology, there is a relatively broad hemodynamic safety margin when patients have a nonsinus rhythm status, such as junctional rhythm, atrial fibrillation, or flutter. Some

patients with an atrial fibrillation maintain a normal life under only medical management for anticoagulation or heart rate control. However, univentricular physiologies, such as heterotaxy syndrome, the maintenance of a normal sinus rhythm with atrioventricular synchrony, or a normofrequent supraventricular rhythm and the avoidance of considerable bradycardia, are the most important factors.<sup>1,2</sup> This is not only because of postoperative complications like tachyarrhythmias, but also to promote quality of life and long-term survival. In Fontan patients, arrhythmias have been noted in up to 40% at 10-year follow-ups, and are most common among those with an atriopulmonary anastomosis.<sup>3,4</sup> The main cause of arrhythmia in these patients is probably right atrial enlargement, which increases systemic venous pressure and can cause atrial arrhythmias, compression of the right pulmonary veins, coronary sinus hypertension, atrial thrombi formation, and pulmonary embolism. This results in a low cardiac output status, and a failing Fontan. Furthermore, increased venous pressure causes progressive right atrial dilation, which in turn causes further fluid energy losses, increases the work required to pump blood to pulmonary arteries and central venous pressure, and decreases cardiac output.<sup>5-8</sup> In this study, there were 18 Fontan conversion patients, and 14 of these, which included 10 atrial flutter patients and two atrial fibrillation patients, had undergone atriopulmonary anastomosis before Fontan conversion and had presented with Fontan failure. We categorized these patients into group 1. We performed the Cox-maze procedure for atrial fibrillation patients, and for atrial flutter patients, right-side maze or only right atrial isthmus ablation procedure. All these failing Fontan patients, except for the single patient who expired, showed improved cardiac function after Fontan conversion with concomitant antiarrhythmic procedures. The one expired patient had progressive liver cirrhosis (Child class C) and protein-losing enteropathy, despite Fontan conversion with a concomitant antiarrhythmic procedure. We have previously reported the outcomes of some part of the patients in Fontan conversion group.<sup>9</sup> Four lateral tunnel Fontan patients with failing Fontan features also showed an improved functional class after a Fontan conversion procedure. With regard to the failure modes of atriopulmonary anastomosis Fontan procedures, lateral tunnel Fontan procedures cannot be free of common atrial cavity dilatation with the passage of time. In addition to atrial dilatation, lateral tunnel Fontan procedures have another important arrhythmic focus—the long suture line in the atrial free wall.<sup>10,11</sup>

In group 2, we performed the Fontan procedure with concomitant procedures, namely, nine permanent pacemaker implantations and two right atrial isthmus cryoablation procedures. With the exception of the patient who expired and one junctional rhythm patient, nine patients maintained atrioventricle synchrony. The patient who showed junctional rhythm had been performed Fontan procedure with right atrial isthmus cryoablation. Just 50% (one of two) of patients who had undergone right atrial isthmus ablation procedure could maintain the normal sinus rhythm after pro-

cedure. Deal et al.<sup>12</sup> reported that the modified right atrial maze procedure is superior to simple anatomic isthmus block for treating atrial tachycardia in postoperative Fontan patients. In this previous study, the limitations of right atrial isthmus ablation for normal sinus conversion or for recurrent atrial tachycardia in single-ventricle patients were explained. In view of the diagnosis of the patient with junctional rhythm encountered in this study, that is, tricuspid atresia, right atrial isthmus ablation was probably insufficient. In cases of tricuspid atresia, ablation of the cavotricuspid isthmus may be incomplete, because the right atrium has a muscular bed at the atrioventricular junction, and the tricuspid valve has no true orifice. As a result, the reentry circuit is unlikely to be completely ablated unless the maneuver is extended to the point of the dimple where atrial musculature is lacking.<sup>13</sup> Thus, we suggest that in tricuspid atresia patients, more aggressive antiarrhythmic surgery might be needed than in other single-ventricle patients. The patient who expired in this group had right isomerism, and underwent right isthmus cryoablation with permanent pacemaker implantation. The cause of death of this patient was protein-losing enteropathy associated with right heart failure.

In group 3, 10 patients were treated medically after the Fontan procedure, and all showed junctional rhythm, sinus bradycardia, and intermittent ectopic beats before medication. After medication, the rhythms of seven patients were converted to a normal sinus rhythm, and three retained a junctional rhythm. Fontan procedure itself may be helpful to restore the normal sinus rhythm with only postoperative medication without any surgical antiarrhythmic procedures in these patients.

## CONCLUSIONS

Various kinds of arrhythmias in Fontan or Fontan candidate patients were well controlled by surgical or medical treatments. Satisfactory midterm results were obtained in these patients using only the Fontan procedure or Fontan plus concomitant antiarrhythmic procedures, such as the maze or modified maze operation and/or permanent pacemaker implantation, the Fontan conversion procedure following the Fontan procedure. In Fontan patients, to improve long-term results and quality of life, aggressive antiarrhythmic procedures are recommended.

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