The Effects of Modification to Lateral Tunnel Fontan Procedure for Prophylactic Arrhythmia Surgery

Hong-Gook Lim, MD, PhD, Jeong Ryul Lee, MD, PhD, and Yong Jin Kim, MD, PhD

Department of Thoracic and Cardiovascular Surgery, Seoul National University Hospital, Seoul, Korea

CONGENITAL HEART

Background. Refractory atrial arrhythmias and hemodynamic abnormalities are responsible for significant morbidity and mortality after the Fontan operation. We evaluated the long-term feasibility, safety, and efficacy of prophylactic atrial arrhythmia surgery performed concomitantly with the lateral tunnel Fontan operation.

Methods. From 1997 August to 2003 December, 27 patients underwent a initial lateral tunnel Fontan with an interventional atrial incision and cryoablation from the atriotomy to the coronary sinus and right atrioventricular valve annulus. This novel surgical technique consists of (1) right atriotomy extending to the coronary sinus to block the slow rate conduction isthmus; (2) cryoablation between right atriotomy and right atrioventricular valve annulus; (3) baffling to avoid injury to the crista terminalis; and (4) use of a sandwich technique with closure of right atriotomy incorporating the Gore-Tex (W. L. Gore & Associates, Flagstaff, AZ) patch to reduce atrial suture line.

Results. There has been no early death after operation and one late death, which was not arrhythmic in etiology. At late follow-up of 15.2 ± 2.9 years (range, 5.5 to 18.0) after Fontan, spontaneous intraatrial reentrant tachycardia occurred in 1 patient, and inducible intraatrial reentrant tachycardia in 1 patient who required beta-blocker medication without ablation attempts. There was no evidence of early or late complications related to the interventional atrial incision and cryoablation. Four patients required late pacemaker implantation for sinus node dysfunction after Fontan operation.

Conclusions. The prophylactic arrhythmia surgery with our novel modification of the lateral tunnel Fontan procedure to reduce the development of intraatrial reentrant tachycardia was feasible and safe. Long-term follow-up results also demonstrated that this novel modification is effective for the prophylaxis of intraatrial reentrant tachycardia.

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Refractory atrial arrhythmias and hemodynamic abnormalities are responsible for significant late morbidity and mortality after the Fontan operation [1, 2]. Lateral tunnel Fontan patients often have extensive atrial scarring and are prone to atrial dilation, hypertrophy, and fibrosis, which are responsible for atrial tachyarrhythmia [3, 4]. Intraatrial reentrant tachycardia, a type of macroreentrant tachycardia, is the most common form of atrial tachyarrhythmia in Fontan patients, and is related to the incision and suture line, atrial remodeling, atrioventricular valve regurgitation, hemodynamic deterioration, cardiac anatomic substrates, sinus node dysfunction, age at Fontan operation, and duration after Fontan operation [5]. The incidence of intraatrial reentrant tachycardia also increases with the duration of the Fontan circulation [3, 4, 6]. The intricacies and mechanisms of the intraatrial reentrant tachycardia circuits have been well established in Fontan patients, but the responses to antiarrhythmic medications and radiofrequency catheter ablation with mapping are not satisfactory because of limited efficacy or side effects [5, 7, 8]. Fontan conversion with arrhythmia surgery may improve arrhythmia control, but it may be appropriate for selected patients only [9, 10]. Intraatrial reentrant tachycardia reduces quality of life and may even cause death in hemodynamically compromised patients [2]. Therefore, intraatrial reentrant tachycardia should be controlled aggressively. It can also be prevented prophylactically at the time of the Fontan operation.

We developed a novel surgical technique because the mechanisms for intraatrial reentrant tachycardia in lateral tunnel Fontan patients are complex. The atrial incisions and suture lines for the lateral tunnel baffling contribute to the occurrence of intraatrial reentrant tachycardia [11, 12]. The interventional atrial incision and cryoablation connecting the right atriotomy to the coronary sinus and right atrioventricular valve annulus could create intraatrial conduction block, interrupt

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Address correspondence to Dr Kim, Department of Thoracic and Cardiovascular Surgery, Seoul National University Hospital, 101 Daehak-ro, Jongno-gu, Seoul 110-744, Korea; email: kyj@plaza.snu.ac.kr.

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intraatrial reentrant tachycardia circuits from developing around the atriotomy, coronary sinus, and right atrioventricular valve annulus, and reduce the overall incidence of intraatrial reentrant tachycardia. Based on this concept, right atriotomy was extended to the coronary sinus to block slow rate conduction isthmus, and cryoablation was performed between the right atriotomy and right atrioventricular valve annulus. Because the location of the intraatrial suture line in relation to the crista terminalis also induces intraatrial reentrant tachycardia, the lateral tunnel baffling was modified to avoid injury to the crista terminalis [12]. In addition, the surgical technique was also modified, and the sandwich technique was used with closure of the right atriotomy incorporating the Gore-Tex (W. L. Gore & Assoc, Flagstaff, AZ) patch to reduce the atrial suture line (Fig 1, Video).

The aim of this study was to evaluate the long-term feasibility, safety, and efficacy of prophylactic atrial arrhythmia surgery performed concomitantly with the lateral tunnel Fontan operation.

Material and Methods

Patients

From August 18, 1997 to December 8, 2003, 27 patients underwent an initial lateral tunnel Fontan operation with an interventional atrial incision/cryoablation from the atriotomy to the coronary sinus/right atrioventricular valve annulus, which might increase the conduction time. Extracardiac conduit Fontan repair was performed for an anticipated reduction in arrhythmia burden. When lateral tunnel Fontan repair was performed to avoid the risk of anticoagulation therapy and no growth potential, prophylactic arrhythmia surgery was performed to reduce the development of intraatrial reentrant tachycardia. There was no selection bias in this population. In particular, the patients who had previous atrial surgery were not eliminated from the selection.

There were 15 male and 12 female patients. Age at Fontan operation was 3.39 ± 3.69 years (range, 2.14 to 17.56). Body weight was 14.04 ± 8.86 kg (range, 11 to 53), height was 93.03 ± 20.70 cm (range, 88 to 160), and body surface area was 0.59 ± 0.24 m^2 (range, 0.53 to 1.54). Diagnoses included double-outlet right ventricle in 8 patients, tricuspid atresia in 7 patients, right ventricle type single ventricle in 7 patients, left ventricle type single ventricle in 4 patients, and pulmonary atresia with intact ventricular septum in 1 patient. The systemic ventricle was a dominant left ventricle in 12 patients, biventricle in 8 patients, and a dominant right ventricle in 7 patients. Three patients had heterotaxy syndrome with right isomerism, and 1 patient had heterotaxy syndrome with left isomerism. The segmental cardiac anatomy was SDS (S = solitus, D = D-loop) in 6 patients, SDD (D = D-transposition) in 6 patients, SDL (L = L-transposition) in 5 patients, ADD (A = ambiguous) in 3 patients, SLD (L = L-loop) in 2 patients, SLL (S = solitus, L = L-loop) in 2 patients, IDD (I = inversus) in 1 patient, ILL (I = inversus, L = L-loop) in 1 patient, and AXD (X = unknown) in 1 patient.

Pre-Fontan balloon atrial septostomy was performed in 1 patient, Blalock-Taussig shunt was performed in 10 patients, and pulmonary artery banding was performed in 6 patients. Coarctoplasty was performed in 1 patient, and total anomalous pulmonary venous return repair was performed in 1 patient before the Fontan operation. Staged Fontan operation was performed in 18 patients. Pre-Fontan echocardiographic data showed that all patients could be candidates for the Fontan operation. Pre-Fontan catheterization showed that the systolic pulmonary arterial pressure was 13.1 ± 3.3 mm Hg (range, 9 to 23), ventricular end-diastolic pressure was 7.8 ± 3.1 mm Hg (range, 2 to 15), transpulmonary pressure gradient was 6.8 ± 3.0 mm Hg (range, 2 to 16), and pulmonary vascular resistance was 2.52 ± 1.23 Wood units · m^2 (range, 0.93 to 5.83).

Surgical Techniques

The novel surgical technique consisted of the following: (1) right atriotomy extending to the coronary sinus to block

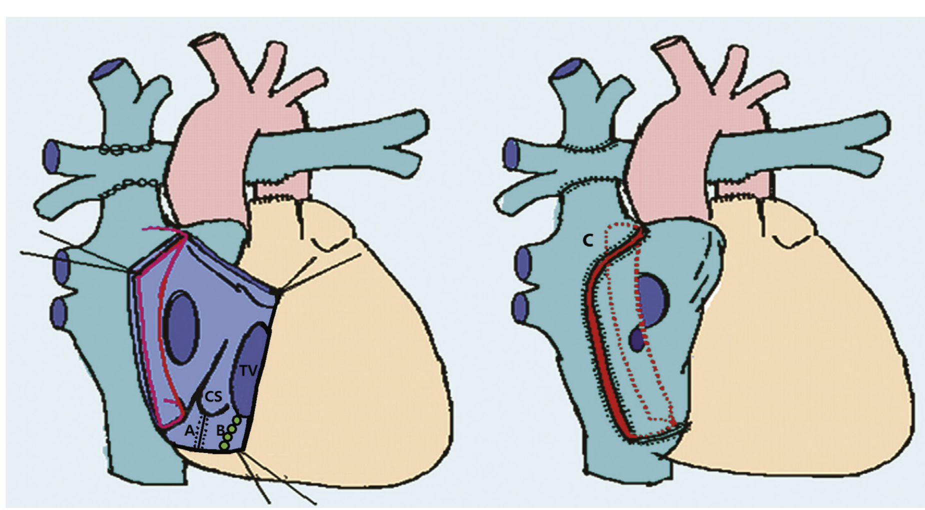

Fig 1. Schematic drawings of prophylactic arrhythmia surgery at the time of lateral tunnel Fontan operation. Right atriotomy extending to the coronary sinus (CS; left drawing, A), and cryoablation between right atriotomy and tricuspid valve (TV; left drawing, B). Sandwich closure of right atriotomy (right drawing, C).

slow rate conduction isthmus; (2) cryoablation between right atriotomy and right atrioventricular valve annulus; (3) baffling to avoid injury to the crista terminalis; and (4) use of sandwich technique with closure of right atriotomy incorporating the Gore-Tex patch to reduce the atrial suture line (Fig 1, Video). At the time of Fontan operation, fenestration was performed in 26 patients, pulmonary artery angioplasty was performed in 12 patients, atrioventricular valvuloplasty was performed in 2 patients, widening of the pulmonary vein was performed in 2 patients, and permanent pacemaker implantation for complete atrioventricular block was performed in 1 patient. Cardiopulmonary bypass time was 129.26 ± 25.98 minutes (range, 72 to 189), and aortic cross-clamp time was 58.22 ± 16.88 minutes (range, 31 to 95).

Evaluations

The study was approved by the Institutional Review Board of Seoul National University Hospital (H-1508-180-703). We reviewed the clinical notes, electrocardiograms, 24-hour Holter monitor records, echocardiograms, cardiac catheterization, electrophysiologic studies, and cardiopulmonary exercise tests. Electrophysiologic studies to assess the inducibility of intraatrial reentrant tachycardia at the time of atrial stimulation were performed with and without isoproterenol intravenous infusion. After the Fontan operation, we performed electrophysiologic studies 21 times in 19 patients (70.4%), and spontaneous or inducible intraatrial reentrant tachycardia was documented by electrocardiograms, 24-hour Holter monitor records, and electrophysiologic studies. Sinus node dysfunction was defined as sinus bradycardia with a resting heart rate more than 2 SD lower than normal for the patient's age, predominant junctional rhythm, or sinus pause of 3 seconds or longer, with or without escape beats [13, 14].

Statistical Analyses

Statistical analyses were performed using IBM SPSS Statistics 22.0 (IBM Corporation, Armonk, NY). Data were expressed as mean ± SD or median with ranges, as appropriate. Freedom from events, including intraatrial reentrant tachycardia and sinus node dysfunction, was determined by the Kaplan-Meier method. Variables were evaluated by using the likelihood ratio test in the Cox proportional hazards regression model. Hazard ratio (HR) with 95% confidence interval was calculated for the significant predictors. A p value less than 0.05 was considered statistically significant.

Results

Mortality

Among 27 patients, there has been no case of early death. There has been 1 late death (3.7%); the patient had right isomerism, right ventricle type single ventricle, and supracardiac type total anomalous pulmonary venous return, and it was necessary to perform correction of total anomalous pulmonary venous return and atrioventricular valvuloplasty before the Fontan operation. This patient underwent repair of severe atrioventricular valve regurgitation at the time of Fontan operation, needed pacemaker implantation for sinus node dysfunction after the Fontan operation, and died of brain damage after pulmonary vein widening at 5.9 years after the Fontan operation.

Intraatrial Reentrant Tachycardia

Nineteen of the 27 patients (70.4%) underwent a diagnostic electrophysiologic study at 6.8 ± 3.8 years (range, 1.2 to 12.6) after the Fontan operation. At late follow-up performed 15.2 ± 2.9 years (range, 5.5 to 18.0) after Fontan, it was observed that spontaneous intraatrial reentrant tachycardia occurred in 1 patient at 12.6 years after the Fontan operation. A beta-blocker drug was needed, but the ablation procedure was not performed. Freedom from development of spontaneous intraatrial reentrant tachycardia requiring a beta-blocker drug was 96.0% ± 3.9% at 18 years after the Fontan operation. On electrophysiologic study performed 11.8 years after Fontan operation, inducible intraatrial reentrant tachycardia occurred in 1 patient and required a beta-blocker drug without ablation attempts. Freedom from development of spontaneous or inducible intraatrial reentrant tachycardia on electrophysiologic study requiring a beta-blocker drug was 92.0% ± 5.4% at 18 years after the Fontan operation. On electrophysiologic study, inducible nonsustained intraatrial reentrant tachycardia occurred in 2 patients at 5.2 years and 10.7 years after the Fontan operation, respectively, but neither a beta-blocker drug nor ablation attempts were required for both patients until 16.8 years after the Fontan operation. Freedom from development of spontaneous, inducible sustained, or nonsustained intraatrial reentrant tachycardia on electrophysiologic study that did or did not require a beta-blocker drug was 84.3% ± 7.2% at 18 years after the Fontan operation (Table 1, Fig 2).

Multivariate analysis demonstrated that systolic pulmonary arterial pressure just before the Fontan operation (HR 1.9, $p = 0.029$) and greater than mild degree of atrioventricular valve regurgitation during follow-up after the Fontan operation (HR 251.4, $p = 0.014$) were statistically significant risk factors for spontaneous, inducible sustained, or nonsustained intraatrial reentrant tachycardia on electrophysiologic study that did or did not require a beta-blocker drug. The segmental anatomy of atrioventricular discordance, such as SLL, SLD, and IDD, has been commonly associated with conduction anomaly and tended to be more associated with intraatrial reentrant tachycardia occurrence in our univariate analysis (HR 5.521, $p = 0.089$).

Surgical Outcomes

The early postoperative evaluations showed that the average heart rate on an electrocardiogram was 98.9 ± 25.1 beats per minute, and the median PR interval on a 12-lead electrocardiogram was 146 ms (range, 106 to 196). The most recent evaluations showed that the average heart rate on an electrocardiogram was 74.0 ± 19.6 beats per minute, and the median PR interval on a 12-lead electrocardiogram was 164 ms (range, 118 to 256). There were two new cases of first-degree atrioventricular block

Table 1. Overview of Fontan Patients With Prophylactic Arrhythmia Surgery

Pt. No.	Sex	Age at Fontan (years)	Segmental Anatomy	Dominant Ventricle	Follow-Up (years)	SND (years)[a]	PR Interval (ms)[b]	IART (years)[c]	EPS (years)[d]
1	Female	3.9	SDD	Biventricle	18.0	…	188 (−)	…	5.4 (−)
2	Male	11.1	ADD	Biventricle	17.8	…	128 (−)	…	12.1 (−)
3	Male	6.1	SDL	LV	17.6	…	136 (−)	…	…
4	Male	1.3	SDS	LV	17.0	…	150 (−)	…	8.1 (−)
5	Male	1.6	SLD	Biventricle	5.5	…	136 (−)	…	2.9 (−)
6	Male	1.3	SLL	LV	16.7	…	180 (−)	+ (12.6)	8.3 (+)[e], 12.6 (+)[e]
7	Male	3.8	SDS	LV	16.8	…	164 (−)	…	5.2 (+)[f]
8	Female	1.4	SDD	Biventricle	16.8	…	150 (−)	…	10.7 (+)[f]
9	Male	1.4	SDS	LV	16.1	…	206 (−)	…	…
10	Female	2.1	SDD	RV	16.6	…	194 (−)	…	…
11	Female	2.2	ILL	RV	16.4	+ (12.9)	190 (−)	…	7.4 (−)
12	Male	1.7	SDL	LV	16.3	+ (12.3)	256 (+)	…	7.3 (−), 12.3 (−)
13	Male	2.0	SDS	LV	14.4	…	174 (−)	…	…
14	Male	2.1	SDD	RV	16.2	…	140 (−)	…	…
15	Male	1.6	SDL	Biventricle	16.2	…	180 (−)	…	3.7 (−)
16	Female	2.7	SDS	LV	16.1	…	128 (−)	…	4.4 (−)
17	Male	2.2	SLL	Biventricle	15.2	…	152 (−)	…	1.1 (−), 6.6 (−)
18	Female	17.6	ADD	RV	14.4	…	204 (−)	…	3.0 (−)
19	Female	9.1	IDD	RV	15.3	…	146 (−)	…	11.8 (+)[g]
20	Female	1.3	SDD	Biventricle	15.1	…	234 (+)	…	8.5 (−)
21	Male	1.7	SDD	LV	15.1	…	118 (−)	…	1.2 (−)
22	Female	2.6	SDS	LV	15.1	…	208 (−)	…	3.2 (−)
23	Male	2.2	SDL	Biventricle	15.2	…	164 (−)	…	…
24	Male	1.1	SLD	LV	15.1	…	192 (−)	…	…
25	Female	1.8	SDL	LV	15.3	+ (8.2)	136 (−)	…	8.0 (−)
26	Female	2.4	AXD	RV	14.3	…	126 (−)	…	…
27	Female	3.4	ADD	RV	5.9	+ (2.3)	184 (−)	…	2.2 (−)

[a] Pacemaker implantation for sinus node dysfunction (SND) after Fontan operation. [b] PR interval on most recent electrocardiogram, first-degree atrioventricular block. [c] Development of spontaneous intraatrial reentrant tachycardia (IART) after Fontan operation. [d] Any inducible IART. [e] Inducible sustained IART. [f] Inducible nonsustained IART without requiring beta-blocker medication. [g] Inducible nonsustained IART.

ADD (A = ambiguous); AV = atrioventricular; AXD (X = unknown); EPS = electrophysiologic study; IDD (I = inversus); ILL (I = inversus, L = L-loop); LV = left ventricle; Pt. No. = patient number; RV = right ventricle; SDD (D = D-transposition); SDL (L = L-transposition); SDS (S = solitus, D = D-loop); SLD (L = L-loop); SLL (S = solitus, L = L-loop).

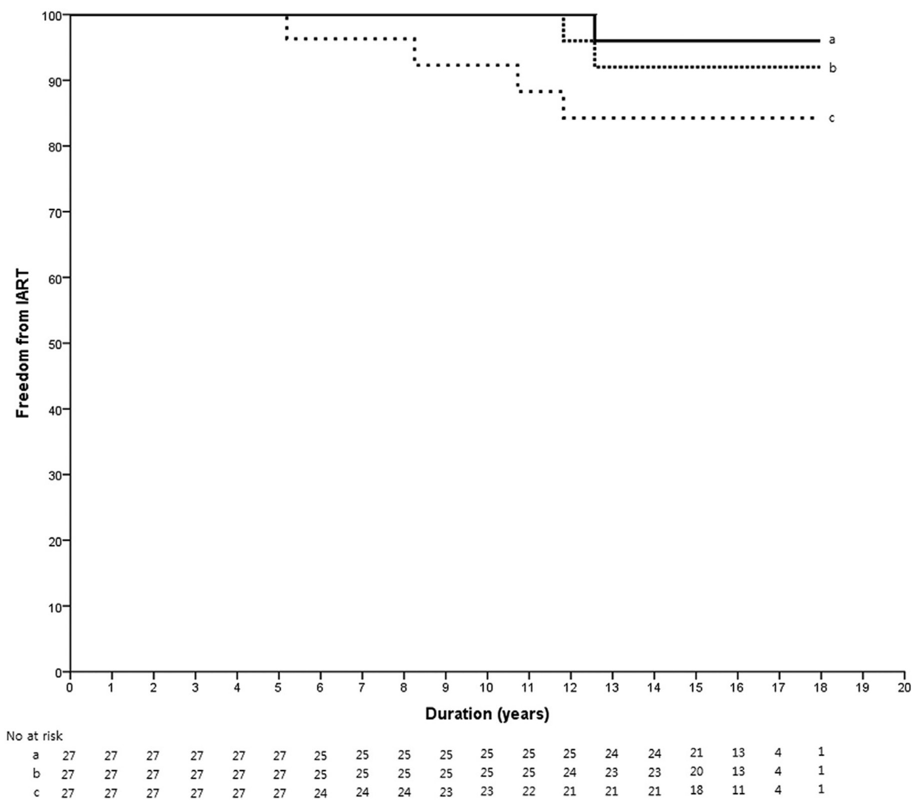

Fig 2. Kaplan-Meier curves for freedom from development of spontaneous or inducible intraatrial reentrant tachycardia (IART) after Fontan operation: (a) spontaneous IART requiring beta-blocker medication; (b) spontaneous or inducible IART on electrophysiologic studies requiring beta-blocker medication; (c) spontaneous, any inducible sustained or nonsustained IART on electrophysiologic studies including no beta-blocker medication. (No = number.)

with PR intervals of 256 ms and 234 ms (Table 1). In Holter data, the average 24-hour heart rate was 78.0 ± 9.9 beats per minute (range, 62 to 96). Peak heart rate was 143.3 ± 25.0 beats per minute, and minimum heart rate was 52.8 ± 12.4 beats per minute. Nonsustained atrial tachycardia occurred in 4 patients. There was no new case of second- or third-degree atrioventricular block.

At late follow-up, there were four new cases of sinus node dysfunction that required pacemaker implantation at 2.3, 8.2, 12.3, and 12.9 years after Fontan, respectively (Table 1). Freedom from sinus node dysfunction was 84.7% ± 7.0% at 18 years after the Fontan operation (Fig 3). On multivariate analysis, none of the factors was found to be statistically significantly associated with the

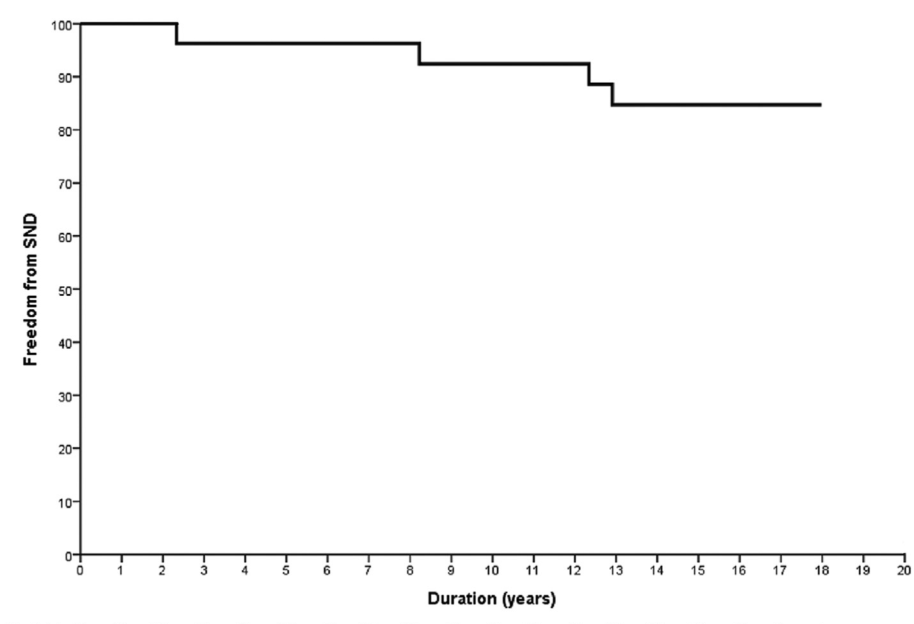

Fig 3. Kaplan-Meier curves for freedom from sinus node dysfunction (SND) after Fontan operation. (No = number.)

development of sinus node dysfunction, but sinus node dysfunction tended to be more associated with decreased maximal oxygen consumption, although statistical significance was not reached (HR 0.731, $p = 0.095$). Intraatrial reentrant tachycardia was also not a statistically significant risk factor for sinus node dysfunction in univariate analysis (HR 0.038, $p = 0.578$).

The most recent echocardiographic data showed that fractional shortening was 34.7% ± 9.1% (range, 23.2 to 56.1), and ejection fraction was 59.5% ± 9.8% (range, 42.0 to 86.8). The degree of atrioventricular valve regurgitation was none in 3 patients, trivial in 8, mild in 12, mild to moderate in 2, and moderate to severe in 2. The latest cardiac catheterization was performed in 23 of the total 27 patients (85.2%) at 8.0 ± 4.7 years (range, 1.2 to 16.3) after the Fontan operation. Systolic pulmonary arterial pressure was 12.8 ± 3.3 mm Hg (range, 7 to 23), ventricular end-diastolic pressure was 9.6 ± 3.8 mm Hg (range, 2 to 17), transpulmonary pressure gradient was 7.6 ± 2.7 mm Hg (range, 5 to 11), pulmonary vascular resistance was 2.60 ± 1.70 Wood units · m^2 (range, 1.40 to 3.80), systemic arterial oxygen saturation was 92.4% ± 2.9% (range, 86.7 to 97.0), and systemic blood flow was 2.30 ± 0.57 L · min^{-1} · m^{-2} (range, 1.9 to 2.7). The latest cardiopulmonary exercise test was performed in 24 of the total 27 patients (88.9%) at 12.6 ± 2.8 yeas (range, 5.7 to 16.3) after the Fontan operation, and maximal oxygen consumption (normal greater than 30 mL · kg^{-1} · min^{-1}) was 31.52 ± 7.29 mL · kg^{-1} · min^{-1} (range, 18.4 to 47.0 [n = 23]). Resting heart rate was 85.6 ± 13.0 beats per minute (range, 59 to 112), peak heart rate was 155.3 ± 22.8 beats per minute (range, 101 to 192), maximum predicted heart rate was 193.1 ± 6.5 beats per minute (range, 184 to 210), percent predicted heart rate (peak heart rate/maximum predicted heart rate) was 80.52% ± 12.17% (range, 53.72 to 97.96), and chronotropic index was 0.65 ± 0.20 (range, 0.22 to 0.96 [n = 24]). There was no evidence of early or late onset complications related to the interventional atrial incision/cryoablation such as injury to the right coronary artery or atrioventricular node. None of our patients underwent late radiofrequency catheter ablation or Fontan revision surgery owing to intraatrial reentrant tachycardia.

Comment

Prophylactic Arrhythmia Surgery

Refractory atrial arrhythmia with hemodynamic abnormalities is a frequent, potentially fatal complication of the Fontan operation. In adult patients with a Fontan-type operation, the presence of atrial tachyarrhythmias is associated with higher morbidity and mortality at midterm follow-up [2]. Radiofrequency catheter ablation is a reasonable option for treatment of intraatrial reentrant tachycardias related to repair of congenital heart disease. Complex atrial surgery limits the success of radiofrequency catheter ablation, and older age is associated with a higher risk of intraatrial reentrant tachycardia recurrence [15]. Acute success rates from catheter ablation in the Fontan patient range from 40% to 75%, with recurrence of tachycardia in 60% of patients during the first year [16]. Therefore, aggressive preventive arrhythmia surgery was performed, particularly at the time of the Fontan operation.

Intraatrial reentrant tachycardia is a well-recognized late sequela of the Fontan operation for complex univentricular congenital heart disease [17]. We have developed and performed a novel prophylactic arrhythmia surgery at the time of the lateral tunnel Fontan operation since August 1997 (Fig 1, Video). Cryoablation was performed between right atriotomy and right atrioventricular valve annulus based on the simulated canine Fontan study, and right atriotomy extending to the coronary sinus was added to block slow rate conduction isthmus. In an acute canine model of the modified Fontan operation, conduction block imposed by the lateral tunnel suture line is an essential component of the atrial flutter circuit. The inducibility of atrial flutter is increased by suture line placement along the crista terminalis. Slow conduction, resulting from injury to the crista terminalis, promotes this increased inducibility. Avoidance of the crista terminalis may reduce the incidence of atrial flutter in children undergoing the modified Fontan operation [18]. Hence, we placed the lateral aspect of the Fontan baffle anterior to the crista terminalis to avoid injury to the crista terminalis. Arrhythmia outcome was further improved by the location of the lateral tunnel suture line in relation to the crista terminalis [18]. After the surgical technique of the lateral tunnel Fontan operation was modified, the incidence of atrial tachyarrhythmia was reported to be low, and it mainly depended on the underlying cardiac morphology and preoperative arrhythmia [14]. In contrast to the canine model of simulated Fontan operation, our surgical technique has been modified several times over the years so that it is less arrhythmogenic, and the sandwich technique was also used with closure of the right atriotomy incorporating the Gore-Tex patch to reduce the atrial suture line, which resulted in much improved arrhythmia outcome.

Intraatrial Reentrant Tachycardia

In this series, none of the patients had spontaneous intraatrial reentrant tachycardia until 12.57 years after prophylactic arrhythmia surgery. However, as the follow-up period exceeded more than 12 years after the Fontan operation, atrioventricular valve regurgitation might have induced spontaneous intraatrial reentrant tachycardia in 1 patient who did not have a long PR interval because isthmus conduction block had not been completely achieved over time. We also demonstrated that hemodynamic factors contribute to intraatrial reentrant tachycardia occurrence because the pre-Fontan systolic pulmonary artery pressure and post-Fontan atrioventricular valve regurgitation are statistically significantly associated with intraatrial reentrant tachycardia occurrence in multivariate analysis. Prophylactic arrhythmia surgery may not have been performed completely. If cryoablation was not transmural and conduction across the cryolesion was not completely interrupted, some atrial tissues were still intact and long-lasting conduction block was not completely achieved across the area over time. If conduction times were not long, PR interval would not be sufficiently long. In this series, 4 patients had intraatrial reentrant tachycardia over time. None of them had a long PR interval (more than 180 ms), suggesting that conduction block has not been completely created at the isthmus over time. To prevent intraatrial reentrant tachycardia occurrence, high-risk patient groups should be identified, such as those with segmental anatomy of atrioventricular discordance, unfavorable hemodynamic factors (including the pre-Fontan systolic pulmonary arterial pressure and post-Fontan atrioventricular valve regurgitation), and insufficient conduction block around the isthmus over time. Particularly, in these high-risk patients, careful follow-up is mandatory, and early surgical management is also required for sinus node dysfunction and associated chronic bradycardia, as well as for atrioventricular valve regurgitation.

Long-Term Results

The long-term results of our prophylactic arrhythmia surgery showed a very low incidence of spontaneous intraatrial reentrant tachycardia and a low incidence of inducible sustained or nonsustained intraatrial reentrant tachycardia induced by atrial stimulation and use of isoproterenol. In contrast to the canine simulated Fontan study, our Fontan suture lines might not completely form transmural block for atrial conduction, which acts as a substrate for intraatrial reentrant tachycardia. However, the electrophysiologic effects can also differ in the diseased hearts with a pathologic substrate for atrial flutter from that in normal canine models [19]. In our series, there was no death that was arrhythmic in etiology, and no surgical complications related to prophylactic arrhythmia surgery such as injury to the right coronary artery and atrioventricular node. Also, none of our patients underwent ablation or revision to the extracardiac conduit. By presenting these long-term results, we have shown that our novel prophylactic arrhythmia surgery can be performed in all lateral tunnel Fontan patients.

Sinus Node Dysfunction

Sinus node dysfunction is reported to be a troublesome complication after various types of Fontan operations [20]. Sinus node dysfunction has also been identified as a correlate and presumptive predisposing factor for atrial flutter [3, 5, 17]. Late postoperative atrial flutter is associated with chronotropic incompetence in pediatric congenital heart disease patients. Permanent pacing strategies based on maximum achievable heart rate and chronotropic index data, and aimed at the restoration of chronotropic competence, could potentially offer improved prevention of late postoperative atrial flutter in this young and active patient population [17]. In this series, sinus node dysfunction was not a statistically significant risk factor for intraatrial reentrant tachycardia in univariate analysis (HR 0.040, $p = 0.613$), because we implanted a permanent pacemaker as early as possible for sinus node dysfunction after the Fontan operation to reduce the occurrence of intraatrial reentrant tachycardia.

Historic Significance

Collins and associates [21] reported an interventional atrial incision, similar to that used in canine models, placed at the time of the initial lateral tunnel Fontan operation performed between October 1999 and May 2001. We performed prophylactic arrhythmia surgery at the time of the lateral tunnel Fontan operation for the first time in August 1997. Prophylactic arrhythmia surgery with our novel modification of the lateral tunnel Fontan procedure to reduce the development of intraatrial reentrant tachycardia was feasible and safe. Long-term follow-up results also demonstrated that this novel modification is effective for the prophylaxis of intraatrial reentrant tachycardia. This is the first report that demonstrates the long-term, 18-year, efficacy and safety of prophylactic arrhythmia surgery at the time of the lateral tunnel Fontan operation.

References

1. Diller GP, Giardini A, Dimopoulos K, et al. Predictors of morbidity and mortality in contemporary Fontan patients: results from a multicenter study including cardiopulmonary exercise testing in 321 patients. Eur Heart J 2010;31:3073–83.
2. Giannakoulas G, Dimopoulos K, Yuksel S, et al. Atrial tachyarrhythmias late after Fontan operation are related to increase in mortality and hospitalization. Int J Cardiol 2012;157:221–6.
3. Fishberger SB, Wernovsky G, Gentles TL, et al. Factors that influence the development of atrial flutter after the Fontan operation. J Thorac Cardiovasc Surg 1997;113:80–6.
4. Weipert J, Noebauer C, Schreiber C, et al. Occurrence and management of atrial arrhythmia after long-term Fontan circulation. J Thorac Cardiovasc Surg 2004;127:457–64.
5. Walsh EP, Cecchin F. Arrhythmias in adult patients with congenital heart disease. Circulation 2007;115:534–45.
6. Kim SJ, Kim WH, Lim HG, Lee JY. Outcome of 200 patients after an extracardiac Fontan procedure. J Thorac Cardiovasc Surg 2008;136:108–16.
7. Delacretaz E, Ganz LI, Soejima K, et al. Multi atrial macro-re-entry circuits in adults with repaired congenital heart disease: entrainment mapping combined with three-dimensional electroanatomic mapping. J Am Coll Cardiol 2001;37:1665–76.
8. Triedman JK, Alexander ME, Love BA, et al. Influence of patient factors and ablative technologies on outcomes of radiofrequency ablation of intra-atrial re-entrant tachycardia in patients with congenital heart disease. J Am Coll Cardiol 2002;39:1827–35.
9. Kim WH, Lim HG, Lee JR, et al. Fontan conversion with arrhythmia surgery. Eur J Cardiothorac Surg 2005;27:250–7.
10. Jang WS, Kim WH, Choi K, et al. The mid-term surgical results of Fontan conversion with antiarrhythmia surgery. Eur J Cardiothorac Surg 2014;45:922–7.
11. Rodefeld MD, Gandhi SK, Huddleston CB, et al. Anatomically based ablation of atrial flutter in an acute canine model of the modified Fontan operation. J Thorac Cardiovasc Surg 1996;112:898–907.
12. Bromberg BI, Schuessler RB, Gandhi SK, Rodefeld MD, Boineau JP, Huddleston CB. A canine model of atrial flutter following the intraatrial lateral tunnel Fontan operation. J Electrocardiol 1998;30:S85–93.
13. Song MK, Bae EJ, Kwon BS, et al. Intra-atrial reentrant tachycardia in adult patients after Fontan operation. Int J Cardiol 2015;187:157–63.
14. Stamm C, Friehs I, Mayer JE, et al. Long-term results of the lateral tunnel Fontan operation. J Thorac Cardiovasc Surg 2001;121:28–41.
15. Yap SC, Harris L, Silversides CK, Downar E, Chauhan VS. Outcome of intra-atrial re-entrant tachycardia catheter

ablation in adults with congenital heart disease: negative impact of age and complex atrial surgery. J Am Coll Cardiol 2010;56:1589–96.
16. Deal BJ, Jacobs ML. Management of the failing Fontan circulation. Heart 2012;98:1098–104.
17. Anand N, McCrindle BW, Chiu CC, et al. Chronotropic incompetence in young patients with late postoperative atrial flutter: a case–control study. Eur Heart J 2006;27:2069–73.
18. Gandhi SK, Bromberg BI, Rodefeld MD, et al. Lateral tunnel suture line variation reduces atrial flutter after the modified Fontan operation. Ann Thorac Surg 1996;61:1299–309.
19. Shinagawa K, Li D, Leung TK, Nattel S. Consequences of atrial tachycardia-induced remodeling depend on the preexisting atrial substrate. Circulation 2002;105:251–7.
20. Bae EJ, Lee JY, Noh CI, Kim WH, Kim YJ. Sinus node dysfunction after Fontan modifications—influence of surgical method. Int J Cardiol 2003;88:285–91.
21. Collins KK, Rhee EK, Delucca JM, et al. Modification to the Fontan procedure for the prophylaxis of intra-atrial reentrant tachycardia: short-term results of a prospective randomized blinded trial. J Thorac Cardiovasc Surg 2004;127:721–9.

INVITED COMMENTARY

I have the greatest admiration for the work presented by Lim and colleagues in this issue of *The Annals* [1]. They not only describe an innovative prophylactic measure to prevent arrhythmia but also include 15 years of follow-up analyzing the benefits of this modification, a rare and welcome fate in our specialty.

Why is this work important? Many would consider that the benefits of the extracardiac conduit have been proven, and that most, but not all, teams have now adopted it and dropped the use of the lateral tunnel. In 2013, a survey from the STS database and EACTS database showed that a quarter of the Fontan procedures performed recently were still lateral tunnels [2]. Despite improved reported outcomes after the Fontan procedure, we all know that this population remains at a high risk of failure of their circulation, and that arrhythmia burden will play a major role in their demise. We, in Australia and New Zealand, have adopted the extracardiac conduit as our sole form of Fontan in the late 1990s. We were comforted in our choice because we subsequently demonstrated the superiority of this form of Fontan in terms of protection against arrhythmia [3], although the differences in outcomes noted with the lateral tunnel were small. Preventing arrhythmia after Fontan surgery seems primordial as it is now accepted that onset of arrhythmia will precipitate failure of the Fontan circulation [4]. For this perspective, the modification described is welcome. I have no doubt that this modification was effective because only 4% of the patient group developed clinically relevant tachyarrhythmia, whereas over the same time period more than 20% of our patients with a lateral tunnel had developed arrhythmia. Their 85% freedom from bradyarrhythmia at 18 years was similar to the results we have reported.

The extracardiac conduit is an attractive technique because of the simplicity of its concept, but it can lead to distortions of the pulmonary arteries and has been linked to higher early morbidity [5]. Twenty-year outcomes show us that the extracardiac technique is superior to the lateral tunnel, but only because it protects patients against the late occurrence of arrhythmia. Could it be that this new modification erases this observed difference? We are unlikely to find the answer to this question. The authors themselves have already switched to the extracardiac conduit and, like most, are unlikely to move back to the lateral tunnel. But we should nonetheless feel inspired by these authors who demonstrated that bold, innovative approaches can bring solutions to clinical issues, provided that clinicians have patience—even if these issues take two decades to develop. Tachyarrhythmia remains an issue even after an extracardiac conduit, but this technique will limit interventional cardiologist access to the atria. This work should push us to investigate new modifications for these patients.

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Yves d'Udekem, MD, PhD

Department of Cardiac Surgery
The Royal Children's Hospital Melbourne
Murdoch Childrens Research Institute
Melbourne, Australia
and
Department of Paediatrics
Faculty of Medicine
The University of Melbourne
50 Flemington Rd
Parkville VIC 3052, Australia
email: yves.dudekem@rch.org.au

References

1. Lim HG, Lee JR, Kim YJ. The effects of modification to lateral tunnel Fontan procedure for prophylactic arrhythmia surgery. Ann Thorac Surg 2017;104:197–204.
2. Jacobs JP, Maruszewski B. Functionally univentricular heart and the Fontan operation: lessons learned about patterns of practice and outcomes from the congenital heart surgery databases of the European association for cardio-thoracic surgery and the society of thoracic surgeons. World J Pediatr Congenit Heart Surg 2013;4:349–55.
3. d'Udekem Y, Iyengar AJ, Galati JC, et al. Redefining expectations of long-term survival after the Fontan procedure: twenty-five years of follow-up from the entire population of Australia and New Zealand. Circulation 2014;130(11 suppl 1):S32–8.
4. Carins TA, Shi WY, Iyengar AJ, et al. Long-term outcomes after first-onset arrhythmia in Fontan physiology. J Thorac Cardiovasc Surg 2016;152:1355–63.
5. Stewart RD, Pasquali SK, Jacobs JP, et al. Contemporary Fontan operation: association between early outcome and type of cavopulmonary connection. Ann Thorac Surg 2012;93:1254–61.

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