Improved Early Morbidity and Mortality After Fontan Operation: The Mayo Clinic Experience, 1987 to 1992

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Objectives. This study sought to evaluate changes in early morbidity and mortality as well as predictors of outcome in our most recent 339 patients undergoing modified Fontan operations.

Background. The Fontan operation is the preferred definitive palliation for patients with functional single ventricles. Previously reported early mortality rates after Fontan operation have been substantial.

Methods. Records of 339 consecutive patients who had a Fontan operation at the Mayo Clinic between 1987 and 1992 (recent cohort) were reviewed. This cohort was compared with the previous 500 patients who had Fontan operations performed between 1973 and 1986 (early cohort).

Results. Recently, overall early mortality after Fontan has decreased significantly compared with that for the early cohort (from 16% to 9%, p = 0.002). This decline occurred despite increased anatomic complexity of patients. Short-term posthospital survival has also improved significantly in recent patients.

Conclusions. Many factors may have contributed to decreased early mortality after Fontan. Improved patient selection, younger age at time of operation, refinements in surgical techniques and postoperative management may all have had important roles. Proposed technical modifications of the Fontan operation must be evaluated in light of these improved results.

In 1971, Fontan and colleagues (1) reported the first successful total right heart bypass that separated the pulmonary and systemic circulations and relieved ventricular volume overload in patients with tricuspid atresia. Recently, revised selection criteria have been suggested, and many technical modifications of the Fontan operation have been proposed (2–14). Some of these modifications (4,5) involve a persistent interatrial communication ("fenestration") that allows an ongoing right-to-left shunt. Because these most recent modifications represent a major change in approach to these patients, it seemed that a reassessment of early postoperative results of the nonfenestrated Fontan operation was indicated. Therefore, we studied the clinical course and outcomes of a recent cohort of patients \((n = 339)\) who had Fontan operations at the Mayo Clinic (operations performed from 1987 to 1992). This cohort was then compared to our previously reported results in patients who had Fontan operations before 1987 \((n = 500)\). In addition, an attempt was made to identify factors that currently were predictive of early mortality.

Methods

Patients. Clinical data from outpatient records and hospital charts were reviewed for all 339 patients who had a Fontan operation at the Mayo Clinic between January 1, 1987, and December 31, 1992 ("recent cohort"). These results were compared with similar data obtained from the first 500 patients who had a Fontan operation at the Mayo Clinic between 1973 and 1986 (this group is referred to as the "early cohort") (3,6–10). These data were available in an existing computer-
ized data base. None of the patients in either cohort had atrial fenestration at the time of the Fontan operation.

**Measurements.** Data from patient records, preoperative echocardiograms and cardiac catheterizations were reviewed to assess factors that have been described previously as possibly being predictive of early mortality after Fontan operation (a list of all variables analyzed appears in a footnote in the Results section). Arrhythmias were documented by surface electrocardiogram, Holter monitoring or rhythm strip tracings. Early mortality was defined as death occurring at any time before hospital discharge or within 30 days after operation (even if the patient had been discharged). The day of operation was considered day 1 for calculating the length of hospital stay, intubation and chest tube drainage. The duration of chest tube drainage was "prolonged" if it continued beyond 14 days. The hospital stay was "prolonged" after 21 days. Postoperative mean right atrial pressure was measured after the discontinuation of cardiopulmonary bypass but before sternal reapproximation. Renal failure was defined as the need for peritoneal dialysis or hemodialysis. Impaired hepatic function was defined as clinical jaundice associated with elevated serum transaminase levels.

**Follow-up.** Follow-up regarding vital status of hospital survivors was obtained by reviewing outpatient records, correspondence letters and vital status questionnaires. Survival beyond the last patient contact was not assumed.

**Statistical analysis.** All the factors described in previous Mayo Clinic publications (3,6-10) as being predictive of early mortality were analyzed as possible predictors of outcome in the recent cohort. In addition, type of Fontan connection and pacemaker placement were also assessed as possible predictors of outcome. Univariate assessment of the association of variables with mortality was performed using chi-square and Wilcoxon rank-sum tests. Univariate comparisons of variables that differentiated the recent cohort from the early cohort were performed with the same tests. Cumulative actuarial survival was estimated using the Kaplan-Meier method (15). For the multivariate assessment of factors potentially associated with early mortality, the preoperative and perioperative or postoperative variables used were those indicated to be univariately associated with early mortality. Multivariate analysis was performed using the logistic regression procedures available in the SAS statistical analysis system (16). A stepwise, backward elimination of the nonsignificant variables was performed using a p value cut point to retain variables with p < 0.02. For all other comparisons, two-tail p values ≤0.05 were taken as evidence of findings not attributable to chance.

## Results

**Patients.** The recent cohort consisted of 339 patients (202 boys [60%], 137 girls [40%]). Median age at time of surgery was 7 years (range 9 months to 38 years; mean 9.3 years). Sixty-three patients (19%) had tricuspid atresia; 72 (21%) had double-inlet left ventricle; 62 (18%) had a heterotaxy syndrome (asplenia 34 [10%]; polysplenia 28 [8%]); and 142

<table>
<thead>
<tr>
<th>Lesion</th>
<th>No. (%) of Patients</th>
<th>Early Mortality*</th>
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<tbody>
<tr>
<td>Tricuspid atresia</td>
<td>63 (19%)</td>
<td>3/63 (5%)</td>
</tr>
<tr>
<td>Double-inlet left ventricle</td>
<td>72 (21%)</td>
<td>4/72 (6%)</td>
</tr>
<tr>
<td>Polysplenia</td>
<td>28 (8%)</td>
<td>4/28 (14%)</td>
</tr>
<tr>
<td>Asplenia</td>
<td>34 (10%)</td>
<td>5/34 (15%)</td>
</tr>
<tr>
<td>Pulmonary atresia, IVS</td>
<td>18 (5%)</td>
<td>1/18 (6%)</td>
</tr>
<tr>
<td>Double-inlet right ventricle</td>
<td>13 (4%)</td>
<td>1/13 (8%)</td>
</tr>
<tr>
<td>DORV or TGA with AVV hypoplasia</td>
<td>36 (11%)</td>
<td>1/36 (3%)</td>
</tr>
<tr>
<td>UVH with AVV atresia</td>
<td>32 (9%)</td>
<td>5/32 (16%)</td>
</tr>
<tr>
<td>Other</td>
<td>43 (13%)</td>
<td>6/43 (14%)</td>
</tr>
<tr>
<td>Total</td>
<td>339 (100%)</td>
<td>30/339 (9%)</td>
</tr>
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</table>

*No significant differences in mortality rate between groups (see text for definition of early mortality). AVV = atrioventricular valve; DORV = double-outlet right ventricle; IVS = intact interventricular septum; TGA = transposition of the great arteries; UVH = univentricular heart.

(42%) had other complex forms of functional single ventricle. Overall, early mortality for the recent cohort was 30 (9%) of 339. Early mortality is described according to each cardiac lesion in Table 1. Patient age, gender and cardiac lesion were not significantly predictive of early mortality after Fontan operation.

**Comparison of early and recent cohorts.** Early mortality after Fontan operation significantly decreased in the recent cohort compared to the early cohort (30 [9%] of 339 vs. 81 [16%] of 500, p = 0.002). Patients in the recent cohort were significantly younger (median 7 years, range 1 to 38) than those in the early cohort (median 10 years, range 1 to 42, p < 0.001). There were more patients with complex anatomy (not tricuspid atresia or double-inlet left ventricle) in the recent cohort than in the early group (60% vs. 36%, p < 0.001). The hemodynamic features of both cohorts were similar (Table 2). The only statistically significant differences in hemodynamic values between the two cohorts were preoperative ventricular end-diastolic pressure and postoperative mean right atrial pressure. The early cohort had more patients in the high extreme of the range for these pressures, although the means and medians were similar.

**Univariate analysis of factors affecting early mortality in the recent cohort.**

**Preoperative factors.** Preoperative factors that were significantly associated with early mortality by univariate analysis are listed in Table 3. Those factors included cardiothoracic ratio >0.60, ambiguous atrial situs, common atrium, common atrioventricular valve, total anomalous pulmonary venous connection and daily diuretic therapy.

**Postoperative factors.** Intraoperative and postoperative factors affecting early mortality by univariate analysis included atrioventricular valve repair or replacement, mean right atrial pressure (immediately postoperative) >20 mm Hg and the postoperative occurrence of any of the following: pericardiotomy, supraventricular tachycardia, renal failure or impaired hepatic function (Table 3). A "lateral tunnel"-type anastomosis was performed in 55% of patients (187 of 339) in
Table 2. Hemodynamic Variables in Early and Recent Cohorts

<table>
<thead>
<tr>
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<tr>
<td>Preoperative</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PAP (mm Hg)</td>
<td>17 (4-65)</td>
<td>17 (5-65)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>19 ± 9</td>
<td>18 ± 8</td>
<td></td>
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<tr>
<td>RPA (U m²)</td>
<td>1.5 (0.1-6.1)</td>
<td>1.3 (0.1-6.6)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>1.6 ± 1.0</td>
<td>1.5 ± 0.9</td>
<td></td>
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<tr>
<td>VEDP (mm Hg)</td>
<td>13 (1-34)</td>
<td>12 (2-27)</td>
<td>0.002*</td>
</tr>
<tr>
<td></td>
<td>13 ± 5</td>
<td>12 ± 4</td>
<td></td>
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<tr>
<td>EF (%)</td>
<td>58 (6-82)</td>
<td>55 (30-84)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>56 ± 12</td>
<td>55 ± 10</td>
<td></td>
</tr>
<tr>
<td>C/T ratio</td>
<td>0.55 (0.35-0.82)</td>
<td>0.57 (0.36-0.81)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>0.56 ± 0.08</td>
<td>0.57 ± 0.07</td>
<td></td>
</tr>
<tr>
<td>Operative</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RAP (mm Hg)</td>
<td>18 (8-34)</td>
<td>18 (10-28)</td>
<td>&lt; 0.001*</td>
</tr>
<tr>
<td></td>
<td>18 ± 3</td>
<td>17 ± 2.5</td>
<td></td>
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<tr>
<td>Bypass time (min)</td>
<td>122 (50-384)</td>
<td>133 (57-346)</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td></td>
<td>130 ± 43</td>
<td>142 ± 49</td>
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</table>

*Significantly different, Wilcoxon rank sum test; statistical significance was achieved because the high extreme of the range for these variables was lower in the recent cohort, although the mean and median values in the early and recent cohorts were similar. Data presented are median (range) or mean value ± SD. C/T = cardiothoracic ratio; EF = ejection fraction; PAP = pulmonary artery pressure; RAP = right atrial pressure; RPA = pulmonary arteriolar resistance; VEDP = ventricular end-diastolic pressure.

the recent cohort. This type of connection did not affect early mortality in either a positive or negative way, nor did prolonged hospitalization or prolonged chest tube drainage. Numerous other preoperative and postoperative factors were analyzed and were not found to significantly influence early mortality.

Multivariate analysis of factors affecting early mortality in recent cohort. Multivariate assessment was performed as outlined in Methods. Initial candidate predictors were all of the variables found to be associated with early death by univariate analysis (Table 3). By use of logistic regression, a stepwise, backward elimination of the nonsignificant variables was performed. The p value cut point to retain variables was <0.02. This resulted in three variables being identified as multivariate risk factors for early death: postoperative renal failure (p < 0.001), postoperative right atrial pressure greater than 20 mm Hg (p = 0.008) and the presence of a common atrioventricular valve (p = 0.017).

There were 230 patients with none of these three risk factors. Early mortality was 3% (7 of 230) in this group. Of 48 patients with common atrioventricular valve but normal renal function and right atrial pressure less than 20 mm Hg, there was no early mortality (0%). Twelve patients had right atrial pressure elevated above 20 mm Hg as a sole multivariate risk; there was one early death (8%) in this group. Twenty patients had renal failure alone, and there were five early deaths (25%). Nineteen had renal failure and at least one of the two other variables present, and 14 (74%) of these patients died. Of five patients with a common atrioventricular valve and elevated right atrial pressure, there were two early deaths (40%). Presence of postoperative supraventricular tachycardia met the usual criteria of significance (p < 0.05), but in light of the number of variables considered, and because this factor was only marginally significant by univariate analysis (p = 0.044, Table 3), it was not included as a significant multivariate predictor.

In order to assess which preoperative factors were independently associated with early mortality, a separate multivariate analysis was performed. This analysis included only the “preoperative” factors (see Table 3) found to be significantly associated with early mortality by univariate analysis (i.e., information known to the clinician during the preoperative evaluation). Statistical methods used were identical to those used for the overall analysis described above. Two of the preoperative variables were found to be independently associated with an increased risk of early mortality. They were the presence of a common atrioventricular valve (p < 0.001) and daily diuretic therapy (p = 0.007). There were 225 patients who had neither of these risk factors, and early mortality in this group was 4.9% (11 of 225). Forty-five patients were on daily diuretic therapy but did not have common atrioventricular valves; early mortality in this group was 11.1% (5 of 45). Fifty-nine patients had common atrioventricular valves but were not on daily diuretic medication; early mortality in these patients was 15.3% (9 of 59). When both risk factors were present in the same patient, early mortality was 50% (5 of 10).

Influence of age on early mortality. In the recent cohort, 20% of patients (69 of 339) were under 4 years old, and early mortality in this subgroup was 13% (9 of 69); 34 (10%) of 339 patients were younger than 3 years old, and early mortality for these patients was 9% (3 of 34). This was a significant decrease in mortality for patients less than 4 years of age compared with the early cohort (14 [27%] of 51, p = 0.047) (10). In the recent cohort, early mortality for patients <4 years old was not significantly different from early mortality for older patients (21 [8%] of 270).

In the recent cohort, 46 of 339 patients (14%) were adults...
Table 3. Preoperative and Postoperative Factors Associated With Early Mortality After Fontan Operation in the Recent Cohort

<table>
<thead>
<tr>
<th>Factor</th>
<th>No. (%)</th>
<th>EM (%)</th>
<th>p Value</th>
<th>Preoperative Analysis</th>
<th>Postoperative Analysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preoperative</td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>C/T ratio &gt;0.60</td>
<td>232 (68%)</td>
<td>11%</td>
<td>0.026</td>
<td>NS</td>
<td>NS</td>
</tr>
<tr>
<td>Ambiguous atrial situs</td>
<td>52 (15%)</td>
<td>17%</td>
<td>0.020</td>
<td>NS</td>
<td>NS</td>
</tr>
<tr>
<td>Common atrium</td>
<td>57 (17%)</td>
<td>19%</td>
<td>0.002</td>
<td>NS</td>
<td>NS</td>
</tr>
<tr>
<td>Common AV valve</td>
<td>69 (20%)</td>
<td>20%</td>
<td>&lt; 0.001</td>
<td>0.017</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Total anomalous pulmonary venous connection</td>
<td>41 (12%)</td>
<td>20%</td>
<td>0.010</td>
<td>NS</td>
<td>NS</td>
</tr>
<tr>
<td>Daily diuretic therapy</td>
<td>55 (16%)</td>
<td>18%</td>
<td>0.008</td>
<td>NS</td>
<td>0.007</td>
</tr>
<tr>
<td>Postoperative</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>AV valve repair/replacement</td>
<td>68 (20%)</td>
<td>16%</td>
<td>0.017</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Mean RAP &gt;20 mm Hg</td>
<td>23 (7%)</td>
<td>30%</td>
<td>&lt; 0.001</td>
<td>0.008</td>
<td></td>
</tr>
<tr>
<td>Pericardiocentesis</td>
<td>31 (9%)</td>
<td>26%</td>
<td>&lt; 0.001</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Supraventricular tachycardia*</td>
<td>107 (32%)</td>
<td>13%</td>
<td>0.044</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Renal failure</td>
<td>39 (12%)</td>
<td>49%</td>
<td>&lt; 0.001</td>
<td>&lt; 0.001</td>
<td></td>
</tr>
<tr>
<td>Impaired hepatic function</td>
<td>17 (5%)</td>
<td>35%</td>
<td>&lt; 0.001</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>339 (100%)</td>
<td>9%</td>
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</tbody>
</table>

*atrial flutter, atrial fibrillation, atrial tachycardia or accelerated junctional rhythm. AV = atrioventricular; EM = early mortality; other abbreviations as in Table 2.

Arrhythmias. Tachyarrhythmias. Preoperatively, 46 (14%) of 339 patients had a history of tachyarrhythmia. Preoperative history of tachyarrhythmia did not correlate with increased early mortality. During the postoperative hospitalization, 32% of patients (107 of 339) had postoperative supraventricular tachyarrhythmias (atrial flutter, atrial fibrillation, atrial tachycardia or accelerated junctional rhythm), and only 8% of patients (27 of 339) had a postoperative ventricular arrhythmia. In the multivariate analysis, postoperative supraventricular tachyarrhythmia was weakly correlated with increased risk of early mortality after Fontan operation (p = 0.033, Table 3). However, the degree of significance was considered to be borderline because of the number of variables considered and the variable selection process that was employed. Although 40% of patients in the recent cohort had transient postoperative tachyarrhythmias, only 41 (13%) of 311 required an antiarrhythmic medication other than digoxin at the time of hospital discharge.

Arrhythmias. Bradyarrhythmias. In the recent cohort, complete heart block was present in 11 (3%) of 339 patients before the Fontan operation. Complete heart block developed in an additional 12 (4%) of 328 patients during the Fontan hospitalization. Twenty-two percent of patients (73 of 339) required prolonged (>5 days) temporary pacing for sinus node dysfunction or junctional rhythm in order to maintain adequate cardiac output. A permanent pacemaker was placed or replaced in 17 patients (6%) during the Fontan hospitalization. Neither pace-
Figure 1. Survival curves after Fontan operation comparing the early cohort and the recent cohort. The recent cohort demonstrated significantly improved early survival (p = 0.006).

Figure 2. Survival curves after Fontan operation for heterotaxy patients, comparing the early and recent cohorts. The recent cohort demonstrated significantly improved early survival (p < 0.001).

Figure 3. Survival curves after Fontan operation for children <4 years old, comparing the early and recent cohorts and showing significantly improved early survival in the recent cohort (p = 0.011).

Risk factor analysis. In 1977, Choussat and colleagues (11) described 10 criteria for selecting patients for the Fontan operation. Subsequent experience has shown that many of these criteria should be considered to be risk factors for but not absolute contraindications to the Fontan operation. Our recent experience demonstrated that patients with common atrioventricular valves, those requiring daily diuretic therapy and patients with specific postoperative problems (mean right atrial pressure >20 mm Hg or renal failure) were at significant risk for increased early mortality. No other factors were significantly predictive of early mortality in the multivariate analyses performed in this study. However, this does not imply that traditional preoperative risk factors (such as pulmonary artery hypoplasia, decreased ventricular ejection fraction, increased ventricular end-diastolic pressure, increased pulmonary artery pressure and increased pulmonary arteriolar resistance) are unimportant. In fact, they are likely to be at least as important as the risk factors identified in this study. Indeed, few patients with these previously defined “high risk” characteristics were selected for Fontan operation in the recent cohort.

Younger age at the time of Fontan operation may contribute to the improved survival observed in this study (17). Young patients, specifically those under 4 years old, have been reported to experience increased early mortality after Fontan operation (10). However, during the last 20 years, early mortality after Fontan for young patients has decreased pro-

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gressively to the current rate of 13% for those <4 years old and to 9% for those <3 years old. Many factors may have contributed to the improved early outcome in young patients. For example, in the early cohort, 37% of patients younger than 4 years had a Fontan operation on a nonelective basis because of progressive clinical deterioration. In the recent cohort, all patients were operated electively and, therefore, represent better candidates and had fewer complicating factors. Previous staging operations (i.e., cavopulmonary connections) may favorably influence early mortality after Fontan operation (18,19). However, the number of patients in the recent cohort who had previous cavopulmonary connections was small (24 years had a Fontan operation on a nonelective basis because of without heterotaxy syndromes. Patients with heterotaxy syndromes. In the recent cohort, early mortality after Fontan for heterotaxy patients has decreased dramatically compared to the early cohort (14.5% vs. 43%). Many factors, including the use of an intraluminal catheter to route hepatic venous and inferior vena caval blood flow to the pulmonary arteries, may have contributed to these improved results. In the early cohort, patients with asplenia had increased early mortality compared with patients with polysplenia (61% vs. 27%). But in the recent cohort, early mortality after Fontan operation is similar in these two subsets of patients (asplenia, 15% vs. polysplenia, 14%).

Atioventricular valve repair. Atioventricular valve repair or replacement, a recognized risk factor for early mortality, was required more frequently in heterotaxy than nonheterotaxy patients (45% vs. 14%). If one controls for atioventricular valve repair or replacement, then the outcomes observed in recent patients with heterotaxy are similar to those in patients without heterotaxy syndromes. Patients with heterotaxy syndromes who did not have atioventricular valve procedures had early mortality of 9%, compared to 21% in heterotaxy patients who had atioventricular valve procedures.

Nonfenestrated versus fenestrated approach. The length of the hospital stay (median 14 days) observed in the recent cohort is similar to that reported by medical centers that routinely perform atrial fenestration (4,20–22). However, no patient in the present series had an atrial fenestration performed. In the recent cohort, 36% of patients had chest tube drainage that was defined as prolonged (>14 days), and 24% of patients were hospitalized for longer than 21 days. It appears that centers that routinely perform fenestration have lower rates of prolonged chest tube drainage and fewer prolonged postoperative hospital stays (4,20–22). However, comparison of results from different centers is difficult for many reasons, including differences in surgical techniques and postoperative care and, most importantly, patient selection and heterogeneity. It is unclear whether the reduction in initial hospital stay and duration of chest tube drainage justifies the potential morbidity (including risk of paradoxical embolization and desaturation) from a persistent postoperative right-to-left shunt and the added economic cost of subsequent atrial baffle fenestration closure.

It seems that it would still be difficult to predict which patients would benefit from atrial baffle fenestration, as most patients have a satisfactory result without fenestration. In most instances, postoperative management can be successfully accomplished with hemodynamic adjustments, prevention of acidosis and pharmacologic manipulation of the pulmonary vasculature, including the potential benefits of inhaled nitric oxide. Perhaps for these reasons, atrial baffle fenestration is best reserved for patients who have right atrial pressures ≥20 mm Hg after cessation of cardiopulmonary bypass and optimization of hemodynamic variables.

Short-term follow-up. Information regarding vital status was available for 92% of patients in the recent cohort. Although there is ongoing risk for late morbidity and mortality after Fontan operation, the short-term survival for recent Fontan patients has improved markedly. Current 1-year survival was 88%, and 5-year survival was 81%. By comparison, 5-year survival in the early cohort was 79% and 73%, respectively. Also, short-term survival for heterotaxy patients, a recognized “high-risk” subgroup, has also improved dramatically (1-year survival 78%; 5-year survival 73%) compared with the early cohort (1-year survival 53%; 5-year survival 42%).

Conclusions. Early and short-term survival after the nonfenestrated Fontan operation have improved dramatically despite the increased anatomic complexity and younger age of the recent cohort of patients. In the recent cohort, early survival, even for traditionally high risk heterotaxy syndrome patients, has improved significantly. Also, young age (between 2 and 4 years of age) no longer increases operative risk for Fontan patients. Many factors may have contributed to the improvement in early and short-term survival that was observed. Because many of these changes were subtle and tended to evolve continuously, the factors most responsible for these improvements could not be specifically identified by retrospective statistical analysis. It is possible that improved patient selection, younger age at time of operation, refinements in surgical techniques and changes in postoperative management have all contributed. Any major technical modification of the Fontan operation must be evaluated in light of the improved outcomes observed after nonfenestrated Fontan operations in the recent cohort.

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