Fontan fenestration closure and event-free survival

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Objectives: The purpose of the present study was to evaluate the association of open and closed Fontan fenestration status with event-free survival.

Methods: All patients who underwent a fenestrated Fontan procedure at our institution from January 1994 through June 2007 were reviewed. Patient information was obtained from the medical records. The patients were assigned to 1 of 2 study groups, open or closed, according to their most recent fenestration status. Clinically relevant morbid events were tabulated, and Kaplan-Meier event analysis was used to create event-free probability curves with log-rank comparisons.

Results: A total of 161 patients were classified as open and 51 as closed. The median interval to an event was 1.1 years (interquartile range, 0.1-3.3 years) after the Fontan procedure. The median interval to closure was 1.2 years (interquartile range, 0.7-3.3 years). The median interval to an event was 1.5 years (interquartile range, 0.1-4.6 years) in the closed group and 1.1 years (interquartile range, 0.1-3.3 years) in the open group. Event-free probability analysis revealed no significant difference between the 2 groups ($P = .15$). The median follow-up arterial oxygen saturation was greater in the closed group (96.0%; interquartile range, 94.0%-97.0%) than in the open group (91.0%; interquartile range, 86.0%-95.0%; $P < .0001$).

Conclusions: Fenestration closure was associated with greater arterial oxygen saturation but not greater event-free survival. The interval to an event was slightly less than the interval to fenestration closure, suggesting potential merit in the evaluation of earlier fenestration closure. Adoption of specific fenestration management guidelines might help improve the overall outcomes and enhance the quality of future studies. (J Thorac Cardiovasc Surg 2013;145:183-7)

Fenestration of the Fontan circuit has been shown to improve early outcomes, including a decreased duration and quantity of chest tube drainage, a shorter duration of mechanical ventilation, and a shorter postoperative length of stay.[1-5] Even in the present era in which fenestration is applied selectively by some groups, it continues to be used in high-risk patients, such as those with hypoplastic left heart syndrome.

Although a benefit in the early postoperative period, Fontan fenestration has theoretical long-term risks, including cyanosis and systemic thromboembolic events due to a persistent right-to-left shunt combined with an increased risk of thrombus formation due to venous stasis and hypercoagulability. Alternatively, persistent fenestration might be a benefit because the lower central venous pressure might decrease the risk of exercise intolerance, protein-losing enteropathy (PLE), plastic bronchitis, and bradyarrhythmias. The question of whether and when to intentionally close a fenestration remains unanswered. Management protocols have tended to vary, ranging from active fenestration closure at predetermined intervals to a ''hands-off'' approach in which the natural history of fenestration status is allowed to progress. A cross-sectional study of a large cohort of Fontan patients by the Pediatric Heart Network found that 20% of patients had patent fenestrations at a median of 8.6 years after the Fontan procedure.[6] Of those that were closed at follow-up, approximately 50% had been closed by active intervention (catheter based or surgical).

Therefore, we performed an analysis of a cohort of Fontan patients to determine the association between fenestration status and morbid clinical events.

METHODS

Subjects

The records of all patients who had undergone a fenestrated Fontan procedure at Children's Hospital of Wisconsin from January 1994 through June 2007 were reviewed. The patients were assigned to 1 of 2 study

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This study was supported by an American Association for Thoracic Surgery Summer Intern Scholarship.

Disclosures: Authors have nothing to disclose with regard to commercial support.

Read at the 38th Annual Meeting of The Western Thoracic Surgical Association, Maui, Hawaii, June 27-30, 2012.

Received for publication May 31, 2012; revisions received Aug 15, 2012; accepted for publication Sept 12, 2012; available ahead of print Oct 11, 2012.

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0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2012.09.006

Abbreviations and Acronyms
EFP = event-free probability
IQR = interquartile range
PLE = protein-losing enteropathy

groups, open or closed, according to their most recent fenestration status as assessed by echocardiography. The presence of a detectable shunt by echocardiography, not the intervention history, determined the categorization as open or closed. Of a total of 218 patients, 6 were excluded because of either undifferentiated ventricular morphology (1 patient) or intraoperative conversion to a nonfenestrated status at the original Fontan procedure (5 patients).

The human research review board at the Children's Hospital of Wisconsin authorized the collection of data from the existing medical records and the waiver of the Health Insurance Portability and Accountability Act for the present retrospective study.

Data Collection

The study participants were identified from the Herma Heart Center Cardiology and Cardiothoracic Surgery database. Patient information was obtained from the medical records, echocardiograms, cardiac catheterization findings, and operative reports. De-identified patient information was maintained using the REDCap web-based research data capture application.

Fenestrated Fontan Technique

Fenestrations were routinely created in all patients undergoing the Fontan procedure. Earlier in the study interval, certain patients were selected for fenestration closure before leaving the operating room according to the hemodynamics and echocardiographic findings. For most patients in the present study (and routinely in the present era), the fenestrations were left open. Our technique of performing the lateral tunnel or extracardiac fenestrated Fontan procedure has been previously described.[7] For all patients, a coronary punch was used to create a fenestration with a diameter of 3.5 to 4.0 mm. For the lateral tunnel Fontan procedure, the fenestration was placed at the center of the Gore-Tex polytetrafluoroethylene baffle (WL Gore & Associates, Flagstaff, Ariz). For the extracardiac Fontan procedure, the atriotomy that results from detaching the inferior vena cava from the right atrium was sewn to the Gore-Tex tube (WL Gore & Associates) as a large circle to centrally encompass the fenestration and prevent the adjacent atrial wall from affecting the size of the fenestration. For the extracardiac Fontan procedure, a fenestration pursestring suture and snare (left open) were placed in an accessible subcutaneous pocket below the sternotomy to permit subsequent potential closure of the snare and fenestration.

Fenestration Closure Technique

Our general approach was to evaluate patients for fenestration closure at 1 to 3 years after the Fontan procedure. The patients were brought to the cardiac catheterization laboratory to first determine their suitability for closure. The decision to close the fenestration was individualized for each patient and was determined from a multidisciplinary assessment of test closure-induced changes in systemic venous pressure, cardiac output, and oxygen saturation. Closure was then completed during the same anesthetic session with echocardiographic and/or angiographic confirmation of the closure. For most patients undergoing closure, the fenestration snare was exposed through a limited skin incision, and the snare was closed and secured with hemoclips. A few patients, including those with residual shunts after attempted snare closure, underwent closure with an Amplatzer septal occluder (AGA Medical, Golden Valley, Minn).

Outcome Variables

Morbid events after Fontan palliation were defined as death, transplantation, Fontan takedown, New York Heart Association functional classification III or IV, pacemaker placement, PLE, stroke, thrombus, plastic bronchitis, subsequent thoracic surgery, or post-Fontan mechanical circulatory support. The interval during which a patient was free from an event determined the event-free survival. Events (but not patients) were excluded from the present analysis if they had occurred within 1 calendar day of the Fontan procedure. Such events were regarded as being more related to the overall impact of the surgical procedure and less to the fenestration. The follow-up duration was calculated from the date of the Fontan procedure to the last known clinical visit, if no events had occurred, or from the date of the Fontan procedure to the date of an event.

Statistical Analysis

Descriptive characteristics of the sample are summarized using the median and interquartile range, because the variables were skewed. The Wilcoxon-Mann-Whitney test was used to compare continuous variables and the chi-square test or Fisher exact test to compare fenestration groups (open vs closed) for patient characteristics. Kaplan-Meier survival analysis was used to assess the event-free survival, with Wilcoxon log-rank comparisons between the open and closed groups. All analyses were done in SAS, version 9.2 (SAS Institute, Cary, NC).

RESULTS

Group Characteristics

Of the 212 patients in the present cohort, 161 were categorized as open and 51 as closed. Included in the open group were 10 patients who had undergone fenestration closure interventions with subsequent echocardiographically determined residual shunts and 19 patients who had undergone fenestration closure on the same day as their last known clinical follow-up visit. Included in the closed group were 18 patients with spontaneously closed fenestrations. The patient characteristics are summarized in Table 1. No significant differences were found in the patient characteristics between the open and closed groups.

The mean and median duration of follow-up after the Fontan procedure was 3.8 and 3.5 years (interquartile range [IQR], 0.9-6.1 years) for the entire cohort, 3.1 and 2.4 years (IQR, 0.7-4.9 years) for the open group, and 5.9 and 6.0 years (IQR, 4.4-8.0 years) for the closed group, respectively. The mean and median interval from Fontan to fenestration closure was 2.1 and 1.2 years (IQR, 0.7-3.3 years), respectively.

Events and Event-Free Probability

Of a total of 92 events, 72 occurred in the open group (50 of 161 patients, 31%) and 20 in the closed group (15 of 51 patients, 29%). In the closed group, 7 patients had events before fenestration closure and 9 after fenestration closure. The events are summarized according to closure status in Table 2. The most notable differences between the open and closed groups occurred with the event of New York Heart Association class III or IV, which occurred in 16.8% of the open compared with 2.0% of the closed group; and the event of PLE, which occurred in 4.3% of the open group compared with 0% in the closed group.