149 Fontan Conversions

Carl L. Backer, MD[a]; *Constantine Mavroudis,* MD[b,c]

[a]ANN & ROBERT H. LURIE CHILDREN'S HOSPITAL OF CHICAGO, NORTHWESTERN UNIVERSITY FEINBERG SCHOOL OF MEDICINE, CHICAGO, ILLINOIS; [b]FLORIDA HOSPITAL FOR CHILDREN, ORLANDO, FLORIDA; [c]JOHNS HOPKINS UNIVERSITY SCHOOL OF MEDICINE, BALTIMORE, MARYLAND

ABSTRACT: Patients with a functionally univentricular heart who have had an atriopulmonary Fontan are at risk for atrial dilatation, atrial arrhythmias, and progressive circulatory failure. Between 1994 and 2018, we performed 149 Fontan conversions with arrhythmia surgery and epicardial pacemaker placement at Ann & Robert H. Lurie Children's Hospital of Chicago. This operation converts the atriopulmonary Fontan to an extracardiac Fontan that improves hemodynamics and controls the atrial arrhythmias. Operative mortality during that time was 2%, and freedom from death or heart transplant at 10 years is 84%. For properly selected patients, Fontan conversion improves both survival and quality of life.

Patients with an atriopulmonary Fontan constitute an eroding population, as they face many comorbidities and have a decreased life expectancy without treatment; therefore, all patients with an atriopulmonary Fontan should be evaluated for this procedure.

BACKGROUND

The operation now referred to as "Fontan conversion" was first described by Dr. Hilel Laks and colleagues from the University of California, Los Angeles in 1994.[1] They reported three patients with atriopulmonary Fontan connections who developed atrial arrhythmias and atrial thrombus. All three patients were successfully treated with conversion of the atriopulmonary Fontan to a lateral tunnel cavopulmonary Fontan.

In 1994, we performed our first Fontan conversion at Ann & Robert H. Lurie Children's Hospital of Chicago (formerly Children's Memorial Hospital). Our contribution to the cavopulmonary conversion was to add arrhythmia surgery in the form of either a right-sided maze or biatrial Cox-maze III procedure.[2] In addition, we preferred to do an extracardiac Fontan with a polytetraflouroethylene (PTFE) graft rather than a lateral tunnel procedure. Our Fontan conversion series now totals 149 operations, and this manuscript is based on our experience and outcomes with this patient population.

One of the most intriguing aspects of the late Professor Fontan's initial description of his procedure was that he actually predicted the complication that has led to the Fontan conversion technique. In 1971, he reported the first "Fontan" operation in three patients.[3] In the discussion portion of the manuscript, he noted, "One element remains unpredictable—the haemodynamic consequences of an eventual atrial rhythm disturbance such as an atrial fibrillation or flutter." We now know that most patients with an atriopulmonary connection develop slow progressive dilatation of the right atrium (Figure 1),[4] thickening of the right atrial wall, and eventual atrial arrhythmias.[5] As a result, the impact of atrial arrhythmias on patients with an atriopulmonary Fontan has been well described. In a multicenter study, Diller and colleagues reviewed the outcomes of 321 adult Fontan patients and found that the presence of atrial arrhythmias led to a 6-fold increase in risk for death or need for transplantation. The 3-year mortality in the group with arrhythmias was 25%, and the 6-year mortality was greater than 40% (Figure 2).[6] We and others believe that the natural history of these patients has been improved by early

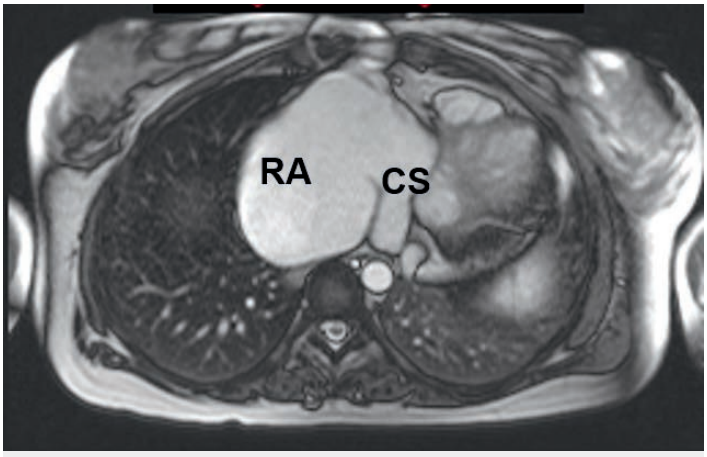

Figure 1.
Magnetic resonance imaging shows a dilated right atrium in a 40-year-old male with double inlet left ventricle and L-transposition of the great arteries. The patient had patch exclusion of the atrial septal defect and right atrioventricular valve and a direct right atrium to pulmonary artery connection. He presented with a massively dilated right atrium and atrial fibrillation. CS: coronary sinus; RA: right atrium. Reprinted with permission.[4]

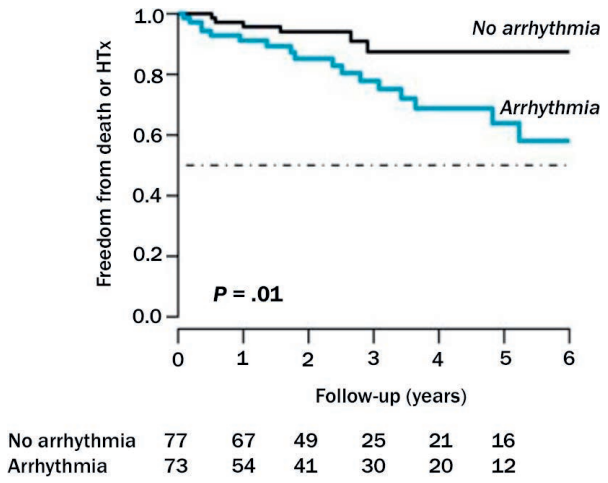

Figure 2.
Kaplan-Meier plot for freedom from cardiac-related hospitalization and from death or cardiac transplantation (HTx) in patients with atriopulmonary or atrioventricular Fontan. Reprinted with permission.[6]

Fontan conversion.[7-9] Poh and colleagues from the Australian and New Zealand Fontan registry reviewed patients undergoing Fontan conversion between 1990 and 2014. They found that an earlier surgical conversion strategy is associated with better survival free from transplantation (Figure 3).[10]

PATIENT CHARACTERISTICS

The majority of patients undergoing Fontan conversion in our series had either tricuspid atresia or a double inlet left ventricle.

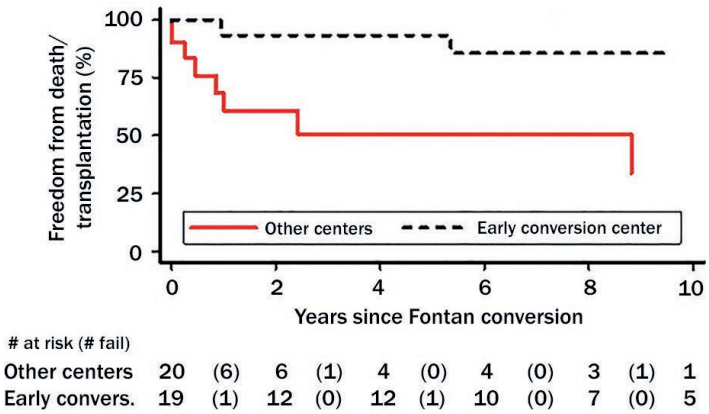

Figure 3.
Kaplan-Meier survival curve of freedom from death and transplantation in the Australian and New Zealand Fontan Registry for Fontan conversion versus other centers. Reprinted with permission.[10]

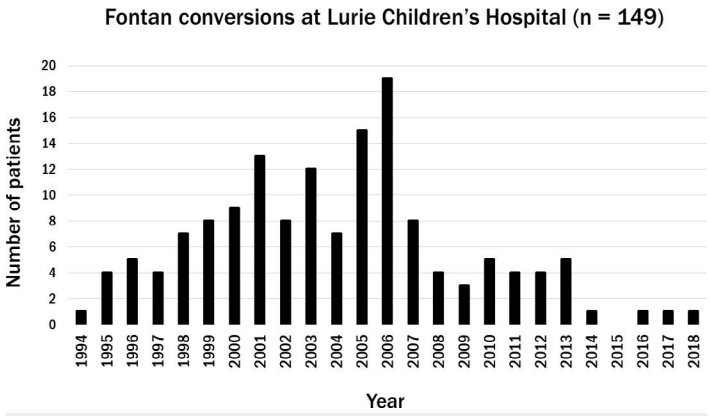

Figure 4.
The number of patients who underwent Fontan conversion at Ann & Robert H. Lurie Children's Hospital of Chicago from 1994 to 2018.

The number of patients undergoing Fontan conversion by year at Lurie Children's is seen in Figure 4. This demonstrates a rapidly eroding population of patients who are candidates for conversion, since most patients with an atriopulmonary connection have either had a conversion or heart transplant, or they have died.

In our series, the mean patient age at initial Fontan repair was 5.6 years, the median interval from initial Fontan repair to conversion was 16.7 years, and the median age at Fontan conversion was 22 years with a range from 2.6 to 47 years. All patients had a prior atriopulmonary Fontan except for 19 who had a Bjork modification and 11 who had a lateral tunnel-type Fontan. Thirty percent of patients were New York Heart Association (NYHA) class I to II, while 70% were class III to IV. Preoperative arrhythmias were present in all but four patients, with the most common being atrial fibrillation followed by right atrial tachycardia and left atrial tachycardia.

The preoperative evaluation varies depending on whether the patient has atrial reentry tachycardia or atrial fibrillation. Patients with atrial reentry tachycardia undergo both pre- and intraoperative electrophysiologic (EP) studies and undergo a modified right atrial maze procedure based on the results of the EP study. The patients with atrial fibrillation all have a biatrial Cox-maze III procedure. The patients with atrial fibrillation receive postoperative amiodarone for 3 to 6 months.

SURGICAL PROCEDURE

Careful preoperative imaging with either computed tomography or magnetic resonance imaging is needed to predict reentry problems at the time of repeat sternotomy. The primary steps

Resect enlarged right atrium
Atrial septectomy
Biatrial maze – cryoablation
Extracardiac polytetraflouroethylene graft – inferior caval vein to pulmonary artery
Bidirectional Glenn
Epicardial pacemaker

Table 1.
Surgical steps when performing a Fontan conversion.

of the Fontan conversion procedure are noted in Table 1. Most patients are initially cannulated with a single venous cannula in the very enlarged right atrium. Once the patient is safely on cardiopulmonary bypass, a full dissection of the right atrium, superior caval vein, inferior caval vein, and right upper pulmonary vein can be performed. We then convert to direct bicaval venous cannulation. If the patient has no intracardiac shunts, a generous portion of the enlarged right atrium can be resected with the heart still beating. The connection of the right atrium to the pulmonary artery can be taken down. The inferior caval vein/right atrial junction is divided, and a 24-mm PTFE graft is sutured to the inferior caval vein.

Next, a vent is placed in the right superior pulmonary vein, cold blood cardioplegia is administered, and an atrial septectomy is performed. A majority of patients referred in the past 10 years had atrial fibrillation, and a full Cox-maze procedure was performed. The left-sided cryoablation lesions (minus 160°C, 1 minute) are shown in Figure 5,[11,12] and the lesions for the right atrial maze are shown in Figure 6.[2] The lesions are modified if the patient has mitral atresia rather than tricuspid atresia. Some of the more challenging anatomic and EP problems that we have encountered with the innovative solutions used to treat them were reviewed in 2007.[13] Once the maze procedure is completed, the right atrium is closed. The right atrium should now be considerably smaller. The only venous blood entering the right atrium is from the coronary sinus, which can now pass freely across the atrial septal defect. Of note, none of the patients in this series were fenestrated.

The cross-clamp is then released, and the remainder of the procedure is performed as the patient is warmed. The PTFE graft that was anastomosed to the inferior caval vein is now carried up to the undersurface of the pulmonary artery, to

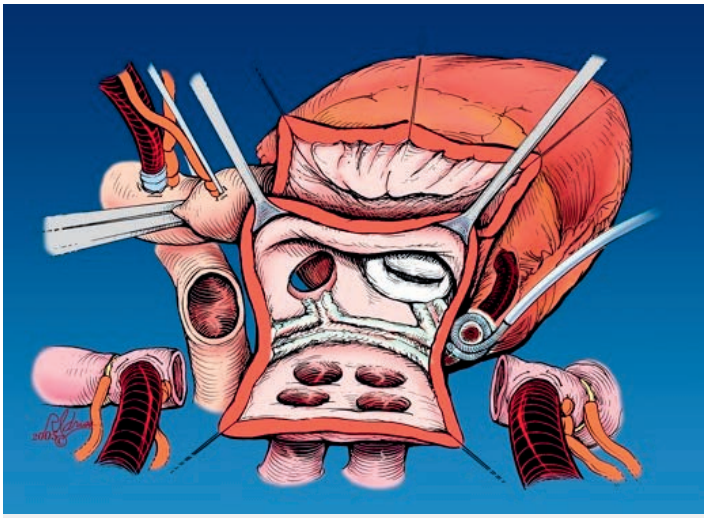

Figure 5.
This illustration demonstrates the lesions placed (with cryoablation at -160°C for 1 minute) to complete the left side of the Cox-maze III procedure in patients with atrial fibrillation. In this illustration, the right-sided pulmonary veins and a portion of the left atrium have been opened surgically. The remaining lesions have been created with the cryoablation probe. The lesion on the coronary sinus is placed for 2 minutes. Reprinted with permission.[12]

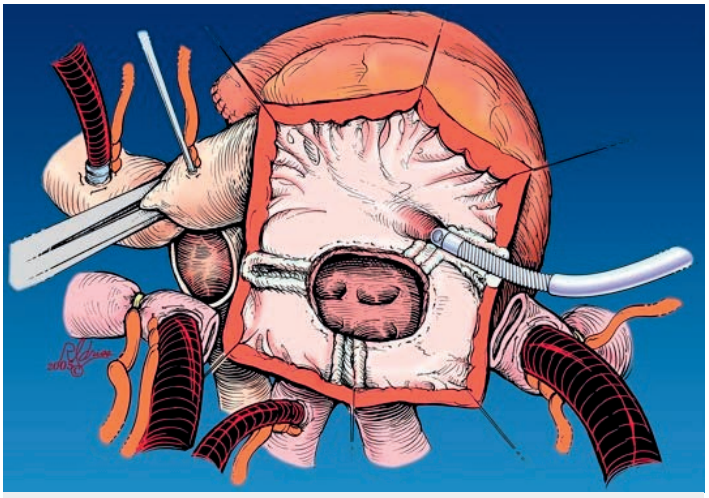

Figure 6.
This illustration shows the cryoablation lesions (-160°C for 1 minute) for the right-sided maze performed in a patient who underwent an atriopulmonary Fontan for tricuspid atresia. Reprinted with permission.[2]

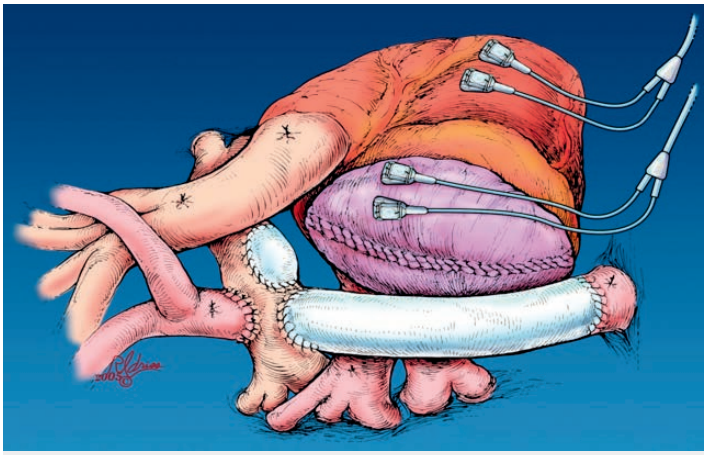

Figure 7.
Completed Fontan conversion. Reprinted with permission.[12]

either the site of the prior atriopulmonary anastomosis or a site more medial, and a patch is placed on the site of the old atriopulmonary anastomosis. As the patient is warmed and started on low-dose inotropic support, the bidirectional superior cavopulmonary anastomosis is performed with running prolene suture. The patient is then ventilated and weaned from cardiopulmonary bypass, and the epicardial pacing system is placed with the heart beating. We have used bipolar steroid eluting leads for the atrial and ventricular lead placement. These are then connected to a dual-chamber device with atrial antitachycardia capabilities.[14] The completed operation is shown in Figure 7.[12]

RESULTS

In our series, the operative mortality was 3 out of 149 patients (2%). One patient underwent reoperation for bleeding (0.7%). The mean length of stay was 14 ± 10 days. Freedom from arrhythmia occurrence is 80% at 15 years. The atrial pacemaker therapy provides atrioventricular synchrony and reduces premature atrial contractions that can trigger atrial reentry tachycardia. In the 24-year time period of our series, 12 patients have undergone heart transplantation after Fontan conversion. The freedom from death or heart transplantation at 10 years is 84%.[15] Six of the transplants were within 1 year of Fontan conversion and would be considered conversion failures. Six transplants occurred more than 3 years post Fontan conversion, and the conversion could be considered a success.

For patients presenting with a failing atriopulmonary Fontan, the typical question is whether they should be referred for conversion or cardiac transplant. Factors favoring conversion are patients with preserved ventricular function but a severely dilated right atrium leading to poor hemodynamics. Also, patients who have not yet had a bidirectional Glenn as part of their cavopulmonary connection have improved hemodynamics after Fontan conversion. A recent decrease in ventricular function related to new-onset atrial arrhythmias often responds very well to Fontan conversion. The Australia/New Zealand group recommends that patients be referred within the first 3 years of their arrhythmia while still in NYHA class II.[10] They also recommend referring patients for conversion before starting a second antiarrhythmic medication. Because of the dramatic thickness of the right atrium in these patients, we do not recommend attempting transcatheter ablation of the atrial arrhythmias.

Risk factors for cardiac death or heart transplantation after Fontan conversion in our series included right or indeterminate ventricular morphology, preoperative ascites or protein-losing enteropathy, cardiopulmonary bypass time greater than 240 minutes, or need for biatrial maze.[15] Patients who have end-stage ventricular failure, protein-losing enteropathy, plastic bronchitis, ascites, ≥ moderate AV valve regurgitation, or arrhythmias continuing for more than 5 to 10 years are generally referred for heart transplantation since, at this point, it is essentially too late for Fontan conversion.[8,15] However, it is important to note that in our experience, prior Fontan conversion was not a risk factor for mortality after heart transplantation.[16] We have now performed 55 heart transplants for failed Fontans, with five operative mortalities and four late deaths.

In 2016, we summarized the international experience with the Fontan conversion operation using 16 published series in addition to our own.[15] In the collective series of 542 patients, there were 31 operative deaths for an overall operative mortality of 6.1%. The number of late deaths or transplants was 56 of 542 patients (10.3%). In the Society of Thoracic Surgery database, the operative mortality of Fontan conversion was 9.7%.[17] The Mayo group reported 70 patients undergoing Fontan conversion from 1994 to 2011.[8] Multivariate analysis revealed age > 27 years, atrioventricular valve regurgitation, and lack of arrhythmia operation to be predictors of perioperative death. The same group reported that a baseline peak VO_2 of ≤ 14 mL/kg/min was predictive of perioperative mortality.[17] Higashida and coauthors demonstrated that cardiac index significantly improved after Fontan conversion and was maintained for at least 5 years.[19] In their 32 patients, new-onset atrial tachycardia was not observed after prophylactic Fontan conversion.

CONCLUSION

A Fontan conversion operation is indicated for patients who had a previous atriopulmonary Fontan and now have atrial

arrhythmias and a dilated right atrium. Fontan conversion improves hemodynamics, decreases arrhythmias, and can be performed with low operative mortality. In our experience and that of others, earlier referral of patients with an atriopulmonary Fontan for conversion will improve the long-term results. Careful selection criteria can help determine which patients should be considered instead for cardiac transplantation. In general, patients with evidence of protein-losing enteropathy or severe ventricular dysfunction should undergo transplant evaluation. Intermediate transplant-free survival and freedom from arrhythmia recurrence are quite favorable (> 80%) at 15 years. Fontan conversion, however, should be done at centers with the institutional experience and expertise to deal with these complex patients. This is an eroding patient population, and the time to act for any patient still surviving with an atriopulmonary connection may be now.

KEY POINTS

- Patients with atriopulmonary Fontan are an eroding population.
- Fontan conversion improves hemodynamics and controls arrhythmias.
- All patients who underwent atriopulmonary Fontan should be evaluated for Fontan conversion.
- Fontan conversion is not recommended for patients with protein-losing enteropathy.

Conflict of Interest Disclosure:
The authors have completed and submitted the *Methodist DeBakey Cardiovascular Journal* Conflict of Interest Statement and none were reported.

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congenital heart surgery, Fontan conversion, atriopulmonary Fontan, arrhythmia, atrial fibrillation, tricuspid atresia, protein-losing enteropathy, epicardial pacemaker, cryoablation

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