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Late Status of Fontan Patients With Persistent Surgical Fenestration

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Objectives This study was undertaken to determine the effects of creating a systemic-to-pulmonary venous atrial-level communication (fenestration) at the time of the Fontan procedure on late outcomes.

Background Fenestrations are frequently performed during Fontan procedures, but late consequences are not well described.

Methods Patient characteristics were compared between those with and without surgical fenestration among 536 subjects (mean age 11.9 years) enrolled in the Pediatric Heart Network Fontan Cross-Sectional Study. The status of the fenestration and the association of a currently patent fenestration with health status and measures of ventricular performance were investigated.

Results Fenestration was performed in 361 patients (67%), and frequency differed by year and center ($p < 0.001$ for each). After adjustment for center, age at Fontan, year of Fontan, and prior superior cavopulmonary surgery, the fenestrated group had shorter length of Fontan hospital stay. At the time of cross-sectional testing 8 ± 3 years after Fontan, the fenestration remained open in 19% of subjects. Among those with confirmed fenestration closure, 59% were by catheter intervention and 1% by surgical intervention, and 40% had apparent spontaneous closure. Compared with those without evidence of a fenestration, subjects with a current fenestration were taking more medications ($p = 0.02$) and had lower resting oxygen saturation (median 89% vs. 95%, $p < 0.001$). Functional health status, exercise performance, echocardiographic variables, prevalence of post-Fontan stroke or thrombosis, and growth did not differ by current fenestration status.

Conclusions Surgical fenestration is associated with well-demonstrated early post-operative benefits. This cross-sectional study found few associations between a persistent fenestration and deleterious later outcomes. (J Am Coll Cardiol 2011;57:2437–43) © 2011 by the American College of Cardiology Foundation

The Fontan procedure has provided excellent surgical palliation for many patients with functional single-ventricle physiology and has evolved over time (1). Creation of a systemic-to-pulmonary venous atrial-level communication (or fenestration) at the time of Fontan completion may benefit patients by limiting Fontan pathway pressure and creating a right to left atrial shunt, which may augment cardiac output and limit central venous pressure in the immediate post-operative period, at the expense of systemic desaturation with potential long-term sequelae. Retrospective, single-center reports suggest that in high-risk patients, those with a fenestrated Fontan have shorter duration of pleural effusion and shorter length of hospital stay (2,3). Subsequently, similar short-term perioperative benefits of fenestration creation have been shown in a single-center randomized study involving standard-risk patients (4). Although this procedure is now commonly performed, long-term benefits, if any, are not documented. Additionally, concerns remain about the need for later intervention to close the fenestration, as well as risks of persistent cyanosis and paradoxical embolization in patients with patent fenestrations.

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Abbreviations and Acronyms

AV = atrioventricular valve
BNP = B-type natriuretic peptide
CHQ P50 = Parent Report Child Health Questionnaire
PHN = Pediatric Heart Network
PLE = protein-losing enteropathy
stage II = cavopulmonary anastomosis

The Pediatric Heart Network (PHN) Fontan Cross-Sectional database (5) provides a very well-characterized cohort of surviving subjects from 7 North American centers in which to examine whether creation of a fenestration at the time of Fontan is associated with long-term health status, medical complications, or laboratory measures of cardiac function. Additionally, comparisons can be made between subjects with a currently patent fenestration and those without.

Methods

Data for analyses described here were obtained from 536 subjects enrolled from 7 clinical centers participating in the National Institutes of Health/National Heart, Lung, and Blood Institute–funded PHN Fontan Cross-Sectional Study. Subjects in this study were survivors of multistage surgical palliation for functional single ventricle culminating in a Fontan procedure who were 6 to 18 years of age at the time of sampling. Medical records were screened for 1,078 patients, of whom 831 (77%) were deemed potentially eligible for participation. After being contacted, 637 patients (60%) were fully eligible, and consent was obtained from 546 (86%). Subjects with a hepatic vein exclusion procedure (n = 10) were excluded from this analysis. The median time between Fontan surgery and study enrollment was 8.1 years. Complete details of the study design have been previously published (6).

Measurements. Demographic data and descriptors of medical history before, during, and after the Fontan procedure were obtained by medical record review. Standardized assessment of functional health status was obtained using the Parent Report Child Health Questionnaire (CHQ P50). Physical performance was assessed using bicycle ergometry (7). Ventricular size and function, atrioventricular valve and semilunar valve regurgitation, and fenestration status were obtained using centrally interpreted echocardiographic measurements. Resting B-type natriuretic peptide (BNP) concentration was obtained and analyzed at a core laboratory.

Statistical analysis. Exploratory analyses incorporating graphical and tabular displays assessed evidence in favor of trends and associations. Sample means, medians, and proportions accompanied by 95% confidence intervals provided descriptive summaries. Crude comparisons of subjects with and without surgical fenestration used Student *t* and chi-square test statistics. Covariate-adjusted comparisons were obtained by multiple linear and logistic regression analyses. Smoothed estimates of the proportion of subjects who underwent surgical fenestration were performed as a function of time and were obtained using generalized additive mixed models with study center entered as a random effect. Graphical analyses of the time to hospital discharge after Fontan surgery for this cohort of Fontan survivors were performed using the Kaplan-Meier method, with significance testing via log-rank tests; covariate-adjusted models for time to discharge were constructed using Cox proportional hazards regression. Analyses were performed using SAS software version 9.2 (SAS Institute, Cary, North Carolina) and the R system version 2.8.1 (R Foundation for Statistical Computing, Vienna, Austria).

Adjustment for covariates. We learned in previous analyses that many of the results of tests performed at the time of cross-sectional evaluation differed among age groups. Exercise performance decreased with age (7). Echocardiographically measured mass-to-volume ratio, dP/dt (rate of rise of left ventricular pressure), and Tei index also varied by age (5). Therefore, for outcomes measured at the time of cross-sectional testing (e.g., CHQ_P50, echocardiography, and exercise testing), age-adjusted comparisons are reported. Because the cohort of patients with a current fenestration were younger and followed for less time since Fontan completion, medical history variables collected since Fontan hospitalization are also reported using age-adjusted comparisons. Medication use among Fontan patients and performance of certain catheter interventions varied considerably across the 7 participating centers in the PHN (8,9). Therefore, where center or era differences might exist, we adjusted for subject age at Fontan cross-sectional enrollment, year that Fontan surgery was performed, and study center. Based on the previously observed association between the performance of cavopulmonary anastomosis (stage II) procedures and fenestration performance, the presence or absence of a prior stage II surgery was also included as a control factor in the post-Fontan medical history outcomes.

The Fontan Cross-Sectional Study protocol was approved by the PHN Protocol Review Committee and an independent Data and Safety Monitoring Board, as well as Institutional Review Boards at each of the 7 clinical centers and the Data Coordinating Center. All centers followed the same protocol and study procedures.

Results

Among this enrolled cohort of Fontan survivors, there was a rapid increase in the performance of a surgical fenestration between 1987 and 1992 (Fig. 1). After 1992, the proportion of Fontan procedures that included surgical fenestration remained approximately 80%. Although the use of fenestration increased over this time period at all centers, the percentage of subjects receiving a fenestration varied widely among centers (13% to 91%, $p < 0.001$).

Medical history before the Fontan procedure. Subject and medical history characteristics before and at the Fontan procedure are shown in Table 1. Age at cross-sectional

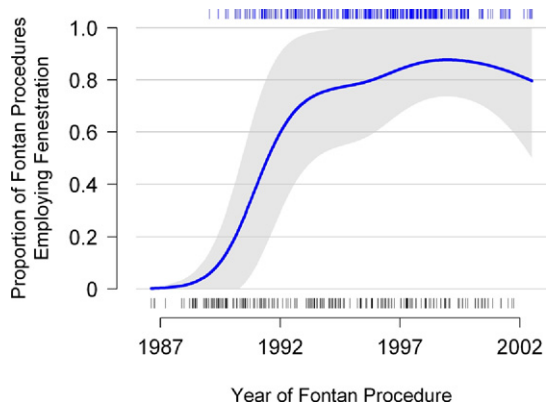

Figure 1 **Smoothed Proportion of Subjects With Surgical Fenestration by Year**

Rug plots at **top and bottom** record the dates of fenestrated and nonfenestrated procedures, respectively. Estimates were obtained using generalized additive mixed models. A 95% confidence interval is displayed as a **shaded region**.

study enrollment was approximately 2 years older among the subjects with no surgical fenestration (13.2 ± 3.6 years vs. 11.2 ± 3.2 years, $p < 0.001$). Subjects who underwent fenestration were more likely to have had prior stage II procedures, (84% vs. 54%, $p = 0.005$), have significant pre-existing atrioventricular valve (AV) regurgitation (40% vs. 24%, $p < 0.001$), and have had a history of thrombosis before the Fontan procedure (6% vs. 2%, $p = 0.04$) than those who did not undergo fenestration. At the pre-Fontan catheterization procedure, subjects who went on to have a fenestration had higher systemic arterial oxygen saturations (85 ± 5% vs. 83 ± 5%, $p < 0.001$) and lower ventricular end diastolic pressures (7.6 mm Hg vs. 8.3 mm Hg, $p = 0.02$) compared with those who did not undergo fenestration. When adjusted for center, age at enrollment, and year of Fontan procedure, there was no difference in numbers of surgical procedures and catheter interventions before Fontan completion between subjects who did and did not undergo surgical fenestration.

Consistent with the association with age and era, surgical fenestration was related to the type of Fontan procedure performed; fenestration was more prevalent among subjects with total cavopulmonary connections and less prevalent among subjects on whom an atriopulmonary connection was performed.

Status after Fontan procedure. Subjects with surgical fenestration had median length of stay approximately 2.5 days shorter than subjects without fenestration ($p < 0.001$) (Fig. 2). This association remained significant after adjusting for study center, age at enrollment, year of Fontan, and performance of a stage II procedure ($p = 0.05$).

Medical history during and after the Fontan hospitalization is shown in Table 2. After adjusting for age, year at Fontan, and performance of a stage II procedure, we found associations between surgical fenestration and increased number of discharge medications and increased number of post-Fontan catheter interventions (excluding procedures related to the fenestration itself). The 2 most common catheter interventions performed in the fenestrated group were coiling of systemic venous collaterals and coiling of aortopulmonary collaterals.

Fenestration was not associated with post-Fontan stroke, ventricular dysfunction, thrombosis, protein-losing enteropathy (PLE), or arrhythmia, even when the analyses were

Table 1 **Patient Characteristics by Surgical Fenestration Status**

	Fenestration (n = 361)	No Fenestration (n = 175)	p Value
Demographics			
Age at study enrollment, yrs	11.2 ± 3.2	13.2 ± 3.6	<0.001
Age at Fontan, yrs	3.5 ± 2.3	3.3 ± 1.8	0.4
Male	60	58	0.6
Medical history before Fontan procedure			
Anatomy			0.3
Double inlet left ventricle	13	18	
Mitral atresia	6	5	
Tricuspid atresia	21	26	
Unbalanced atrioventricular canal	4	5	
Heterotaxy	8	7	
Hypoplastic left heart syndrome	23	15	0.06
Other	26	24	
Stage II performed	84	54	0.005*
Prior surgical procedures	3.3 ± 2.0	2.5 ± 1.7	0.3*
Prior non-stage II surgeries	2.4 ± 1.9	1.9± 1.5	0.3*
Prior catheter interventions	0.9 ± 1.2	0.4 ± 0.7	0.2*
Severity of AV valve regurgitation			<0.001
None/trivial	60	76	
Mild	33	23	
Moderate/severe	7	1	
EDP, mm Hg	7.6 ± 3.4	8.3 ± 3.0	0.02
Mean PA pressure, mm Hg	11.2 ± 3.1	11.8 ± 3.9	0.1
Oxygen saturation, %	84.6 ± 4.9	82.8 ± 5.3	<0.001
Decreased ventricular function	18	17	0.9
Arrhythmia	20	24	0.3
Thrombosis	6	2	0.04
Medical history: Fontan procedure			
Weight for age at Fontan, percentile	26 ± 28	28 ± 27	0.5
Height for age at Fontan, percentile	32 ± 29	32 ± 28	0.8
Type of Fontan			<0.001
AP connection	6	30	
TCPC intracardiac tunnel	65	45	
TCPC extracardiac tunnel	14	12	
TCPC extracardiac conduit	15	8	
Other	1	5	

Data are shown as mean ± SD or %. *Adjusted for center, age at enrollment, and year of Fontan procedure.

AP = atriopulmonary; AV = atrioventricular; EDP = end-diastolic pressure; LV = left ventricle; PA = pulmonary artery; RV = right ventricle; TCPC = total cavopulmonary connection.

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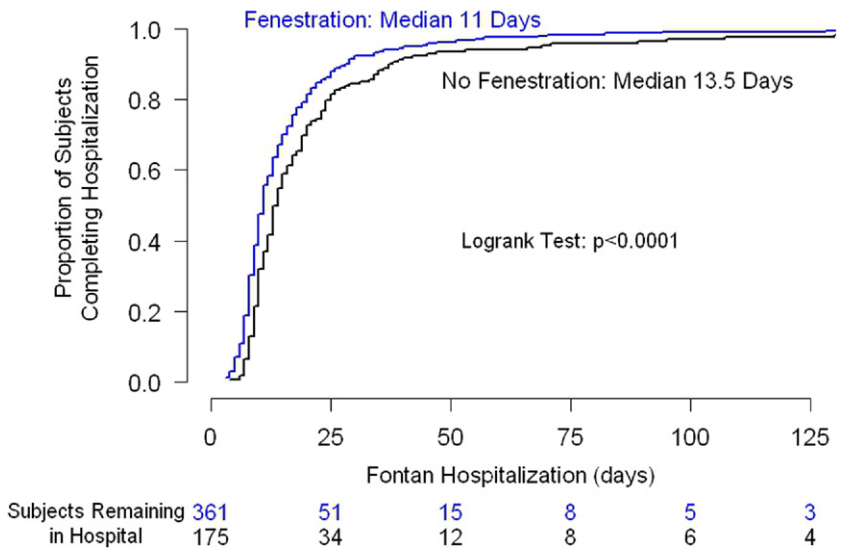

Figure 2 **Proportion of Subjects Achieving Hospital Discharge After the Fontan Procedure as a Function of Time Since the Start of the Fontan Hospitalization**

Proportion estimates were obtained using the Kaplan-Meier method in this cohort of Fontan survivors enrolled in this cross-sectional study.

adjusted for age at enrollment, age at Fontan, center, and performance of a stage II procedure.

Fate of the surgical fenestration. An echocardiographic core laboratory determined "current" fenestration status at study enrollment (a median 8.1 years after Fontan surgery) in 527 of the 536 subjects (98%). Of the 361 subjects who had surgical fenestration at Fontan, 356 had an available echocardiogram (99%). At the time of cross-sectional testing, the fenestration was open in 69 subjects (19%) and closed in 227 (63%). It was not possible to clearly delineate the fenestration status in 60 (17%) subjects.

In the 227 subjects who had a closed fenestration at study enrollment, 91 (40%) are assumed to have had spontaneous closure because no surgical procedures or catheter interventions were performed after the Fontan procedure. Of those subjects with an intervention, 133 (59%) of these underwent closure by catheter intervention, and 3 (1%) underwent closure during a surgical procedure. For subjects who

Table 2 **Status After Fontan Surgery**

Characteristics	Fenestration (n = 361)	No Fenestration (n = 175)	p Value	Adjusted p Value*
Length of hospital stay, days	16.1 ± 20.1	21.9 ± 26.0	<0.001	0.05
Median	11.0	13.5		
Post-operative complications, n	1.4 ± 1.9	2.8 ± 2.6	<0.001	0.3
Median	1	2		
Pericardial effusion	7	21	<0.001	0.3
Pleural effusion	21	35	<0.001	0.3
Discharge medications, n	4 ± 1	3 ± 2	0.003	0.02
Median	4	3		
Antithrombotic	77	50	<0.001	0.02
ACEI	67	24	<0.001	<0.001
Diuretics	96	91	0.02	0.3
Post-Fontan				
Catheter interventions (excluding fenestration-related procedures), n	0.6 ± 1.0	0.3 ± 0.9	0.001	0.03
No. of surgeries	0.3 ± 0.8	0.5 ± 1.1	0.2	0.2
Stroke	2	1	0.5	0.3
Thrombosis	7	7	0.9	0.8
PLE	3	6	0.1	0.2
Arrhythmia	19	24	0.2	0.5

Values are mean ± SD or %. *Adjusted for center, year of Fontan procedure, age at enrollment, and performance of stage II procedure.
ACEI = angiotensin-converting enzyme inhibition; PLE = protein-losing enteropathy.

underwent fenestration closure, the mean interval between Fontan surgery and interventional closure was 8.5 ± 3.5 years (range 1 day to 11 years). Considering only the catheter-related fenestration closures, 116 were closed by a septal occluder device, 3 by a coil, and 14 by closure of a purse string placed around the fenestration at the time of Fontan surgery.

Comparisons of subjects according to current fenestration status. Comparisons of the 69 subjects who had an open fenestration at study enrollment with the 458 subjects without any atrial-level communication assessed by study echocardiogram are shown in Table 3. The group with a current fenestration was younger and had been followed for less time since Fontan (mean 7.1 years vs. 8.7 years, $p < 0.001$). Those with a current fenestration were more likely to have had a prior stage II procedure and to have an intra-cardiac lateral tunnel-type Fontan. Patients with a current fenestration had lower resting oxygen saturation than those without a current fenestration (89% vs. 95%, $p < 0.001$). The 2 groups did not differ with respect to functional health assessed by CHQ P50 summary scores. Height and weight percentile calculated at the time of cross-sectional testing did not differ by fenestration status. BNP levels, angiographic and echocardiographic measures of ventricular performance, and exercise variables did not differ by current fenestration status, nor did they differ in respect to post-Fontan complications, including number of additional interventional procedures, stroke, thrombosis, or PLE.

Table 3 Subject Profiles by Current Fenestration Status

	Fenestration (n = 69)	No Fenestration Detected (n = 458)	p Value
Age at enrollment, yrs	10.7 ± 3.3	12.0 ± 3.4	0.002
Age at Fontan, yrs	3.7 ± 2.3	3.4 ± 2.1	0.3
Years since Fontan	7.1 ± 3.2	8.7 ± 3.5	<0.001
Male	61	59	0.9
Stage II performed	93	80	0.005*
Anatomy			0.3
Double inlet left ventricle	12	15	
Mitral atresia	7	5	
Tricuspid atresia	13	24	
Unbalanced atrioventricular canal	3	4	
Heterotaxy	9	7	
Hypoplastic left heart syndrome	26	19	
Other	30	25	
Type of Fontan			0.001
AP connection	0	16	
TCPC intracardiac tunnel	80	56	
TCPC extracardiac tunnel lateral	9	14	
TCPC extracardiac conduit	10	13	
Other	1	2	
Post-Fontan medical history			
Weight for age at study, percentile	29 ± 28	35 ± 31	0.2
Height for age at study, percentile	36 ± 30	42 ± 33	0.2
Cardiac surgical procedures	0.3 ± 0.6	0.4 ± 1.0	0.8*
Catheter interventions	0.5 ± 1.2	0.5 ± 1.0	0.3*
Stroke	0	2	1.0*
Thrombosis	4	8	0.3*
PLE	3	4	1.0*
Arrhythmia	21	20	0.5*
Functional health assessed by CHQ P50			
Physical summary score	46 ± 10	45 ± 12	0.4*
Psychosocial summary score	46 ± 11	48 ± 11	0.8*
Echocardiography			
Echo EF, %	58 ± 10	59 ± 10	0.6*
Mass:volume ratio z score	1.2 ± 0.5	1.2 ± 0.4	0.5*
dP/dt, mm Hg/s	1,575 ± 1,026	1,374 ± 908	0.3*
Exercise testing			
Resting O_2 saturation (%)	89 ± 5	95 ± 4	<0.001*
Peak VO_2 (ml/kg/min)	25 ± 6	27 ± 6	0.2*
% Predicted peak VO_2	63 ± 14	67 ± 15	0.1*
VO_2 at VAT (ml/kg/min)	16 ± 4	18 ± 6	0.1*
% Predicted VO_2 at VAT	67 ± 14	77 ± 22	0.1*
BNP (pg/ml)	34 ± 72	25 ± 44	1.0*

Data are shown as mean ± SD or %. *Age-adjusted comparisons.

AP = atriopulmonary; BNP = B-type natriuretic peptide; CHQ P50 = Parent Report Child Health Questionnaire; EDV = end-diastolic volume; EF = ejection fraction; ESV = end-systolic volume; LV = left ventricle; PLE = protein-losing enteropathy; RV = right ventricle; TCPC = total cavopulmonary connection; VAT = ventilatory anaerobic threshold; VO_2 = maximal oxygen consumption.

Discussion

We report a large multicenter cohort of single-ventricle patients palliated with a Fontan procedure and demonstrate a dramatic change in surgical practice over time in the use of a surgical fenestration. Fenestration was first introduced in 1990 in an effort to improve the perioperative course for high-risk patients (2); more than 80% of subjects in our cohort had an atrial fenestration at the time of the Fontan operation after 1992. The population evaluated in this cross-sectional study included patients in the pre-fenestration era. Thus, the majority of subjects who did not have a fenestration were from the early time period. They were also somewhat older at the time of evaluation, less likely to have had a stage II operation performed, and more likely to have had an atriopulmonary connection, which was the methodology of the original Fontan operation (9). In the current study, we attempted to control for era effect by adjusting for patient age and year of Fontan and to control for practice variation by adjusting for study center. After adjusting for these factors, surgical fenestration was associated with decreased length of Fontan hospital stay. This cross-sectional study found few associations between a persistent fenestration and deleterious later outcomes.

Pre-operative risk factors do not consistently identify patients who may have poorer outcomes after the Fontan procedure, which has led some to suggest that some patients

undergoing the Fontan do not require routine fenestration (10,11). This large multicenter study supports the numerous single-center reports indicating that an atrial fenestration carried out at the time of the Fontan operation shortens hospital stay (3,4,12). Not appreciated in prior studies, we found an increased use of cardiac medications at discharge in the fenestration group. After adjusting for subject age and center, increased use of angiotensin-converting enzyme inhibitors and antithrombotics in the fenestration group was still observed.

Although considerable data regarding the early postoperative effects of a fenestration at Fontan have been previously reported, the long-term sequelae of a fenestration on clinical outcomes are less well studied. We found that subjects with a history of surgical fenestration at the time of the Fontan operation had significantly greater numbers of catheter interventions, even after excluding procedures carried out for modification of the fenestration itself. One might attribute this finding to the modern era when interventions are more common. However, after correcting for patent age, year of Fontan procedure, and center, we continued to identify a greater number of post-Fontan catheter interventions in the group who underwent surgical fenestration.

Closure of a surgical fenestration has been advocated because of complications associated with persistent cyanosis and increased risk of systemic thromboembolism. As expected, we found that resting arterial oxygen saturation was significantly lower among the 69 subjects who were documented to have a persistent fenestration at a median of 8 years after the Fontan procedure. However, an increased incidence of either stroke or thrombosis was not found among the patients with a persistent atrial communication. This is consistent with smaller single-center reports that did not find a difference in number of strokes (13) or occurrence of thrombosis (12) in Fontan subjects with patent fenestrations. Additionally, we found no differences in the number of surgical or catheter interventions, incidence of PLE, or incidence of arrhythmia between those with a current fenestration and those without. Echocardiographic measurements of ventricular function and mass-to-volume ratio as well as exercise performance, BNP levels, and growth parameters were similar in the 2 groups. Our findings are consistent with previous reports demonstrating that fenestration closure does not positively or negatively affect exercise capacity (14). Functional health status as assessed by the CHQ_P50 also did not differ by current fenestration status. Overall, despite lower resting oxygen saturation, we did not identify any deleterious effects of a persistent fenestration in our cohort.

The long-term fate of the surgical fenestration has not been well described. In our cohort, 40% of subjects with a surgical fenestration had apparent spontaneous closure. Although the timing of such spontaneous closure is unknown, this finding is consistent with prior small reports that have documented spontaneous closure of fenestrations in 29% of subjects within 3 months (15). Multiple centers have reported methods for closing fenestration. Indications for the procedure have also been proposed, and risk factors associated with closure have been described (16,17). Closure of a fenestration improves oxygenation and may reduce the use of anti-congestive medication, but may result in increased use of antiarrhythmic agents after closure (17). Some have suggested that indications for routine fenestration closure should be reconsidered because of potential long-term benefits of increased cardiac output and decreased tachyarrhythmias in patients with patent fenestrations late after Fontan procedures (18).

Study limitations. The study cohort was limited to surviving subjects who agreed to participate in this study. The fates of patients who were not enrolled are necessarily unknown. Furthermore, the effects of fenestration and of interventional closure on early or late mortality, if any, could not be assessed in a cross-sectional study. We utilized a central echocardiography core laboratory to determine current fenestration status, but images were not available in 19 subjects (3%) and were inadequate to determine the status of the fenestration in 60 subjects (17%). This may have limited the power to detect differences between those with and without a current fenestration.

Conclusions

The percentage of Fontan procedures that included surgically created atrial fenestrations increased rapidly over time in subjects enrolled in this multicenter study. Subjects who had surgical fenestration creation at Fontan had a shorter length of stay, but received more medications at hospital discharge. Most fenestrations were later closed in the catheterization laboratory; however, a significant percentage demonstrated spontaneous closure. Subjects with persistent fenestrations at the time of the cross-sectional study had significantly lower resting oxygen saturation. A persistent fenestration, however, was not associated with exercise performance, echocardiographic function, or functional health status. Although patients with a current fenestration are theoretically at ongoing risk for thromboembolic events, there was not an increased incidence of these potentially serious complications during the 8-year follow-up period. Further longitudinal follow-up of this cohort is necessary to determine long-term consequences of a persistent fenestration.

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Key Words: cardiac surgery ■ congenital heart disease ■ Fontan procedure.