Clinical Outcome of Fenestrated Fontan Patients After Closure
The First 10 Years

Donna A. Goff, MS; Elizabeth D. Blume, MD; Kimberlee Gauvreau, ScD; John E. Mayer, MD; James E. Lock, MD; Kathy J. Jenkins, MD, MPH

Background—The late clinical status of Fontan patients after fenestration closure is unknown. Data are now available on all patients who underwent closure from 1989 to 1999.

Methods and Results—All patients who underwent catheter closure of a Fontan fenestration were enrolled in either the Clamshell (1989 to 1994) or CardioSEAL (1996 to 1999) regulatory trials. Physiological values obtained at catheterization helped assess the hemodynamic effects of fenestration occlusion. In addition to survival, outcomes assessed included O2 saturations, medication use, significant clinical findings (eg, heart failure, protein-losing enteropathy, or new arrhythmias), and somatic growth. Of 181 patients who underwent closure, 27 had additional significant leaks. The remaining 154 patients constituted the study group. Median time from closure to latest follow-up was 3.4 years (range 0.4 to 10.3 years). Fenestration closure increased O2 saturation 9.4% on average (P<0.001). The numbers of patients receiving digoxin or diuretics decreased at the most recent follow-up compared with baseline (P<0.001), but use of antiarrhythmic agents increased marginally (P=0.05). Height and weight percentiles rose (medians of 2 and 4, respectively; P<0.001). Clinical decompensation during follow-up of 154 patients was rare (4.5%), with 2 deaths, 3 Fontan revisions, and 1 patient each with protein-losing enteropathy and ascites. No other patient developed chronic congestive symptoms; 21 patients developed new arrhythmias, and 2 had a stroke or transient ischemic attack.

Conclusions—Fenestration closure in Fontan patients was followed by improved oxygenation, reduced need for anticongestive medication, and improved somatic growth at latest follow-up. Death (1.3%) or chronic decompensation (3.2%) was rare. (Circulation. 2000;102:2094-2099.)

Key Words: Fontan procedure ■ outcome ■ fenestration

Surgical procedures based on the Fontan principle separate pulmonary and systemic blood flows in patients with single ventricular physiology.1–4 Death and poor outcomes after Fontan-like procedures may be increased by one or more risk factors, including high pulmonary vascular resistance, poor systolic or diastolic ventricular function, hypoplastic or distorted pulmonary arteries, and systemic outflow obstruction.5–8 Recognizing that some of these risk factors were transient after surgery, Laks et al9 used an adjustable atrial septal defect to preserve cardiac output at the expense of oxygenation in high-risk patients in the immediate postoperative period. Based on a growing experience with transcatheter techniques to close various intracardiac defects,10–12 Bridges et al13 described a technique for late postoperative closure of Fontan baffle fenestrations by use of test occlusion and subsequent permanent closure with an intracardiac device. Both approaches of early closure and late closure after test occlusion have been reported to reduce mortality and morbidity after the Fontan procedure, especially in high-risk patients.9,14–16

Although spontaneous closure of the fenestration can occur,17 in many patients the fenestration remains patent. Despite the resultant cyanosis, patients with a patent fenestration after the Fontan procedure are frequently clinically “well,” with O2 saturations in the 80% to 90% range. Because the fenestration preserves cardiac output at the expense of oxygenation and limits the extent of increases in central venous pressure early and late after surgery, some have argued that fenestration closure is unnecessary and potentially dangerous. Clinical signs of high central venous pressure and/or low cardiac output, such as heart failure, ascites, protein-losing enteropathy, the need for Fontan takedown or

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Dr Lock is a co-inventor of the Clamshell/CardioSEAL device, and his institution receives royalties on its commercial sales; he also participates in regulatory studies of this device.

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transplantation, or death, might result from ill-advised closure.

The lack of availability of approved devices to close post-Fontan fenestrations has limited widespread use of late transcatheter closure of fenestrations. The recent FDA approval of the CardioSEAL device for humanitarian use\(^{18}\) has made device closure easily available in the United States for the first time. However, there is little information regarding indications for and late outcomes after fenestration closure. To provide insight into these issues, we describe clinical outcomes after transcatheter closure of a Fontan fenestration for a large cohort of patients during their first 10 years of follow-up. In this analysis, particular attention was paid to the prevalence of complications that might well be due to chronic low cardiac output or elevated central venous pressures.

**Methods**

**Study Sample**

The study sample consists of all patients who underwent closure of a fenestrated Fontan baffle with use of an intracardiac device as part of a series of studies conducted at Children’s Hospital, Boston, between June 1989 and February 1999, beginning with the first patient.\(^{13}\) The initial group of 111 patients (implanted June 1989 to October 1994) was enrolled in regulatory studies conducted by C.R. Bard, Inc, to evaluate the performance of the Clamshell Septal Occluder (C.R. Bard, Inc). More recent patients (May 1996 to February 1999) are part of an ongoing regulatory study evaluating the CardioSEAL septal occluder (NMT Medical, Inc) in high-risk patients. Three centers (University of San Francisco Medical Center, Children’s Hospital of Philadelphia, and Children’s Hospital, Boston) in the CardioSEAL study have used these devices to close Fontan fenestrations; data from all 3 sites are included. Permission to perform the implantation procedures and to collect follow-up information was obtained from the appropriate institutional review boards. Informed consent for device placement was obtained in all cases.

**Device Description**

The Clamshell is a low-profile double-umbrella device, with hinged arms attached to Dacron fabric.\(^{11}\) The CardioSEAL is a second-generation device, modified to improve long-term device performance.\(^{19}\)

**Implantation Procedure**

Implantation was performed as described previously.\(^{13}\) Briefly, patients underwent elective or, rarely, emergent cardiac catheterization with full heparinization to assess feasibility for fenestration closure on the basis of test occlusion hemodynamics. After baseline hemodynamics were assessed, the location and size of the defect were determined. Small (1- to 2-mm) defects that produced minimal cyanosis (aortic \(O_2\) saturation \(>92\%\)) were not closed. Fenestrations suitable for closure were then occluded with a balloon to determine the effects on hemodynamics. If aortic saturation did not exceed 90\% after 2 minutes of test occlusion, we searched carefully for other sources of right to left shunting. In general, closure was undertaken whenever aortic pressure remained stable, mixed venous saturation rose, and right atrial pressure was acceptable (<20 mm Hg) with test occlusion. After device release, repeat angiography determined the adequacy of position and residual shunting.

**Data Collection**

The Children’s Hospital Department of Cardiology actively maintains databases for the Clamshell Septal Occluder and high-risk CardioSEAL regulatory trials. A retrospective review of these databases, regulatory charts, and medical records yielded surgical, clinical, catheterization, and device-specific information for each patient. The type of Fontan procedure and fenestration size were acquired through review of surgical reports. Patients were grouped into 5 anatomic categories based on echocardiogram reports: hypoplastic left heart syndrome, heterotaxy syndrome, other predominant left ventricle, other predominant right ventricle, and others. Any catheterizations after the implantation procedure were recorded and noted.

**Statistical Analyses**

Because fenestration closure in patients with multiple right to left defects will have less predictable hemodynamic and clinical effects, only patients with a fenestration occurring in isolation, without additional significant right to left shunts, undergoing closure before February 17, 1999, were analyzed. Descriptive information was compared for the Clamshell and CardioSEAL groups by use of the Fisher exact test for categorical variables and the Wilcoxon rank sum test for continuous variables. Data from the 2 cohorts were then combined for analysis. Pretest and posttest occlusion measurements of hemodynamic cardiac catheterization variables were compared by paired \(t\) test. Similar comparisons were made between precatheterization \(O_2\) saturations and those at the most recent follow-up. Changes in height and weight percentiles were evaluated by the Wilcoxon signed rank test, and changes in medication use were evaluated by the McNemar test. Linear regression analysis was used to explore the relationships between change in \(O_2\) saturation and patient characteristics; similarly, logistic regression analysis was used for the outcome variable decompensation in cardiac status. Freedom from death and decompensation were estimated by the Kaplan-Meier method.

**Results**

**Descriptive Information**

Among the 181 (111 Clamshell and 70 CardioSEAL) patients who underwent successful device implantation, 27 had a significant (>2-mm) baffle leak or other cause for cyanosis in addition to the fenestration. The other 154 (91 Clamshell and 63 CardioSEAL) patients had a single defect closed. Descriptive information for this cohort is shown in Table 1. Most patients (128 of 132, 97.0\%) had a lateral tunnel Fontan with a 4-mm fenestration (103 of 130, 79.2\%); about half had a predominant left ventricle (81 of 154, 52.6\%; Table 1).

**Implantation Procedure**

A single device was implanted in 153 of 154 patients; 1 patient had 2 devices. Most devices were 17 mm (n=143, 92.9\%), although 9 were 23 mm, 1 was 28 mm, and 1 was a custom device.

Hemodynamic information for this cohort is presented in Table 2. Baffle pressure and arterial \(O_2\) saturation rose with test occlusion (mean increases of 1.5±2.3 mm Hg [\(P<0.001\)] and 5.3±4.8\% [\(P<0.001\) respectively], whereas cardiac index was lower (mean decrease 0.5±0.9 L \(\cdot\) min\(^{-1}\) \(\cdot\) m\(^{-2}\),
Complications at implantation were uncommon, occurring in 9 of 154 patients (5.8%). They included 2 patients with device embolizations subsequently retrieved, 4 additional patients with acute transient ECG changes requiring medical intervention, 1 patient with an arterial pulse loss requiring heparin therapy, 1 patient with vessel dissection (requiring surgical repair) after device embolization and retrieval (noted above), and 2 patients with hemodynamically silent device malpositions. No complication produced long-term sequelae.

Follow-Up
Median time from catheterization to most recent follow-up was 3.4 years (range 0.4 to 10.3 years). Differences between patients who received a Clamshell versus a CardioSEAL device were primarily due to a shift toward delay of closure in stable patients, once the possibility of spontaneous closure was recognized. Median time from Fontan procedure to first catheterization was longer for the CardioSEAL patients compared with the Clamshell group (33 versus 5 months, \( P < 0.001 \)), and median age at catheterization was older (6.9 versus 4.4 years, \( P < 0.001 \)). Median time from catheterization to last follow-up was longer for Clamshell patients (6.1 versus 1.7 years, \( P < 0.001 \)), and the CardioSEAL patients were slightly younger at the time of the Fontan procedure (2.8 versus 4.0 years, \( P = 0.04 \)).

Improve ment in \( O_2 \) Saturation s
Because information collected for the Clamshell patients focused on safety, not efficacy, \( O_2 \) saturation at latest follow-up was missing from the database for 48 patients. However, in 106 patients with available information, there was a significant mean increase of 9.4 ± 5.8% in \( O_2 \) saturation from baseline to last follow-up (\( P < 0.001 \)). Eight patients had an \( O_2 \) saturation < 90% at last follow-up. The magnitude of the increase in oxygenation was greatest in the patients who had the lowest preprocedure saturations (\( P < 0.001 \)). After adjusting for preprocedure saturation in a linear regression model, there were no significant associations between the amount that the \( O_2 \) saturation improved and anatomy, device size, age at Fontan procedure, age at catheterization, or time from catheterization to last follow-up.

Medication Usage and Growth
Table 3 displays clinical information on patients before the procedure and at the most recent follow-up. Among patients whose medication use changed, use of digoxin, diuretics, and warfarin decreased (\( P < 0.001 \), \( P = 0.001 \), and \( P = 0.03 \), respectively). There was a borderline increase in the use of antiarrhythmics (\( P = 0.05 \)). Age-specific height and weight percentiles both showed a small but significant increase from preprocedure to most recent follow-up (median increases of 2 percentiles [\( P < 0.001 \]) and 4 percentiles [\( P < 0.001 \], respectively).

Clinical Decompensation
No patients died during the placement of the device. Two deaths occurred (2.6 and 8.3 years after closure) during the follow-up period. A 6.5-year-old with double-outlet right

### TABLE 1. Descriptive Characteristics of Patients With Isolated Fenestration Who Underwent Device Closure

| Variables                        | Combined \( n = 154 \) | Clamshell \( n = 91 \) | CardioSEAL \( n = 63 \) | \( P \)  
|----------------------------------|------------------------|------------------------|-------------------------|---------  
| Age at Fontan, y                 | 3.4 (0.8–40)           | 4.0 (0.8–22)           | 2.8 (0.9–40)            | 0.04    
| Time from Fontan to catheterization, mo | 9 (0.2–84)           | 5 (0.2–35)            | 33 (6–84)               | <0.001  
| Age at catheterization, y       | 5.4 (1.3–43)           | 4.4 (1.3–22)           | 6.9 (2.1–43)            | <0.001  
| Sex (male), n (%)                | 96 (62.3)              | 59 (64.8)              | 37 (58.7)               | NS      
| Anatomic categories, n (%)       |                        |                        |                         | NS      
| Hypoplastic left heart syndrome  | 16 (10.4)              | 5 (5.5)                | 11 (17.5)               |         
| Heterotaxy syndrome             | 14 (9.1)               | 8 (8.8)                | 6 (9.5)                 |         
| Other predominant left ventricle| 81 (52.6)              | 49 (53.8)              | 32 (50.8)               |         
| Other predominant right ventricle| 39 (25.3)              | 26 (28.6)              | 13 (20.6)               |         
| Other                           | 4 (2.6)                | 3 (3.3)                | 1 (1.6)                 |         
| Time from closure to last follow-up, y | 3.4 (0.4–10.3) | 6.1 (0.4–10.3) | 1.7 (0.5–2.4) | <0.001  

Values are median (range) or number (percentage) of patients.

### TABLE 2. Hemodynamic Values During Implantation Procedure

<table>
<thead>
<tr>
<th></th>
<th>Before Procedure</th>
<th>After Test Occlusion</th>
<th>After Device Closure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baffle pressure, mm Hg</td>
<td>12.9±3.1 (151)</td>
<td>14.1±3.7 (124)</td>
<td>14.8±3.8 (115)</td>
</tr>
<tr>
<td>Cardiac index, L · min⁻¹ · m⁻²</td>
<td>3.1±1.0 (140)</td>
<td>2.6±0.8 (72)</td>
<td>2.7±1.1 (51)</td>
</tr>
<tr>
<td>Aortic ( O_2 ) saturation, %</td>
<td>87.5±5.7 (154)</td>
<td>92.9±3.6 (134)</td>
<td>91.6±4.7 (140)</td>
</tr>
</tbody>
</table>

Data are mean±SD (n). Before and after comparisons are all \( P < 0.001 \).
ventricle/severe mitral stenosis had an atrial septectomy followed by a fenestrated Fontan procedure at age 3.9 years. Pleural effusions and cyanosis complicated the early postoperative course; the fenestration was closed on postoperative day 12. After 2.5 good years, he developed congestive heart failure and wheezing and was treated with β-agonists. After an unexpected arrest associated with ventricular fibrillation/ventricular tachycardia, his ventricular function was poor; he died on the second hospital day. A second patient with double-inlet left ventricle underwent a fenestrated Fontan procedure at 10 years of age, with elective fenestration closure 1 month after surgery. He developed progressive aortic regurgitation and left ventricular dysfunction resulting in congestive symptoms 8 years later. He acutely decompensated with a pericardial effusion and died at attempted pericardiocentesis.

Three patients underwent subsequent intracardiac surgery. Two patients had right atrial reduction surgery and revision to a lateral tunnel or extracardiac baffle to treat either right pulmonary vein compression or intractable arrhythmias. The fenestration closure devices were explanted at Fontan revision. No other devices were explanted. The third patient, with an atrioventricular connection, had a right Glenn anastomosis. He had a snare-adjustable defect with early closure and (2) fixed fenestrations with late test occlusion and transcatheter device closure. Widespread use of the latter technique has been limited, in the United States, to a few centers that were part of studies to test devices in high-risk patients and in the occasional patient approved on a compassionate-use basis. The recent Food and Drug Administration approval of the CardioSEAL device to allow closure of Fontan fenestrations has made that device widely available in this country, focusing attention on the potential indications for and results of device closure.

Other Complications or Observations
Two patients had cerebral vascular accidents or transient ischemic attacks after device closure. Parenthetically, the number of patients receiving warfarin decreased from 18 to 7 (4%) after fenestration closure (Table 3). In 2 patients, an incidental arm fracture was found on routine follow-up radiograph. Both patients are doing clinically well.

Discussion
The use of fenestrations to reduce mortality and morbidity immediately after a Fontan operation has relied on 2 strategies: (1) snare-adjustable defects with early closure and (2) fixed fenestrations with late test occlusion and transcatheter device closure. Prior studies from several institutions have described the changes in early morbidity and mortality associated with using a fenestration in Fontan patients: the hemodynam-

New-Onset Arrhythmias
New-onset arrhythmias after fenestration closure were noted in 21 patients (13.6%): 11 had sick sinus syndrome or sinus bradycardia, 4 had atrial fibrillation or flutter, 2 patients developed ventricular arrhythmias, and in 4 patients, other dysrhythmias were noted. Pacemaker placement followed device closure after varying intervals in 6 patients, and 4 patients were started on antiarrhythmic agents after closure.

Estimated probabilities of survival (A) and freedom from decompensation of cardiac status (B) after device closure. Upper and lower dashed bands indicate 95% Greenwood confidence limits. Numbers of patients still at risk for failure are shown at 2-year intervals.
ic effects of test occlusion and selection of patients for transcatheter closure, the methodology for transcatheter closure of fenestrations with the use of several different devices, and the short-term follow-up of those patients. The present study, although significantly increasing the number of patients available for analysis, does not substantially alter the conclusions of those prior studies: generally, test occlusion modestly increases O₂ saturations, modestly increases right-sided pressures, and modestly reduces cardiac output. Marked increases in right-sided pressures or falls in cardiac output at test occlusion are taken as signs to defer late closure. Closure is performed percutaneously, with use of a sheath, and is generally safe, with no related mortality or chronic morbidity in 154 patients.

Focus of Study

The present study has been designed to provide insight regarding different questions. The absence of device availability did not stop surgeons from performing fenestrated Fontan procedures. As these patients were, by necessity, managed medically, cardiologists discovered that their clinical courses were generally benign, despite persistent cyanosis in those cases who did not spontaneously close their fenestrations. Several questions arose from that experience: Is fenestration closure really necessary or even beneficial? Will closure produce a significant increase in the need for anticongestive therapy? Will it increase the prevalence of ascites, effusions, or protein-losing enteropathy? Finally, does fenestration closure increase mortality in Fontan patients? Given the frequency with which the Fontan procedure is performed and the high morbidity and mortality of a failed Fontan procedure, these are important questions.

The present study is uniquely suited to address many of these issues. The very first patients to undergo fenestration with late test occlusion and closure are included in the data set, thus providing the longest possible follow-up. The surgical techniques, catheter techniques, device characteristics, and patient selection have been largely constant for the entire 10-year period. Finally, the fact that all patients were enrolled in FDA-supervised trials allowed for prospective information gathering in all patients and follow-up in each of 154 patients.

Mortality, Morbidity, and Failure

The first striking finding in the present study is the infrequency of deaths (2 of 154 patients), a finding that contrasts sharply with prior large series reporting late survival after the Fontan procedure. Because our sample includes only patients who underwent successful closure and is thus highly selective, we would not interpret these data to mean that the lateral tunnel fenestrated Fontan is the procedure of choice for single ventricle. However, these data do support a conclusion that fenestration closure in suitable patients does not increase the risk of death in the first 5 to 10 years of follow-up.

Also striking is the infrequency of morbidity or onset of failure of the Fontan circulation. There were only 3 reoperations, each apparently unrelated to fenestration closure, and each was successful. Two other patients had features of chronic decompensation: one developed protein-losing enteropathy, and one developed ascites. As above, these data support a conclusion that fenestration closure in suitable patients has a minimal impact on the prevalence of complications associated with elevated right heart pressures or low cardiac output. Our general criteria for closure include all patients who do not experience a fall in blood pressure or mixed venous saturation or a rise in right atrial pressure >20 mm with test occlusion.

Beneficial or Adverse Effects of Fenestration Closure

A persistent rise in O₂ saturation is expected after fenestration closure, and the present study confirms that expectation. However, the small but significant growth improvement seen in these children was not expected. Although a return toward normal growth may have been due to the original fenestrated Fontan procedure, the average delay between surgical performance of the fenestrated Fontan and closure of the fenestration (9 months) argues against that hypothesis. Similarly, the reduced need for anticongestive therapy after fenestration closure was unanticipated. This may reflect an improvement in O₂ delivery and/or ventricular function, or it may represent (warranted or unwarranted) a sense of clinical well-being on the part of patients and caregivers. Classic studies have demonstrated that children aged >2 years with persistent cyanosis (mean saturation 86%) will have depressed ventricular function that may persist after restoration of an acyanotic circulation. Regardless of the explanations, the reduced use of digitalis and diuretics provides strong support for the conclusion noted above: fenestration closure does not increase the prevalence of right heart failure and may well improve overall cardiovascular status.

The possible adverse effects of fenestration closure include new arrhythmias or strokes. Both are known to occur in patients after a Fontan procedure, with or without fenestration. The present study provides little new insight into the issue of whether fenestration closure increases or decreases the prevalence of late arrhythmias. In contrast, only 2 strokes occurred after fenestration closure, despite the fact that only 7 of 154 patients are receiving warfarin.

Indications for and Timing of Closure

Although not specifically addressed by these data, the indications for closure were, in our practice, an aortic saturation <90% and tolerance of test occlusion, as noted above. These imprecise guidelines reflect, accurately, the imprecise nature of patient selection for fenestration closure. However, because the vast majority of patients either underwent closure or had fenestrations too small to warrant closure, further efforts to improve the precision of patient selection criteria are unlikely to yield significant clinical insights.

In contrast, the optimal timing of fenestration closure remains an important unanswered question. The marked difference in timing of closure between the Clamshell versus CardioSEAL groups (5 months versus 33 months after surgery) did not produce any noticeable adverse clinical effects; indeed, it is likely that a significant number of fenestrations closed spontaneously. Supporting the thesis that

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closure should be delayed is the finding that early closure was associated with cardiac decompensation. However, the adverse effects of persistent cyanosis are too important to ignore. Careful noninvasive and/or invasive studies on the time course of spontaneous fenestration closure would help define a recommendation for planning fenestration closure.

Conclusions
In summary, we have found that test occlusion and subsequent transcatheter closure of Fontan fenestrations constitute a successful clinical strategy in the management of patients with single-ventricle physiology. Late closure is followed by improved oxygenation, reduced use of antiarrhythmic medications, and improved somatic growth. Failure to tolerate late test occlusion is rare. Death, protein-losing enteropathy, and ascites are also rare after fenestration closure, with a follow-up as long as 10 years. Early closure may predispose to cardiac decompensation or Fontan takedown and would be unnecessary in cases of spontaneous closure. These findings support a recommendation that patients with fenestrated Fontan procedures should undergo late (>6 months after surgery) transcatheter closure if O2 saturations are <90% and test occlusion is tolerated. More precise recommendations on the timing of late closure await further studies.

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