JACC Vol. 19, No. 4
March 15, 1992:828–33

# The Fontan Operation in Infants Less Than 2 Years of Age

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**Young age remains a reported risk factor for a successful Fontan operation despite improved survival rates. Since March 1978, the Fontan operation has been performed in 47 patients. To avoid a primary or secondary palliative shunt, an early Fontan procedure (Group 1: mean age 1.5 ± 0.5 years, range 0.6 to 2) has been performed in 17 children with the outcome similar to that of the remaining 30 older patients (Group 2: mean age 7.5 ± 5 years, range 2.4 to 23 years). Preoperatively both groups had acceptable hemodynamic status for a successful Fontan result.**

**Operative variables including cardiopulmonary bypass time, aortic cross-clamp time and core temperature were similar between groups and did not affect mortality. The postoperative mortality rate including early surgical (0% vs. 13%, respectively), late (18% vs. 12%) and total (18% vs. 23%) was similar between Groups 1 and 2 ($p > 0.05$). Immediate postoperative arrhythmias were more frequent in Group 1 (71% vs. 25%, $p < 0.01$) with no related mortality, while late arrhythmias occurred with equal frequency (29% vs. 39%, $p > 0.05$). Group 1 infants required a longer hospital stay (22 ± 9 vs. 14 ± 5 days, $p < 0.01$).**

**Thus, young age is not a risk factor for successful outcome of the Fontan operation in patients with acceptable preoperative hemodynamic status. An early Fontan operation may also avoid prolonged palliative procedures and their potential deleterious effects.**

*(J Am Coll Cardiol 1992;19:828–33)*

The Fontan operation or its modification remains the primary surgical procedure for many children with complex congenital heart disease. In the early 1970s, Choussat, Fontan and coworkers (1) proposed 10 preoperative criteria, including age between 4 and 16 years, that, if satisfied, would be expected to result in a successful Fontan operation. The particular criterion of age has been further substantiated by a recent study (2) in which age <4 years was judged to be an independent risk factor for poor survival after the operation. However, performing the Fontan operation at an older age is associated with chronic volume loading of the functioning systemic ventricle and may require multiple (two or more) palliative aortopulmonary shunt procedures with their associated operative risks.

Since 1988 we have performed an early Fontan operation in 17 infants whose preoperative anatomy and hemodynamic status were acceptable according to previously established criteria (Table 1). All 17 infants were demonstrating evidence of increasing cyanosis as a result of inadequate pulmonary blood flow and would have required either a primary or a secondary palliative shunt procedure. This study examines our results with this group.

## Methods

**Study patients.** We retrospectively reviewed the hospital and clinic charts of all 47 patients who underwent a Fontan-type operation at the Milton S. Hershey Medical Center between March 1978 and June 1990. During these 12 years, two surgeons have performed all of the Fontan operations, which were performed with cardiopulmonary bypass, direct caval cannulation and deep or moderate hypothermia. Cold crystalloid cardioplegia was used in 44 patients when aortic cross-clamping was required. Three patients had a period of induced ventricular fibrillation during hypothermia and cardioplegia to prevent any ejection of air during intraatrial suturing. Since April 1988, 17 infants have undergone a Fontan-type operation before 2 years of age to avoid further palliative procedures. Nine were male and 8 were female; the mean age was 1.5 ± 0.5 years (range 0.6 to 2). The preoperative anatomy of the 17 infants consisted of single ventricle of left ventricular morphology in 4, complex double-outlet right ventricle in 4, tricuspid atresia or stenosis with right ventricular hypoplasia in 3, hypoplastic left heart syndrome in 3 and complex atrioventricular (AV) canal defect in 3. Fourteen infants had a palliative aortopulmonary shunt procedure before the Fontan operation; three did not. In addition, four infants had a Norwood stage I operation, one infant had a pulmonary valvotomy and Blalock-Hanlon

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Manuscript received May 22, 1991; revised manuscript received September 23, 1991, accepted October 4, 1991.

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0735-1097/92/$5.00

**Table 1. Established Hemodynamic Criteria for the Fontan Operation**

| |
|---|
| Normal ventricular function |
| Ventricular end-diastolic pressure <12 mm Hg |
| Mean pulmonary artery pressure <15 mm Hg |
| No significant atrioventricular valve regurgitation |
| Pulmonary vascular resistance <4 U·m² |
| McGoon ratio of pulmonary arteries >1.8 |

atrial septectomy, one had repair of total anomalous pulmonary venous return and pulmonary artery reconstruction, one had repair of isolated total anomalous pulmonary venous return and one had a pulmonary valvotomy.

We compared these 17 patients with the 30 older patients who underwent a Fontan type operation between March 1978 and June 1990. Twenty-one of these older patients were operated on before and nine were operated on after April 1988. The group had a mean age of 7.5 ± 5 years (range 2.4 to 23); 21 patients were male and 9 female. The majority (n = 16) had various forms of tricuspid atresia or stenosis with right ventricular hypoplasia. Eight patients had single ventricle (six with left ventricular and two with right ventricular morphology), five had complex double-outlet right ventricle and one patient had a complex AV canal defect. Sixteen patients had one or more palliative aortopulmonary shunt procedures, one patient had a Glenn shunt and seven patients had pulmonary artery banding before the Fontan operation to control pulmonary blood flow. Six patients did not require any palliative procedure. In addition, two patients had repair of coarctation and two had resection of subaortic stenosis before their Fontan operation.

**Preoperative variables.** Preoperative variables compared between groups included 1) the age at Fontan operation; 2) the presence of sinus rhythm; 3) the cardiac anatomy, which was classified as tricuspid atresia or stenosis with right ventricular hypoplasia, single ventricle, hypoplastic left heart, complex double-outlet right ventricle and complex AV canal malformation; 4) the main ventricular morphology, defined angiographically as right or left or both right and left if two adequately sized ventricles were present; 5) the type and number of palliative procedures to control pulmonary blood flow including no palliation, one or more aortopulmonary shunts, Glenn shunt, pulmonary artery banding; and 6) the duration in years of palliative aortopulmonary shunts.

*Hemodynamic variables obtained during the preoperative cardiac catheterization and compared between groups included* 1) the presence of subaortic stenosis; 2) the main ventricular end-diastolic pressure; 3) the mean pulmonary artery and right atrial pressures; 4) the indexed pulmonary vascular resistance; 5) the pulmonary and systemic flow indexes; 6) the pulmonary to systemic flow ratio; 7) the percent systemic saturation; 8) the percent hematocrit; 9) the amount of systemic AV valve regurgitation (quantified angiographically from 0 to 4+); and 10) the McGoon ratio of pulmonary artery dimensions (3).

*Operative variables compared between groups included* 1) total cardiopulmonary bypass time; 2) aortic cross-clamp time; and 3) core temperature. Also compared was 4) the type of Fontan operation performed (either a total cavopulmonary, Kreutzer right atrial to pulmonary, or Björk right atrial to right ventricle connection). No fenestrated Fontan procedures or bidirectional Glenn shunts were performed during this time period. In addition we examined 5) the use of a conduit in the repair; 6) the need for pulmonary artery reconstruction or enlargement; 7) the need for additional surgery not involving the Fontan anastomosis (excluding takedown of an aortopulmonary shunt); 8) the duration of the hospital stay; and 9) the need for additional surgery late postoperatively.

*Postoperative variables compared between groups included* 1) early (<30 days postoperatively), late and total mortality; and 2) the incidence of immediate (<30 days postoperatively) and late postoperative arrhythmias. Late arrhythmias were documented by 12-lead surface electrocardiograms (ECGs) or 24-h Holter ambulatory ECG obtained at least yearly in all patients. These included bradyarrhythmias, tachyarrhythmias and the tachycardia-bradycardia syndrome. Bradyarrhythmias included junctional rhythm, nonsinus atrial rhythm and sinus bradycardia defined according to previously established guidelines for the patient's age (4). Tachyarrhythmias included all accelerated rhythms except sinus tachycardia.

*Late echocardiographic variables compared between groups included* 1) qualitative main ventricular systolic function (normal vs. abnormal); and 2) the degree of systemic AV valve and semilunar valve regurgitation quantified by the area of the regurgitant flow defined by Doppler color flow mapping (0 = none, 1 = mild, 2 = moderate and 3 = severe). Also examined were 3) New York Heart Association functional class, and 4) the duration of follow-up.

**Statistical analysis.** Data are expressed as the mean values ± SD. Numeric data were analyzed with an unpaired Student *t* test. Categoric data were analyzed by using chi-square analyses with continuity correction. The Mantel-Haenszl test was performed to test for the effect of age on mortality while accounting for the type of surgical procedure performed. A p value < 0.05 was considered significant.

## Results

**Preoperative variables (Tables 2 to 4).** Group 1 infants were significantly younger at the time of the Fontan operation (Tables 2 and 3). Before the Fontan operation more Group 2 patients had tricuspid atresia or stenosis with right ventricular hypoplasia and there was a tendency for more Group 1 infants to have hypoplastic left heart syndrome. Thus, more Group 1 infants had a morphologic right ventricle as the systemic ventricle. Among patients requiring either no palliation or aortopulmonary shunting, more patients in Group 1 had a single shunt before the Fontan operation. However, the duration of shunt palliation was

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**Table 2.** Preoperative Variables in 47 Patients

| | Group 1 (n = 17) | Group 2 (n = 30) | p Value |
|---|---|---|---|
| Age at Fontan operation (yr) | 1.5 ± 0.5 | 7.5 ± 5 | 0.001* |
| Rhythm (sinus) | 17 (100%) | 26 (87%) | 0.30 |
| Cardiac anatomy | | | |
| TA | 3 (18%) | 16 (53%) | 0.04* |
| Single ventricle | 4 (24%) | 8 (27%) | 0.90 |
| HLHS | 3 (18%) | 0 (0%) | 0.08 |
| DORV | 4 (24%) | 5 (17%) | 0.85 |
| Complex AV canal | 3 (18%) | 1 (3%) | 0.83 |
| Main ventricular morphology | | | |
| RV | 6 (35%) | 2 (7%) | 0.04* |
| LV | 8 (47%) | 22 (73%) | 0.19 |
| RV/LV | 3 (18%) | 6 (20%) | 0.85 |
| Previous palliation | | | |
| 0 AoP shunt | 3/17 (18%) | 6/23 (26%) | 0.80 |
| 1 AoP shunt | 14/17 (82%) | 10/23 (44%) | 0.03* |
| 2 AoP shunts | 0 (0%) | 6/23 (26%) | 0.07 |
| Glenn shunt | 0 (0%) | 1/23 (4%) | 0.77 |
| PA band | 0 | 7 | 0.08 |
| Shunt dur (yr) | 1.4 ± 0.5 | 7.1 ± 4.1 | <0.001* |
| Hemodynamic status | | | |
| Subaortic stenosis | 0 (0%) | 8 (27%) | 0.05 |
| EDP (mm Hg) | 8.6 ± 2 | 9.6 ± 4 | 0.37 |
| PAPm (mm Hg) | 16.1 ± 5.1 | 13.4 ± 4.8 | 0.12 |
| RAPm (mm Hg) | 5.5 ± 1.7 | 7.1 ± 3.2 | 0.06 |
| Sat (%) | 77 ± 6 | 83 ± 6 | 0.002* |
| PVR (RU·m²) | 2.4 ± 1.2 | 1.4 ± 0.6 | 0.001* |
| Qp (liters/min per m²) | 3.8 ± 2.8 | 5.6 ± 3.9 | 0.11 |
| Qs (liters/min per m²) | 4 ± 2 | 3.9 ± 1.7 | 0.89 |
| Qp/Qs | 1 ± 0.6 | 1.8 ± 1.7 | 0.10 |
| McGoon ratio | 2.2 ± 0.6 | 2.4 ± 0.8 | 0.46 |
| Hematocrit (%) | 49 ± 6 | 48 ± 7 | 0.67 |
| AV valve regurgitation (1 to 4) | 0.4 ± 0.6 | 0.2 ± 0.5 | 0.26 |

*Significant difference between groups. AoP = aortopulmonary; AV = atrioventricular; DORV = complex double-outlet right ventricle; EDP = main ventricular end-diastolic pressure; HLHS = hypoplastic left heart syndrome; LV = left ventricle; McGoon ratio = ratio of pulmonary artery dimensions; PA = pulmonary artery; PAPm = mean pulmonary artery pressure; PVR = indexed pulmonary vascular resistance; Qp = indexed pulmonary flow; Qs = indexed systemic flow; RAPm = mean right atrial pressure; RV = right ventricle; RV/LV = right and left ventricle if two chambers of adequate size present; Sat = systemic saturation; Shunt dur = length of palliation in years before the Fontan operation in patients requiring one or more aortopulmonary shunts; TA = tricuspid atresia or stenosis with right ventricular hypoplasia.

**Table 3.** Operative and Postoperative Variables in 47 Patients

| | Group 1 (n = 17) | Group 2 (n = 30) | p Value |
|---|---|---|---|
| Operative variables | | | |
| Fontan (type) | | | |
| Caval | 15 (88%) | 5 (17%) | <0.0001* |
| Björk | 0 (0%) | 3 (10%) | 0.47 |
| Kreutzer | 2 (12%) | 22 (73) | <0.0001* |
| CPB time (min) | 200 ± 34 | 155 ± 39 | 0.009* |
| Cross clamp time (min) | 78 ± 36 | 79 ± 39 | 0.95 |
| Core temp (°C) | 18.2 ± 0.9 | 18.7 ± 1.4 | 0.17 |
| Conduit | 0 (0%) | 5 (17%) | 0.20 |
| Add. proc. | 1 (6%) | 2 (7%) | 0.61 |
| PA repair | 7 (41%) | 6 (20%) | 0.22 |
| Hosp dur (days) | 22 ± 9 | 14 ± 5 | 0.001* |
| Postoperative variables | | | |
| Mortality | | | |
| Early | 0/17 (0%) | 4/30 (13%) | 0.30 |
| Late | 3/17 (18%) | 3/26 (13%) | 0.91 |
| Total | 3/17 (18%) | 7/30 (23%) | 0.93 |
| Arrhythmias (survivors) | | | |
| Early | 12/17 (71%) | 7/26 (27%) | 0.01* |
| Late | 4/14 (29%) | 10/23 (43%) | 0.58 |
| Late operation | 0/14 (0%) | 3/23 (13%) | 0.47 |
| Echo follow-up (mo) | 10.9 ± 9.4 | 25.2 ± 24.1 | 0.05 |
| Normal function | 12/14 (86%) | 15/20 (75%) | 0.74 |
| AV regurg | 0.9 ± 0.9 | 0.5 ± 0.6 | 0.09 |
| Sv regurg | 0.4 ± 0.7 | 0.5 ± 0.5 | 0.60 |
| Follow-up (mo) | 12 ± 7 | 33 ± 34 | 0.04* |
| NYHA functional class | 1.2 ± 0.4 | 1.6 ± 0.6 | 0.09 |

*Significant difference between groups; Add. proc. = additional surgical procedures required at the Fontan operation; AV regurg = echocardiographic quantitation of systemic atrioventricular valve regurgitation; Core temp = central venous temperature during cardiopulmonary bypass; CPB time = duration of cardiopulmonary bypass; Cross clamp = duration of aortic cross clamp time; Echo = echocardiographic; Hosp dur = duration of hospital stay; Normal function = echocardiographic evaluation of systemic ventricular function as normal or abnormal; NYHA = New York Heart Association; PA repair = pulmonary artery reconstruction at Fontan operation; SV regurg = echocardiographic quantitation of systemic semilunar valve regurgitation.

significantly longer for Group 2 patients. There was a trend for older patients before the Fontan procedure to have had two palliative shunts although this difference did not reach statistical significance.

Table 4 illustrates the shunt procedures performed before the Fontan operation, which have changed over time. More Group 1 patients had a 4-mm modified Blalock-Taussig shunt and more Group 2 patients had a classic Blalock-Taussig shunt. No Group 1 patients compared with seven Group 2 patients required pulmonary artery banding. The incidence of subaortic stenosis was greater in Group 2 and was directly related to the increased use of pulmonary artery banding early in our experience ($p < 0.001$). However the presence of a pulmonary artery band or the development of subaortic stenosis did not increase the risk of mortality in Group 2 patients ($p > 0.05$). The systemic oxygen saturation was lower and the pulmonary vascular resistance was higher in Group 1 patients even though the pulmonary flow, systemic flow and pulmonary to systemic flow ratio were similar in the two groups. All other preoperative variables were similar between groups and not associated with increased mortality.

Operative variables. The total cavopulmonary connection was performed more frequently in Group 1, whereas the Kreutzer right atrial to pulmonary artery direct anastomosis was performed more frequently in Group 2 because of changes in technique over time. There was no effect of age on mortality when the type of operative procedure was taken into account ($p = 0.50$, Mantel-Haenszel). The cardiopulmonary bypass time was longer in Group 1 patients whereas cross-clamp time and core temperature were similar in the two groups and also had no effect on mortality. In five Group

Table 4. Preoperative Palliative Shunt Procedures in 47 Patients

| Shunt Type | Group 1 (n = 14) | Group 2 (n = 33) | p Value |
|---|---|---|---|
| Waterston | 0 (0%) | 5 (22%) | 0.17 |
| Classic BT | 0 (0%) | 11 (48%) | 0.007* |
| Mod BT, 3 mm | 2 (14%) | 0 (0%) | 0.27 |
| Mod BT, 4 mm | 10 (72%) | 1 (4%) | <0.0001* |
| Mod BT, 5 mm | 2 (14%) | 5 (22%) | 0.90 |
| Glenn | 0 (0%) | 1 (4%) | 0.80 |

*Significant difference between groups. BT = Blalock-Taussig; Mod = modified.

2 patients early in our experience a conduit (valved in four patients, nonvalved in one patient) was used in the Fontan anastomosis. There was no difference between the groups, but the use of a conduit was associated with increased mortality ($p = 0.003$). Procedures in addition to the Fontan operation were performed with equal frequency in the two groups and included mitral valve repair in one infant in Group 1 and relief of subaortic stenosis in two Group 2 patients. Pulmonary artery reconstruction or enlargement was also performed with equal frequency in the two groups and was not associated with increased mortality ($p > 0.05$). The Group 1 infants required a longer hospital stay. No other operative variables were associated with an increase in mortality.

**Postoperative variables.** Postoperative mortality (early, late and total) did not differ between the groups. The four early deaths in Group 2 were related to low cardiac output: the three late deaths were in patients with subaortic stenosis ($n = 2$) or bacterial endocarditis ($n = 1$). The three deaths in Group 1 were late and primarily respiratory in nature, resulting in a low cardiac output state.

Our most recent experience since April of 1988, which includes all 17 infants <2 years of age and 9 older patients, also yielded similar mortality results in the two groups (early postoperative 0% vs. 11% [$p > 0.05$] and late postoperative 18% vs. 0% [$p > 0.05$]).

*Immediate postoperative arrhythmias* were more frequent in Group 1 and not associated with mortality in either group. Of the 12 Group 1 infants who developed an immediate postoperative arrhythmia, 6 had junctional rhythm that required only a brief period of atrial pacing at physiologic rates to maintain atrial-ventricular synchrony. The remaining six patients had supraventricular tachycardia (paroxysmal atrial tachycardia in four and junctional ectopic tachycardia in two) causing hemodynamic deterioration. All were treated successfully with antiarrhythmic medications, overdrive atrial pacing or moderate hypothermia. The use of the total cavopulmonary connection was not associated with a lower incidence of immediate postoperative arrhythmias in either Group 1 or Group 2 ($p > 0.05$ for all comparisons). However, the incidence of late arrhythmias in long-term survivors was similar although the duration of follow-up of Group 1 infants has been shorter. Of the four Group 1 infants with late arrhythmias, two had a bradyarrhythmia (junctional rhythm not requiring treatment) and two had a tachy-brady arrhythmia (one supraventricular tachycardia with junctional rhythm and one atrial flutter with junctional rhythm). Of the 10 Group 2 patients with late arrhythmias, 6 had a bradyarrhythmia (nonsinus atrial in 4, junctional in 1 and sinus bradycardia in 1); 3 had a tachy-brady arrhythmia (atrial flutter with junctional rhythm) and 1 patient had a tachyarrhythmia (atrial fibrillation).

*Late arrhythmias* also have not resulted in mortality in either group and appear to be well controlled with antiarrhythmic medication when indicated. One Group 1 infant with supraventricular tachycardia early postoperatively continues to take digoxin and two infants with late onset atrial flutter and supraventricular tachycardia are taking digoxin with flecainide and digoxin with procainamide, respectively. Three of the four Group 2 patients with late onset tachyarrhythmias are currently taking digoxin and procainamide. Two patients (one in each group) required a ventricular demand pacemaker for bradyarrhythmia. The occurrence of immediate arrhythmias did not predict the development of late arrhythmias ($p > 0.05$). Three patients in Group 2 required an additional operation after the Fontan procedure: in two for subaortic stenosis (one of these patients died) and in one patient for aortic valve replacement. The incidence of additional operations after the Fontan operation was similar in the two groups.

*The latest echocardiographic evaluation available for Group 1 (n = 14) and Group 2 (n = 20) patients*, obtained at similar postoperative intervals, did not differ with respect to ventricular function, systemic AV valve regurgitation and systemic semilunar valve regurgitation. In addition, the incidence of moderate or more severe AV valve or semilunar valve regurgitation was similar in Groups 1 and 2 (14% vs. 20% and 7% vs. 0%, $p > 0.05$ respectively). The New York Heart Association functional class of all patients was I or II although more Group 1 patients are receiving digoxin, diuretics and afterload-reduction since they were operated on more recently. One patient in Group 1 has a mild neurologic deficit and one patient in Group 2 has a right hemiparesis.

## Discussion

**Selection criteria for the Fontan operation.** Since Fontan et al. (5) first described the successful surgical treatment of tricuspid atresia in 1968, there has been a dramatic improvement in both early and late survival rates. This improvement has been related not only to improved techniques of surgical repair and myocardial preservation, but also to the development of more stringent selection criteria. Of the 10 preoperative criteria originally proposed by Choussat, Fontan and coworkers (1), there is to date little controversy over the view that lower pulmonary vascular resistance, less pulmonary artery distortion, larger pulmonary artery dimensions, less systemic AV valve regurgitation and better systemic ventricular function improve long-term survival (6,7). However, other preoperative selection criteria remain controver-

sial. The optimal age for the Fontan operation was initially suggested to be between 4 and 16 years (8). This suggestion was based on a report of 100 patients with tricuspid atresia who underwent the operation in the early 1980s. There was no apparent explanation why the younger patients did less well. Since that original description, controversy persists as to whether young age is deleterious for a successful outcome.

In 1983, Ishikawa et al. (9) described the successful hemodynamic results of the Kreutzer (direct right atrial to pulmonary anastomosis) procedure in three infants with tricuspid atresia who were <2 years of age. They commented that when a conduit is not used in the repair, the age could be ≤12 months. Kirklin et al. (10) reported on 102 patients with a wide variety of cardiac malformations who underwent the Fontan operation between 1975 and 1985; they noted that younger age (<4 years) was a risk factor for early postoperative mortality but that the risk was neutralized by a more recent date of operation. They concluded that, in the current era, the Fontan operation should be performed at a young age (2 to 4 years) to avoid the increasing main ventricular hypertrophy seen in older patients. With regard to infants <2 years of age they expressed the requirement for additional studies. In contrast, Bartmus et al. (2) recently reported that age <4 years is an independent risk factor for mortality (39% mortality at 1 year and 44% mortality at 4 years postoperatively). Nevertheless, in a select subgroup of these patients with one or no risk factors, survival was not detectably different from that of older patients.

**Risk factors.** Our overall and most recent experience suggests that young age (<2 years), independent of the type of Fontan repair performed, does not increase the risk of mortality when the preoperative anatomy and hemodynamic status are acceptable. Although the pulmonary vascular resistance was somewhat higher in our younger patients, it was generally comparable to previously reported values of ≤2 U·m$^2$, which are associated with a lower operative risk (11). The higher resistance may be a function of the different types of shunts performed or due to primary differences in the pulmonary vascular bed in younger patients with congenital heart disease. Despite these differences, the pulmonary artery dimensions, expressed as the McGoon ratio, were similar. According to a recent large series of 334 patients reported by Fontan et al. (7), a ratio <1.8 sharply increased the risk of death or takedown of the Fontan anastomosis within 30 days of operation. In our study, infants <2 years of age had a mean McGoon ratio of 2.2 (range 1.9 to 3.3) with no early mortality or Fontan anastomosis takedown required. Therefore, adequate pulmonary artery dimensions were not dependent on older age or a longer duration of palliation. The only variable associated with increased mortality was the use of a conduit in the atriopulmonary anastomosis, which is no longer used in the Fontan operation.

*Our rationale for performing an earlier Fontan operation* was to avoid the chronic hypoxemia, excess volume loading of the systemic ventricle and the associated operative risks that may occur with multiple and prolonged palliative aortopulmonary shunt procedures. Snyder et al. (12) demonstrated, in 40 children with cyanotic congenital heart disease amenable to a Fontan operation, that an aortopulmonary artery shunt was commonly associated with elevated pulmonary artery pressure and resistance, congestive heart failure and arterial distortion. They found that these factors did in turn result in a greater risk for a subsequent Fontan operation. We currently believe that the Fontan operation can be performed without undue risk in infants as young as 7 months in order to avoid a primary or secondary palliative aortopulmonary shunt when there are no other anatomic or hemodynamic contraindications. Assessment of whether an early Fontan procedure will improve long-term survival and ventricular performance remains hypothetical and requires longer follow-up. However, a recent report of outcome after a perfect Fontan procedure in a large series stated that only "older age" at the time of operation led to the late decline in survival and functional status (13). At this time, all our young patients remain in functional class I or II, and long-term follow-up of systemic ventricular function with echocardiography, cardiac catheterization and exercise testing is in progress.

**Arrhythmias.** A higher incidence of immediate postoperative arrhythmias in our younger patients is of concern. The use of the total cavopulmonary connection in 88% of the Group I infants appears not to have reduced the incidence of immediate postoperative arrhythmias as has been previously speculated (14). This may be related to operating on a less mature and structurally smaller heart with a greater risk of injury to the sinus node and intraatrial conduction. However, only 6 of the 12 patients with immediate postoperative arrhythmias were in hemodynamically unstable condition and all 6 were treated successfully with no mortality. Nevertheless, the higher incidence of immediate postoperative arrhythmias in the younger patients did contribute to their longer hospital stay. Although the incidence of late arrhythmias is currently somewhat greater in the older group (though not significant), the duration of follow-up is shorter in the younger patients and thus evaluation of the benefit of the total cavopulmonary connection will require longer follow-up. A previous study (15) has shown that as follow-up duration increases the percent of arrhythmia-free patients declines and thus differences between our groups may emerge.

**Conclusions.** A successful Fontan operation can be performed in young children without increased risk. Our findings suggest that infants <2 years of age may be suitable candidates for an early Fontan operation if the preoperative hemodynamic status is good and that an early operation may avoid complications associated with multiple and prolonged palliative procedures. Although immediate postoperative arrhythmias occur more frequently in infants and most are hemodynamically insignificant, the use of the total cavopul-

monary connection appears not to have reduced the incidence of this complication. Whether the introduction of the fenestrated Fontan operation (16) will reduce the incidence of early arrhythmias and further decrease morbidity remains speculative. With regard to ventricular performance, late arrhythmias and long-term survival, assessment of the benefit of early Fontan repair will require long-term follow-up.

## References

1. Choussat A, Fontan F, Besse P, Vallot F, Chavc A, Bricaud H. Selection criteria for Fontan's procedure. In: Anderson RH, Shinebourne EA, eds. Pediatric Cardiology. Edinburgh: Churchill Livingstone, 1977:559–66.
2. Bartmus DA, Driscoll DJ, Offord KP, et al. The modified Fontan operation for children less than 4 years old. J Am Coll Cardiol 1990;15: 429–35.
3. Piehler MJ, Danielson GK, McGoon DC, et al. Management of pulmonary atresia with ventricular septal defect and hypoplastic pulmonary arteries by right ventricular outflow construction. J Thorac Cardiovasc Surg 1980;80:552–67.
4. Garson A. The Electrocardiogram in Infants and Children: a Systematic Approach. Philadelphia: Lea & Febiger, 1983:61–79.
5. Fontan F, Mounicot FB, Baudet E, Simonneau J, Gordo J, Gouffrant JM. Correction de l'atresie tricuspidenne: Raport de deux cas "corriges par l'utilisation d'une technique chirurgicale nouvelle". Ann Thorac Cardiovasc 1971;10:39–45.
6. Myers JL, Waldhausen JA, Weber HS, et al. A reconsideration of risk factors for the Fontan operation. Ann Surg 1990;211:738–44.
7. Fontan F, Fernandez G, Costa F, et al. The size of the pulmonary arteries and the results of the Fontan operation. J Thorac Cardiovasc Surg 1989;98:711–24.
8. Fontan F, Deville C, Quaegebeur J, et al. Repair of tricuspid atresia in 100 patients. J Thorac Cardiovasc Surg 1983;85:647–60.
9. Ishikawa T, Neutze JM, Brandt PW, Barratt-Boyes BG. Hemodynamics following the Kreutzer procedure for tricuspid atresia in patients under 2 years of age. J Thorac Cardiovasc Surg 1984;88:373–9.
10. Kirklin JK, Blackstone EH, Kirklin JW, Pacifico AD, Bargeron LM. The Fontan operation: ventricular hypertrophy, age, and date of operation as risk factors. J Thorac Cardiovasc Surg 1986;92:1049–64.
11. Mayer JE, Helgason H, Jonas RA, et al. Extending the limits for modified Fontan procedures. J Thorac Cardiovasc Surg 1986;92:1021–8.
12. Snyder MM, Lang P, Mayer JE, Jonas RA, Castenada AR, Lock JE. Childhood systemic-pulmonary shunts: subsequent suitability for Fontan operation. Circulation 1987;76(suppl III):III-39–44.
13. Fontan F, Kirklin JW, Fernandez G, et al. Outcome after a "perfect Fontan operation". Circulation 1990;81:1520–36.
14. De Leval MR, Kilner P, Gewillig M, Bull C. McGoon DC. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. J Thorac Cardiovasc Surg 1988; 96:682–95.
15. Weber HS, Hellenbrand WE, Kleinman CS, Perlmutter RA, Rosenfeld LE. Predictors of rhythm disturbances and subsequent morbidity after the Fontan operation. Am J Cardiol 1989;64:762–7.
16. Bridges ND, Lock JE, Castanada AR. Baffle fenestration with subsequent transcatheter closure. Circulation 1990;82:1681–9.