Follow-Up of a Prospective Surgical Strategy to Prevent Intra-Atrial Reentrant Tachycardia After the Fontan Operation

Ian H. Law, MD; Osman Alam, BS; Edward L. Bove, MD; Richard G. Ohye, MD; David J. Bradley, MD; Sunkyung Yu, MS; Macdonald Dick II, MD

Background—Intra-atrial reentrant tachycardia (IART) after the Fontan operation had an early reported incidence of 10% to 35% during early and intermediate follow-up and posed substantial management challenges.

Methods and Results—To reduce the incidence of IART after the Fontan procedure, we performed a randomized, double-blind study to evaluate the impact of an incision in the right atrium joining the lateral tunnel suture line and the tricuspid valve annulus. Between March 1998 and September 2003, 134 subjects (median age: 1.8 years; range: 1.3–5.2 years; 91 men) were randomly assigned to receive the incision. All 134 patients had a form of single ventricle pathological anatomy. The clinical course, electrocardiograms, and Holter monitoring were available for review in 114 subjects at a median of 8.2-year follow-up (range: 0.9–11.9 years). There were 2 late deaths, neither subject had IART. The combined incidence of sustained IART was 3.5% (4/114). There was no difference in the occurrence of sustained IART between those subjects receiving the incision and those who did not (2 in each group) during follow-up. No patients of either group experienced short-term complications.

Conclusions—Despite the fact that the primary outcome of this trial was not reached, the most significant finding was that with current management, the incidence of IART is considerably lower than the early retrospective, observational studies suggested. **(*Circ Arrhythm Electrophysiol*. 2016;9:e004478. DOI: 10.1161/CIRCEP.116.004478.)**

Key Words: cardiovascular surgery ■ congenital heart disease ■ Fontan procedure ■ suture ■ tachycardia

The Fontan operation is one of the most common cardiac surgical procedures performed in children and is the final operation in the 3-stage palliative repair for ≈20% of children with congenital heart defects, specifically those with univentricular malformations. The original Fontan operation[1] was designed to bypass the rudimentary right ventricle in patients with tricuspid atresia by creating a right atrial appendage to left pulmonary artery connection in patients with tricuspid atresia. Several different modifications have altered the original design.[2–12]

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Despite these modifications, the most common postoperative complicating arrhythmia has been intra-atrial tachycardia, a reentry arrhythmia similar to atrial flutter.[13–16] The 1980s and early 1990s reports of the incidence of intra-atrial reentrant tachycardia (IART) after the Fontan operation approached 40%. This experience was largely confined to the patients who had direct right atrial to pulmonary anastomosis with or without conduits or right atrial to the rudimentary right ventricle anastomosis with or without a conduit.[13,16–24] In the early 1990s reports, the incidence of IART in patients with the lateral tunnel technique decreased to 10% to 20% of patients over a relatively short follow-up interval.[16,18,21,22,24,25] Nonetheless, it was deemed important to minimize as much as possible this postoperative arrhythmia as it can significantly compromise the physiology of the Fontan repair.[25–30]

The purpose of this study was to investigate whether a strategically placed surgical linear incision combined with cryosurgical lesions (Figure 1) applied at the completion of the fenestrated lateral tunnel Fontan was safe and would prevent the postoperative atrial reentrant arrhythmias. A randomized prospective study, similar to other studies,[26,31] was designed in which patients would receive either the fenestrated Fontan repair or a fenestrated Fontan repair with a surgical incision and cryosurgery lesions placed at the site that, similar to the canine models,[27] so as to create a line of block between the anterior suture line of the lateral tunnel and the atrioventricular ring to disrupt potential reentrant intra-atrial circuits. Previously, in a subset of this cohort of patients, we demonstrated that inducible IART by electrophysiological study is present in ≈11% of patients who have completed stage 1 and 2 of the Fontan sequence[32] before the Fontan operation (stage 3).

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From the Division of Pediatric Cardiology, Department of Pediatrics, Carver College of Medicine, University of Iowa, Iowa City (I.H.L.); and Division of Pediatric Cardiology, Department of Pediatrics and Communicable Diseases (O.A., D.J.B., S.Y., M.D.) and Department of Cardiac Surgery (E.L.B., R.G.O.), University of Michigan Medical School, Ann Arbor.

Correspondence to Macdonald Dick II, MD, UM Congenital Heart Center, C.S. Mott Children's Hospital, 1540 E Hospital Dr, Ann Arbor, MI 48109. E-mail mdick@umich.edu

WHAT IS KNOWN

- To reduce the known 10% to 35% incidence of intra-atrial reentrant tachycardia (IART) after the Fontan procedure, we performed a randomized, double-blind study to evaluate the impact of an incision in the right atrium joining the lateral tunnel suture line and the tricuspid valve annulus.
- Of the 134 patients with a form of single ventricle pathological anatomy enrolled in the study, the clinical course, electrocardiograms, and Holters were available for review in 114 subjects at a median 8.2 years (range: 0.9–11.9 years) follow-up.

WHAT THE STUDY ADDS

- No difference in the occurrence of sustained IART was found between those subjects receiving the incision and those who did not (2 in each group) during follow-up. No patients of either group experienced short-term complications from the intervention.
- In contrast to early retrospective, observational studies, this trial found that, with current management, the incidence of IART is low in the first 1 to 2 decades after the Fontan operation.

In contrast to our earlier accounts about pre-Fontan IART inducibility, this report summarizes a retrospective clinical follow-up of the incidence of IART along with a comparison of the intraoperative data and the early pre- and postoperative electrophysiological characteristics' between the 2 groups of Fontan patients.

Methods

This study was initially designed as a prospective, randomized controlled clinical trial. Between March 1, 1998, and September 30, 2003, all patients with univentricular anatomy who were scheduled for the fenestrated lateral tunnel Fontan operation were potential participants in this investigation. Patients who were found to be high-risk candidates (diagnosis of hypoplastic left ventricle with intact atrial septum at birth) for a fenestrated Fontan or who had a palliative procedure other than the Fontan operation were excluded. The patients were randomized to the fenestrated lateral tunnel Fontan (the standard-of-care procedure, no incision group) or the fenestrated lateral tunnel Fontan with the surgical incision and cryo lesions (incision group). Patients who had an extended intraoperative course and were deemed to be at increased risk by the surgeon in the operating room were excluded from the study. Patients with preexisting clinical arrhythmias were also excluded from the study.

Randomization of the patients was performed after at least one parent (or guardian) of each patient consented to and signed an informed consent document covering all components of the study. The patient, investigators, and the follow-up cardiologists (but not the surgeons) were blind to the patient's group status. The study was approved by the Institutional Review Board of the University of Michigan Medical School.

During the previous decade, the Michigan Congenital Heart Center surgeons performed 40 to 50 fenestrated Fontan operations per year. On the basis of the experience at our center, and on the published incidence of IART in the post-Fontan population, our total enrollment of 134 patients (67 in each group) would detect a reduction from the contemporary (1990s) incidence of IART, reported to be ≈30%,[14] to an IART incidence of 10% to 11% with a power of 80% and a significance level of 5%.

Surgical Procedure

The surgeon was informed of the patient's randomization assignment immediately before surgery in the operating room (in a sealed envelope). All subjects received a hemi-Fontan procedure as the stage 2 operation except one, who underwent a bidirectional Glenn.

A standard fenestrated Fontan procedure was performed using a lateral tunnel within the right atrium inserted between the internal orifice of the inferior vena cava and the previously placed patch that was inserted at the time of the hemi-Fontan operation.[11,28] This patch was excised to allow an unobstructed pathway to the pulmonary arteries to complete the total cavopulmonary anastomosis. In all patients, a 3- to 4-mm fenestration was placed in the lateral tunnel patch. In the incision group, a linear incision was created between the tricuspid annulus and the anterior suture line of the intercaval lateral tunnel (Figure 1), as suggested by canine studies.[29] Cryosurgical lesions were applied at the tricuspid end of the incision to ensure complete interruption of excitable tissue along the incision line.

Follow-Up Procedures

For this long-term retrospective component of the study, patient clinical course, ECG, and dynamic 24-hour ECG recording (Holter) data were performed and collected at 1 year and whenever the attending cardiologist deemed necessary thereafter. During the decade after the surgery, each patient and their cardiologist were contacted via telephone for follow-up information. In addition, public web-based databases (Archives.com and Spokeo.com) were queried for patient status and location in an effort to complete the follow-up data. The primary end point was the incidence of sustained documented IART (>30 seconds).

Statistical Analysis

Data are presented as frequency with percentage for categorical variables and median (interquartile range or range) or mean±SD, as appropriate, for continuous variables. Patient and clinical characteristics

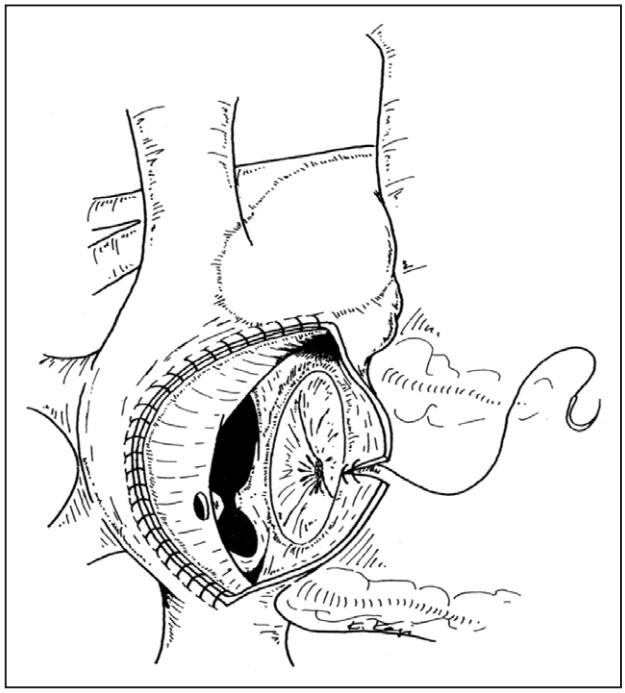

Figure 1. Artist's rendering of the incision from the triuspid annulus to the atriotomy–lateral tunnel suture line used to eliminate potential intra-atrial rentrant circuit.

Table 1. Patient Data (N=134)

	All (N=134)	Randomization	
		Incision (n=67)	No Incision (n=67)
Male sex	91 (67.9)	45 (67.2)	46 (68.7)
Cardiac diagnosis			
HLHS or HLV	92 (68.7)	48 (71.6)	44 (65.7)
AVSD/HLV or DORV	14 (10.4)	8 (11.9)	6 (9.0)
DILV	10 (7.5)	3 (4.5)	7 (10.4)
PA/IVS	9 (6.7)	4 (6.0)	5 (7.5)
TA/TGA	8 (6.0)	4 (6.0)	4 (6.0)
Ebstein	1 (0.7)	0 (0.0)	1 (1.5)
Age at stage I palliation, d	6 (5–9)	7 (5–9)	6 (5–10)
Type of stage I palliation			
Norwood/DKS	102 (76.1)	52 (77.6)	50 (74.6)
BT or central shunt	22 (16.4)	11 (16.4)	11 (16.4)
PA band	4 (3.0)	2 (3.0)	2 (3.0)
None	6 (4.5)	2 (3.0)	4 (6.0)
Age at stage II palliation, d	164 (140–196)	164 (143–192)	161 (136–198)
Type of stage II palliation			
Hemi-Fontan	133 (99.3)	67 (100.0)	66 (98.5)
Bidirectional Glenn	1 (0.7)	0 (0.0)	1 (1.5)
Age at Fontan, y	1.8 (1.6–2.1)	1.7 (1.6–2.1)	1.8 (1.6–2.2)
Weight at Fontan, kg	11.2±1.6	11.0±1.7	11.3±1.5

Data are presented as N (%) for categorical variables and median (interquartile range) or mean±SD for continuous variables. AVSD indicates atrioventricular septal defect; BT, Blalock–Taussig; DILV, double inlet left ventricle; DKS, Damus–Kaye–Stansel; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; HLV, hypoplastic left ventricle; PA, pulmonary artery; PA/IVS, pulmonary atresia with intact ventricular septum; and TA/TGA, tricuspid atresia/transposition of the great arteries.

were compared between the 2 groups (incision versus no incision groups), using χ^2 test or Fisher exact test for categorical variables and Wilcoxon rank-sum test or 2-sample *t* test for continuous variables. Post-Fontan electrophysiology data were compared with the pre-Fontan data using McNemar test for the incidence of IART and Wilcoxon signed-rank test for other continuous variables. Repeated-measures ANOVA accounting for intrasubject correlation was used to compare change in heart rates (measured by Holter) over time between the 2 groups. All analyses were performed using SAS version 9.3 (SAS Institute Inc, Cary, NC). A *P* value of <0.05 was considered statistically significant.

Results

One hundred and thirty-four subjects were initially enrolled into the study and were randomized at surgery to either the incision or not (1:1). There was one early postsurgery death (no incision), leaving 133 participants available for follow-up. Of the 133, long-term data were available in 114 patients (median follow-up: 8.2 years; range: 0.9–11.9 years). There are 19 patients who lost to follow-up (6 incision and 13 nonincision).

There was no significant difference (Tables 1 through 5) in any of the patient characteristics, pre-Fontan catheterization and electrophysiological data, Fontan clinical and intraoperative characteristics, intensive care length of stay, hospital length of stay, and follow-up in years between the 2 groups. However, intraoperative cross-clamp time in the incision group was slightly longer (6 minutes) than in the nonincision group. Interestingly, 30 patients (22%) had inducible IART during the pre-Fontan electrophysiological study, all by burst pacing, with no significant group difference (*P*=0.08). Note that 6 patients did not have stage 1 palliation. There was no significant difference in inducible IART after Fontan surgery between the subjects in the incision group (12 inducible, 18%) and those in the nonincision group (10 inducible, 15%; Table 3). In addition, there was no significant difference (Table 5) in any of the electrophysiological characteristics by postoperative programed extrastimulation through transthoracic epicardial wires between the 2 groups or in comparison of pre-Fontan and post-Fontan (<1 week postsurgery; Table 6) electrophysiological data except total sinoatrial conduction time and increased paced atrial effective refractory period in post-Fontan. Both of these measures were within the normal range.[30] Finally, there was no significant difference in non-IART rhythms observed on ECG in the follow-up period between the 2 groups.

Four patients (3.5%; Table 3) had IART during the postoperative period, 2 in each group. One additional patient (not included in this group of 4) had supraventricular tachycardia mediated through a concealed accessory pathway which was successfully ablated. All 4 subjects had their tachycardia between 18 days to 22 months after surgery. One patient had 2 episodes. Follow-up for the 4 patients was 10 years in 2, 5 years in the third, and 6 years 10 months in the fourth. None had any further reported or documented IART during that follow-up period.

In a comparison of inducible IART between the pre-Fontan study and the post-Fontan study (<1 week postsurgery; Table 4), 7 patients had IART induced at both studies; 39 had no IART induced in either study. Eighteen patients who had IART induced at the pre-Fontan study had no IART at the post-Fontan study. Eight patients had IART induced in the post-Fontan study but not in the pre-Fontan study.

Figure 2 plots the average, maximal, and minimal heart rate by the study groups across time. There was no significant difference in the heart rates observed in these Holters between the groups during the follow-up intervals. The acquisition of 24-hour Holters tracings during the 5- to 10-year follow-up period varied by individual provider practices.

Four patients had pacemakers implanted. An 18-month-old girl with double inlet left ventricle developed intermittent complete heart block 15 months after the hemi-Fontan procedure. She received a dual-chamber pacemaker at the time of her Fontan operation. An 18-month-old boy demonstrated a junctional rhythm between a heart rate of 56 to 80 beats per minute 3 days after the Fontan operation. Fourteen days after surgery, his heart rate was 53 with a low atrial origin. Six months later, his ventricular rate was 51 beats per minute with no discernible P waves. A dual-chamber pacemaker was placed for sick sinus syndrome. The third patient, a 2-year-old girl with hypoplastic

Table 2. Fontan and Post-Fontan Data (N=134)

Characteristics	All (N=134)	Randomization		*P* Value*
		Incision (n=67)	No Incision (n=67)	
Pump time, min (n=132)	66 (58–75)	67.5 (60–79)	63.5 (56–74)	0.10
Cross-clamp time, min (n=132)	29 (26–34)	32 (29–36)	26 (25–29)	<0.0001
Circulatory arrest	47 (35.1)	20 (29.9)	27 (40.3)	0.21
If yes, circulatory arrest time, min	20 (16–25)	21.5 (19.5–25.5)	18 (14–22)	0.09
Concurrent surgery	24 (17.9)	11 (16.4)	13 (19.4)	0.65
ICU length of stay, d (n=125)	2 (1–3)	2 (1–3)	1 (1–2)	0.11
Hospital length of stay post-Fontan repair, d	9 (7–13)	9 (7–14)	8 (6–12)	0.17
Duration of follow-up post-Fontan repair, y	8.2 (6.8–9.7)	7.7 (6.8–9.1)	8.4 (6.9–10.0)	0.23
Post-op rhythm				N/A
Pacemaker	4 (3.0)	1 (1.5)	3 (4.5)	
Ectopic (JXN, LAR, RAR)	11 (8.2)	8 (11.9)	3 (4.5)	
Sinus tachy/bradycardia	113 (84.3)	56 (83.6)	57 (85.1)	
Not performed	3 (2.2)	1 (1.5)	2 (3.0)	
No data	3 (2.2)	1 (1.5)	2 (3.0)	
Holter arrhythmia	16 (11.9)	9 (13.4)	7 (10.4)	0.77
Atrial ectopic	7 (5.2)	5 (7.5)	2 (3.0)	0.45
Sustained >30 s	5 (3.7)	3 (4.5)	2 (3.0)	1.00
Nonsustained	2 (1.5)	2 (3.0)	0 (0.0)	0.50
Fenestration closed	39 (29.1)	22 (32.8)	17 (25.4)	0.96
Dead	3 (2.2)	2 (3.0)	1 (1.5)	1.00

Data are presented as n (%) for categorical variables and median (interquartile range) for continuous variables. IART indicates intra-atrial reentrant tachycardia; ICU, intensive care unit; JXN, junctional escape rhythm; LAR, left atrial rhythm; N/A, not applicable; RAR, right atrial rhythm; and TCPC, total cavopulmonary connection.

*P value from χ^2 test or Fisher exact test variables and Wilcoxon rank-sum test for continuous variables.

left heart syndrome, developed complete heart block with the Fontan operation. A dual-chamber pacemaker was implanted. The fourth patient, a 2-year-old girl, demonstrated sinus rhythm at a rate of 150 beats per minute immediately after the Fontan operation. Five years later, her rhythm was junctional at ≈60 beats per minute with the atrial beats normally conducted

Table 3. IART Data (N=134)

	Characteristics	All (N=134)	Randomization		*P* Value*
			Incision (n=67)	No Incision (n=67)	
Pre-Fontan					
	Inducible IART	30 (22.4)	13 (19.4)	17 (25.4)	0.08
	None	57 (42.5)	36 (53.7)	21 (31.3)	0.08
	Not performed/outside study	47 (35.1)	18 (26.9)	29 (43.3)	
Post-Fontan					
	Inducible IART	22 (16.4)	12 (17.9)	10 (14.9)	0.70
	None	92 (68.7)	46 (68.7)	46 (68.7)	0.70
	Not performed/outside study	19 (14.2)	9 (13.4)	10 (14.9)	
	Unknown	1 (0.7)	0 (0.0)	1 (1.5)	
IART at follow-up		4/114 (3.5)	2/61 (3.3)	2/53 (3.8)	1.00

Data are presented as n (%). IART indicates intra-atrial reentrant tachycardia.

*P value from χ^2 or Fisher exact test.

Table 4. Pre-/Post-Fontan EP Study (N=134)

	Post-Fontan EP Study				*P* Value
	Inducible IART	None	Not Done	Unknown	
Pre-Fontan EP study					
Inducible IART	7 (5.2)	18 (13.4)	6 (4.5)	0 (0.0)	
None	8 (6.0)	39 (29.1)	8 (6.0)	1 (0.7)	0.10*
Not done	7 (5.2)	35 (26.1)	5 (3.7)	0 (0.0)	

Data are presented as n (%). IART indicates intra-atrial reentrant tachycardia.
*Comparison was made as inducible IART vs none, and *P* value came from MacNemar test.

at an atrial rate rhythm of 40 beats per minute. A dual-chamber pacemaker was placed because of sick sinus syndrome. All 4 patients were alive at the last follow-up; they all had undergone several pacemaker replacements.

Discussion

This study was designed to evaluate the efficacy and safety of a right atrial incision placed between the tricuspid annulus and the anterior suture line of the lateral tunnel to prevent postoperative IART in Fontan patients. On the basis of anatomic and electrophysiological studies, the incidence of postoperative IART was expected to decrease in the incision group. However, no difference between the groups was observed. Importantly, the most significant finding was that, with current surgical management, the incidence of IART was considerably lower than the previous retrospective, observational studies.

The electrophysiological data also indicated that there was no acute deleterious effect generated by the incision itself. Importantly, there was no difference in sinus node or atrioventricular node function as determined by programmed atrial extrastimulation between the 2 groups. In addition, the follow-up data suggest that this incision itself was not associated with the development of arrhythmia in the intermediate term.

Surgical approaches for the management, including prevention of cardiac arrhythmias, are well established. In contrast to the design of surgical strategies,[33,34] our plan, like that of Collins et al,[31] was to provide a simple approach that would not significantly add to the cardiopulmonary bypass or operating time, indicating minimal if any impact on the surgical procedure. Although cross-clamp time was statistically significantly longer in the incision group compared with the

Table 5. Post-Fontan Wire Data (N=134)

	All (N=134)	Randomization				*P* Value*
		Incision, n=67		No Incision, n=67		
Age at wire study, y	1.8 (1.6–2.2)	1.8 (1.6–2.1)		1.9 (1.7–2.3)		0.41
Weight at wire study, kg	11.3±1.5	11.1±1.4		11.5±1.6		0.27
Wire study arrhythmia						
IART	22 (16.4)	12 (17.9)		10 (14.9)		0.73
None	95 (70.9)	48 (71.6)		47 (70.1)		
Not performed	14 (10.4)	6 (9.0)		8 (11.9)		
Unknown	3 (2.2)	1 (1.5)		2 (3.0)		
			n		n	
Wire-MCSNRT (N=115)	142 (95–214)	149 (88–240)	59	131 (95–199)	56	0.35
Wire-TSACT (N=84)	99.1 (80.8–122)	102 (85–122)	41	98.2 (77–121)	43	0.39
Wire-AVCERP SCL (N=95)	200 (180–240)	200 (180–244)	48	200 (180–240)	47	0.69
Wire-AVCERP_PCL (N=110)	210 (180–240)	220 (190–240)	58	200 (180–230)	52	0.17
Wire-AERP SCL (N=100)	190 (180–215)	190 (180–220)	51	190 (180–200)	49	0.48
Wire-AERP PCL (N=108)	190 (180–220)	190 (180–220)	56	190 (180–210)	52	0.75
Wire-AV Wenckebach (N=112)	250 (230–300)	260 (230–286)	57	250 (230–300)	55	0.80

Data are presented as N (%) for categorical variables and median (interquartile range) or mean±SD for continuous variables. AV indicates atrioventricular; AVCERP, atrioventricular conduction system effective refractory period; IART, intra-atrial reentrant tachycardia; MCSNRT, maximal corrected sinus node recovery time; PCL, coupled to 400 ms paced cycle length; and SCL, coupled to sinus cycle length; and TSACT, total sinoatrial conduction time.
**P* value from χ^2 test for categorical variables and Wilcoxon rank-sum test or *t* test for continuous variables.

Table 6. Pre-/Post-Fontan Wire Data

	Pre-Fontan	Post-Fontan	*P* Value*
MCSNRT (n=70)	137 (83–200)	139 (95–190)	0.62
TSACT (n=47)	113 (92–153)	95 (80–121)	0.003
AVCERP (n=58)	200 (170–237)	195 (180–220)	0.73
AVCERP_PCL (n=61)	190 (170–240)	200 (180–240)	0.43
AERP (n=60)	190 (160–204)	190 (180–204)	0.19
AERP_PCL (n=60)	170 (150–200)	190 (180–210)	0.002
AV Wenckebach (n=62)	240 (210–290)	250 (230–300)	0.49

Data are presented as median (interquartile range). AERP indicates atrial effective refractory period; AV, atrioventricular; AVCERP, atrioventricular conduction system effective refractory period; MCSNRT, maximal corrected sinus node recovery time; PCL, coupled to 400 ms paced cycle length; and TSACT, total sinoatrial conduction time.

**P* from Wilcoxon signed-rank test.

no incision group, the duration of a median of 6 minutes was clinically insignificant (Table 2).

Many recent studies have also demonstrated that the incidence of the IART after the Fontan operation is considerably less than originally observed, at least for the intermediate follow-up period (10–15 years).[2,31,35–39] Collins et al[31] reported the results of a similar study design; mean follow-up was 2.4 years. There was no statistical difference in spontaneous or induced arrhythmia between the 2 groups in the early postoperative period at follow-up. A 10-year follow-up of this cohort showed no incidence of IART or other arrhythmia; there was a 31% incidence of sinus node dysfunction (no group difference) but only one pacemaker implanted.[36]

Data from the Pediatric Heart Network reviewing arrhythmias in a large Fontan cohort indicate that the incidence of IART was also low (7.3%; mean follow-up 8.6±3.4 years). The hazard for IART decreased until 4 to 6 years post-Fontan, and then increased with age thereafter (likely because of short follow-up time for the lower risk groups, ie, lateral tunnel group).

Lasa et al[37] reported a cohort of patients receiving either intracardiac lateral tunnel or extracardiac conduit between 1995 and 2005. There was no difference in early (<30 days) or in late (>30 days) bradyarrhythmias or tachyarrhythmias including IART. The incidence of tachycardias was low at 4%, confirming the low incidence of IART in contemporary cohorts.

Balaji et al[2] reported a 10% late (median follow-up: 9.2 years) incidence of tachyarrhythmias in patients with an intracardiac lateral tunnel, considerably less than that reported earlier. Interestingly, the extracardiac patients had

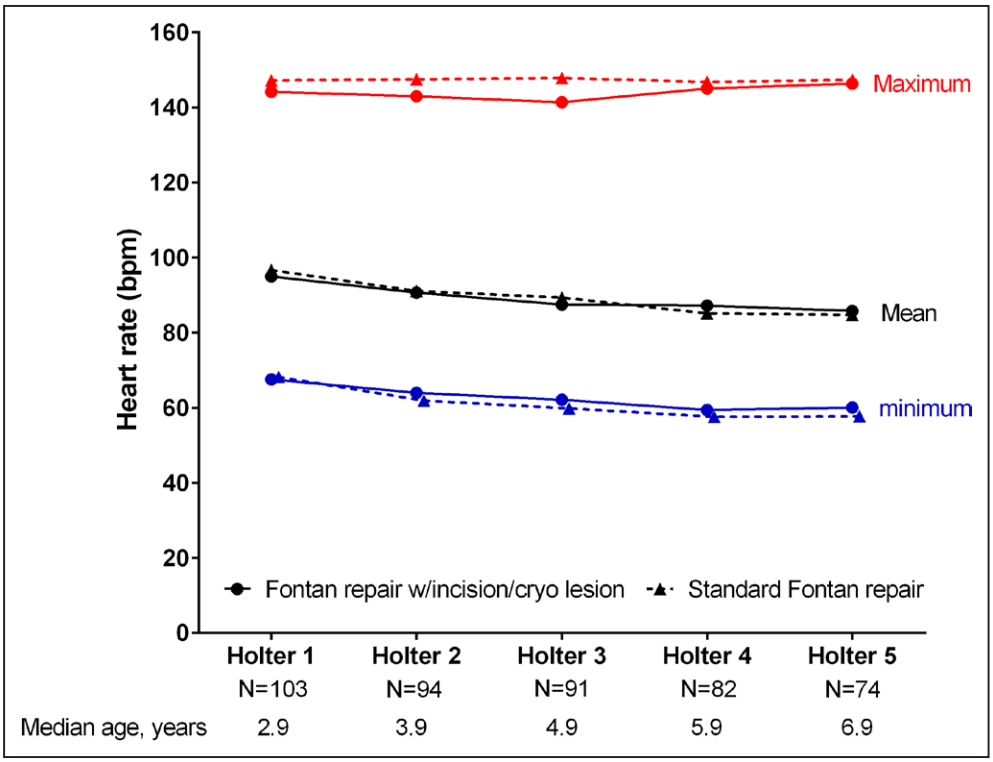

Figure 2. Average, maximal, and minimal heart rate by the study groups across time. The 4 pacemaker patients are not included in this analysis.

a nonsignificantly lower incidence of tachyarrhythmias (3%) than did the intracardiac lateral tunnel patients, perhaps related to a shorter follow-up.

A large, single-center study from the Mayo Clinic[38] encompassing 40-year follow-up demonstrated that 41% of patients had one or more arrhythmia at least 30 days after the Fontan operation. Among those patients with arrhythmias, atrial flutter was found in 74%; other arrhythmias found were atrial fibrillation, atrial tachycardia, reentrant supraventricular tachycardia, and ventricular tachycardia. Of the 41% of patients with late arrhythmias, an arrhythmia was present at 10, 20, and 30 years after the Fontan in 29%, 58%, and 76% of patients, respectively. In this study, in contrast to our study and that of Collins et al,[31] only 2% of the patients had hypoplastic left heart syndrome. In addition, the patients who had surgery during the early years of the study received an atriopulmonary connection as the surgical technique, that is, the patients with the longest follow-up and highest incidence of post-Fontan and arrhythmias.

A cross-sectional multicenter study from Europe[39] compared incidence of arrhythmias at follow-up between an intra-atrial lateral tunnel (baffle or prosthetic material) and the extracardiac conduit. They found that there was a greater incidence of atrial tachycardia in the intracardiac lateral tunnel group (15%) when compared with extracardiac group (1%). However, in contrast to the surgical techniques used in our patients,[28] their lateral tunnel baffle technique involved more incisions as well as a flap of the atrial wall to create the Fontan pathway. Interestingly, all of these patients were >8 years of age when enrolled in the study and therefore not comparable to our group who were all enrolled at Fontan operation when they were <5 years of age.

Many of the studies indicated that there was a significant incidence of sick sinus syndrome (27%)[37] requiring pacemaker implant in ≤20% patients.[2,36–39] In this series, 3 patients eventually came to the diagnosis of sick sinus syndrome requiring a pacemaker. In addition, in contrast to the usual patient with sick sinus syndrome, the mean heart rate of a cohort altogether (minus the pacemaker patients) was 90 to 100 beats per minute (Figure 2). Given the wide confidence limits of these data and the variability in individual provider practices, some of these patients might have had slow enough heart rates that would have triggered a pacemaker implantation at different centers.

Limitations of the study are the intermediate duration of follow-up (≈10 years) and the absence of follow-up data points (lost to follow-up) despite querying last known addresses, the patient's physicians and cardiologists, public databases, and death registries. Further follow-up of this cohort may reveal both an increased incidence of IART and possibly a difference between the 2 groups. The findings of this study are also potentially applicable only to the specific type and timing of the stage 2 hemi-Fontan and the stage 3 lateral tunnel Fontan performed at this single institution.

In conclusion, this randomized trial showed no difference in the incidence of IART after the Fontan operation between the study incision and nonincision groups during a 10-year period. Second, there was no difference in the inducibility of arrhythmia between the pre-Fontan and early post-Fontan regardless of the intervention. Third, the intervention of incision does not seem to have an arrhythmogenic effect in the short or medium term. Fourth, perhaps the most significant finding was that the incidence of IART is markedly less than previously reported and is congruent with more contemporary reports. These findings suggest that improvements in surgical management have, in general, reduced the risk of IART in the early and intermediate term for patients after the Fontan operation. Nevertheless, long-term follow-up as reported in several contemporary reports[38,39] indicates that after a 6- to 10-year postoperative period, IART may yet again emerge as a significant arrhythmia complicating the Fontan operation. Continued late monitoring for cardiac arrhythmias is, therefore, warranted.

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Disclosures

None.

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Follow-Up of a Prospective Surgical Strategy to Prevent Intra-Atrial Reentrant Tachycardia After the Fontan Operation

Ian H. Law, Osman Alam, Edward L. Bove, Richard G. Ohye, David J. Bradley, Sunkyung Yu and Macdonald Dick II

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