Use of institutional criteria for transcatheter device closure of Fontan fenestration – Midterm outcomes

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ABSTRACT

Background : **There are no established criteria to decide suitability for Fontan fenestration closure. Our institution has the following criteria: an unobstructed Fontan pathway with no significant decompressing venovenous collaterals, baseline Fontan pressure ≤15 mmHg, baseline cardiac index ≥2 L/min/m², and a decrease in cardiac index ≤20% with test occlusion of the fenestration.**

Objective : **The objective of the study was to review midterm outcomes following device closure of Fontan fenestration using institutional criteria.**

Materials and Methods : **A retrospective review was performed of patients who underwent catheterization with prior fenestrated Fontan procedure between May 2005 and January 2015. Patients were classified as those who underwent successful closure (A), had closure deferred due to failure to meet criteria (B), or were not referred for closure (C).**

Results: : **There were 42 patients in Group A, 10 in Group B, and 150 in Group C. The mean Fontan pressure increased from 13.1 ± 2.1 to 14.5 ± 2.1mmHg in Group A and 14.6 ± 1.5 to 15.7 ± 2.2 mmHg in Group B (*P* = not significant). With test occlusion, cardiac index fell by 18.12% ± 15.68% in Group A and 33.75% ± 14.98% in Group B (*P* = 0.019). At a median of 46 month follow-up, oxygen saturation increased significantly from 85.15% ± 6.29% at baseline to 94.6% ± 4.43% (*P* < 0.001) in Group A but with no statistically significant difference in the rates of plastic bronchitis, protein-losing enteropathy, stroke, or heart transplantation between the three groups.**

Conclusions : **Using institutional criteria, transcatheter device closure of Fontan fenestration was followed by significant increase in oxygen saturations and no statistically significant difference in morbidity or mortality between closure and nonclosure groups.**

Keywords : Catheterization, congenital, interventional devices, percutaneous

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.APC_154_19

How to cite this article: Thatte N, Dimas V, Nugent A, Zellers T, Forbess J, Zabala L, *et al*. Use of institutional criteria for transcatheter device closure of Fontan fenestration – Midterm outcomes. Ann Pediatr Card 2020;13:327-33.

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Submitted: 08-Aug-2020 **Revised:** 25-Mar-2020 **Accepted:** 23-Jul-2020 **Published:** 21-Sep-2020

INTRODUCTION

The Fontan procedure[1] has become the conventional palliative operation in patients with single-ventricle congenital heart disease. Creation of a fenestration in the Fontan pathway allowing right-to-left shunting of blood was a novel modification which has been successful in decreasing the incidence of acute postoperative pleural effusions and low cardiac output state.[2-4] However, this benefit comes at the cost of long-term reduced systemic oxygen saturations and a theoretical increased risk of stroke due to the iatrogenically created right-to-left shunt. Once the fenestration has outlived its utility, there is marked practice variation in deciding candidacy for transcatheter fenestration device closure. The indications and timing for such closure still remain a matter of debate.[5-7] For the subset of fenestrated Fontan patients who were taken to the catheterization laboratory for an attempted closure, our institution has used a set of criteria to decide suitability for closure. The objectives of this study were to review midterm outcomes in our cohort of patients who underwent attempted Fontan fenestration closure based on our catheter-based patient selection criteria and to determine if there are any precatheterization clinical characteristics that predict failure to meet these criteria.

MATERIALS AND METHODS

Study population

The study design was reviewed and approved by the Institutional Review Board at our institution. All patients who underwent a fenestrated Fontan procedure at our institution between May 2005 and January 2015 were included in a retrospective chart review. It has been our institutional practice to place a fenestration in all patients undergoing Fontan procedure unless there existed a specific reason to avoid fenestration such as the presence of pulmonary arteriovenous malformations. Patients who underwent a trial of fenestration closure in the catheterization laboratory were further classified into two groups: those who underwent successful closure (Group A) and those in whom closure was deferred due to failure to meet hemodynamic or other parameters as noted below (Group B). Fenestrated Fontan patients who were not referred for catheterization and/or did not undergo any attempt at closure were included as controls (Group C).

Criteria to decide suitability for closure

The following criteria were assessed to determine suitability for fenestration closure:

(1) anatomically unobstructed Fontan pathway with no significant decompressing systemic to pulmonary veno-venous collaterals; (2) baseline Fontan pressure ≤15 mmHg; (3) baseline cardiac index ≥2 L/min/m^2; and (4) decrease in cardiac index ≤20% from baseline with test occlusion of the Fontan fenestration.

Data collection

Baseline demographic data were collected for all groups. Single-ventricle anatomy type was classified as left ventricle or right ventricle morphology. Surgical data included the type of Fontan pathway (extracardiac conduit or lateral tunnel). For the patients who had an attempted intervention, baseline clinical data were collected from a precatheterization clinic visit. This included a symptomatic assessment of exercise capacity (coded as presence or absence of exercise intolerance by reviewing the clinic visit documentation), oxygen saturation by pulse oximetry, and the following echocardiographic findings: estimated Fontan fenestration gradient, atrioventricular (AV) and semilunar valve regurgitation, and qualitative ventricular function. For all patients, these data were also collected from their latest follow-up clinic visit. Outcomes data collected included mortality or heart transplantation, need for subsequent device removal or recreation of fenestration, prevalence of cerebrovascular accidents, protein-losing enteropathy (PLE), plastic bronchitis, and pacemaker status.

For patients in Groups A and B, catheterization data included pressures and oxygen saturations at baseline and at test occlusion of the fenestration or postintervention as detailed below. If multiple Fontan pathway pressures were noted in the catheterization report post-test occlusion, the highest pressure was included for this analysis. The type of device used to close the fenestration was documented.

Fenestration occlusion procedural details

Written informed consent was obtained from all patients undergoing attempted closure. The procedure was performed under general anesthesia and endotracheal intubation with baseline hemodynamics obtained in room air. Hemodynamic and oxygen saturation measurements at baseline were taken from the right and left heart circulations. The cardiac index was calculated using the Fick principle. Selective angiograms were performed in the left innominate vein, right superior vena cava, Fontan pathway, and at the inferior vena cava to Fontan junction to assess for patency of the entire Fontan circuit and branch pulmonary arteries and to check for the presence of systemic to pulmonary veno-venous collaterals. A balloon-tipped catheter was then advanced through the fenestration into the pulmonary venous atrium. A test occlusion of the fenestration was performed. Oxygen saturations and hemodynamic measurements were repeated in the Fontan pathway, superior vena cava, and aorta after 10 minutes of test occlusion. If the parameters for suitability for closure were met, then the fenestration was closed. All children received antibiotic

prophylaxis and heparin anticoagulation per institutional protocol. Patients were admitted to the cardiology ward for overnight observation and discharged the following morning. Post-device closure, patients resumed routine anticoagulation per their primary cardiologist's preference, typically monotherapy with aspirin.

Statistical analysis

The results are reported as mean ± standard deviation or median (interquartile range [IQR]) for continuous variables and frequencies and percentages for categorical variables. Descriptive information and outcomes data for the three groups were analyzed using the Fisher's exact test or Chi-square test for categorical variables and unpaired *t*-test or Wilcoxon signed-rank test for continuous variables. Echocardiographic variables were divided into two dichotomous groups for severity of valve regurgitation and severity of ventricular dysfunction as follows: AV valve and semilunar valve regurgitation was classified as none-to-mild and moderate-to-severe regurgitation and qualitative ventricular dysfunction was classified as none-to-mild and moderate-to-severe dysfunction. Catheterization laboratory hemodynamic and saturation data for baseline and post-test occlusion conditions were compared using paired *t*-test or Wilcoxon signed-rank test as appropriate. Statistical significance for the major clinical outcomes such as mortality and strokes was tested by grouping together patients in Groups B and C and comparing against Group A. The intention was to compare major outcomes for patients who underwent device closure of the fenestration (Group A) against all other patients. All data were analyzed using standard statistical software (SAS Institute Inc., Cary, NC, USA).

RESULTS

A total of 203 patients who underwent fenestrated Fontan procedures between May 2005 and January 2015 were included in the review. The median length of follow-up was 46.73 (24.2–90.95) months post-catheterization in Group A, 26.5 (12.67–54.1) months post-catheterization in Group B, and 30.2 (1.66–64.88) months post-Fontan surgery in Group C. One patient underwent an attempted closure that was deferred but was subsequently taken back to the catheterization laboratory 2 years later and had successful closure of the fenestration. This patient was included in both Groups A and B with pre-catheterization data referring to the last clinic visit prior to each catheterization. One patient had their Fontan fenestration closed during a surgical AV valve repair procedure and was not included in this analysis. This patient went on to undergo heart transplantation due to recurrent severe AV valve regurgitation and heart failure. One patient who moved out of state immediately after their Fontan operation was lost to follow-up and was excluded.

Patient characteristics

Table 1 describes the baseline characteristics for all patients. For patients in Groups A and B, further details are provided about their baseline clinical condition from a pre-catheterization clinic visit [Table 2]. There were 42 patients in Group A, 10 patients in Group B, and 150 patients in

Table 1: Patient characteristics

	Group A (*n*=42), *n* (%)	Group B (*n*=10), *n* (%)	Group C (*n*=150), *n* (%)	*P*
Male sex	27 (64.29)	8 (80)	84 (56)	NS
Age at Fontan, years	2.98±0.71	3.88±2.21	3.82±1.67	<0.001 (A vs. B). Others NS
Age at fenestration closure, years	5.81±2.11	7.97±3.67	N/A	NS
Interval between Fontan and closure, months	28.03 (14.98-46.17)	59.97 (23.57-90.2)	N/A	NS
Fontan type				
Extracardiac	21 (50)	7 (70)	100 (66.67)	NS
Lateral tunnel	21 (50)	3 (30)	50 (33.33)	NS
Single-ventricle morphology				
LV type	19 (45.24)	5 (50)	61 (40.67)	NS
RV type	23 (54.76)	5 (50)	89 (59.33)	NS

Values are mean±SD, median (IQR) or number of patients (%). *P* values indicate all intergroup comparisons unless specifically indicated. NS: Not significant, LV: Left ventricle, RV: Right ventricle, IQR: Interquartile range, SD: Standard deviation

Table 2: Baseline clinical characteristics of patients undergoing catheterization

	Group A (*n*=42), *n* (%)	Group B (*n*=10), *n* (%)	*P*
Exercise intolerance, *n*	11/36 (30.56)	3/10 (30)	NS
O_2 saturation, %	85.1±6.2	90.7±4.11	0.0113
Fenestration gradient by echocardiography, mmHg	5.58±2.4	6.7±1.08	NS
Moderate-to-severe AV valve regurgitation	3/36 (8.33)	2/8 (25)	NS
Moderate-to-severe semilunar valve regurgitation	2/32 (6.25)	0/8 (0)	NS
Moderate-to-severe ventricular dysfunction	0/35 (0)	1/8 (12.5)	NS

Values are mean±SD, or number of patients (%). NS: Not significant, AV: Atrioventricular, SD: Standard deviation

Group C. Among patients in Group C, 12/150 (8%) patients were noted to have spontaneous closure of the fenestration. Pre-catheterization oxygen saturation by pulse oximetry and age at Fontan operation were the only clinical variables that were significantly different at baseline between Groups A and B. Patients in Group A had lower baseline oxygen saturation (85.15% ± 6.29% vs. 90.7% ± 4.11%, $P < 0.05$) and were younger at the time of Fontan operation (2.98 ± 0.71 years vs. 3.88 ± 2.21 years, $P < 0.001$) compared to patients in Group B, although there was no statistically significant difference in age at attempted fenestration closure between the two groups. Transthoracic echocardiography demonstrated a Fontan fenestration gradient of 5.58 ± 2.40 mmHg in Group A and 6.7 ± 1.08 mmHg in Group B (P = not significant [NS]). No statistically significant difference was found with respect to AV valve and semilunar valve regurgitation and qualitative ventricular function at baseline between Groups A and B.

Data from fenestration closure procedure

The procedure was performed at a median 28.03 months (IQR 14.98–46.17 months) after the Fontan operation in Group A and at a median 59.97 months (IQR 23.57–90.2 months, P = NS) in Group B. A variety of devices were utilized for closure, the majority being Amplatzer™ Septal Occluder devices (22/42). Covered stents were used if a baffle leak was noted (Covered Cheatham Platinum™ Stents n = 4; iCAST™ Covered Stent n = 1). Arterial oxygen saturation increased with test occlusion from 85.70% ± 5.67% to 94.89% ± 2.95% ($P < 0.0001$) in Group A and from 87.35% ± 7.25% to 93.7% ± 3.27% ($P < 0.01$) in Group B [Table 3]. The mean Fontan pathway pressure increased with test occlusion from 13.15 ± 2.17 mmHg to 14.46 ± 2.15 ($P < 0.001$) in Group A and from 14.56 ± 1.51 mmHg to 15.75 ± 2.25 mmHg (P = NS) in Group B. There was no statistically significant difference between the baseline or post-test occlusion Fontan pathway pressures between Groups A and B. In Group A, the cardiac index fell from 3.77 ± 1.09 L/min/m^2 at baseline to 2.88 ± 0.52 L/min/m^2 with test occlusion ($P < 0.001$). In Group B, cardiac index fell from 3.24 ± 1.01 L/min/m^2 at baseline to 2.08 ± 0.9 L/min/m^2 with test occlusion ($P < 0.01$). The average percentage fall in cardiac index with test occlusion was 18.12% ± 15.68% in Group A, while it was significantly larger at 33.75% ± 14.98% in Group B ($P = 0.019$).

Procedural complications

Complications at the time of implantation were rare (2/52 cases). One patient developed transient heart block during long sheath positioning which resolved without intervention within a few seconds. One patient developed a worsening of their baseline junctional rhythm to junctional bradycardia with rate 30-35 beats per minute overnight. They were eventually brought back for permanent pacemaker placement 6 months after fenestration closure.

Follow-up clinical data

A total of 21 patients were lost to follow-up from the complete initial cohort of 203. Clinical characteristics at follow-up are summarized in Table 4. Outcomes data are presented in Table 5. Oxygen saturations by pulse oximetry rose from 85.15% ± 6.29% at baseline to 94.6% ± 4.43% ($P < 0.001$) at last follow-up in Group A, whereas it stayed unchanged from 90.7% ± 4.11% at baseline to 91.67% ± 1.97% (P = NS) at last follow-up in

Table 3: Hemodynamic data from cardiac catheterization for attempted fenestration closure

	Group A (n=42)	Group B (n=10)	P (Group A vs. Group B)
Aortic saturation (%), baseline	85.70±5.67	87.35±7.25	NS
Aortic saturation (%), test occlusion	94.89±2.95*	93.7±3.27*	NS
Qp:Qs, baseline	0.70±0.14	0.75±0.18	NS
Qp:Qs, test occlusion	0.96±0.07*	0.96±0.07*	NS
Fontan mean pressure (mmHg), baseline	13.15±2.17	14.56±1.51	NS
Fontan mean pressure (mmHg), test occlusion	14.46±2.15*	15.75±2.25	NS
Cardiac index (L/min/m^2), baseline	3.77±1.09	3.24±1.01	NS
Fall in cardiac index with test occlusion (%)	18.12±15.68	33.75±14.98	0.02

Values are mean±SD. *P<0.05 for within-group comparison of baseline against test-occlusion values. NS: Not significant. Qp:Qs: Pulmonary-to-systemic blood flow ratio. SD: Standard deviation

Table 4: Clinical characteristics of patients at the last follow-up

	Group A	Group B	Group C	P
Exercise intolerance, n (%)	8/38 (21.05)	2/7 (28.57)	29/124 (23.39)	NS
O_2 saturation, %	94.59±4.43	91.67±1.97	90.84±5.53	<0.001 (A vs. B and A vs. C); NS (B vs. C)
Fenestration gradient by echocardiography, mmHg	N/A	6.3±1.89	5.64±1.75	NS
Moderate-to-severe AV valve regurgitation, n (%)	3/37 (8.11)	0/6 (0)	19/123 (15.45)	NS
Moderate-to-severe semilunar valve regurgitation, n (%)	0/33 (0)	0/6 (0)	2/113 (1.77)	NS
Moderate-to-severe ventricular dysfunction, n (%)	2/36 (5.56)	0/6 (0)	20/126 (15.87)	NS

Values are mean±SD, or number of patients (%). NS: Not significant, AV: Atrioventricular, SD: Standard deviation

Table 5: Clinical outcomes

	Group A	Group B	Group C	*P**
Follow-up since catheterization, months	46.73 (24.2-90.95)	26.5 (12.67-54.1)	30.2 (1.66-64.88)†	NS
Death or transplantation	1/40 (2.5)	0/10 (0)	12/149 (8.05)	NS
PLE	2/39 (5.13)	0/10 (0)	7/148 (4.73)	NS
Stroke	5/39 (12.82)‡	1/10 (10)	11/149 (7.38)	NS
Plastic bronchitis	1/39 (2.56)	0/10 (0)	2/149 (1.34)	NS
Pacemaker placement	5/40 (12.5)§	3/10 (30)	9/150 (6)	NS

Values are median (IQR), or number of patients (%). **P* values are for comparisons between Group A and pooled patients in Groups B and C, †Follow-up period for Group C patients indicates period since Fontan operation, ‡5 patients had strokes in Group A at the last follow-up, but four of these patients had strokes that had occurred prior to device closure. Only one stroke occurred after device closure, which occurred 2 days after the patient underwent ventricular assist device placement, §Five patients had permanent pacemakers in Group A at the last follow-up, but three of these patients already had a pacemaker in place prior to the device closure procedure. Only two pacemakers were placed after device closure. NS: Not significant. PLE: Protein-losing enteropathy, IQR: Interquartile range

Group B. Oxygen saturation at follow-up was significantly higher among Group A patients compared to those in Group B or Group C (90.88% ± 5.58%, $P < 0.001$ for Group A vs. Group B and Group A vs. Group C).

There were one transplant and no deaths in Group A, no transplants or deaths in Group B, and four transplants and ten deaths in Group C (P = NS for all comparisons). There was no statistically significant difference in the rates of PLE, plastic bronchitis, stroke, and pacemaker placement between the patients in the three study groups. Five patients in Group A had strokes at the last follow-up, but four of the five had suffered strokes prior to the fenestration closure procedure. Only one patient suffered a stroke post-device closure, which occurred 2 days following a ventricular assist device (VAD) placement. Besides this patient, there were no new or recurrent strokes in Group A. Five patients in Group A had permanent pacemakers in place at the last follow-up, but three of these patients already had pacemakers in place prior to the device closure procedure with only 2 new pacemaker placements in Group A after device closure.

Adverse outcomes requiring device removal

One patient required removal of a fenestration closure device. The patient is a 3-year-old male with hypoplastic left heart syndrome status post staged palliation to a lateral tunnel fenestrated Fontan completed at 2 years of age. He underwent uncomplicated device closure of the fenestration 16 months after the Fontan procedure. About 6 weeks after fenestration closure, he developed clinical signs and symptoms consistent with PLE. Following medical management for about 3 months, he was brought back to the catheterization laboratory. This catheterization showed pulmonary-to-systemic blood flow ratio of 1 with elevated Fontan pathway mean pressures at 22 mmHg and normal pulmonary capillary wedge pressure of 9–10 mmHg. The fenestration closure device (18 mm Amplatzer™ Cribriform Occluder) was removed via a 12-French 80 cm Amplatzer™ Torque Vue sheath, using a 7-French MPA catheter with a 25 mm snare and the residual fenestration was balloon dilated. At the last follow-up 4 years after device removal, the patient is doing well with resolution of PLE and plastic bronchitis symptoms and most recent echocardiogram showing a patent fenestration.

DISCUSSION

Transcatheter closure of a surgically created fenestration in Fontan pathways has been shown to be safe and effective in raising arterial oxygen saturations.[8-11] However, the optimal timing and indications for such closure remain a matter of debate.[7] The benefits of device closure are increased oxygen saturation, prevention of exercise-induced desaturation, and theoretical decrease in stroke risk. However, this latter benefit has not been demonstrated in cross-sectional studies to date.[5,12] Fenestration closure does not acutely improve nor worsen exercise capacity.[13] Occluding a patent fenestration leads to loss of a "pop-off" on the venous limb of the single-ventricle Fontan circuit which leads to increased venous congestion, especially notable in the splanchnic and hepatic venous beds. This raises the concern for adverse outcomes such as the poorly understood PLE and hepatic fibrosis.

Given these conflicting risks and benefits of fenestration closure, optimal patient selection for transcatheter intervention is of the utmost importance. Prior to catheterization, it is difficult to predict which patients will have suitable hemodynamics for fenestration closure, and there is a lack of consensus to guide patient selection or intraprocedural decision-making. At our institution, we have evolved a set of parameters that are founded in physiologic principles to determine which patients should be considered suitable candidates for fenestration closure. A complete anatomic survey showing patency of the entire Fontan pathway and ruling out other sources of right-to-left shunting is essential. Test occlusion of the fenestration provides valuable hemodynamic details. Occluding a fenestration in a Fontan pathway leads to several physiologic changes. The most striking immediate impact is a rise in arterial oxygen saturation accompanied by a fall in mixed venous oxygen saturation (due to acute loss of preload to the ventricle along with a slight rise in central venous

pressure). The net result, in the acute setting, is a fall in systemic oxygen delivery (as the rise in arterial saturation is not adequate to compensate for the acute fall in cardiac output). However, over the course of hours to days, the circulation achieves a new homeostasis with changes in vascular tone compensating for some of the acutely lost preload to the ventricle. Patients who experience a large fall in cardiac index with test occlusion (which we have arbitrarily chosen to be a fall of >20%) are felt to be bad candidates for closure of the fenestration as they will struggle to achieve a new homeostasis without significant elevation in central venous pressure. A previously reported cutoff by Bridges *et al.*[11] was a fall in the cardiac index of >33%, which our data are in close agreement with as patients in Group B had a 34% fall in the cardiac index compared to 18% in Group A. We show here that the major difference between the group that underwent successful closure of the fenestration and the group that had an attempted but deferred closure of the fenestration was the degree of fall in cardiac index with test occlusion of the fenestration. The acute hemodynamic outcomes of fenestration occlusion in our study are otherwise in close agreement with prior reports.[11,14,15]

Utilizing our institutional parameters, we were able to safely perform fenestration closure without an increase in midterm adverse outcomes such as pacemaker placement, PLE, plastic bronchitis, or other failure of the Fontan circulation when comparing the successful closure group to both those who had no intervention as well as those who had an attempted but deferred intervention. There were no new or recurrent strokes in our successful closure group, with the exception of one patient that suffered a stroke in the context of a VAD placement, though our data regarding strokes did not reach statistical significance possibly due to the limited sample size and a follow-up period in years rather than decades.

Limitations

This study being retrospective in nature carries all of the drawbacks associated with a review of this type. It is likely that patients who are not doing well clinically are not referred for attempted fenestration closure by their primary cardiologists, introducing a selection bias. One of the goals of this review was to study if the cohort of patients in whom fenestration closure was attempted but deferred in the catheterization laboratory due to failure to meet suitability criteria had any predictive pre-catheterization clinical signs. Although this group had significantly higher oxygen saturation at baseline compared to those who underwent successful closure, we are unable to add any definitive pre-catheterization evaluation recommendations due to the small sample size of this group of patients. A multi-institutional review looking at this particular cohort with attention paid to pre-procedural clinical data may shed light on this question.

CONCLUSIONS

Using predefined institutional criteria, transcatheter device closure of Fontan fenestration was followed by significant increase in oxygen saturations and no significant increase in complications such as PLE or plastic bronchitis between the closure and non-closure groups. The indications and timing of Fontan fenestration closure remain controversial. This retrospective single-institution dataset adds weight to the argument that fenestration closure is not harmful in appropriately selected patients with respect to midterm outcomes such as PLE and plastic bronchitis, with the benefit of improved oxygen saturations and possibly prevention of new or recurrent strokes. Further studies are needed to identify the patients who are most likely to benefit from device closure of patent Fontan fenestrations.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax 1971;26:240-8.
2. Bridges ND, Mayer JE Jr., Lock JE, Jonas RA, Hanley FL, Keane JF, *et al*. Effect of baffle fenestration on outcome of the modified Fontan operation. Circulation 1992;86:1762-9.
3. Lemler MS, Scott WA, Leonard SR, Stromberg D, Ramaciotti C. Fenestration improves clinical outcome of the fontan procedure: A prospective, randomized study. Circulation 2002;105:207-12.
4. Airan B, Sharma R, Choudhary SK, Mohanty SR, Bhan A, Chowdhari UK, *et al*. Univentricular repair: Is routine fenestration justified? Ann Thorac Surg 2000;69:1900-6.
5. Atz AM, Travison TG, McCrindle BW, Mahony L, Quartermain M, Williams RV, *et al*. Late status of Fontan patients with persistent surgical fenestration. J Am Coll Cardiol 2011;57:2437-43.
6. Ono M, Boethig D, Goerler H, Lange M, Westhoff-Bleck M, Breymann T. Clinical outcome of patients 20 years after Fontan operation--effect of fenestration on late morbidity. Eur J Cardiothorac Surg 2006;30:923-9.
7. Hijazi ZM. Extracardiac fenestrated Fontan operation: To close or not to close the fenestration? Catheter Cardiovasc Interv 2001;54:93-4.
8. Al-Qbandi M, Hijazi Z. Device closure of fenestrations post fontan operation. In: Hijazi Z, Feldman T, Mustafa H, Al-Qbandi M, Sievert H, editors. Transcatheter Closure

of ASDs and PFOs: A Comprehensive Assessment. USA: Cardiotext Publishing; 2010. p. 243-9.

9. Goff DA, Blume ED, Gauvreau K, Mayer JE, Lock JE, Jenkins KJ. Clinical outcome of fenestrated Fontan patients after closure: The first 10 years. Circulation 2000;102:2094-9.
10. Bridges ND, Lock JE, Castaneda AR. Baffle fenestration with subsequent transcatheter closure. Modification of the Fontan operation for patients at increased risk. Circulation 1990;82:1681-9.
11. Bridges ND, Lock JE, Mayer JE Jr., Burnett J, Castaneda AR. Cardiac catheterization and test occlusion of the interatrial communication after the fenestrated Fontan operation. J Am Coll Cardiol 1995;25:1712-7.
12. Imielski BR, Woods RK, Mussatto KA, Cao Y, Simpson PM, Tweddell JS. Fontan fenestration closure and event-free survival. J Thorac Cardiovasc Surg 2013;145:183-7.
13. Meadows J, Lang P, Marx G, Rhodes J. Fontan fenestration closure has no acute effect on exercise capacity but improves ventilatory response to exercise. J Am Coll Cardiol 2008;52:108-13.
14. Hijazi ZM, Fahey JT, Kleinman CS, Kopf GS, Hellenbrand WE. Hemodynamic evaluation before and after closure of fenestrated Fontan. An acute study of changes in oxygen delivery. Circulation 1992;86:196-202.
15. Lloyd TR, Rydberg A, Ludomirsky A, Teien DE, Shim D, Beekman RH 3rd, *et al*. Late fenestration closure in the hypoplastic left heart syndrome: Comparison of hemodynamic changes. Am Heart J 1998;136:302-6.