Effect of Baffle Fenestration on Outcome of the Modified Fontan Operation

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Background. The "fenestrated Fontan" (surgical baffle fenestration followed by transcatheter test occlusion and permanent closure after postoperative recovery) was adopted in an effort to reduce perioperative mortality and morbidity. This study assesses the effect of baffle fenestration on outcome.

Methods and Results. Patients having a modified Fontan operation with a cavocaval baffle and cavopulmonary anastomosis were retrospectively selected for study. Those with baffle fenestration (n=91) were compared with those without baffle fenestration (n=56) with respect to preoperative risk factors, age, anatomy, surgical date, and presence or absence of a previous bidirectional cavopulmonary anastomosis. Outcome variables were failure (death or take-down) and duration of postoperative pleural effusions and hospitalization. Survival and clinical status after hospital discharge were ascertained. The two groups did not appear to differ with respect to age or anatomic diagnosis. Patients having baffle fenestration were at significantly greater preoperative risk by univariate and multivariate analysis (p<0.01). Operative failure was low in both groups (11% without and 7% with baffle fenestration, p=NS). Durations of pleural effusions and hospitalization were significantly shorter with baffle fenestration (p<0.01). Neither date of surgery nor a previous bidirectional cavopulmonary anastomosis appeared to contribute to improved outcome. Patients with baffle fenestration had lower postoperative systemic venous pressure (p<0.01). There were no late deaths. Functional status in both groups is good (82% in New York Heart Association class I).

Conclusions. Baffle fenestration is associated with low mortality, significantly less pleural effusion, and significantly shorter hospitalization among high-risk patients having a modified Fontan operation.

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KEY WORDS • congenital heart surgery • single ventricle • pleural effusions

Several approaches to "staging" of the Fontan operation have been proposed, including variations on the bidirectional cavopulmonary anastomosis as preliminary palliation1-7 and the adjustable atrial septal defect.8-9. Modification of the Fontan operation by surgical fenestration of the atrial baffle and subsequent closure of the fenestration at cardiac catheterization was introduced in 1989.10 There are several theoretical benefits to this approach. First, a right-to-left shunt at the atrial level should maintain cardiac output at the expense of oxygenation when conditions that limit pulmonary blood flow are present. Second, baffle fenestration might limit the postoperative increase in right atrial pressure, thus reducing the incidence and/or duration of postoperative pleural effusions. Finally, postoperative assessment in the cardiac catheterization laboratory affords the opportunity to test the patient's tolerance of "Fontan physiology" before permanent closure of the fenestration, and transcatheter techniques can be used to correct certain conditions (such as residual distal pulmonary artery stenosis or hemodynamically important aortopulmonary collateral vessels) that may be detrimental to patients without a pulmonary ventricle. This report presents the results of a retrospective study designed to address the effect of baffle fenestration on outcome after a modified Fontan operation.

Methods

Patient Selection

The study population consisted of all patients who have undergone a modified Fontan repair at our institution by the surgical technique shown schematically in Figure 1. The atria were partitioned with a partial tube made of Gore-tex placed along the lateral atrial wall, connecting the superior and inferior cava-atrial junctions. Systemic venous return was brought to the pulmonary arteries via unilateral or bilateral cavopulmonary anastomoses. This specific modification of the Fontan procedure was first used at our institution in October 1987; the study period dates from that time through June 1991. Pulmonary artery angioplasty, atrial septectomy, and ligation and division of aortopulmonary shunts were performed when indicated; patients having any other additional surgical interventions (such
as replacement of the atrioventricular valve, end-to-side pulmonary artery–to-aortic anastomosis, or repair of anomalous pulmonary venous connection) were excluded. Patients were assigned to one of two groups, depending on whether they had nonfenestrated or fenestrated atrial baffles. Fenestration size among the first 20 patients has been reported previously; in all remaining patients having baffle fenestration in this report, a 4-mm fenestration was created by use of a coronary punch.

These criteria were chosen to produce two groups of patients whose surgical management was uniform except for the presence or absence of baffle fenestration.

**Comparison of Preoperative Characteristics**

Patients considered for a Fontan operation are a heterogeneous population. The primary variables used in this study to compare the two groups with regard to preoperative risk were those that have the most influence on preoperative clinical decision-making at our institution: presence or absence of pulmonary artery distortion, pulmonary vascular resistance, mean pulmonary artery pressure, ventricular filling pressure, and age at surgery.

Pulmonary artery distortion was defined as peripheral pulmonary artery stenosis or hypoplasia or discontinuity of the pulmonary arteries. This was a subjective assessment based on angiographic and operative findings. Presence of pulmonary artery distortion was designated a risk factor. Mean pulmonary artery pressure was measured with fluid-filled catheters either directly or indirectly as the pulmonary vein wedge pressure. In patients having different pressures in the right and left pulmonary arteries, the higher mean pulmonary artery pressure was used. A mean pulmonary artery pressure >15 mm Hg was designated a risk factor. Pulmonary vascular resistance was calculated by use of either directly measured pulmonary artery pressures or pulmonary vein wedge pressures. When the pulmonary arterics were discontinuous or if there was clearly disparate pulmonary flow caused by branch pulmonary artery stenosis and no lung perfusion scan was available, the pulmonary vascular resistance was considered impossible to calculate. A pulmonary vascular resistance greater than 2 Wood’s units (mm Hg·L⁻¹·min⁻¹·m⁻²) was designated a risk factor. Ventricular filling pressure was taken as the mean pressure in the atrium that emptied into the systemic ventricle. A ventricular filling pressure ≥12 mm Hg was arbitrarily designated a risk factor.

In addition, a multivariate comparison of preoperative characteristics was performed using the results of a study that has been reported previously. In this analysis, 27 candidate variables drawn from a data base of 334 patients who underwent a modified Fontan operation between July 1, 1984, and June 30, 1990, were entered into a stepwise logistic regression analysis. The regression coefficients of significant preoperative variables (atrioventricular valve anatomy, pulmonary artery pressure, completeness of preoperative hemodynamic data, and pulmonary artery distortion) were used to generate a “risk score” for each of the patients in the current study. The risk score is expressed as the probability of failure, with failure defined as perioperative death or take-down of the Fontan operation.

Finally, other preoperative factors that may be considered to influence outcome after a Fontan operation were noted and compared, including anatomic diagnosis, presence of a previous bidirectional cavopulmonary anastomosis, and date of surgery.

**Assessment of Operative Outcome**

Outcome variables were operative failure, duration of pleural effusions, and duration of hospitalization. Operative failure was defined as death or take-down of the Fontan operation to some other form of palliation. Deaths and take-downs that occurred within 30 days of surgery or that were associated with prolonged, unremitting pleural effusions were included. In determining duration of hospitalization, the day of surgery was counted as day 1, and the day of discharge counted as the final day. In the event of a separate hospitalization for fenestration closure, those hospital days (usually 2) were included in the total. The duration of perioperative pleural effusions was determined by designating the day of surgery as day 1 and the last day of drainage (e.g., the day the chest tubes were removed or the day of the final percutaneous thoracostentesis) as the final day. Effusions that began more than 30 days after the last day of perioperative drainage are classified as late effusions (see below). The duration of pericardial effusions was defined as for pleural effusions.
Fenestration Closure

Patients underwent elective postoperative cardiac catheterization for test occlusion and permanent transcatheter closure of the fenestration at various time intervals after complete recovery from surgery (that is, after any pleural or pericardial effusions appeared to have resolved); the timing of referral for postoperative catheterization was left to the judgment of the patient’s cardiologist and cardiac surgeon. Early in our experience (i.e., during the first year), fenestration closure was generally performed within 2–3 weeks of surgery. By the second year, it became evident that some patients had spontaneous closure of the fenestration during the first few months after surgery, and others had qualitative improvement in ventricular function. As a result, we now wait 3–6 months after surgery before carrying out fenestration closure in most cases. At the time of cardiac catheterization, test occlusion of the fenestration is carried out as previously described. Briefly, baseline measurements of right atrial pressure and saturation and of aortic saturation are taken. Then, the fenestration is occluded with a balloon-tipped catheter for 10 minutes, and the measurements are repeated. Guidelines for criteria for permanent transcatheter closure of the fenestration include 1) with test occlusion, arterial oxygen saturation should increase above 92%; 2) systemic venous pressure should not increase above 18 mm Hg; and 3) the arteriovenous difference in oxygen saturation should not increase by more than about 30% compared with the preocclusion value. Formal calculation of changes in cardiac output or systemic oxygen transport was not performed in the catheterization laboratory at the time of test occlusion; rather, the decisions were based on the saturations and pressures. Patients who did not fit within the guidelines generally did not undergo transcatheter fenestration closure.

There was, however, no strict protocol for management in the catheterization laboratory. Test occlusion was performed routinely, and the data were used in a judgment made by the catheterizing cardiologist, referring cardiologist, and surgeon.

When the data were reviewed, cardiac output (Fick method) and systemic oxygen transport were calculated for all patients undergoing test occlusion. Calculations were based on measured oxygen consumption (using a Waters Instruments metabolic rate meter) whenever available; otherwise, values were taken from standard tables derived from the formulas of LaFarge and Miettinen.

Follow-up

The patients’ clinical course (survival, New York Heart Association [NYHA] class, cardiac medications, occurrence of late effusions) after hospital discharge was determined by assessment by the primary cardiologist.

Statistical Analysis

Preoperative and outcome variables are expressed as the mean followed by the SEM and the range. The following statistical tests were performed with Stata software (Computing Resource Center, Los Angeles, Calif.): comparison of means by a two-tailed t test (paired or unpaired, as appropriate); comparison of proportions by χ²; a stepwise multiple regression with preoperative and operative variables as independent variables and duration of pleural effusions as the dependent variable; and an actuarial analysis of survival (Kaplan-Meier method with Greenwood confidence limits). Several of the preoperative and outcome variables are interdependent (e.g., pulmonary artery pressure and pulmonary vascular resistance or duration of pleural effusions and duration of hospitalization); for this reason, a value of p ≤ 0.01 was taken as significant.

Analysis of the data is carried out according to “intention to treat.” Thus, for example, two patients who developed pleural effusions after closure of a fenestration (but within the defined 30-day period) as well as five patients who had a fenestration performed but had little or no evidence of right-to-left shunt after surgery (arterial oxygen saturations of 92–96%) remained within the fenestrated group for analysis. Similarly, two patients who had unintentional baffle leaks that were subsequently closed at cardiac catheterization remained in the nonfenestrated group.

Results

A total of 147 patients met criteria for entry into the study. There were 56 patients with nonfenestrated atrial baffles who underwent surgery between October 1987 and January 1991 and 91 patients with fenestrated atrial baffles who had surgery between June 1989 and June 1991. Anatomic diagnoses are given in Table 1.

Preoperative Characteristics

The results of the univariate and multivariate assessment of preoperative risk are shown in Table 2. The two groups did not differ significantly with respect to age. The preoperative mean pulmonary artery pressure was significantly higher among patients having a fenestrated procedure than in those having a nonfenestrated procedure; 40% of the former versus 14% of the latter had a mean pulmonary artery pressure >15 mm Hg. Pulmonary artery distortion was significantly more prevalent among patients having a fenestrated procedure. The preoperative ventricular filling pressure was slightly higher among patients having a fenestrated procedure; however, the difference was not significant. Preoperative pulmonary vascular resistance was significantly higher among patients having a fenestrated procedure; 30% of these patients versus 16% of those having a nonfenestrated procedure had a pulmonary vascular resistance >2 Wood’s units. A significantly greater proportion of patients having a fenestrated procedure had multiple risk factors, whereas none of the patients having a nonfenestrated procedure had more than two risk factors. The preoperative predicted multivariate “probability of failure” was significantly higher among patients having a fenestrated procedure. Thus, the group in which baffle fenestration was performed appears to have been at higher risk because of their preoperative pulmonary artery pressure, resistance, and distortion, number of individual risk factors, and multivariate assessment of preoperative risk. The two groups are not demonstrably different with regard to age or ventricular filling pressures. In no category that we evaluated did the patients having a nonfenestrated procedure appear to be at higher risk than those having a fenestrated procedure.
TABLE 1. Cardiac Anatomic Diagnosis

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Total (n=147)</th>
<th>Nonfenestrated (n=56/147)</th>
<th>Fenestrated (n=91/147)</th>
<th>Failed (n=12/147)</th>
<th>Effusions (n=32/135)</th>
</tr>
</thead>
<tbody>
<tr>
<td>DILV, n (%)</td>
<td>39 (27)</td>
<td>20 (36)</td>
<td>19 (21)</td>
<td>1 (8)</td>
<td>8 (25)</td>
</tr>
<tr>
<td>DORV/MA, n (%)</td>
<td>18 (12)</td>
<td>5 (9)</td>
<td>13 (14)</td>
<td>3 (25)</td>
<td>3 (9)</td>
</tr>
<tr>
<td>DORV/other, n (%)</td>
<td>10 (7)</td>
<td>3 (5)</td>
<td>7 (8)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>HLHS, n (%)</td>
<td>13 (9)</td>
<td>7 (13)</td>
<td>6 (7)</td>
<td>3 (25)</td>
<td>4 (13)</td>
</tr>
<tr>
<td>TA/TGA, n (%)</td>
<td>13 (9)</td>
<td>3 (5)</td>
<td>10 (11)</td>
<td>0</td>
<td>1 (3)</td>
</tr>
<tr>
<td>TA/NRGA, n (%)</td>
<td>7 (5)</td>
<td>1 (2)</td>
<td>6 (7)</td>
<td>0</td>
<td>2 (6)</td>
</tr>
<tr>
<td>Heterotaxy, n (%)</td>
<td>9 (6)</td>
<td>3 (5)</td>
<td>6 (7)</td>
<td>0</td>
<td>2 (6)</td>
</tr>
<tr>
<td>Other, n (%)</td>
<td>38 (26)</td>
<td>14 (25)</td>
<td>24 (26)</td>
<td>5 (42)</td>
<td>12 (38)</td>
</tr>
</tbody>
</table>

DILV, double inlet left ventricle; DORV/MA, double outlet right ventricle with left atrioventricular valve stenosis or atresia; DORV/other, other types of non septatable double outlet right ventricle; HLHS, hypoplastic left heart syndrome; TA/TGA, tricuspid atresia with transposition of the great arteries; TA/NRGA, tricuspid atresia with normally related great arteries; other, all other cardiac anatomic diagnoses.

By χ² with seven degrees of freedom, the following comparisons are made: diagnosis, nonfenestrated vs. fenestrated, p=0.3; diagnosis, total vs. failed, p=0.1; diagnosis, total vs. prolonged effusions, p=0.4.

Among patients having a nonfenestrated procedure, two (4%) had previous bidirectional cavopulmonary shunts. Among those having a fenestrated procedure, 21 patients (23%) had previous bidirectional cavopulmonary shunts.

Operative Outcome

Outcomes for the two groups are shown in Table 3. There were six failures in each group (five deaths and one take-down among patients without baffle fenestration, for a failure rate of 11%, and four deaths and two take-downs among patients with baffle fenestration, for a failure rate of 7%; p=NS). The duration of pleural effusions was significantly shorter among patients having a fenestrated procedure. Prolonged effusions (requiring drainage more than 14 days after surgery) were seen in both groups but were much more common among those having a nonfenestrated procedure (38% of survivors) than those having a fenestrated procedure (13% of survivors). Prolonged pleural effusions occurred in 29% of patients who had a previous cavopulmonary shunt. Drainage of pericardial fluid was deemed necessary in only a small number of patients (four patients in each group). Hospital stay was significantly longer for patients having a nonfenestrated procedure; in that group, 30% of survivors remained in the hospital for more than 21 days, whereas only 12% of patients having a fenestrated procedure did so.

To assess the combined effect of preoperative characteristics, date of surgery, and presence or absence of baffle fenestration on the duration of pleural effusions, these variables were entered into a stepwise multiple regression analysis. The only variable that demonstrated a significant effect on duration of effusions was baffle fenestration (Table 4).

Additional Observations

Other data collected are shown in Table 5. The preoperative mean pressure in the systemic venous atrium was similar in the two groups (6.5 versus 7.7 mm Hg); however, the mean postoperative pressure in the systemic venous atrium was higher in patients having a nonfenestrated procedure. The postoperative increase in right atrial pressure (compared with the preoperative right atrial pressure measured at catheterization) was also higher in patients having a nonfenestrated procedure. Postoperative arterial oxygen saturation was predictably lower in patients who had a fenestrated baffle (mean, 87%) than in those with a nonfenestrated baffle (mean, 94%) in the immediate postoperative period. One patient early in the series (previously reported10) had too large a fenestration, which resulted in excessive cyanosis. Although some

TABLE 2. Preoperative Risk Assessment

<table>
<thead>
<tr>
<th></th>
<th>Group 1 (nonfenestrated) [mean±SEM (range)]</th>
<th>Group 2 (fenestrated) [mean±SEM (range)]</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>6.5±0.8 (0.9–30)</td>
<td>5.0±0.45 (0.7–22.5)</td>
<td>0.09</td>
</tr>
<tr>
<td>Mean PAP (mm Hg)</td>
<td>11.9±0.48 (5–26)</td>
<td>15±0.6 (7–35)</td>
<td>0.0004</td>
</tr>
<tr>
<td>Fraction with PAD</td>
<td>0.19</td>
<td>0.42</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Ventricular FP (mm Hg)</td>
<td>6.5±0.42 (1–13)</td>
<td>7.7±0.33 (2–17)</td>
<td>0.02</td>
</tr>
<tr>
<td>Rp (mm Hg·1·min⁻¹·m⁻²)</td>
<td>1.4±0.09 (0.4–3.3)</td>
<td>1.9±0.13 (0.5–7.0)</td>
<td>0.007</td>
</tr>
<tr>
<td>Fraction with &gt;1 risk factor (PAP&gt;15, PAD, FP&gt;12, Rp&gt;2)</td>
<td>0.09</td>
<td>0.38</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Multivariate risk score, expressed as probability of failure</td>
<td>0.15±0.02 (0.01–0.84)</td>
<td>0.30±0.03 (0.02–0.98)</td>
<td>0.0004</td>
</tr>
</tbody>
</table>

PAP, pulmonary artery pressure; PAD, pulmonary artery distortion; FP, filling pressure; Rp, pulmonary vascular resistance.
TABLE 3. Postoperative Outcome

<table>
<thead>
<tr>
<th></th>
<th>Group 1 (nonfenestrated)</th>
<th>Group 2 (fenestrated)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Failure</td>
<td>6/56 (11%)</td>
<td>6/91 (7%)</td>
<td>NS</td>
</tr>
<tr>
<td>Duration of PL (days)</td>
<td>23.8±5.1 (2–187)</td>
<td>10.7±2.3 (1–176)</td>
<td>0.009</td>
</tr>
<tr>
<td>Fraction with PL lasting &gt;14 days</td>
<td>20/52 (38%)</td>
<td>11/87 (13%)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Length of hospitalization (days)</td>
<td>21.4±2.9 (6–119)</td>
<td>13.3±1.3 (4–94)</td>
<td>0.005</td>
</tr>
<tr>
<td>Fraction hospitalized for &gt;21 days</td>
<td>17/57 (30%)</td>
<td>11/91 (12%)</td>
<td>&lt;0.01</td>
</tr>
</tbody>
</table>

PL, pleural effusions. Values given are the mean±SEM and range where applicable.

Clinical Course After Hospital Discharge

Among the 50 patients who had a successful nonfenestrated Fontan operation, three (6%) were lost to follow-up; all three reside outside the United States. One additional non-US patient was known to be alive as of March 1992, but information regarding NYHA class could not be obtained. There were no late deaths or take-downs among the remaining 46 survivors. The average duration of follow-up was 34±12 months (range, 6–53 months). Actuarial analysis of survival is shown in Figure 2. Late effusions occurred in five of 46 patients (11%). NYHA class was I in 36 patients (78%) and II in 10 patients (22%). No cardiac medications were taken by 35 patients (41%); 11 (13%) took digoxin only; 34 took a diuretic and/or captopril or enalapril.

Fenestration Closure

The outcome among 85 patients having a successful fenestrated Fontan repair with regard to closure of the fenestration is shown in Table 6. Of 52 patients who had transcatheter closure of the fenestration, 10 had hemodynamics that were deemed unacceptable at the first test occlusion, and their fenestrations were left open at that time. At a subsequent catheterization with test occlusion, hemodynamics were found to have improved considerably; interim interventions included transcatheter coil embolization of aortopulmonary collaterals in five patients, transcatheter pulmonary artery balloon angioplasty of residual pulmonary artery stenosis in three patients, and medical management in the remaining two patients. The interval between surgery and transcatheter closure ranged from 4 days to 19 months (mean, 3.8 months). Table 7 shows the hemodynamic effects of fenestration closure in these patients: there was a consistent increase in arterial oxygen saturation and systemic venous pressure, with no significant change in systemic venous saturation or systemic oxygen transport. Complications occurred in association with four of 52 transcatheter closures (8%); these included intracardiac air in two patients, leading to transient hypotension in one and seizures in the other; a transient brachial plexus injury (previously reported in one patient; and transient complete heart block with retrograde passage of a wire into the left ventricle in one patient. None of these complications had permanent sequelae. The mean arterial oxygen saturation in the 69 patients whose fenestrations are closed is 94%. No patient had a deterioration in clinical status after fenestration closure; one patient, as mentioned above, had a late pericardial effusion after fenestration closure.

Discussion

Methodology

Horwitz et al18 describe the principles that, if applied, allow an observational cohort study to approximate the results of an experimental trial. These include specifi-

TABLE 4. Impact of Date of Surgery

<table>
<thead>
<tr>
<th>Variable</th>
<th>F</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary vascular resistance</td>
<td>0.1142</td>
<td>NS</td>
</tr>
<tr>
<td>Ventricular filling pressure</td>
<td>0.3651</td>
<td>NS</td>
</tr>
<tr>
<td>Pulmonary artery pressure</td>
<td>1.0754</td>
<td>NS</td>
</tr>
<tr>
<td>Date of surgery</td>
<td>2.7469</td>
<td>NS</td>
</tr>
<tr>
<td>Age at surgery</td>
<td>1.9988</td>
<td>NS</td>
</tr>
<tr>
<td>Fenestration</td>
<td>5.4002</td>
<td>0.001</td>
</tr>
</tbody>
</table>

To address the difference in treatment period between the two groups (October 1987 to January 1991 vs. June 1989 to June 1991), a stepwise regression analysis was performed, with "duration of pleural effusions" as the outcome variable and the independent variables listed above. Among these variables, the only one with a significant impact on duration of effusions is baffle fenestration.

TABLE 5. Postoperative Filling Pressures

<table>
<thead>
<tr>
<th></th>
<th>Nonfenestrated</th>
<th>Fenestrated</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Postoperative pressure in systemic venous atrium</td>
<td>14.3±0.45</td>
<td>12.6±0.36</td>
<td>0.004</td>
</tr>
<tr>
<td>Postoperative increment in systemic venous pressure</td>
<td>9.3±1.5</td>
<td>5.4±0.5</td>
<td>0.0001</td>
</tr>
</tbody>
</table>

Values are in mm Hg, mean±SEM.
cation of a zero time for determining eligibility, application of inclusion and exclusion criteria as in a randomized trial, classification of patients according to suitable clinical criteria, and statistical methods that include testing of a predetermined hypothesis and use of intention-to-treat procedures. We attempted to apply all of these principles in this study. Nevertheless, this study has those limitations that are inherent in all retrospective studies; even with the best effort, a retrospective study does not achieve the force of a prospective trial.

We intentionally chose a study sample in which surgical management was uniform. The overall failure rate in this sample of 12 of 147 patients (8%) is nearly identical to that of the concurrent group of Fontan patients from which the study sample was drawn (22 of 241, 9%). In addition, we attempted to address the problem of comparability as completely as possible.

The two patient groups are not demonstrably different with regard to age or anatomic diagnosis. In addition, as shown in Table 1, there were no particular anatomic diagnoses that were significantly overrepresented among patients whose operation failed or who had prolonged effusions. This is in accord with our view that anatomic diagnosis per se does not predict outcome among patients having a Fontan operation.11

Within the study period of October 1987 to June 1991, the treatment periods for the two groups were not identical (October 1987 to January 1991 for patients having a nonfenestrated procedure and June 1989 to June 1991 for patients having a fenestrated procedure). If there were other, time-related changes in management that contributed significantly to improvement in outcome, one would expect that “date of surgery” would emerge as a significant variable when analyzed along with risk factors, age, and fenestration. It did not. The problem of comparing two groups with overlapping but not identical treatment periods cannot be dealt with in an entirely satisfactory fashion, especially in a retrospective study; we think it unlikely, however, that other, unidentified interventions have had an important effect on the results of the study.

Finally, a greater proportion of patients having a fenestrated procedure had a previous bidirectional cavopulmonary shunt. This difference reflects the fact that patients in the fenestrated group were perceived to be at higher risk, and thus a greater number underwent bidirectional cavopulmonary anastomosis as interim palliation. This procedure has been reported to have an association with decreased duration of pleural effusions in patients having a subsequent Fontan repair.2,19 However, in our study sample, 29% of patients with a previous cavopulmonary shunt had prolonged pleural effusions, compared with 38% of all patients having a nonfenestrated procedure, 13% of all patients having a fenestrated procedure, and 22% of the study sample as

<table>
<thead>
<tr>
<th>Table 6. Outcome of Patients Having a Successful Fenestrated Fontan Operation (n=85) With Regard to Fenestration Closure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Outcome</td>
</tr>
<tr>
<td>Spontaneous closure of fenestration</td>
</tr>
<tr>
<td>Surgical closure at time of surgery for closure of a large marginal baffle leak</td>
</tr>
<tr>
<td>Transcatheter closure</td>
</tr>
<tr>
<td>Total with closed fenestration</td>
</tr>
<tr>
<td>Catheterized, with fenestration left open</td>
</tr>
<tr>
<td>No postoperative catheterization</td>
</tr>
<tr>
<td>Total with open fenestration</td>
</tr>
</tbody>
</table>

FIGURE 2. Graph showing freedom from death or take-down among patients having a nonfenestrated Fontan operation. The number of patients potentially available for follow-up at a given duration and the number for whom follow-up was actually obtained are indicated below the curve.

FIGURE 3. Graph showing freedom from death or take-down among patients having a fenestrated Fontan operation. The number of patients potentially available for follow-up at a given duration and the number for whom follow-up was actually obtained is indicated below the curve.
a whole. Removing this subset of patients from the study would increase the difference in duration of effusions between the two groups of patients. Thus, the presence of a greater proportion of these patients in the nonfenestrated group tends to bias the study against the result obtained and does not diminish the validity of the result.

The two groups cannot be considered comparable with regard to follow-up. Patients having a nonfenestrated procedure have a longer period of follow-up; on the other hand, surveillance of patients having a fenestrated procedure has been considerably more vigilant. In addition, formal follow-up is mandated in a Food and Drug Administration protocol for patients who have had transcatheter device closure of a fenestration. For these reasons, although all follow-up data are presented, no statistical comparisons are made. The follow-up period for both groups of patients is relatively short.

The multiple preoperative patient characteristics that may influence Fontan outcome and the various surgical adaptations of the operation made to accommodate specific cardiac anatomies make it difficult to ascertain the influence of baffle fenestration per se on the results of these procedures. This problem would not disappear if the study were performed prospectively. Any decisions made regarding the requirements for comparability of the two groups would be, to some degree, arbitrary and subjective; if all possible patient characteristics were included in a stratification/randomization scheme, the resulting sample size requirement might well make the study impossible to carry out in a reasonable time period.

**Outcomes**

Both by univariate assessment of those characteristics that most strongly influence clinical decision making and by the multivariate risk score, the patients having a fenestrated procedure were at higher preoperative risk. Thus, the fact that success among the patients having a fenestrated procedure was as great as among those having a nonfenestrated procedure supports (but does not definitively prove) the notion that fenestration of the atrial baffle decreases the risk of the Fontan procedure.

Patients who had baffle fenestration had significantly shorter hospitalizations and duration of pleural effusions than did patients in whom the atrial baffle was not fenestrated. An association between increased postoperative atrial pressure and poor outcome has been reported. Our observations of lower postoperative right atrial pressures and better outcome in patients having a fenestrated Fontan procedure support that notion and also suggest that the incremental increase in perioperative systemic venous pressure may be of prognostic significance, particularly with regard to pleural effusions. Nevertheless, the specific mechanism whereby baffle fenestration contributes to the reduction in postoperative pleural effusions is not addressed by this study. Duration of hospitalization is closely tied to duration of pleural effusions. It was included separately as an outcome variable to assess the possibility that the added hospital days necessary for transcatheter fenestration closure might balance those that were the result of prolonged effusions among the patients having a nonfenestrated procedure.

Driscoll et al have reported a considerable late morbidity and mortality among patients at increased risk for a Fontan operation. Our patients are a smaller group, with shorter follow-up; nevertheless, it is worth noting that the perioperative and 2-year survival is considerably higher among patients who had a "risk score" (as defined by Driscoll) of at least 1 or 2 and had 1 or 2 years of follow-up after a fenestrated Fontan operation than was observed in their study (93% versus 78%). Whatever its origin, this difference in early survival is such that we are optimistic about an improved long-term prognosis in our survivors.

Of the 85 patients who had a successful fenestrated Fontan operation, 81% have had closure of the fenestration without clinical deterioration. Patients selected for transcatheter closure were those who, by our arbitrary criteria, "tolerated" test occlusion; that is, test occlusion did not result in an increase in either systemic venous pressure or arteriovenous difference in oxygen content, which we deemed to be clinically important. There was no significant change in systemic oxygen transport. The remaining patients remain mildly to moderately cyanotic while they await elective or spontaneous closure of the fenestration. Whether all of these patients will tolerate fenestration closure or whether some will remain cyanotic but well palliated (that is, without ventricular volume overload and with acceptable arterial oxygen saturation) because of inability to tolerate fenestration closure remains to be seen. Complications (none of which resulted in permanent injury) occurred in four patients in association with the postoperative catheterization. In three, the complication was directly associated with transcatheter closure. The use of large venous sheaths to deliver devices carries the risk of air embolism, with potentially serious consequences. Introduction of air into the sheath, a preventable complication, is most likely to occur when the inner dilator is removed, before introduction of the umbrella;
it is avoided by constant flushing of the sheath through the dilator (with a 60-cm³ syringe and a side-arm adapter) as the dilator is removed.

Summary and Conclusions

In a patient population selected for similarity of surgical management, baffle fenestration at the time of a modified Fontan repair was associated with significantly shorter hospitalization and duration of pleural effusions than were seen in patients undergoing a nonfenestrated procedure. Although patients in the group with baffle fenestration were at significantly higher preoperative risk than were those without baffle fenestration, they had comparably high operative success rates. This degree of success in high-risk patients is in marked contrast to our previously reported experience. Among patients with a nonfenestrated baffle, one late death and one late take-down occurred, both in association with unremitting pleural effusions; there were no other late failures. At present, 72% of survivors of a nonfenestrated operation and 88% of survivors of a fenestrated Fontan operation are in NYHA class I. We conclude that baffle fenestration followed by postoperative assessment in the cardiac catheterization laboratory and transcatheter closure of the fenestration is beneficial in selected patients undergoing a modified Fontan operation in that it is associated with low mortality, significantly less pleural effusion, and significantly shorter hospitalization. These results justify an aggressive and optimistic approach to their management.

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