Long-term results after the Fontan operation for tricuspid atresia

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ABSTRACT Between 1968 and 1977, 32 patients who underwent the Fontan operation for tricuspid atresia at Bordeaux, France, or Leiden, The Netherlands, survived at least 1 year after surgery. These patients were evaluated clinically 7 to 16 years (mean 8.9 years) after surgery by review of clinical records, questionnaire, or direct examination. There were five deaths: one during arrhythmia, the second sudden and unexplained, and three others after reoperation. Four of the five patients had evidence of obstruction of the atroventricular or atroventricular conduit. There were eight reoperations. Two were for residual atrial septal defects, one for an atrial septal defect and an intrapulmonary arteriovenous fistula, and one for a residual ventricular septal defect. The other four were reoperated for severe conduit or homograft obstruction. One of these four also had a residual shunt, severe mitral regurgitation, and reduced left ventricular function. One patient was found to have left ventricular dysfunction 3 months after surgery, which persisted. Another patient had left ventricular dysfunction unmasked after successful mitral annuloplasty for severe mitral regurgitation. No other patients had clinical evidence of left ventricular dysfunction. Recurrent supraventricular tachycardia occurred in four patients and was eventually fatal in one patient with associated conduit obstruction. Of the 27 survivors, 13 are in NYHA class I, 13 in class II, and one is in class III. We conclude that clinical results of the Fontan operation remain good in most patients 7 to 16 years after surgery. Conduit or homograft obstruction has been associated with mortality, reoperation, and poor hemodynamic tolerance of arrhythmia.


THE FONTAN OPERATION has been used with increasing success in the treatment of patients with tricuspid atresia.1-4 The operative mortality is relatively low, ranging from 7% to 17% in recent reports.1-4 Most survivors have experienced symptomatic improvement.1-4 The Fontan operation results in a circulation characterized by absent or normal right ventricular function, right atrial and systemic venous hypertension, and dependence on left ventricular function. Because of these features there is continuing concern about intermediate and long-term results.5

The purpose of this report is to present the follow-up status of 32 patients who underwent the Fontan operation between April 1968 and December 1977 and who survived at least 1 year. This study includes the first patients6,7 who had this operation and thus provides the longest available follow-up.

Patients and methods

From April 1968 to December 1977, 43 patients with tricuspid atresia underwent surgical repair at the Hôpital Cardiologique du Haut-Lévêque, l’Université de Bordeaux, and at the University Hospital of Leiden. Thirty-two patients survived at least 1 year after surgery. Their course and present status form the basis for this report.

At the time of surgery, the patients’ ages ranged from 4 to 36 years (mean 11.1 years). There were 16 male and 16 female patients. All patients were cyanotic and had decreased exercise tolerance. All were in sinus rhythm. All had undergone measurement of intracardiac and pulmonary arterial pressures at a preoperative catheterization or at operation before institution of extracorporeal circulation. The pulmonary vascular resistance was estimated to be less than 4 units/m² in all patients. Twenty patients had had “shunt” operations, including five with a Glenn anastomosis, seven with a Blalock-Taussig shunt, three with both a Glenn anastomosis and a Blalock-Taussig shunt, two with a central aorta to main pulmonary artery anastomosis, and three with a Waterston anastomosis.

Twenty-five patients had tricuspid atresia with absence of the right atroventricular connection, left atrium connecting to left
ventricle, and ventriculoarterial concordance. In 24 of these, an
atrioventricular connection was established and in the other
patient a direct atropulmonary anastomosis was done. Eight
patients had a Glenn anastomosis as part of the final con-
nexions. In one case, the Glenn was done simultaneously and in the
other seven it had been done previously. An inferior vena caval
valve was used in two cases.

Seven patients had an absent right atrioventricular con-
nection, a left atrium connecting to a left ventricle, and ventricu-
loarterial discordance. A right atrium–to–pulmonary artery con-
nexion was made with an aortic valve homograft in all seven
patients. A simultaneous Glenn operation and placement of an
inferior vena caval valve was performed in one.

Because this study was designed to determine intermediate
and long-term results of the Fontan operation, we have studied
only those patients who underwent surgery before December
31, 1977. We have limited our study to patients who survived at
least 1 year after surgery. The details of surgery and of the early
postoperative period are included in previous reports.2, 6-8

For all 32 patients, current status and the clinical course from
1 year after the operation to the present were obtained by review
of the clinical records at the Hôpital Cardiologique de the Uni-
versité de Bordeaux and at the University Hospital of Leiden, by
a questionnaire sent to the patient’s physician or by direct ex-
amination. The patients were evaluated during 1984 and early
1985, providing a 7 to 16 year follow-up (mean 8.9 years) after
surgery. Angiographic evaluation of left ventricular and mitral
valve function was available in 16 patients. Mitral valve pro-
lapse was diagnosed by standard angiographic criteria.9 Con-
firmation by echocardiographic criteria10 was not possible because
the echocardiograms were not available for review. Other re-
cently proposed criteria11 were not available during the study.

Results (table 1)

Mortality. There were five deaths. A 13-year-old girl
had undergone repair with placement of a 20 mm atrio-
ventricular Dacron conduit at 6 years of age. Cardiac
catheterization initially showed no conduit obstruc-
tion, but a repeat cardiac catheterization 5 years after
operation because of chronic peripheral edema showed
a 10 mm Hg mean gradient between the right atrium
and pulmonary artery. The patient died suddenly 6½
years after the operation.

A 19-year-old man had undergone surgical repair
that included placement of a 20 mm atrioventricular
Dacron conduit at 10 years of age. In the early postop-
erative period the patient began to experience recur-
rent supraventricular tachycardia. Cardiac catheterization 8
years postoperatively revealed a right atrial mean pres-
sure of 9 mm Hg and a mean pulmonary arterial pres-
sure of 4 mm Hg. The angiogram showed a somewhat
narrow conduit lumen. At 8½ years after surgery the
patient developed atrial flutter associated with low car-
diac output and hypotension. The patient died during
attempted cardioversion.

The third death occurred in a 51-year-old woman
who was the second patient to undergo the Fontan
operation. The operation included placement of both
atriopulmonary and inferior vena caval homograft
TABLE 1
Results after the Fontan operation for tricuspid atresia in 32
patients surviving more than 1 year (7 to 16 year follow-up)

<table>
<thead>
<tr>
<th></th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mortality (n = 5)</td>
<td></td>
</tr>
<tr>
<td>Sudden death (known conduit obstruction)</td>
<td>1</td>
</tr>
<tr>
<td>Arrhythmia (known conduit obstruction)</td>
<td>1</td>
</tr>
<tr>
<td>After reoperation (2/3 had known conduit or homograft obstruction)</td>
<td>3</td>
</tr>
<tr>
<td>Reoperation (n = 8)</td>
<td></td>
</tr>
<tr>
<td>Conduit or homograft obstruction</td>
<td>4</td>
</tr>
<tr>
<td>Intrapulmonary AV fistula, ASD</td>
<td>1</td>
</tr>
<tr>
<td>Residual ASD</td>
<td>2</td>
</tr>
<tr>
<td>Residual VSD</td>
<td>1</td>
</tr>
<tr>
<td>Systemic venous congestion</td>
<td>8</td>
</tr>
<tr>
<td>LV dysfunction</td>
<td>2</td>
</tr>
<tr>
<td>Arrhythmias (n = 4)</td>
<td></td>
</tr>
<tr>
<td>Atrial fibrillation</td>
<td>1</td>
</tr>
<tr>
<td>Recurrent supraventricular tachycardia</td>
<td>3</td>
</tr>
<tr>
<td>Cyanosis (n = 6)</td>
<td></td>
</tr>
<tr>
<td>Residual ASD, obstructed homograft</td>
<td>1</td>
</tr>
<tr>
<td>Intrapulmonary shunt related to preexisting Glenn anastomosis</td>
<td>2</td>
</tr>
<tr>
<td>Unexplained</td>
<td>3</td>
</tr>
<tr>
<td>MV abnormalities (n = 4)</td>
<td></td>
</tr>
<tr>
<td>MV prolapse with MR</td>
<td>2</td>
</tr>
<tr>
<td>MV prolapse without MR</td>
<td>2</td>
</tr>
</tbody>
</table>

AV = arteriovenous; ASD = atrial septal defect; VSD = ventricular septal defect; LV = left ventricular; MV = mitral valve; MR = mitral regurgitation.

valves. The patient did well initially but later devel-
oplated atrial fibrillation, ascites, and peripheral edema.
Catheterization 14 years postoperatively showed an
obstructed atropulmonary homograft, residual shunts
to the left atrium and pulmonary artery, and severe
mitral regurgitation. At reoperation, the homograft
was replaced, the shunts were closed, and a mitral
valvuloplasty was performed. The patient initially im-
proved but died 6 months after reoperation.

The fourth death occurred in a 15-year-old girl with
ventriculoarterial concordance and previous Glenn an-
astomosis who underwent repair with placement of an
atrioventricular conduit. Two and one-half years later,
hemoptysis and cyanosis prompted studies that indi-
cated a right lower lobe arteriovenous fistula and a
persistent atrial septal defect. A right lower lobectomy
was performed without complication. Six days later
the atrial septal defect was closed through a right tho-
racotomy. The patient died of cerebral complications
thought to be caused by air embolism.

The fifth death occurred in an 18-year-old woman
with ventriculoarterial discordance who had under-
went repair with placement of an 18 mm atrioventricu-
lar Dacron conduit at 7 years of age. The patient did very well during the first 8 years after surgery but then developed symptoms related to conduit obstruction. The patient died after reoperation to replace the conduit 10½ years after the first operation.

**Reoperation.** There were eight reoperations performed in patients surviving more than 1 year after the initial surgery. Three of these patients died (see above), and five are living. Two of these required closure of residual atrial septal defects. A third patient had closure of a residual ventricular septal defect 1 month postoperatively. The remaining two patients developed severe obstruction of the atroventricular Dacron conduit and underwent reoperation 8 and 9 years, respectively, after the initial surgery. The former developed a protein-losing enteropathy. Cardiac catheterization revealed a mean gradient across the conduit of 9 mm Hg, and angiography showed severe obstruction. At reoperation the conduit was almost totally occluded by neointima and was replaced with an aortic homograft. After reoperation the patient’s exercise tolerance improved markedly and there were no further signs of the protein-losing enteropathy. The other patient underwent replacement of the atroventricular conduit with an aortic homograft with complete relief of symptoms.

**Functional status.** Of the 27 survivors, 13 are in NYHA class I, 13 are in class II, and one is in class III (table 2).

**Reproduction.** Only one of our patients is known to have offspring. A 39-year-old woman with a small residual atrial septal defect and a small left ventricle–to–pulmonary artery shunt was mildly cyanotic and delivered vaginally a small (1000 g) but otherwise normal infant at 36 weeks gestation, 3 years after surgery. The pregnancy resulted in moderate fluid retention, easily controlled with diuretics, and terminated in premature spontaneous labor and delivery. The infant is now a healthy 12-year-old girl.

**Systemic venous congestion.** Eight patients have significant hepatomegaly and/or peripheral edema or ascites. Six patients still require diuretics and three receive digitalis.

**Clinical assessment of left ventricular function.** One patient has clinical findings indicative of reduced left ventricular function. This patient had normal left ventricular function with a left ventricular ejection fraction of 58% before surgery. Cardiac catheterization 3 months postoperatively showed an ejection fraction of 41%. One year later, the ejection fraction had fallen to 34%. The cause for the reduced left ventricular contractility is unknown. A second patient, described above, showed evidence of reduced left ventricular function after reoperation for a residual atrial septal defect, residual left ventricle–to–pulmonary artery shunt, obstructed aortic homograft, and severe mitral regurgitation.

**Arrhythmias.** Four patients have experienced cardiac arrhythmias. One patient, described above, began to experience intermittent atrial fibrillation 4 years after surgery. The arrhythmia was controlled with digoxin and disopyramide. The patient later developed atrio-pulmonary homograft obstruction and mitral regurgitation but continued to tolerate recurrent atrial fibrillation relatively well. A second patient, also discussed above, began to experience recurrent supraventricular tachycardia shortly after surgery. The arrhythmia appeared to be relatively well controlled on disopyramide. Cardiac catheterization showed a 5 mm Hg gradient across the atroventricular Dacron conduit and angiography showed moderate narrowing of the conduit lumen. The patient developed atrial flutter that resulted in hypotension. Attempts at cardioversion were unsuccessful and the arrhythmia terminated fatally. Two additional patients have had recurrent supraventricular tachycardia. One has benefited from therapy with amiodarone and the other has not received medication.

**Cyanosis.** Six patients were cyanotic. In one of these, the cyanosis is explained by the presence of a residual atrial septal defect and obstructed valved homograft. In two others, cyanosis has persisted despite closure of residual atrial septal defects and is unexplained. In the fourth patient, cardiac catheterization did not reveal any right-to-left shunt and the cyanosis remains unexplained. In the remaining two patients, arteriovenous shunting in the right lung was related to a preexisting Glenn anastomosis.

**Hepatic function.** Seven patients had laboratory determination of total bilirubin, SGOT, and total protein (table 3). Four of the patients had at least one abnormal

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**TABLE 2**

<table>
<thead>
<tr>
<th>Year</th>
<th>No. of patients</th>
<th>NYHA classification</th>
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<tbody>
<tr>
<td>1968</td>
<td>1</td>
<td>I II III IV</td>
</tr>
<tr>
<td>1973</td>
<td>1</td>
<td>I II III IV</td>
</tr>
<tr>
<td>1974</td>
<td>5</td>
<td>I II III IV</td>
</tr>
<tr>
<td>1975</td>
<td>3</td>
<td>I II III IV</td>
</tr>
<tr>
<td>1976</td>
<td>9</td>
<td>I II III IV</td>
</tr>
<tr>
<td>1977</td>
<td>8</td>
<td>I II III IV</td>
</tr>
<tr>
<td>Total</td>
<td>27</td>
<td>13 13 1 0</td>
</tr>
</tbody>
</table>

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TABLE 3
Hepatic function in seven patients

| Patient | Follow-up (yr) | Total bilirubin (NL <1.2 mg/100 ml) | SGOT (NL <40 IU/l) | Total protein (NL >5.5 g/100 ml) |
|---------|----------------|-----------------------------------|-------------------|---------------------------------
| 1       | 6              | 0.7                               | 97                | 3.9                              |
| 2       | 7              | 1.2                               | 22                | 6.5                              |
| 3       | 8              | 0.8                               | 7                 | 7.1                              |
| 4       | 9              | 0.7                               | 15                | 7.0                              |
| 5       | 9              | 2.0                               | 9                 | 6.3                              |
| 6       | 10             | 1.6                               | 11                | 7.4                              |
| 7       | 14             | 1.3                               | 15                | 7.3                              |

NL = normal.

value. One of these, whose course was complicated by severe edema and hepatomegaly, had an elevated SGOT and decreased total proteins. The total serum bilirubin was mildly elevated in three patients. Two of these had obstruction of an atriocentric conduit and one had significant ventriculoatrial regurgitation.

Mitral valve abnormalities. Four of 16 patients for whom postoperative angiograms were available showed mitral valve prolapse. Two of these had moderately severe associated mitral regurgitation. Mitral valve prolapse was first noted in one of these at catheterization 1 year after surgery and in the other at catheterization 14 years after surgery. In the latter patient, mitral valve prolapse was confirmed at surgery. The two other patients had no associated mitral regurgitation. In one of these the mitral valve was normal at catheterization 1 year after surgery but was noted to prolapse at catheterization 5 years postoperatively. Mitral valve prolapse was noted at catheterization in the other patient 1 year after surgery.

Discussion

Several studies have described early and midterm follow-up results in patients with tricuspid atresia who have undergone the Fontan operation. These reports include patients followed for up to 9 years, and there is agreement that most patients benefit from the operation within this period of follow-up. Our study describes the status of 32 patients operated on between 1968 and 1977 who survived at least 1 year, giving a 7 to 16 year follow-up.

Five deaths occurred during the follow-up period. In four patients there was obstruction between the right atrium and right ventricle or the pulmonary artery. In three of these there were signs of systemic venous hypertension. In the other, moderate obstruction was well tolerated when the patient was in sinus rhythm, but fatal hemodynamic deterioration occurred when the patient developed atrial flutter.

Reoperation was performed because of residual atrial septal defects, a left ventricle-to-pulmonary artery shunt through an incompletely closed proximal pulmonary artery in a patient with ventriculoarterial discordance, severe mitral regurgitation, obstructed Dacron conduits, an obstructed aortic homograft, and an intrapulmonary arteriovenous fistula in a patient who had a previous Glenn anastomosis.

Residual atrial septal defect, a troublesome complication early in the series, was prevented later by careful technique and frequent use of patches. A residual left ventricle-to-pulmonary artery shunt was observed in only one patient. The possibility for this shunt to occur has been eliminated by a modification of technique whereby the proximal pulmonary artery is divided and the proximal portion is closed by direct suture while the distal portion is used for establishment of atrio pulmonary continuity. Furthermore, by dividing instead of ligating the pulmonary trunk, mobility and freedom are given to the pulmonary arteries and more space is available for a wide pulmonary arteriomy. This maneuver facilitates the placement of the aortic valve homograft or permits a wider direct atrio pulmonary anastomosis.

Obstruction of a Dacron conduit required reoperation in four patients and contributed to the death of two other patients. Development of a thick fibrin peel within Dacron valved and nonvalved conduits has been reported after Fontan operation and has prompted some surgeons, as well as us, to eliminate their use. Obstruction of the Dacron conduit usually occurs years after the operation but has been observed as early as 2 months postoperatively. It must be remembered that a population of patients with such conduits exists and must be evaluated regularly by clinical examination, noninvasive laboratory studies, and, if necessary, by cardiac catheterization to determine the possible need for reoperation.

Homograft valve obstruction occurred in one patient 14 years after surgery. This is the only homograft among seven in this series that has become obstructed. In this patient the wall of the aortic homograft was heavily calcified and calcifications appeared to extend onto the base of the homograft valve leaflets. In the same patient, an aortic homograft valve in the inferior vena cava to which was attached only a very narrow rim of aortic wall appeared entirely normal. Thus it appeared that homograft valve calcification did not occur as a primary process of the valve but rather occurred by extension of calcification from the homograft valve leaflets.
graft wall. The durability of homografts has been excellent, although experience extends only to 15 years. 19

Twenty-six of our 27 patients are in NYHA class I or II. This distribution is similar to that reported in series with shorter follow-up. 1-4 Within our study group, the distribution of functional classification was similar among those with shorter vs longer follow-up, suggesting stability in functional classification.

The only pregnancy we have observed resulted in the birth of a small but otherwise normal infant. In this patient the increased intravascular volume and increased demand on cardiac output 20 that normally accompany pregnancy were tolerated satisfactorily, even though this patient had an imperfect repair.

Since left ventricular function is a major determinant of the results after the Fontan operation, 21 we were especially interested to find whether any patients showed evidence of increasing left ventricular dysfunction during long-term follow-up. One of our patients had left ventricular dysfunction contributing to a poor result. This had developed by 3 months after surgery. Intraoperative impairment of the myocardium may have occurred, possibly related to the unavailability at that time of current techniques of intraoperative myocardial protection. In the other patient, long-standing mitral regurgitation and left-to-right shunting could have resulted in poor left ventricular function that was masked by the severe mitral regurgitation but became obvious after the mitral valve was repaired. Also, long-standing high coronary venous pressure could have contributed to left ventricular dysfunction, a possibility that can be obviated by left atrial translocation of the coronary sinus.

Four of our patients have had recurrent supraventricular arrhythmias. Two of our patients tolerated the arrhythmia well with little change in symptoms. The third patient showed signs of congestive heart failure that were easily controlled. However, in one patient with associated conduit obstruction, atrial flutter resulted in low cardiac output and death. This may be similar to a patient described by Laks et al. 1 who had increased right atrial pressure caused by increased pulmonary vascular resistance and who died during an episode of supraventricular tachycardia. Thus supraventricular arrhythmias, often well tolerated, may be fatal in the presence of obstruction to pulmonary flow. We suspect that the high incidence of supraventricular tachycardia seen in these patients is related to high right atrial pressure. It should be recalled, however, that an increased incidence of atrial arrhythmia also is part of the natural history of tricuspid atresia. 22

Six of our patients had residual cyanosis. This was explained by a residual atrial septal defect in one patient and by pulmonary arteriovenous fistulas related to a preexisting Glenn anastomosis in two patients. In three patients, cyanosis was present shortly after surgery but was not explained by intracardiac shunting or pulmonary arteriovenous fistulas. Late onset of cyanosis suggestive of development of arteriovenous fistulas, a theoretic complication of the Fontan operation, 23 was not observed in this series.

None of our patients had clinical evidence of hepatic dysfunction, although four had some abnormality of laboratory studies. This is encouraging, but we recognize that significant hepatic fibrosis can occur in the absence of clinical findings and with little abnormality of liver enzymes or serum bilirubin. 24 We do not have biopsy results from our surviving patients, and we did not perform histologic examinations of the livers of the patients who died.

Two of our patients had clinical evidence of mitral regurgitation associated with mitral valve prolapse. This finding prompted a review of all available cineangiograms. Two additional patients were noted to have a prolapsing mitral valve without regurgitation. Although the significance of this finding is uncertain, the early identification and careful follow-up of such patients is important, especially since left ventricular function is critical in post-Fontan patients. Also, mitral valve dysfunction may occur in some cases related to congenital abnormalities of the mitral valve. 25

This report includes results from patients operated on in 1977 or earlier. Patients undergoing the Fontan operation currently will undoubtedly have better long-term results than those reported here. Left ventricular function is better preserved by improved methods of intraoperative myocardial protection, and earlier surgery should reduce the detrimental effects of chronic left ventricular volume overload. Problems related to atrioventricular or atrio pulmonary obstruction, prevalent in this series of patients, should be minimized by using direct, wide atrioventricular or atrio pulmonary anastomoses and by avoiding valved or nonvalved conduits and homograft valves whenever possible. Pulmonary resistance can be minimized by planning preceding palliative surgery judiciously and perhaps by earlier Fontan surgery. At the time of operation, the arterial blood supply to the sinus node should be preserved carefully, possibly reducing the incidence of postoperative atrial arrhythmia.

Results of the Fontan operation remain good for most patients 7 to 16 years after surgery. Further careful follow-up studies are necessary because of the
markedly altered physiology related to this type of repair.

References