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Sinus node dysfunction after Fontan modifications—influence of surgical method

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Abstract

Background: Sinus node dysfunction (SND) is reported to be a troublesome complication following various types of Fontan operations. The correlation of post-Fontan SND with surgical methods was evaluated in this study. *Methods:* By reviewing the medical records, surface ECGs, and Holter monitoring, the range of heart rate (HR) and the risk of SND at intermediate term after Fontan type operation (follow up: 41.3±13.1 months) were analyzed between two age matched groups of patients, consisting of the extracardiac conduit group (EC, $n=33$) and the lateral tunneling group (LT, $n=35$). *Results:* Junctional rhythm was observed in nine out of 35 patients in LT and five out of 33 patients in EC during the follow-up period. Resting HR was faster in EC than that in LT (108±15 vs. 82±21, $P<0.001$). Average and maximal HR in Holter monitoring were also faster in EC than those in LT. SND was found in 13 cases (10 in LT, three in EC) during follow-up and one required pacemaker implantation. In the case of situs solitus heart, SND was less frequent in EC than in LT (0/16 vs. 8/26, $P=0.01$). In the case of heterotaxy syndrome, SND occurred in similar number of cases (3/17 vs. 2/9). The staged approach to Fontan completion did not influence SND. LT repair was the only factor causing sinus node dysfunction according to multivariate logistic regression ($P=0.03$, OR 5.96). *Conclusions:* Lateral tunnel type surgical repair was more likely to lead to the development of sinus node dysfunction than extracardiac conduit operation. In the case of heterotaxy syndrome, surgical method had no significant influence.

Keywords: Fontan operation; Sinus node; Extracardiac conduit

1. Introduction

Sinus node dysfunction (SND) is reported to be a troublesome problem following various kinds of Fontan modifications [1–4]. The lateral tunnel Fontan operation (LT) is known to lead to a higher incidence of sinus node dysfunction than the atriopulmonary connection procedure [1], and the concerns about atrial tachyarrhythmia remain because of the extensive suture lines [5,6]. In recent years, the extracardiac conduit Fontan procedure (EC) has been tried in order to prevent such risks of rhythm disturbance [7]. In this study, we compared the results of ECG and 24 h ambulatory ECG monitoring following lateral tunneling and extracardiac conduit Fontan operations, at intermediate term in order to evaluate the impact of extracardiac conduit Fontan modification on the prevention of sinus node dysfunction. The influence

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of the anatomical aspect of the atria on rhythm changes following Fontan operation was also evaluated.

2. Materials and methods

We performed this retrospective evaluation by reviewing all medical records and surface ECGs and 24 h ambulatory ECGs.

2.1. Patient populations

The study population involves 68 consecutive cases of patients having survived either type of Fontan modification performed by two particular surgeons in two hospitals between July 1996 and June 1998, and who had been followed up for more than 2 years. Those patients who had the Fontan type operation when they were more than 5 years old were excluded (three patients with age of 14, 10 and 7 years, respectively, who underwent lateral tunnel type repairs). Among 68 patients, 35 had the lateral tunnel type Fontan operations (LT group) and 33 had the extracardiac conduit Fontan operations (EC group). The demographic data are summarized in Table 1. There were no patients with hypoplastic left heart syndrome (HLHS), but 26 out of 68 patients (38%) had heterotaxy syndrome. The diagnosis of heterotaxy syndrome (asplenia syndrome and polysplenia syndrome) was made according to previously reported criteria [8,9]. Neonatal palliations were performed in 15 out of 35 cases for the LT group (pulmonary artery banding in six, modified BT shunt in eight, Norwood type palliation in one) and 12 out of 33 cases for the EC group (pulmonary artery banding in six and modified BT shunt in six). For the staged approach, the bi-directional Glenn operation was performed in 18 out of 35 cases for the LT group and 30 out of 33 cases for the EC group. The Hemi-Fontan operation was performed in 3 out of 35 cases for the LT group. The sex ratio, disease pattern, age at the time of operation, duration of follow up and age at the last ECG were not different between the two surgical groups.

Table 1
Demographic data of two surgical groups

	LT (n=35)	EC (n=33)	P value
Male:female	20:15	16:17	ns
Situs			
Solitus/inversus	22/4	15/1	ns
Right isomerism	7	12	ns
Left isomerism	2	5	ns
Single RV (not HLHS)	12	18	ns
Single LV	2	5	ns
Tricuspid atresia	7	2	ns
Others	14	8	ns
Age at BDG (months)	11±7	11±9	ns
Age at Fontan (months)	30±14	32±16	ns
Staged operation	21	30	ns
Fenestrated Fontan	23/35	20/33	ns
Duration of follow-up	43±16	40±9	ns
Age of latest ECG (months)	73±22	73±19	ns

The lateral tunnel type Fontan modification consists of a longitudinal right atriotomy incision and the making of a polytetrafluoroethylene (PTFE) baffle connecting the inferior vena cava along the lateral atrial wall to the superior vena cava entrance into the pulmonary artery. The extracardiac conduits were made of PTFE tube grafts sized 18±1 (16–20) mm. The conduit was connected to the IVC in an end-to-end fashion and to the pulmonary artery in an end-to-side fashion. A fenestration was made by means of a single punch in the LT group (23/35, 3.7±0.7 mm sized), and by means of a single side-to-side fenestration or placement of small tube graft in the EC group (20/33, 4.0±0.5 mm sized).

Prolonged pleural effusion for more than 2 weeks was complicated in six out of 35 patients in the LT group and three out of 33 patients in the EC group. One patient had been suffering from protein losing enteropathy following EC operation. To date, no death has occurred among the total of 68 patients during the follow-up period.

2.2. ECG evaluation

For all patients a standard 12 lead ECGs were reviewed, from the initial diagnosis to the latest follow-up, after the Fontan operation. An early postoperative ECG was taken in 1 month of the Fontan operation performed. All the ECGs were taken in the resting condition. P wave axis and heart rates were analyzed. The rhythm and P wave axis change were analyzed before and after Fontan operation. The resting awake heart rates of late postoperative period were also analyzed and compared between

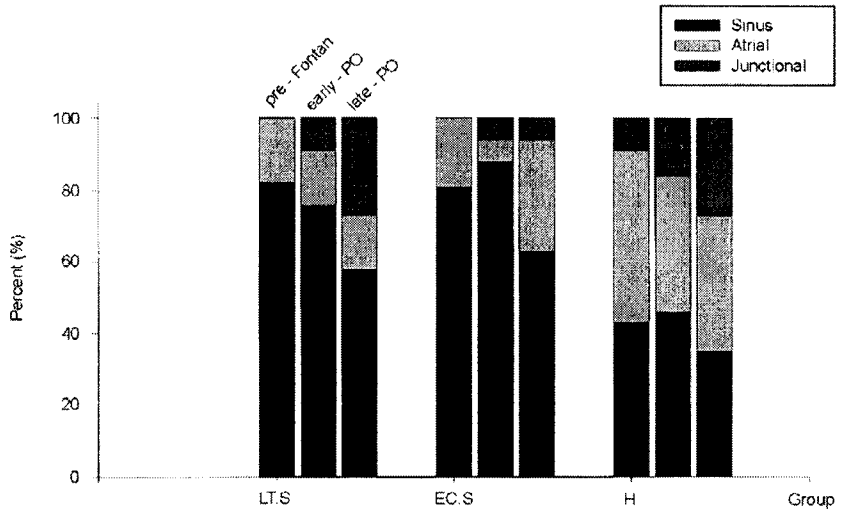

Fig. 1. The proportions of the rhythm at pre-Fontan period, early after Fontan operation and late after Fontan operation. First group with situs solitus/inversus heart and LT repair (LT.S), second group with situs solitus/inversus heart and EC repair (EC.S) and last group with heterotaxy syndrome and either LT or EC repair (H).

two surgical groups. The changed P wave axis was considered if the P axis differed by more than 45 degrees from the initial preoperative tracing. Immediate postoperative bradycardia was defined as a slow HR, requiring temporary atrial pacing or a slow HR of more than 2 S.D. below the age-adjusted mean for a period of more than 12 h at the postoperative ICU setting. Ambulatory 24 h ECGs were taken and analyzed for basic rhythm and for the presence of arrhythmia. Sinus rhythm was diagnosed, when present in two or more consecutive surface ECG recordings, or for >90% of the 24 h period in the ambulatory 24 h ECGs, and junctional rhythm was diagnosed when present in two or more consecutive surface ECG recordings or >10% of the 24 h period in the ambulatory 24 h ECGs. SND was diagnosed as previously reported [11] when more than one of the following three items were found in the resting ECGs or 24 h ECGs: (1) junctional rhythm present >50% of the 24 h period, (2) sinus pause of 3 s or more in duration with or without escape beats and (3) slow resting heart rate more than 2 S.D. below the age-adjusted mean at the two or more consecutive resting ECGs recorded [10–12].

2.3. Statistical analysis

Values are reported as mean±standard deviation. Univariate comparisons of continuous variables were made using the Student's t-test. Univariate analysis of the differences in proportion between the two groups was analyzed by the χ^2 test. For multivariate analysis, multiple logistic regression or the GLM (generalized linear model) procedure were used.

3. Results

3.1. Rhythm changes

By comparison of the P waves between the preoperative ECG and the post-palliative surgery ECG, the bi-directional Glenn shunt or Hemi-Fontan (n= 51) was found to be associated with the changed P wave axis in 17 out of 51 patients. Heterotaxy syndrome was found to be more prone to rhythm change than the heart with either situs solitus or inversus (solitus/inversus) following bi-directional Glenn shunt operation (11/22 in heterotaxy syndrome, 6/29 in solitus/inversus, P=0.026) (Fig. 1).

Immediate postoperative bradycardia after a Fontan type operation was found in seven out of 35 cases in the LT group and five out of 33 cases in the EC group. Transient junctional ectopic tachycardia or atrial tachycardia was found in three out of 35 cases

in the LT group and three out of 33 cases in the EC group.

In the early period, following the Fontan operation, there was no significant difference between the two surgical groups (Table 2). After a longer period of time following the operation, nine out of 35 patients in the LT group (7/26 with solitus/inversus, 2/9 with heterotaxy) showed junctional rhythm. However, five out of 33 patients in the EC group (1/16 with solitus/inversus, 4/17 with heterotaxy) had junctional rhythm. There was a tendency for junctional rhythm to occur more frequently following the lateral tunnel operation than following the extracardiac conduit Fontan operation, particularly in the case of solitus/inversus heart (7/26 vs. 1/16; $P=0.09$). In the patients with heterotaxy syndrome, there was no difference in the late term rhythm according to surgical types (2/9 vs. 4/17, P=ns) (Fig. 1).

3.2. Comparison of heart rate

The resting heart rates at the intermediate term after Fontan operation were obtained. The resting heart rate was 84±22 per min in LT group and 91±24 per min in EC group. In the case of situs solitus/inversus heart, the heart rate was slower in the LT group than in the EC group (83±22 vs. 108±15, $P<0.001$). In the case of heterotaxy syndrome, there was no difference in heart rate between the two surgical groups (Table 3). The lateral tunnel type operation was the only factor found to be affecting the heart rate at intermediate term follow-up, according to multivariate regression analysis ($P=0.035$). Age at Fontan operation ($P=0.118$), staged approach to Fontan completion ($P=0.350$) and cardiac situs ($P=0.926$) were found not to be associated with intermediate term resting heart rate, according to multivariate regression analysis.

Table 2
Rhythm change

	LT	EC	P value
Immediate bradycardia	7/35	5/33	ns
Sinus rhythm, early	24/35	22/33	ns
Sinus rhythm, late	18/35	16/33	ns
Junctional rhythm, late	9/35	5/33	ns
Solitus/inversus	7/26	1/16	0.09
Heterotaxy	2/9	4/17	ns

Table 3
Heart rate after operation at intermediate term follow-up

	LT	EC	P value
Resting HR			
Total	84±22	91±24	0.11
(age, months)	(73±22)	(73±19)	(0.97)
Solitus/inversus	83±28	108±15	<0.0001
(age, months)	(71±19)	(64±11)	(0.24)
Heterotaxy	86±23	84±18	0.32
(age, months)	(77±26)	(80±22)	(0.7)
Holter monitoring			
Minimum HR	57±22	61±14	ns
Average HR	83±19	97±18	0.02
Maximum HR	138±25	167±20	0.0005
(age, months)	57±21	65±20	0.2

3.3. Comparison of the results of the 24 h ambulatory ECGs

Twenty-four hour ambulatory ECGs were recorded 24 out of 35 patients (68%) in the LT group and 20 out of 33 patients (60%) in the EC group (Table 3). In the LT group, we found sinus pause in eight patients, sustained atrial tachycardia and persistent first degree AV block in one patient. In the EC group we found sinus pause in two patients and sustained atrial tachycardia in one patient. The minimum heart rates did not differ between the two surgical types. However the average and maximum heart rate were faster in the EC group than in the LT group.

3.4. Sinus node dysfunction

Sinus node dysfunctions at intermediate term follow up were found in 10 out of 35 patients in the LT group (8/26 in solitus/inversus heart and 2/9 in heterotaxy syndrome) and three out of 33 patients in the EC group (0/16 in solitus/inversus heart and 3/17 in heterotaxy syndrome). One patient required a permanent pacemaker at intermediate term follow-up

Table 4
Sinus node dysfunction

	LT	EC	P value
Total	10/35	3/33	0.041
Solitus/inversus	8/26	0/16	0.013
Heterotaxy	2/9	3/17	ns
Pacemaker	1/35	0/33	ns

(Table 4). Extracardiac conduit operation was effective to preserve sinus node function and lateral tunnel type surgical repair was the only factor causing sinus node dysfunction ($P=0.031$, odds ratio 5.952 and 95% confidence limits 1.175–30.157) according to multivariate logistic regression. Immediate bradycardia was found to be a weak risk factor ($P=0.072$) and higher age at Fontan repair was also found to be a weak risk factor ($P=0.061$). Sex ($P=0.928$), situs solitus/inversus or heterotaxy syndrome ($P=0.701$), and staged approach to Fontan completion ($P=0.584$) were not related to postoperative sinus node dysfunction according to multivariate logistic regression.

4. Discussion

The development of early sinus node dysfunction, with or without junctional escape beats or accelerated junctional rhythm, after a Fontan type operation, can have an adverse effect on marginal postoperative hemodynamics of Fontan physiology, resulting in low postoperative cardiac output, high left atrial pressure, high central venous pressure and prolonged chest tube drainage. Late term sinus node dysfunction has also been reported to be progressive, with occurrence rates of almost 10–12% after atriopulmonary connection and 20–25% after LT type operation [1,11,12]. Moreover, sinus node dysfunction or slow sinus rate may predispose patients to the development of atrial flutter or fibrillation, which is one of the major causes of late term morbidity and mortality [2–4,13].

The procedures have been modified to prevent the occurrence of arrhythmias and to obtain improved hemodynamics. The lateral tunnel type Fontan modification and extracardiac conduit Fontan operation are the more commonly used methods in recent times. It has been argued that the extracardiac conduit operation has an advantage in terms of rhythm conditions, but this is at the expense of depletion of the growth potential of the Fontan pathway and possible thromboembolic complications [12,14–16]. There have been a few reports comparing the effects on rhythm status following these types of surgery. However, these previous studies had some limitations, with one report offering a comparison of only early postoperative sinus node dysfunction, while another report contained a comparison based on different follow-up intervals [12,17]. Because of the increasing incidence of sinus node dysfunction as time passes, any comparison of surgical methods must be adjusted according to the duration of follow-up. This study presents a comparison of rhythm status, heart rate range and development of sinus node dysfunction between two popular types of surgery, after an average of 43 and 40 months follow-up, respectively.

In the normal heart, the sinus node is located near the junction of the superior vena cava and the right atrium in the sulcus terminalis. In the case of heterotaxy syndrome, the structure of the cavo-atrial junction is usually anomalous, such as in the case of a bilateral vena cava with bilateral SA node in asplenia syndrome ("right atrial isomerism"). In polysplenia syndrome, there is no specific right atrial structure and the structure of sinus node may not be recognizable [18,19]. Although the real primary pacer(s) within the atrium may not be simple, multiple P waves and sinus node dysfunctions have been observed in the case of heterotaxy syndrome [20–22]. Our patient population is noteworthy because of relatively large number of patients with heterotaxy syndrome. This study showed clearly that heterotaxy syndrome tends to have multiple P wave morphology, even before the Fontan operation, and tends to give rise to a slower resting heart rate after the Fontan operation, irrespective of the surgical method employed, although severe sinus node dysfunction or predominant junctional rhythm did not occur very frequently.

However, the sinus node function, in the groups with normal visceroatrial situs, was influenced by the surgical method being used. This study shows that the extracardiac conduit Fontan operation is more effective in preserving intermediate term sinus node function than the lateral tunnel operation, in the heart with either situs solitus or inversus. This advantage, in terms of the preservation of sinus node function following the extracardiac conduit Fontan operation, may lead to the prevention of the late-term complication of atrial tachyarrhythmia, that has been known to be associated with sinus node dysfunction and the presence of extensive atrial suture lines. Moreover, in this study, the average and maximum heart rates of

the patients having undergone an extracardiac Fontan operation were faster than those of the patients having undergone a lateral tunnel operation. Such a difference in heart rate may be associated with a difference in cardiac output, especially in the period of early childhood.

Our study contains a certain number of limitations. Firstly, this is a non-randomized, retrospective study. Although we tried to obtain more Holter monitor recordings for all of the patients, with or without any rhythm problems, we were only able to obtain these data for 60 and 68% of each group, respectively. This factor may be certain limitation to know the correct incidence of sinus node dysfunction. Moreover, the changes from the preoperative to postoperative state could not be assessed exactly as the preoperative Holter monitoring was not available. The surgical methods were also not randomized and the decision to perform either a lateral tunnel type operation or extracardiac conduit placement depended on surgeon's preference. Secondly we have no direct electrophysiologic data regarding sinus node function such as sinus node recovery time or sinoatrial conduction time. The correlation between the rhythm status and hemodynamic conditions could not be assessed as the postoperative cardiac catheterization was not performed. Thirdly the study populations are relatively small and the postoperative follow-up durations were not long enough to define clearly the postoperative rhythm abnormality. However, we were able to compare the rhythm change, heart rates and the occurrence of sinus node dysfunction for two different surgical methods, and this for two age-matched groups.

5. Conclusion

In this study, we found that the occurrence of sinus node dysfunction after a Fontan type operation may be influenced by surgical method. The extracardiac Fontan operation leads to a lower incidence of sinus node dysfunction than does the lateral tunneling procedure, during intermediate term follow-up, in the

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