The Fontan Procedure in Adults

Richard N. Gates, MD, Hillel Laks, MD, Davis C. Drinkwater, Jr, MD, Lydia Lam, BS, Arie Blitz, MD, John S. Child, MD, and Joseph K. Perloff, MD

Division of Cardiothoracic Surgery, Department of Surgery, and Division of Cardiology, Department of Medicine, University of California, Los Angeles, School of Medicine, Los Angeles, California

Background. A retrospective clinical study was performed to document the course of adult patients undergoing the Fontan procedure.

Methods. Between 1982 and 1994, 21 adults aged 18 to 40 years (mean age, 27 ± 7 years) underwent a Fontan procedure. Anatomic diagnosis was tricuspid atresia in 9, double-inlet left ventricle in 4, and various single ventricles in 8. Four underwent a right atria–right ventricle connection, 13 had a right atria–pulmonary artery connection, and 4 had a lateral-tunnel Fontan. Three of these 4 had a snare-adjustable atrial septal defect. Preoperative risk factors assessed were left ventricular end-diastolic pressure greater than 10 mm Hg, ejection fraction lower than 0.45, mean pulmonary artery pressure higher than 15 mm Hg, transpulmonary gradient greater than 10 mm Hg, pulmonary artery abnormalities, and atrioventricular valve regurgitation. Mean preoperative risk score was 1.6 ± 1.1. Mean New York Heart Association class was 2.6 ± 0.5.

Results. The operative mortality rate was 5% (1/21). Six patients (30%) had a major complication, four being prolonged effusions. One patient was lost to follow-up; the remaining 20 have been followed for a mean of 7.4 ± 3.8 years. At follow-up, mean New York Heart Association class was 1.7 ± 0.5. There has been one late death (5%) at 9½ years, which was probably due to ventricular arrhythmia. Three patients (16%) have required and survived reoperation. During follow-up, 7 patients (37%) have had development of atrial arrhythmias requiring medication, and 2 have been treated for ventricular arrhythmias.

Conclusions. These results indicate that properly selected adults can undergo the Fontan procedure with low morbidity and mortality. However, late-developing arrhythmias, need for reoperation, and decreasing ventricular function are serious problems that mandate careful follow-up.

Table 1. Summary of Patient Data

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<th>Patient No.</th>
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<th>Operations</th>
<th>LVEDP (mm Hg)</th>
<th>EF &lt;0.45</th>
<th>PAP (mm Hg)</th>
<th>AVV Regurgitation</th>
<th>PA Abnormalities</th>
<th>TPG mm Hg</th>
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AA = atrial arrhythmia; AVV = atrioventricular valve; B-T = Blalock-Taussig; DILV = double-inlet left ventricle; DORV = double-outlet right ventricle; EF = ejection fraction; LVEDP = left ventricular end-diastolic pressure; NA = not available or not applicable; NYHA = New York Heart Association; PA = pulmonary artery; PAB = pulmonary artery banding; PAP = pulmonary artery pressure; TA = tricuspid atresia; TPG = transpulmonary gradient; VA = ventricular arrhythmia.
volume left lung. This is anatomically the opposite of the classic Glenn shunt.

In a retrospective manner, an attempt was made to assess the operative risk of these patients. As risk assessment for the Fontan procedure has evolved substantially over the last decade, the selection factors were not specifically used at the time of each patient's surgical procedure to determine operability. Although the selected factors are somewhat arbitrary, they could be reliably assessed for all patients over the 13-year span of this review. As such, the following risk factors were assessed: left ventricular end-diastolic pressure higher than 10 mm Hg, ejection fraction lower than 0.45, mean pulmonary artery pressure greater than 15 mm Hg, transpulmonary gradient greater than 10 mm Hg (defined as preoperative mean pulmonary artery pressure minus left atrial pressure), pulmonary artery abnormalities requiring operative repair, and atrioventricular valve regurgitation of a moderate degree or greater. One point was given for each preoperative risk factor identified, and the mean score was 1.6 ± 1.1 (range, 0 to 4).

Age range was 18 to 40 years (mean age, 27 ± 7 years), and there were 12 men and 9 women. Mean preoperative New York Heart Association (NYHA) class was 2.6 ± 0.5. Indications for operation were variable and included major or increasing cyanosis with marked polycythemia, increasing functional limitation, or concern about chronic volume overload with increasing ventricular size in patients with a previously placed shunt.

A summary of the patient data is shown in Table 1.

Results

Early Mortality

There was one early death (5%) in this series. The patient was a 23-year-old woman with heterotaxia/asplenia syndrome, double-outlet right ventricle with unbalanced atrioventricular canal, malpositioned great vessels, and total anomalous pulmonary venous connections. She had undergone no prior palliative procedures and had a preoperative room air saturation of 89% and was in NYHA class III. Risk factors included a left ventricular end-diastolic pressure of 11 mm Hg, and an ejection fraction of approximately 0.45. Pulmonary artery pressure was never directly measured. She underwent a lateral-tunnel, snare-adjustable atrial septal defect Fontan operation. Postoperatively she had markedly elevated right atrial pressures. Low cardiac output syndrome persisted despite atrial septal defect decompression. A ventricular assist device was emergently placed prior to Fontan takedown. However, multiple organ system failure developed, and the patient did not recover. Because of the small series size and the low number of deaths, no statistically proper risk-factor analysis could be performed.

Late Mortality

There has been one late death among the 20 early survivors. A 32-year-old woman died suddenly and alone 9 3/4 years after operation. The patient had tricuspid atresia and had undergone a right atria–right ventricle conduit type of Fontan repair. One year postoperatively, she was noted to have atrioventricular node reentry tachycardia and was treated with amiodarone hydrochloride and an AAI pacemaker. Subsequently, amiodarone had to be withdrawn, and radiofrequency ablation of the atrioventricular node with placement of a DDD pacemaker was performed. She continued to have both atrial and ventricular arrhythmias and was being medically treated for them at the time of death. The actuarial survival rate at 3, 6, and 12 years was 95%, 95%, and 81%, respectively (Fig 1).

Morbidity

Six patients (6/20 or 30%) sustained a major complication in the postoperative period. Prolonged pleural effusions requiring extended chest tube drainage developed in 4 patients. Two were treated with chest tube pleural sclerosis. All prolonged effusions occurred early in the series. Of the 3 patients treated with a snare-adjustable atrial septal defect, lateral-tunnel Fontan, none had substantial pleural drainage. Complete heart block developed in 1 patient after operation. This patient had essentially congenitally corrected transposition of the great vessels with
a hypoplastic right ventricle. A DDD pacemaker was placed without event. A symptomatic superior vena cava thrombus, which was probably related to an internal jugular line, developed in another patient and was treated with streptokinase and heparin sodium with resolution.

Reoperations

Three patients (3/19 or 16%) have required reoperation. Early in the series, 1 patient had development of rapid obstruction of a right atria–right ventricle Dacron conduit. It was replaced 1 year postoperatively, and the patient has done well since. The second patient had a late atrial baffle leak. This led to slowly progressive desaturation, which required reoperation 4 years after the initial right atria–pulmonary artery type of Fontan procedure. At revision, the patient was converted to a lateral-tunnel, snare-adjustable atrial septal defect Fontan. The third patient had originally undergone a right atria–pulmonary artery type of Fontan and late after operation had development of an enlarged atrium with atrial fibrillation. Right atrial pressure was elevated, and moderate atrioventricular valve regurgitation was present. The patient was converted to a lateral-tunnel, snare-adjustable atrial septal defect Fontan 11 years after the original procedure. All patients requiring reoperation survived and had an uncomplicated postoperative course. The rate of actuarial freedom from reoperation at 3, 6, and 12 years was 94%, 87%, and 69%, respectively (Fig 2).

Arrhythmias

At the time of the initial Fontan operation, no patient had a history of major atrial arrhythmias requiring medical therapy. Postoperatively, at varying intervals, 7 patients (37%) experienced such arrhythmias. In patients treated with a right atria–pulmonary artery type of Fontan procedure, atrial enlargement has been associated with atrial fibrillation. One patient, as already mentioned, underwent conversion to a lateral tunnel with valve repair. Two patients have had ventricular arrhythmias, which have been treated medically. One of them, already discussed, died suddenly 9 1/4 years after operation, and the other has progressively deteriorating ventricular function and will likely undergo heart transplantation within 1 or 2 years. An additional 2 patients have had complete heart block remote from the Fontan procedure. In 1 of them it was therapeutically induced to treat reentrant atrioventricular node tachycardia, and in another it occurred spontaneously. Both patients had DDD pacemakers placed.

NYHA Class

At mean follow-up of 7.4 ± 3.8 years, the mean NYHA class of the survivors was 1.7 ± 0.5. Of the 6 patients followed up less than 5 years, 3 were in NYHA class I and 3, NYHA class II. Of the 13 patients followed up 5 years or more, 3 were in NYHA class I and 10, NYHA class II.

Comment

Fontan originally suggested that his procedure was best performed on children between the ages of 4 and 15 years [10]. More recently, the prevailing view has been that the procedure ideally is performed between 18 months and 6 years of age [5, 6, 9]. In general, the move toward earlier definitive palliation has been prompted by the appreciation of risk factors for the procedure and their associated contributing factors. Of most significance appear to be ventricular hypertrophy with elevated left ventricular end-diastolic pressure [6, 11] and increased pulmonary vascular resistance owing to many causes [4, 5, 8, 10]. These two conditions contribute to elevated postoperative right atrial pressure, which is a significant determinant of early death [12] as well as a predictor of long-term complications such as atrial enlargement with thrombosis or arrhythmia and protein-losing enteropathy. Thus, it is generally believed that conversion to a total cavopulmonary circulation with early removal of necessary systemic–pulmonary artery shunts or pulmonary artery bands will improve both the early mortality and the long-term results of the Fontan procedure.

Nevertheless, occasional adult patients are encountered who are anatomic candidates for the Fontan procedure but have not undergone definitive palliation.
we chronicle our experience with 21 such patients palliated with various modifications of the Fontan procedure. The mean NYHA class before operation was 2.6 ± 0.5, indicating that the patients were fairly symptomatic and limited at the time of operation. At mean follow-up of 7.4 ± 3.8 years, the NYHA class of survivors was 1.7 ± 0.5, suggesting that the majority of patients were enjoying substantial symptomatic improvement. Of the 6 patients followed up less than 5 years, 3 were in NYHA class I and 3, in class II. Of the 13 patients followed up 5 years or more, 3 were in NYHA class I and 10, in class II. This suggests that increasing disability is to be anticipated in this group as time from operation is extended and is similar to the findings of Fontan and co-workers [7] in a larger and more age-diverse group.

Early mortality in this series was 1 (5%) of 21 patients. This is similar to the experience of Mayer [5], Kirklin [6], Mair [13], Humes [14], Mair [15], and their associates, who reported a mortality rate of 8% for a group of 25 patients older than 15 years, 7% for a group of 14 patients older than 20 years, 6% for a group of 16 patients more than 18 years old, 6% for a group of 77 patients older than 17 years, and 10% for a group of 121 patients greater than 17 years old, respectively. However, it is difficult to directly compare these studies with our study, as specific and individual patient risk factor were not provided in the other studies.

To present our results in this age group with reference to some currently accepted risk factors for the Fontan operation, we developed a system for assessing risk that could be applied over the 13 years of the study. One point each was given for left ventricular end-diastolic pressure greater than 10 mm Hg, ejection fraction of less than 0.45, mean pulmonary artery pressure higher than 15 mm Hg, transpulmonary gradient greater than 10 mm Hg (as previously defined), pulmonary artery abnormalities requiring operative repair, and atrioventricular valve regurgitation of a moderate degree or greater. Mean score was 1.6 ± 1.1 with a range of 0 to 4 (see Table 1). These data may serve as a benchmark for anticipated results for patients with similar profiles and imply that low operative mortality can be expected in this age range in a well-selected moderate-risk group. The only early death that occurred involved a 23-year-old woman with heterotaxia/asplenia syndrome, double-outlet right ventricle with unbalanced atrioventricular canal, malpositioned great vessels, and total anomalous pulmonary venous connections. She was at increased risk with a left ventricular end-diastolic pressure higher than 10 mm Hg, an ejection fraction lower than 0.45, and pulmonary artery pressures that were not directly measured. Although not assessed as a risk factor in this study, the asplenia/polyasplenia syndromes also convey increased risk [16].

The actuarial survival rate in this series at 3, 6, and 12 years was 95%, 95%, and 81%, respectively. The one late death occurred 9½ years postoperatively and was unwitnessed and sudden. However, this patient had a history of ventricular arrhythmia and declining ventricular function, and her death was quite likely cardiac related. With a mean follow-up of 7.4 ± 3.8 years and 6 patients followed longer than 10 years, this study suggests that the modified Fontan procedure does provide very good palliation and systematic improvement in this older age group over a 10-year period. Nonetheless, a significant incidence of late atrial arrhythmias and a decline in the functional status and ventricular function of some patients followed lead us to believe that the survival curve will continue to decline in the 10- to 20-year interval. Such a trend would be similar to the one in the report of Fontan and colleagues [7] on a heterogeneous group of 334 patients.

Morbidity in this series was related primarily to prolonged pleural drainage, which occurred in 4 patients (19% overall). This complication was seen only early in the series; none of the 3 recent patients treated with a lateral-tunnel, snare-adjustable atrial septal defect Fontan procedure and neither of the 2 having reoperation with a lateral-tunnel, snare-adjustable atrial septal defect Fontan sustained prolonged drainage. We believe this is a direct result of the lower right atrial pressure, which can be individually tailored with the snare-adjustable atrial septal defect [17]. At a time remote from operation, the snare-adjustable atrial septal defect can be completely closed with elevation of right atrial pressure but without the frequent occurrence of pleural effusion [18].

Three patients (16%) required reoperation, and the rate of actuarial freedom from reoperation was 94%, 87%, and 69% at 3, 6, and 12 years, respectively. The cause of two reoperations was related to technique—a late atrial baffle leak and rapid obstruction of a right atria–right ventricle Dacron conduit. The final reoperation was of most interest and was pursued electively. Eleven years previously, this patient had undergone a right atria–pulmonary artery type of Fontan procedure and had subsequently had development of a grossly dilated atrium, atrial dysrrhythmias, atrioventricular valve regurgitation, and elevated right atrial pressure. The patient was converted to a lateral-tunnel, snare-adjustable atrial septal defect Fontan with resection of excess atrial tissue and valve repair. This approach, which has been used in other patients with late atrial dilatation and arrhythmias [19], was helpful in controlling the atrial rhythm and improving the patient’s functional status. Whether this approach will yield long-term improvement has yet to be determined, but early results are encouraging.

The most disturbing long-term follow-up problem encountered has been atrial arrhythmias. At varying intervals, 7 patients (37%) have had atrial arrhythmias requiring medical therapy. This incidence is similar to the 32% and 57% incidence reported by Peters and Somerville [20] and Cromme-Dijkstra and colleagues [21], respectively, in their mid-term and long-term follow-up of Fontan patients. It is important to note, however, that this high incidence of atrial arrhythmias is essentially no different from that reported for adult patients with tricuspid atresia and normally related or transposed great arteries who have not undergone a Fontan type of procedure [22, 23]. Thus, there is no clear evidence that these arrhythmias are a direct consequence of Fontan palliation.
Nonetheless, it is clear that atrial enlargement, elevated pulmonary artery pressure, elevated right atrial pressure, and older age are risk factors for the development of late atrial arrhythmias [24]. Operative strategies, such as the lateral-tunnel Fontan or partial Fontan, that attempt to limit atrial enlargement and maintain low right atrial pressures should be aggressively pursued. This is particularly true in the older age group. More recent reports suggesting that atrial incisions themselves may be in part responsible for late atrial arrhythmias argue for the extracardiac conduit approach to the lateral-tunnel Fontan concept [25]. Such an approach may be ideally suited to the low-risk adult patient, as outgrowth of the conduit is not a concern. Nonetheless, in higher risk patients, if an extracardiac conduit is used, an approach that can incorporate an adjustable atrial septal defect is recommended.

Older patients in whom atrial arrhythmias develop should be screened for atrial enlargement, atrial thrombosis, ventricular function, and valvular regurgitation. Serious consideration should be given to valvular repair or atrial reduction with conversion to a lateral-tunnel Fontan procedure in patients with acceptable operative risk and decreased functional status related to the recent onset of atrial arrhythmia. Ventricular function should be carefully noted in this group of patients. Our experience has been that patients with ventricular enlargement, a significantly depressed ejection fraction, and atrial arrhythmias represent a high-risk group for reoperation and that cardiac transplantation should be considered.

References