Impact of Fontan conversion with arrhythmia surgery and pacemaker therapy

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Abstract

Objective: In the long-term period after Fontan operation, atrial arrhythmia was one of the important factors to decide the postoperative quality of life. We reviewed the impact of Fontan conversion with arrhythmia surgery and pacemaker therapy.

Methods: Thirty-eight patients underwent Fontan conversion using extracardiac conduit from 1992, and 22 patients with atrial arrhythmia underwent maze procedure simultaneously using cryoablation or radiofrequency ablation and epicardial DDD pacemaker implantation and 16 patients had regular ‘sinus’ rhythm before Fontan conversion. Mean follow-up period was 52 months. Pre- and postoperative clinical course were analyzed. Average weight, age at Fontan conversion, and years after first Fontan operation were 49.0 kg, 25.8 years old, 14.7 years, respectively. Nineteen percent of patients were in New York Heart Association class I (NYHA I), and 74% of patients were in NYHA II, and 7% were in NYHA III, respectively.

Results: Except three early deaths, actual survival rate at 1 year and 5 years were 80% and 64%, respectively. In survivors, 80% of the patients obtained regular heart rhythm including artificial pacemaker rhythm, although only 43% of the patients had regular ‘sinus’ rhythm before the Fontan conversion. Postoperative average cardiothoracic ratio and oxygen saturation (SpO₂) were 50% and 94%, and 74% of patients were in NYHA I and 26% were in NYHA II, respectively, after Fontan conversion.

Conclusions: Mid-term results of Fontan conversion with arrhythmia surgery and pacemaker therapy were acceptable. Restoration of regular rhythm might improve the postoperative NYHA status and the activity of the daily life.

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Keywords: Fontan conversion; Arrhythmia surgery; Pacemaker therapy

1. Introduction

Despite several improvements in surgical techniques and postoperative treatment, late deterioration in functional status could be observed with longer duration of follow-up after the Fontan operation. Late postoperative complications include obstruction of venous pathways, atrial tachyarrhythmia, cyanosis secondary to systemic venous collaterals, thrombus formation, and protein-losing enteropathy (PLE). These complications are associated with high central venous pressure (CVP) and systemic ventricular dysfunction [1—5]. Atrial arrhythmia was also one of the important factors to decide the postoperative quality of life. Recently, Fontan conversion and thrombus removal with concomitant arrhythmia surgery and pacemaker placement were performed for the patients who had dilated right atrium, large thrombus in the right atrium and atrial tachyarrhythmia after Fontan completion [6—9]. Fontan conversion has been performed to rescue the failed Fontan circulation as a palliative strategy [10], and its indication for Fontan conversion is still controversial and surgical outcomes are uncertain. In this study, we observed medical records from the patients who underwent Fontan conversion, and investigated early and midterm outcome after the Fontan conversion, and reviewed the impact of Fontan conversion with arrhythmia surgery and pacemaker therapy.

2. Patients and methods

Thirty-eight patients underwent Fontan conversion using extracardiac conduit from 1992, and 22 patients with atrial arrhythmia underwent maze procedure simultaneously using cryoablation or radiofrequency ablation and epicardial DDD pacemaker implantation and 16 patients had regular rhythm before Fontan conversion. Primary diagnosis was tricuspid atresia in 21 patients, single right ventricle in 10 patients, single left ventricle in three patients, atrioventricular (AV) septal defect in three patients, and Ebstein’s anomaly in one patient. There were 10 Bjork procedure (right atrial to right ventricular connection) and 28 atropulmonary connections.
Mean follow-up period was 52 months. Pre-and postoperative clinical course were analyzed. Average weight, age at Fontan conversion, and years after first Fontan operation were 49.0 kg, 25.8 years old, 14.7 years, respectively. Nineteen percent of patients were in New York Heart Association class I (NYHA I), and 74% of patients were in NYHA II, and 7% were in NYHA III, respectively. NYHA class was determined by the pediatrician at the outpatient clinic. Average cardio-thoracic ratio (CTR) was 58 ± 9%, and oxygen saturation (SpO₂) was 91 ± 5%. Thrombus in the right atrium was found in 16 patients (16/38, 42%).

The criteria for surgery were atrial arrhythmias (22/38, 58%), right atrial thrombus (16/38, 42%), dilated right atrium (38/38, 100%), and hemodynamic abnormalities (38/38, 100%). All patients underwent preoperative cardiac catheterization with hemodynamic assessment and angiography for anatomic definition. Most underwent preoperative intracardiac electrophysiological mapping; patients with atrial fibrillation and poor vascular access were omitted. In the nine patients with atrial arrhythmia, isthmus catheter ablation was used for arrhythmia control. The 12 cases had variations of the modified right atrial maze for macro re-entrant atrial tachycardia and the 10 cases had the Cox maze III procedure for atrial fibrillation and left macro-re-entrant atrial tachycardia.

Average preoperative CVP was 15 ± 4 mmHg, and ejection fraction in systemic ventricle was 54 ± 9%. Regarding AV valve regurgitation grade I was two patients (5/38, 13%), grade II was seven patients (7/38, 18%), grade III was two patients (2/38, 5%), and grade IV was one patient (1/38, 3%).

2.1. Operation

The femoral vessels were usually dissected before the resternotomy for possible rapid cannulation and institution of cardiopulmonary bypass when there is high risk for cavitary entry. As an extracardiac conduit, 20–24 mm Gore-Tex (W.L. Gore & Associates, Flagstaff, AZ) tube graft was used except initial three patients (Dacron graft: 1, coated kitted graft: 2).

In the cases of conversion from a Bjork–Fontan modification to total cavopulmonary artery extracardiac connection, we preserved right ventricular to main pulmonary artery continuity, and performed the total cavopulmonary extracardiac connection.

Regarding arrhythmia surgery, in the initial period, cryoablation lesions were placed with a Surgifrost device (CryoCath Inc., Kirkland, Quebec, Canada) applied at −160 °C for 10–15 min, and, recently, radiofrequency ablation was placed with AtriCure (AtriCure®, Inc. 6217 Centre Park Drive West Chester, Ohio, USA) for 2–3 min. For the patients who underwent the maze procedure, the epicardial DDD pacemaker implantation was performed simultaneously to prevent possible bradyarrhythmia in the future. It was sometimes time consuming to find a suitable pacing position, because these patients were multi-operated, but a good position could be found in all patients after careful dissection on the surface of the atrium and ventricle. The steroid-eluting type electrodes were implanted, which contributed to maintain good pacing threshold postoperatively. We have not performed pacemaker implantation for the 16 patients with preoperative regular rhythm after Fontan conversion. We have also performed valvar repair simultaneously in six patients (19%) with AV valve regurgitation more than moderate.

3. Results

There were three (3/38, 7.9%) early and five (5/38, 13.2%) late deaths. Actual survival rate at 1 year and 5 years were 80% and 64%, respectively (Fig. 1). The early mortality in all three patients was related to unexpected cardiac laceration during the revision of median sternotomy and dissections, which might require longer duration of cardiopulmonary bypass (CPB) and operative time, leading to large volume of blood transfusion and deterioration of lung condition which influences postoperative hemodynamics. The causes of late death were cardiac dysfunction: two, pneumonia: one, liver cirrhosis: one, and rupture of esophageal varix: one. The average postoperative cardiothoracic ratio and SpO₂ were 50.3 ± 5.8% and 94 ± 1%.

In the midterm follow-up of the survivors, 80% of the patients obtained regular heart rhythm including artificial pacemaker rhythm (40%) and sinus rhythm (40%), although 19% of the patients had regular rhythm before the Fontan conversion (Fig. 2).

![Fig. 1. Survival curve after Fontan conversion.](https://academic.oup.com/ejcts/article-abstract/40/4/1007/446939/1008)

![Fig. 2. Ratio of regular rhythm before and after Fontan conversion. SR: sinus rhythm, PM: pacemaker rhythm.](https://academic.oup.com/ejcts/article-abstract/40/4/1007/446939/1010)
Regarding the patient's quality of life from NYHA classification, 74% of patients (22/30) were in NYHA class I and 26% (8/30) were in NYHA class II, respectively, after Fontan conversion, although 7% were in NYHA class I and 74% were in NYHA class II preoperatively (Fig. 3).

The freedom rate from atrial tachycardia or atrial fibrillation after the Fontan conversion was 91% (19/21), although it was 43% (6/14) in the patients without maze procedure ($p < 0.05$ vs with maze).

4. Discussion

The strategy of the conversion from failed Fontan to extracardiac total cavopulmonary connection was first reported by Laks et al. [6]. Although there are some indications for Fontan conversion (including a giant atrium with thrombus, arrhythmias refractory to medical or simple electrophysiological treatment, pulmonary venous obstruction by the enlarged right atrium and ventricular dysfunction), optimal timing for this approach is not clearly defined. Since arrhythmia surgery was not performed at the Fontan conversion initially, the reduction of the energy loss was reported to improve the disadvantage in hemodynamics. Marcelletti et al. reported that the Fontan conversion reduced the risk for arrhythmia and thrombo-embolism [7]. Especially when anti-arrhythmic surgical intervention was performed at the Fontan conversion simultaneously, this procedure decreased arrhythmia resulting in overall improvement in quality of life and functional class [8–10]. In our series, the improvement from 19% to 74% of NYHA class I was recognized, which could be attributed to restoration of regular rhythm. However, this improvement might be from the better hemodynamic performance of an extracardiac conduit as well.

Our early and late mortality rate was relatively higher compared with Mavroudis's group. Early mortality might be related with severe adhesions resulting from the multiple previous operations. Unexpected cardiac laceration during the revision of median sternotomy and dissections might require longer duration of CPB and operative time, leading to large volume of blood transfusion and deterioration of lung condition which influences postoperative hemodynamics. Consequently, it might affect early mortality. Another concern is preoperative condition such as cardiac function or hepatic function, which may affect late mortality. Since the interval from initial Fontan to Fontan conversion was relatively longer in our cases, the cardiac dysfunction and liver cirrhosis were observed preoperatively in many patients, and actually five patients died after discharge due to heart failure or complication of hepatic failure. Severe cardiac dysfunction and hepatic dysfunction are deemed as high risk factors for late death. There was no PLE in our patients, and PLE might not be a contraindication to performing Fontan conversion with arrhythmia surgery [11]. However, PLE could cause malnutrition leading to sepsis postoperatively, and was thought to be one of high-risk factors for late death.

To decide optimal timing of the Fontan conversion is important and the Fontan conversion should be undertaken at the early phase when patients are asymptomatic and their occult organ failure is reversible. Preoperative careful assessment is mandatory for the Fontan conversion and careful follow-up is also necessary to avoid overlooking the timing of the Fontan conversion. Recently, we started earlier Fontan conversion, when the enlargement of right atrium was detected without specific complications such as arrhythmia or thrombus formation, and there has been no early or late death since then.

This series represents a window onto long-term outcomes of patients with atrтопulmonary Fontan connections. Despite the introduction of lateral tunnel and extracardiac connection techniques, it is likely that the problems we encountered will arise with some regularity in the future. Cardiac transplantation will be an important mode of therapy for the failed Fontan. However, the number of pediatric cardiac transplantation is limited it has never been performed in Japan. Then, Fontan conversion with arrhythmia surgery continues to be an efficacious procedure for patients with increasingly complex anatomic lesions, and atrial arrhythmia.

5. Conclusion

Fontan conversion to total cavopulmonary artery extracardiac Fontan with concomitant arrhythmia surgery and pacemaker placement is an excellent therapy for patients whose Fontan repair has failed. Complex reconstructive techniques ensure unobstructed pathways. Evolution of pacemaker technology has provided more options for postoperative electrophysiological management. Fontan conversion does not preclude cardiac transplantation, which will continue to be an important mode of therapy for these patients.

References

Initially we did not perform the Maze procedure for the patient with transient atrial arrhythmia. For the patient where some arrhythmia recurrence has occurred, then we change to a Maze procedure for the patient with transient atrial tachycardia. This is the reason why the number is changing.

Regarding the second question, mainly pediatricians prescribe anti-arrhythmic drugs for patients with postoperative arrhythmia. But patients without significant arrhythmia did not take any anti-arrhythmic drugs.

Dr Cikirikcioglu: The third question was about the preventative Maze operation. The first group of patients showed a better quality of life and better sinus rhythm ratio compared to the second group. Do you use preventative Maze operation for sinus rhythm?

Dr Hiramatsu: Yes. Recently we have performed preventative Maze procedure for patients with significant arrhythmia, even if the basic rhythm is sinus.

Dr R. Hosseinpour (Seville, Spain): First of all, I would like to congratulate you for your courage, and I mean that profoundly, for dealing with such a difficult group of patients, nightmare patients really, very, very difficult.

I would like to express my cynicism, if I may. I have never done this operation personally, but I have assisted at it very many times in different centers done by different surgeons in different countries, and I have been consistently unimpressed by this operation.

You are starting with a Fontan, and at best, you are ending up with a Fontan. And to me, a failing Fontan is a failing Fontan. You improve things slightly.

And the ones that I have seen, they have either died, or, at best, you have to put them under the loop to convince yourself that they are better.

NYHA class is crude. It says, yes, NYHA class has gone up one point. You could argue that. That is not a very refined way of looking at it.

And I look at the literature. The literature is very rosy about it, and you read a large series with zero mortality. I do not know, maybe we do not know how to operate. But I have seen it in many centers, and I have been consistently underwhelmed by it to the point that I am thinking anyone who needs this, a Fontan conversion for sinus rhythm, they need is a transplant. But that is just my cynicism. It is more of a comment than a question.

Dr G. Ziemer (Tuebingen, Germany): My comment would be that, as with all surgery, the result of surgery, at least in major part, is a matter of who you choose to operate upon. Obviously you were unlucky to participate only in those cases where ventricular function was the main problem and not the rhythm. Fontan conversion surgery will not work if your main problem is ventricular dysfunction.

However, if your problem is arrhythmia, patients have a much better outcome. So the patients that even I operated upon got better, but I never would do a conversion Fontan with poor ventricular function as the main indication for surgery. You will not help those Fontan patients by conversion surgery.

And the same is true, and this was discussed earlier in this room, with reconstruction of AV valves in Fontan patients because they almost always are a sign of poor ventricular function. You may be lucky in certain cases with valve replacement, and this is even said in the good series you were citing.