International Journal of Cardiology 187 (2015) 157–163

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Intra-atrial reentrant tachycardia in adult patients after Fontan operation

Mi Kyoung Song [a], Eun Jung Bae [a,1,*], Bo Sang Kwon [a], Gi Beom Kim [a], Chung Il Noh [a], Jung Yun Choi [a], Woong Han Kim [b], Jeong Ryul Lee [b], Yong Jin Kim [b]

[a] Department of Pediatrics, Seoul National University Children's Hospital, Seoul, Republic of Korea
[b] Department of Thoracic and Cardiovascular Surgery, Seoul National University Children's Hospital, Seoul, Republic of Korea

ARTICLE INFO

Article history:
Received 4 September 2014
Received in revised form 17 December 2014
Accepted 15 March 2015
Available online 17 March 2015

Keywords:
Fontan procedure
Tachyarrhythmia
Supraventricular tachycardia
Congenital heart disease

ABSTRACT

Background: : Atrial tachyarrhythmia is a major late complication in adult Fontan patients. This study examined the clinical features and risk factors of late intra-atrial reentrant tachyarrhythmia (IART) in adult patients after Fontan surgery and the mid-term outcome of Fontan conversion with or without antiarrhythmic surgery in these patients.
Methods: : We conducted a retrospective study on adult patients who were born before 1994 and survived at least 3 months after a Fontan operation at Seoul National University Children's Hospital.
Results: : We followed 160 patients over 20.9 ± 4.1 years. Sustained atrial tachycardia was identified in 51 patients, and IART was found in 41, appearing a mean 13.6 years after surgery. By the 25 year follow-up, 40% had developed IART. The incidence of IART significantly increased over time. Patients with an atriopulmonary connection (APC) (n = 65) had significantly longer follow-up duration and higher incidence of IART than patients with a lateral tunnel (n = 86) or extracardiac conduit Fontan (n = 9). On multivariate analysis, APC, sinus node dysfunction, and nonsustained atrial tachycardia were found to be significantly associated with IART. Twenty-four patients with IART underwent Fontan conversion. Over the follow-up period, IART severity scores in the 22 patients who survived after Fontan conversion decreased significantly, and New York Heart Association functional class significantly improved. On multivariate analysis, protein losing enteropathy and ventricular dysfunction were found to be significant risk factors for mortality.
Conclusions: : IART was common in adult Fontan patients, and Fontan conversion with or without antiarrhythmic surgery and pacemaker placement helped to control it.

1. Introduction

Atrial tachycardia is a troublesome problem, affecting morbidity and mortality in patients after Fontan operation. Fontan patients often have extensive atrial scarring and are prone to right atrial dilatation and fibrosis, which may act as arrhythmogenic substrates. Indeed, up to 50% of Fontan patients develop atrial tachyarrhythmia within 20 years of the operation [1,2]. Intra-atrial reentrant tachycardia (IART), a type of macroreentrant tachycardia, is the most common form of atrial tachyarrhythmia in Fontan patients [3,4]. In adult Fontan patients (a growing population), IART reduces quality of life and may even cause death in hemodynamically compromised patients [5]. Several studies have recommended that these patients be treated with Fontan conversion to the lateral tunnel (LT) or extracardiac conduit (ECC) Fontan with concomitant antiarrhythmic surgery [6–8]; however, outcomes of such a procedure have not been well identified. The aim of this study was to evaluate clinical features of and risk factors for late IART over an extended period and to assess mid-term outcomes of Fontan conversion with antiarrhythmic surgery.

2. Methods

2.1. Patients and definitions

We conducted a retrospective cohort study of 160 patients born before May 1994 who have been followed up at Seoul National University Children's Hospital after Fotan operation. Patients who were lost to follow-up, died within 3 months of their Fontan operation, or had incomplete medical records were excluded. We reviewed clinical notes, electrocardiograms, 24-hour Holter monitor records, echocardiograms, images and reports obtained through cardiac catheterization, electrophysiologic studies, and computed tomography scans.

Late IART was defined as sustained IART, documented by electrocardiography, Holter monitoring, or pacemaker monitoring, that occurred 3 months or more after Fontan surgery. Sinus node dysfunction (SND)

* Corresponding author at: Department of Pediatrics, Seoul National University Children's Hospital, Republic of Korea.
E-mail address: eunjbaek@snu.ac.kr (E.J. Bae).
[1] Mailing address: Eun Jung Bae, MD, Department of Pediatrics, Seoul National University Children's Hospital, 101 Daehang-Ro, Jongno-gu, Seoul 110-744, Republic of Korea.

http://dx.doi.org/10.1016/j.ijcard.2015.03.157
0167-5273/

was defined as sinus bradycardia with a resting heart rate more than 2 standard deviation lower than normal for the patient's age, predominant junctional rhythm, or sinus pause of ≥3 s, with or without escape beats [9–11]. Ventricular dysfunction was defined as ejection fraction < 50% of systemic ventricle on echocardiogram.

To evaluate clinical outcomes of Fontan conversion, we used a modified version of the multiscale IART severity score developed by Triedman et al. [12]. The instrument was composed of 4 categories; documented IART, IART severity, antiarrhythmic medications, and frequency of cardioversion for prior 3 months, which was modified to frequency of cardioversion for prior one year in this study. Scores on these 4 scales were summed to yield a total IART severity score ranging from 0 to 12 points, which was calculated for baseline (before Fontan conversion) and last follow-up. A favorable long-term outcome was defined as a score at last follow-up that was ≤3 or had decreased by ≥3 from the preoperative score. An unfavorable long-term outcome was defined as a follow-up score that was ≥4 or had decreased by ≤2 from the preoperative score.

The study was approved by the Institutional Review Boards of Seoul National University Hospital.

2.2. Statistical methods

Continuous variables were recorded as mean ± standard deviation or as median and range, and values of the normally distributed ones were compared between groups by *t*-test and analysis of variance. Univariate analysis of categorical variables was performed using the log-rank test, Pearson's chi-square test, Fisher's exact test, and Cox regression to determine which ones might be risk factors for late IART and mortality. The Cox proportional-hazards model was used to estimate the hazard ratio for occurrence of late post-Fontan IART in relation to years of survival after the operation. Cumulative probability of freedom from IART was analyzed using the Kaplan–Meier estimator, both for the study sample as a whole and for each Fontan configuration. Multivariate logistic regression was conducted to determine which factors were associated with late post-Fontan IART and death. Statistical analyses were performed using SPSS version 21.0 (IBM, Armonk, NY, USA). A p-value of <0.05 was considered statistically significant.

3. Results

3.1. Fontan patients

Demographic and clinical characteristics of the patients included in this study are shown in Table 1. Age at Fontan operation ranged from 8 months to 36.3 years, and 3351.8 patient-years of follow-up were conducted over a median period of 20.9 years (range 6.3–31.6). Atriopulmonary connection (APC) Fontan was performed in 65 patients (40%) since 1982, lateral tunnel (LT) Fontan in 86 (53%) since 1990 and extracardiac conduit (ECC) Fontan in 9 (6%) since 1997.

The initial diagnosis of patients was categorized into 7 groups; tricuspid atresia in 29 patients (18%), common inlet ventricle with unbalanced atrioventricular septal defect in 39 (24%), mitral atresia in 29 (46%), double inlet ventricle in 33 (21%), complicated double outlet right ventricle in 9 (6%), complicated transposition of the great arteries in 7 (4%), and pulmonary atresia with intact ventricular septum in 2 (1%) (Table 1). Heterotaxy syndrome was diagnosed in 41 (25.6%) patients, of whom 21 had right isomerism and 20 had left isomerism. Most patients had a single ventricle of right ventricular type (88/160, 55%); 53 (33%) had a single ventricle of left ventricular type, and 19 (12%) had both ventricles type.

3.2. Supraventricular tachycardia

Sustained supraventricular tachycardia was identified in 51 patients (31.8%; Table 2), of whom 41 (25.6%) had late IART, including 6 patients with both IART and sustained atrial fibrillation. Paroxysmal atrial fibrillation without IART occurred in 1 patient, 23.1 years after Fontan

Table 1
Demographics and clinical characteristics.

Demographics and clinical variables		All patients (n = 160)	Patients with IART (n = 41)	Patients without IART (n = 119)	p
Age at Fontan (years)[a]		4.8 ± 4.7 3.2 (0.8–36.3)	5.0 ± 3.3 4.1 (0.8–15.4)	4.8 ± 5.1 3.0 (1.2–36.3)	NS
Follow-up time after Fontan (years)[a]		20.9 ± 4.1 20.9 (6.3–31.6)	23.3 ± 4.1 23.7 (12.8–30.6)	20.1 ± 3.8 20.2 (6.3–31.6)	<0.001
Gender, male		96 (60%)	25 (61%)	71 (60%)	NS
	APC	65 (41%)	31 (76%)	34 (29%)	
Fontan type	LT	86 (54%)	9 (22%)	77 (65%)	<0.001
	ECC	9 (6%)	1 (2%)	8 (7%)	
Type of single ventricle					NS
Right ventricle		88 (55%)	20 (49%)	68 (57%)	NS
Left ventricle		53 (33%)	14 (34%)	39 (33%)	NS
Both ventricles		19 (12%)	7 (17%)	12 (10%)	NS
Heterotaxia		41 (26%)	12 (29%)	19 (16%)	NS
Right isomerism		21 (13%)	5 (12%)	16 (13%)	NS
Left isomerism		20 (13%)	7 (17%)	13 (11%)	NS
Initial diagnosis					NS
Tricuspid atresia		29 (18%)	7 (17%)	22 (19%)	
Common inlet ventricle with Unbalanced AVSD		39 (24%)	9 (22%)	30 (25%)	
Mitral atresia		41 (26%)	11 (27%)	30 (25%)	
Double inlet ventricle		33 (21%)	6 (15%)	27 (23%)	
Complicated DORV		9 (6%)	3 (7%)	6 (5%)	
Complicated TGA		7 (4%)	4 (10%)	3 (3%)	
PA with IVS		2 (1%)	1 (2%)	1 (1%)	
Previous BT shunt		62 (39%)	20 (49%)	42 (35%)	NS
Sinus node dysfunction		38 (24%)	23 (14%)	15 (13%)	<0.001
Non-sustained atrial tachycardia		57 (39%)	28 (76%)	29 (27%)	<0.001
Protein losing enteropathy		9 (6%)	3 (2%)	6 (5%)	NS
Ventricle dysfunction		37 (24%)	13 (32%)	24 (20%)	NS
Moderate-to-severe AVVR		11 (7%)	5 (12%)	6 (5%)	NS
Death		13 (8.1%)	6 (14.6%)	7 (5.9%)	0.08

IART indicates intra-atrial reentrant tachycardia; APC, atriopulmonary connection Fontan; LT, lateral tunnel.
Fontan; ECC Extracardiac connection Fontan; AVSD, atrioventricular septal defect; DORV, double outlet of right ventricle; TGA, transposition of great artery; BT shunt, Blalock–Taussig shunt; AVVR, atrioventricular regurgitation.
[a] Data expressed as mean ± standard deviation and median value (range).