The failing Fontan circulation: Successful conversion of atriopulmonary connections

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Objectives: Symptoms from low cardiac output or refractory atrial arrhythmias are complicating atriopulmonary (classical) Fontan connections. We present our experience of converting such patients to total cavopulmonary connections with and without arrhythmia surgery.

Methods: Between 1997 and 2002, 15 patients (mean age, 19.7 ± 7.0 years) underwent conversion operations 12.7 ± 3.5 years after atriopulmonary Fontan operations. Preoperative New York Heart Association functional class was I in 2 patients, II in 2 patients, III in 6 patients, and IV in 5 patients. Four patients underwent intracardiac lateral tunnel conversion alone, and 11 received extracardiac total cavopulmonary connection, right atrial reduction, and cryoablation.

Results: No mortality occurred. One patient had conduit obstruction in the immediate postoperative period requiring replacement, and another required a redo operation for endocarditis. Average hospitalization was 17.9 ± 9.38 days; chest drains were removed on median day 4 (range, 1-29; mean, 7.4 ± 7.58 days). At follow-up (mean, 42.6 ± 22.1 months), late atrial arrhythmias had recurred in 3 of 4 patients with intracardiac total cavopulmonary connections (without ablation) and 1 of 11 patients with extracardiac total cavopulmonary connections with ablation. All patients are in New York Heart Association class I or II. Exercise ability (Bruce protocol) improved 69% from a mean of 6.18 ± 4.01 minutes to 10.45 ± 2.11 minutes ($P < .05$). Need for antiarrhythmic agents decreased postoperatively (patients receiving ≤ 1 antiarrhythmic: 9 preoperatively vs 15 at long-term follow-up, $P < .05$). No patient has required transplantation. Protein-losing enteropathy, which was present in 1 patient, improved transiently with conversion. There was 1 late death from gastrointestinal hemorrhage.

Conclusions: Fontan conversion can be achieved with low mortality and improvement in New York Heart Association class and exercise ability. Concomitant arrhythmia surgery reduces the incidence of late arrhythmias.

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Some of the data displayed in this article were presented at the EACTS Meeting, Frankfurt, Germany, October 7-11, 2000.

Received for publication Sept 9, 2003; revisions received Feb 9, 2004; accepted for publication Feb 26, 2004.

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J Thorac Cardiovasc Surg 2004;128:60-6022-5223/\$30.00

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doi:10.1016/j.jtcvs.2004.02.011

The Fontan procedure was described in 1971 for treatment of tricuspid atresia, and through its atriopulmonary modifications, it has been extended to palliate complex congenital heart defects that preclude biventricular repair.¹ Late postoperative complications after an atriopulmonary connection (APC) have limited both survival and functional state. These complications include pathway obstruction, right atrial dilatation, atrial arrhythmias, poor exercise capacity, failing ventricular function, and protein-losing enteropathy (PLE).²⁻⁵ These problems tend to occur together and have been described as “the Fontan state,” which many authors believe is inevitable with the systemic venous hypertension necessary to drive this type of circulation.⁶

Total cavopulmonary artery connection (TCPC) with intracardiac baffle was introduced by de Leval and colleagues⁷ and resulted in improved early outcomes.

Table 1. Patient details

Age at time of revision	Mean, 19.7 ± 7.0 y; median, 18 y (range, 14-43 y)
Time since original Fontan operation	Mean, 12.7 ± 3.5 y; median, 11 y (range, 8-20 y)
Sex	6 male, 9 female
No. of previous surgeries	Median, 3 (range, 2-4)
Original diagnoses, n (%)	Tricuspid atresia: 7 (47); double-inlet left ventricle: 6 (40); pulmonary atresia with intact ventricular septum: 2 (13)
Preoperative NYHA class, n (%)	
I	2 (13.3)
II	2 (13.3)
III	6 (40)
IV	5 (33.3)
Preoperative LVEDP, n = 10 (mean ± SD)	8.1 ± 2.77

NYHA, New York Heart Association; LVEDP, left ventricular end-diastolic pressure.

Late arrhythmias are still a problem with this type of connection. More recently, many have moved to using an extracardiac TCPC, thus minimizing the interference with the right atrium and theoretically reducing the incidence of late arrhythmias.

For recipients of APCs who have mechanical obstruction or poor functional capacity, conversion to TCPC has helped in selected reported cases to relieve obstruction, optimize hemodynamics, and improve functional status, although with significant morbidity and mortality.⁸ However, the theoretic potential to reduce or abolish atrial arrhythmias by eliminating right atrial hypertension was not universally realized. Indeed, both clinical and experimental data suggest that atrial arrhythmias can persist and develop anew after conversion to intracardiac TCPC. Additional surgical measures become necessary to overcome late atrial arrhythmias because they are usually refractory to both pharmacologic and ablation therapies. Work by Mavroudis and Deal and their associates^{9,10} suggests that conversion to TCPC with an extracardiac conduit, electrophysiologically guided cryoablation of the atrial arrhythmic pathways, atrial reduction, and use of an atrial antitachycardia pacemaker might be the most promising solution. We present our experience with Fontan conversion at the Wessex regional cardiac unit.

Patients and Methods

Patient Population

A total of 15 patients underwent conversion operations for late complications after the Fontan procedure between June 1997 and October 2002. The demographics and native cardiac pathology are summarized in Table 1. The mean interval from the initial Fontan procedure was 12.7 ± 3.5 years, with an average age at the time of redo operation of 19.7 ± 7.0 years. Preoperatively, most patients were in New York Heart Association (NYHA) class III or IV

TABLE 2. Preoperative antiarrhythmic drugs

No. of antiarrhythmic medications	No. of patients, n (%)
0	2 (13.3)
1	7 (46.7)
2	5 (33.3)
3	1 (6.7)

Digoxin: 6; amiodarone: 4; sotalolol: 4; propranolol: 2; verapamil, procainamide, disopyramide, atenolol: 1 each (n = 15).

with poor exercise tolerance and received multiple antiarrhythmic drug therapy (Table 2). The congenital cardiac anomalies included tricuspid atresia (n = 7), double-inlet left ventricle (n = 6), and pulmonary atresia with intact ventricular septum (n = 2). All patients had previously undergone palliative Blalock-Taussig shunt procedures or pulmonary artery banding to control pulmonary blood flow leading up to the Fontan operation, with additional surgical procedures in appropriate cases. Atrial tachyarrhythmia was diagnosed on resting electrocardiography (ECG) and 24-hour ambulatory ECG recording in 13 of 15 patients. Among these, multiple preoperative radiofrequency ablation procedures were attempted, with unsatisfactory results in 3 patients who demonstrated persistent atrial flutter. All patients had a markedly dilated right atrium with venous stasis and spontaneous contrast on echocardiography. Ventricular function was assessed on echocardiography and angiography. All patients had good systolic function, except 2 patients in whom ventricular function was moderate. All patients underwent cardiac catheterization. Mean left ventricular end-diastolic pressure was 8.1 ± 2.77 mm Hg (n = 10). Two patients showed significant gradients across the surgical Fontan pathway.

Effort intolerance and recurrent atrial arrhythmias were the indications for conversion in 10 patients; of these, one had PLE, and another had a moderate pulmonary paravalvular leak (having had previous pulmonary disconnection and subsequent pulmonary valve replacement). A further 3 patients primarily had debilitating arrhythmias, one of whom had mechanical obstruction (homograft with a gradient of 5 mm Hg). One patient mainly had effort intolerance, which was also associated with mechanical obstruction (gradient of 5 mm Hg across the homograft). A calcified narrowed homograft (gradient of 2 mm Hg) was the indication for conversion in one patient.

In this series 2 patients were active on transplant lists, and a further 3 were under consideration.

Electrophysiologic Study

Preoperative electrophysiologic assessment for each patient included 12-lead ECG, 24-hour Holter monitoring, and (in 9 patients) electrophysiologic study with noncontact mapping. The use of noncontact mapping in patients having undergone the Fontan procedure has been reported previously.¹¹ Briefly, a 64-electrode array is positioned in the right atrial chamber through the femoral venous route. The array detects a low-frequency locator signal that is passed through a steerable, moveable mapping catheter. As the catheter is dragged around the endocardial surface of the right

atrium, a 3-dimensional reconstruction of chamber geometry is created. Anatomic and surgical landmarks can be identified by using operation records, fluoroscopy, and electrogram characteristics and labeled on the geometry. After induction of the arrhythmia, the array detects far-field electrical potentials from the endocardial surface. By using inverse solution mathematics, more than 3300 virtual unipolar electrocardiograms are simultaneously recorded, allowing high-density real-time isopotential and isochronal maps of global activation that are superimposed on the chamber geometry. These are analyzed offline to determine arrhythmia circuits and their relationship to surgical anatomy and to guide placement of cryoablative lesions.

Surgical Techniques

Cardiopulmonary bypass was established by using either aortobiventricular cannulation ($n = 9$) or femorofemoral cannulation with subsequent superior vena cava cannulation on re sternotomy ($n = 6$). In one patient femoral arterial cannulation failed to achieve adequate arterial bypass flow because of a small femoral artery and required conversion to aortic cannulation. We now increasingly favor femorofemoral cannulation with vacuum-assisted venous drainage to decompress the grossly dilated right atrium before re sternotomy to avoid undesired cavity entry. Inadvertent cavity entry occurred in 3 patients: 2 involved the right atrial appendage (aortic cannulation in one and femoral cannulation in the other), and the third involved a right atrium–pulmonary artery homograft adherent to the sternum, where precautionary femoral artery cannulation was undertaken before re sternotomy.

The procedure was conducted during moderate hypothermia (core temperature, 28°C) by using the alpha-stat cooling strategy, whereas myocardial preservation was achieved by combining intermittent antegrade cold blood cardioplegia with topical cooling. After dissection of adhesions to obtain adequate exposure, the previous APC was taken down, and a modified bidirectional Glenn shunt was fashioned by anastomosing the superior vena cava to the cranial aspect of the right pulmonary artery. In the first 4 patients, the TCPC was completed through the construction of an intracardiac lateral tunnel. This was fashioned from a bovine pericardial patch (polytetrafluoroethylene* in one patient) tailored and sutured to the lateral wall of the right atrium. A 4-mm fenestration was used in the first 2 patients in our series, but routine fenestration was discontinued as the surgical method evolved.

Because of concern that the right atrial wall contributing to the lateral tunnel was still prone to dilatation and a focus for arrhythmias, departmental surgical policy changed to favor the extracardiac TCPC for first-time Fontan procedures, as well as for conversion patients. In the latter 11 patients, an extracardiac TCPC was performed by using a polytetrafluoroethylene conduit (Dacron in 2 patients) in combination with right atrial reduction, cryoablation of the right atrial arrhythmic pathways (plus left-sided maze procedure if prior atrial fibrillation), and implantation of bipolar steroid-eluting endocardial atrial pacemaker. The atrial pacing leads were placed through the atrial suture line while the cross-clamp was still on, testing thresholds later when off bypass. We have found that with steroid-eluting pacing leads, in the ensuing

postoperative period, pacing thresholds improve from those recorded intraoperatively as the electrodes embed into the myocardium and myocardial edema abates. Most of the dilated redundant right atrial wall was excised to reduce atrial arrhythmogenic mass.

Cryoablation sites covered areas of atrial slow conduction and reentry pathways. Cryoablation lesions were made in the fashion described by Mavroudis and associates.¹⁰ Briefly, the right-sided lesions were created after excision of a large portion of the right atrial free wall, including the sinoatrial node. By using standard linear cryoprobes, lesions were made from the coronary sinus to the inferior vena cava orifice and where a tricuspid valve exists from the inferior vena cava to the tricuspid valve annulus. Further lesions were created from the atrial septal defect toward the crista terminalis and the cut edge of the atrium. A lesion was then created from the superior border of the atrial septal defect to the superior remnant of the atrial wall. The right atrial appendage was partially excised. The left-sided lesions were created in the manner of the Cox-Maze III procedure. The cryoprobe was used at -60°C for 2 minutes for each linear lesion.

Statistical Methods

Means are expressed with SDs, and medians are expressed with ranges. Comparison of groups with normal distribution was analyzed by using the Student paired t test. Categorical data were analyzed by using χ^2 techniques and, when necessary, application of Fisher's exact test.

Results

Electrophysiologic Study

Noncontact mapping was performed without complication in 9 (82%) of 11 patients who underwent arrhythmia surgery. In each case the majority of the right atrial mass was electrically inert, either because of the presence of baffle or conduit material or because of fibrosis and scarring caused by hypertrophy and longstanding hemodynamic abnormalities. Visible electrical activity was often of low amplitude, with slow conduction velocities and fractionated electrocardiograms. A total of 9 arrhythmias (of which 5 were deemed clinically relevant) were mapped. All were macroreentrant circuits involving the right atrial mass, apart from one patient in whom the circuit was biatrial. Typically, the circuits used surgical barriers to circle around with a critical isthmus between the inferior vena cava and the inferior margin of the atriotomy incision or the inferior vena cava and a tricuspid valve patch (or coronary sinus in patients with tricuspid atresia).

Surgical Intervention

Intraoperative parameters of the surgical cohort are detailed in Table 3. There was no surgical mortality. No patients required exploration for bleeding. Postoperative parameters are shown in Table 4. Patients were extubated 17.8 ± 22.8 hours after arrival in the intensive care unit. All but 3 were discharged to the ward within 48 hours. Transient postoperative inotropic support was required in 11 patients. Total hospital stay was 17.9 ± 9.38 days, and predischarge echo-

* Gore-Tex patch, registered trademark of W. L. Gore & Associates, Inc, Newark, Del.

TABLE 3. Operative details

Cardiopulmonary bypass time, min (n = 14)	242.5 ± 75.7 (120-320)
Crossclamp time, min (n = 14)	44.3 ± (20-129)
Circulatory arrest was used in 1 patient in whom the total arrest time was 57 minutes.	

TABLE 4. Postoperative course

	Mean ± SD	Median (range)
Time to extubation, h	17.8 ± 22.8	12 (4-94)
Length of stay in PICU, d	2.2 ± 1.37	2.0 (1-5)
Time to removal of final chest drains, d	7.4 ± 7.58	4 (1-26)
Length of hospital stay, d	17.9 ± 9.38	14.0 (8-39)
Early complications (n = 15), n (%)	Pleural effusions: 6 (40); infective complications: 4 (27); pacing failure: 2 (13); Dacron graft thrombosis: 1 (7)	

PICU, Pediatric intensive care unit.

cardiography confirmed well-preserved left ventricular function in all but one patient, in whom it was moderate preoperatively and remained moderate postoperatively. In one patient left ventricular function improved from moderate preoperatively to good postoperatively.

Complications

Of the in-hospital complications, the most common was pleural effusion (6 patients). This was managed with intercostal chest tube drainage, which was used in all but 1 patient, and the final drain was removed after a mean 7.4 ± 7.58 days. Four infective complications were seen (2 wound infections, 1 pneumonia, and 1 bacteremia), and all responded to antibiotics. Two pacing complications (1 pacemaker failure and 1 lead displacement) required replacement. There was thrombus formation in the extracardiac conduit within hours of return to the intensive care unit in 1 patient in whom Dacron had been used as the conduit. This patient was returned straight to the operating theater, where replacement of the conduit with polytetrafluoroethylene was uneventful. All other minor complications responded well to conservative treatment.

During medium-term follow-up, one 20-year-old patient had infective endocarditis 6 months after the Fontan conversion, requiring replacement of his extracardiac conduit with bovine valved venous conduit, after which he made a full recovery. The patient remains well.

Postoperative Arrhythmias

In-hospital, significant atrial arrhythmias occurred in 1 patient and required DC cardioversion. During follow-up,

TABLE 5. Change in need for antiarrhythmic medication: Complete series (n = 15)

No. of antiarrhythmic agents	Preoperative (%)	Short-term postoperative (%)*	Long-term postoperative (%)†
0	2 (13.3)	5 (33.3)	4 (26.7)
1	7 (46.7)	9 (60)	11 (73.3)
2	5 (33.3)	1 (6.7)	0
3	1 (6.7)	0	0

Short-term drugs: propranolol, 8; amiodarone, 1; atenolol, 1; disopyramide, 1. Long-term drugs: propranolol, 7; amiodarone, bisoprolol, sotalol, digoxin, 1 each. β -Blockers used for anti-cardiac failure therapy have been included in this analysis.

* $P = .08$, χ^2 (Fisher's exact test): comparing number of patients taking 1 or fewer antiarrhythmic agents preoperatively versus postoperatively at the time of discharge.

† $P = .017$, χ^2 (Fisher's exact test): comparing number of patients taking 1 or fewer antiarrhythmic agents preoperatively versus postoperatively at latest follow-up.

significant atrial dysrhythmia occurred in 3 of the 4 patients who underwent lateral tunnel conversion without cryoablation, requiring multiantiarrhythmic therapy.

Of the 11 patients undergoing extracardiac TCPC, right atrial reduction, cryoablation, and pacemaker implantation, the first patient had multiple episodes of asymptomatic atrial arrhythmias. After some time, those disappeared, and he remains in a paced sinus rhythm. A further 2 patients were stimulated into a nonsustained atrial flutter during postsurgical electrophysiologic studies. Clinically, they have remained in paced sinus rhythm. During our experience, practice has changed, and all patients undergoing conversion Fontan operations are treated postoperatively with low-dose propranolol or some other β -blocker prophylactically. This change occurred from our eighth patient onward and reflects our protocol of managing patients undergoing the Fontan procedure as "heart failure" patients with a combination of angiotensin-converting enzyme inhibitors and β -blockers. All patients received warfarin postoperatively.

The number of antiarrhythmic agents patients required preoperatively, on discharge, and at long-term follow-up is shown in Table 5 for the complete series. There was a reduction in the number of antiarrhythmics at the time of discharge compared with preoperatively ($P = .08$, Fisher's exact test). Patients continued on less antiarrhythmics at long-term follow-up compared with preoperatively ($P < .05$). Subgroup analysis was carried out for the 11 patients undergoing extracardiac TCPC, right atrial reduction, cryoablation, and pacemaker implantation (Table 6). Similarly, less antiarrhythmics were required at discharge compared with preoperatively ($P = .06$, Fisher's exact test), a trend that was further accentuated at long-term follow-up ($P < .05$).

TABLE 6. Change in need for antiarrhythmic medication: Antiarrhythmia surgery group (n = 11)

No. of antiarrhythmic agents	Preoperative (%)	Short-term postoperative (%)*	Long-term postoperative (%)†
0	0	1 (9)	1 (9)
1	5 (45.5)	9 (82)	10 (91)
2	5 (45.5)	1 (9)	0
3	1 (9)	0	0

β -Blockers used for anti-cardiac failure therapy have been included in this analysis.

* $P = .06$, χ^2 (Fisher's exact test): comparing number of patients taking 1 or fewer antiarrhythmic agents preoperatively versus postoperatively at the time of discharge.

† $P = .012$, χ^2 (Fisher's exact test): comparing number of patients taking 1 or fewer antiarrhythmic agents preoperatively versus postoperatively at latest follow-up.

TABLE 7. Exercise tolerance testing (Bruce protocol)

Patient	Preoperative, min	Postoperative, min
1	11	11
2	9.77	13
3	1.5	11
4	6.5	6.5
5	8.16	9.5
6	0.3	3.16
7	6	12.16
Mean time: whole group $\pm$ SD, min (n = 7)*	6.18 $\pm$ 4.01	10.45 $\pm$ 2.11
Mean time: arrhythmia surgery subgroup $\pm$ SD, min (n = 5)†	4.49 $\pm$ 3.40	9.83 $\pm$ 2.12

* $P < .05$, paired t test.

† $P = .57$, paired t test; the arrhythmia surgery subgroup comprised patients 3 to 7.

Exercise Ability

Exercise tolerance testing (Bruce protocol) both preoperatively and postoperatively was available in 8 patients. One patient was in atrial flutter when undergoing postoperative exercise testing, and the test was discontinued after 49 seconds. This result has been excluded from the analysis. Table 7 shows the exercise testing results for the other 7 patients. A significant improvement in exercise ability of almost 70%, with a preoperative mean of 6.18 ± 4.01 minutes increasing to 10.45 ± 2.11 minutes ($P < .05$), was seen (Figure 1).

Of these 7 patients, 5 had undergone extracardiac TCPC, right atrial reduction, cryoablation, and pacemaker implantation. A subgroup analysis carried out on these 5 patients demonstrated an increase in exercise tolerance, with a preoperative mean exercise time of 4.49 ± 3.40 minutes increasing to 9.83 ± 2.12 minutes postoperatively ($P = .057$).

Figure 1. Graphic display showing exercise stress test results before and after Fontan conversion surgery (Bruce protocol).

A significant improvement in NYHA functional class ($P < .001$) corroborated the improvement in exercise ability (n = 14; Table 8) seen in the series as a whole, as well as in the extracardiac TCPC antiarrhythmia surgery subgroup ($P < .001$). No patients have worsened since surgical intervention, and none remain in NYHA functional class IV.

No patients in this series have subsequently required cardiac transplantation.

Late Death

Follow-up was a mean of 42.6 ± 22.1 months. One patient died 3 years after his operation in a district referral center as a result of gastrointestinal hemorrhage.

Discussion

Our results demonstrate that conversion of an atriopulmonary Fontan connection to a TCPC can be accomplished safely and successfully, with resultant improvement in exercise ability and arrhythmia control. The most common presentation of the failing atriopulmonary Fontan connection in our patient population was with both poor functional ability and atrial dysrhythmia. There were no operative deaths and only one late death, which was caused by lower gastrointestinal hemorrhage, most likely related to warfarin, although there was no postmortem examination to confirm this. All other patients continue to do better than preoperatively or at least no worse. Left ventricular function was well preserved with the operation, as demonstrated with preoperative and postoperative echocardiography.

In one patient in whom Dacron was used for the extracardiac TCPC, acute thrombosis within hours of the conversion necessitated emergency replacement of the conduit with polytetrafluoroethylene. We advocate against the use of Dacron in this position; polytetrafluoroethylene is our conduit of choice, with all patients subsequently receiving warfarin.

Recurrent Arrhythmias

It is evident from our results that there was an improvement in arrhythmia control in our group as a whole, demonstrated by the few patients who had arrhythmic events after surgical intervention, as well as the reduction in the number of antiarrhythmic medications required compared with the multiagent treatment most patients needed preoperatively. Conversion to extracardiac TCPC, with atrial reduction and antiarrhythmia surgery (cryomaze), is particularly effective in arrhythmia control because only 1 of 11 patients had significant arrhythmias after revision compared with 3 (75%) of the 4 undergoing lateral tunnel conversion. This latter finding is in keeping with the literature, in which intracardiac TCPC conversions without arrhythmia surgery have a collective arrhythmia recurrence rate of 76%.¹⁰

Electrophysiologic studies, which were performed in 9 of the 11 patients undergoing cryoablation surgery, currently help in deciding which dysrhythmias might cause future problems. As our practice evolves, we anticipate that they will not be routinely required as part of the work-up for Fontan conversions.

Exercise and Functional Ability

Improved exercise ability was seen in most patients, as assessed by using the Bruce protocol. Improvement was also seen in the latter group that underwent extracardiac TCPC, with a 119% improvement in mean exercise time from 4.49 minutes to 9.83 minutes, although this was just short of statistical significance. Although our numbers are small, it seems that improvement is gradual but steadily occurring, mainly within the first year in most cases. After conversion surgery, all patients are in NYHA functional class I or II. All our patients have been able to return to full-time employment or college, and no patient has worsened in NYHA functional class.

Our current indications for Fontan conversion are mechanical pathway obstruction, or atrial dilatation with symptomatic dysrhythmias uncontrolled by multiple therapy, or controlled at the expense of cardiac output or amiodarone side effects; if favorable pulmonary and left ventricular hemodynamics are present, then impaired ventricular function is not a contraindication in itself. Two patients were active on transplant lists, and another 3 were under consideration before Fontan conversion. Left ventricular function before conversion surgery was good in 4 of these patients and moderate in 1, as determined on the basis of echocardiographic criteria. However, all 5 had profound low cardiac output states when dysrhythmic, with severe debility sometimes necessitating intensive care unit admission. Antiarrhythmic agents were contributing to low cardiac output states or causing intolerable side effects. After conversion surgery, none of the patients has needed transplantation, and

TABLE 8. Exercise functional capacity (NYHA class)

NYHA class	Total group*		Arrhythmia surgery subgroup†	
	Preoperative (%)	Postoperative (%)	Preoperative (%)	Postoperative (%)
I	2 (14)	9 (64)	1 (10)	6 (60)
II	2 (14)	5 (36)	1 (10)	4 (40)
III	5 (36)	0	3 (30)	0
IV	5 (36)	0	5 (50)	0

* $P < .001$, χ^2 (Fisher's exact test; $n = 14$): comparing combined number of patients in NYHA classes I and II preoperatively versus postoperatively for the group as a whole.

† $P < .001$, χ^2 (Fisher's exact test; $n = 10$): comparing combined number of patients in NYHA classes I and II preoperatively versus postoperatively for the arrhythmia surgery subgroup.

all made sufficient improvements to be removed from the transplantation waiting lists.

One patient, who was initially active on a transplant list, had PLE. This patient had very poor exercise ability preoperatively and had atrial arrhythmias, gross peripheral edema, and ascites, running a serum albumin level of approximately 19 g/L. He responded well initially to extracardiac TCPC conversion surgery. His preoperative exercise time (Bruce protocol) doubled from 6 minutes to 12.16 minutes postoperatively. Edema and ascites completely abated. The serum albumin level increased to 22 g/L at its peak. The PLE recurred some 6 months later, and the albumin level has decreased to the high teens.

In conclusion, conversion of the failing APC Fontan circulation to extracardiac TCPC with concomitant arrhythmia surgery can be undertaken safely with low morbidity. It can improve a patient's exercise and functional ability, as well as provide effective relief or control of arrhythmias.

References

- Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax*. 1971;26:240-8.
- Cromme-Dijkhuis AH, Hess J, Hahlen K, Henkens CMA, Bink-Boelkens EME, Eygelaar AA, et al. Specific sequelae after Fontan operation at mid- and longterm follow-up. *J Thorac Cardiovasc Surg*. 1993;106:1126-32.
- Gewillig M, Wyse RK, de Leval MR, Deanfield JE. Early and late arrhythmias after the Fontan operation: predisposing factors and clinical consequences. *Br Heart J*. 1992;67:72-9.
- Driscoll DJ, Offord KP, Feldt RH, Schaff HV, Puga FJ, Danielson GK. Five- to fifteen year follow-up after Fontan operation. *Circulation*. 1992;85:469-96.
- Mertens L, Hagler DJ, Sauer U, Somerville J, Gewillig M. Protein-losing enteropathy after the Fontan operation: an international multicenter study. *J Thorac Cardiovasc Surg*. 1998;115:1063-73.
- Fontan F, Kirklin JW, Fernandez G, Costa F, Naftel DC, Tritto F, et al. Outcome after a "perfect" Fontan operation. *Circulation*. 1990;81:1520-36.
- de Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations—experimental studies and early clinical experience. *J Thorac Cardiovasc Surg*. 1988;96:682-95.

8. Marcelletti CF, Hanley FL, Mavroudis C, McElhinney DB, Abella RF, Marianeschi SM, et al. Revision of previous Fontan connections to total extracardiac cavopulmonary anastomosis: a multicenter experience. *J Thorac Cardiovasc Surg.* 2000;119:340-6.
9. Mavroudis C, Backer CL, Deal BJ, Johnsrude CL. Fontan conversion to cavopulmonary connection and arrhythmia circuit cryoablation. *J Thorac Cardiovasc Surg.* 1998;115:547-56.
10. Mavroudis C, Backer CL, Deal BJ, Johnsrude CL, Strasburger J. Total cavopulmonary conversion and maze procedure for patients with failure of the Fontan operation. *J Thorac Cardiovasc Surg.* 2001;122:863-71.
11. Betts TR, Roberts PR, Allen SA, Salmon AP, Keeton BR, Haw MP, et al. Electrophysiological mapping and ablation of intra-atrial reentry tachycardia after Fontan surgery with the use of noncontact mapping system. *Circulation.* 2000;102:419-25.

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