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Ten-year outcomes of Fontan conversion in Australia and New Zealand demonstrate the superiority of a strategy of early conversion†

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Abstract

OBJECTIVE: To investigate the benefits of a strategy of early Fontan conversion.

METHODS: Using the Australia and New Zealand Fontan Registry, retrospective analysis of their long-term follow-up data was performed.

RESULTS: Between 1990 and 2014, a total of 39 patients underwent surgical conversion in 6 centres at a median age of 23.8 years (IQR: 19.3–28.2), 18.7 ± 5.0 years post-Fontan. One centre tended to perform conversion earlier: interval since first documented arrhythmia 2.9 ± 4.0 vs 4.0 ± 4.2 years, average NYHA Class 2 ± 0.4 vs 3 ± 0.9 ($P = 0.008$), mean number of preop anti-arrhythmics 1 ± 0.4 vs 2 ± 0.6 ($P = 0.05$). Two patients underwent conversion to an extracardiac conduit only, while 36 had concomitant right atrial cryoablation, of which 16 also had pacemaker implantation. Nine patients suffered major cardiac-related complications (7 low output syndrome, 3 ECMO, 3 acute renal failure, one stroke) (2/17 from the early conversion centre and 7/22 of the others; $P = 0.14$). Four patients died in hospital (10.3%) and 4 late after a median of 0.9 years [95% confidence interval (CI): 0.5–1] after conversion. An additional 2 patients needed transplantation at 1 and 8.8 years after conversion, respectively. The 10-year freedom from heart transplantation was 86% (95% CI: 51–97%). Outcomes from the centre with an early conversion strategy were significantly better: 8-year freedom from death or heart transplantation was 86% (95% CI: 53–96) vs 51% (95% CI: 22–74; log-rank $P = 0.007$). Eight additional patients required pacemaker implantation and 5 had arrhythmia recurrence.

CONCLUSIONS: Fontan conversion is associated with lasting survival outcomes up to 10 years after conversion. A strategy of surgical conversion at earlier stage of failure may be associated with better survival free from transplantation.

Keywords: Fontan conversion • Atriopulmonary Fontan • Long-term outcomes • Fontan procedure

INTRODUCTION

The first modification of the Fontan procedure, the lateral tunnel, was described in 1984 and in a matter of a few years, this modification was embraced by our community [1, 2]. The original technique, the atriopulmonary Fontan connection, has been replaced by the extracardiac Fontan connection and the lateral tunnel as the ideal surgical palliation, making up less than 1% of Fontan palliation in Europe and North America [3]. The population of patients who have undergone the atriopulmonary Fontan connection are now entering their second and third decade. The management of this population of young active adults is one of the biggest challenges faced by our community as we are now observing their progressive attrition. Heart transplantation after Fontan remains associated with high technical challenges even though favourable outcomes have been recently reported [4, 5]. Its practice continues to be impeded in many places by the scarcity of donors

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[6]. Fontan conversion has been offered since 1991 [7]. The Chicago team has been a strong advocate of this strategy, but we recently demonstrated in the follow-up data from the Australia and New Zealand Fontan Registry that Fontan conversion remained a rare practice in our region [8]. The mortality of conversion has been noted to be high, between 8 and 15% [9–13]. It is suspected that some of the survivors of the operation are still subjected to ongoing deterioration of their clinical status, eventually still requiring transplantation [14]. The poor quality of these outcomes has deterred many of us from performing the Fontan conversion at an early stage. The recently established Australia and New Zealand Fontan Registry has allowed us to identify all Fontan conversions in both countries. In order to better delineate the indication for this procedure and its late outcomes, we reviewed our regional experience and compared the impact of the variation of its indications.

PATIENTS AND METHODS

Retrospective analysis of the data of all patients who have undergone a Fontan procedure in Australia and New Zealand was approved nationally and for each state in Australia, and nationally in New Zealand. As required, the design of the audit was approved by at least one hospital in each state in Australia and in one hospital in New Zealand. The Australia and New Zealand Fontan Registry is a regional initiative that centrally collects a limited set of health data on all patients of the region who have undergone a Fontan procedure. The organization of the Registry has been previously described [15].

Follow-up data were collated from the Australia and New Zealand Fontan Registry and missing data from hospital databases and clinic letters. Copies of all available reports of echocardiograms, exercise stress testing and cardiac MRI were gathered.

Special effort was made to identify the timing of the first occurrence of arrhythmias. The time-point of first diagnosis of atrial arrhythmia was set as the date of first documentation describing its occurrence, either in hospital admission documentation or outpatient follow-up letters, whichever preceded.

Early postoperative complications included morbidity that occurred to patients during the same admission of surgical conversion or within 30 days of hospital admission. Late postoperative complications defined all events that occurred subsequently.

Patients were defined as having atrial dilatation when such a finding was described in the most recent cardiac imaging available including echocardiogram and magnetic resonance imaging.

Statistical analysis

Continuous data were expressed as mean (standard deviation) or median (interquartile range, IR) as appropriate. Categorical data were summarized as counts and proportions (percentage). This was further evaluated with χ^2 analysis. Survival analysis was performed with the Kaplan–Meier method. A log-rank test of survival functions was used to evaluate the effect of conversion approach on the risk of mortality or cardiac transplantation. Analyses were performed in Stata 12 (Statacorp., College Station, TX, USA).

As it became clear that one centre in the region had a lower threshold for Fontan conversion, we decided to compare the results obtained in this centre with the remaining centres of the region. In this centre, patients were offered conversion surgery if they had dilated atria and if they had sustained supraventricular tachyarrhythmias even if they were isolated episodes whereas all patients operated in the other centres of the region were symptomatic at the time of surgery. The right atrial maze performed in the centre with lower threshold for intervention varied from the original technique described by Mavroudis: no line of ablation was performed in the right atrial roof in order to preserve the pathway between the sinus node and the atrioventricular node [14, 16]. Accordingly, while pacemaker leads were implanted in all patients, only those requiring a pacemaker at the time of surgery were immediately implanted with a pacemaker chamber.

Additionally, outcomes after surgeries performed in adult hospitals were compared with surgeries performed in paediatric hospitals or in adults hospitals attached to a paediatric hospital.

RESULTS

At the end of 2013, a total of 1328 patients were found to have undergone a Fontan operation in the region: 236 atriopulmonary connections, 300 lateral tunnel and 792 extracardiac conduits. At last follow-up, the Registry data cross-checked with the National death index of Australia and the automatic reporting of all deaths to treating hospital in New Zealand identified the following number of deaths within each Fontan techniques: 63 deaths after an atriopulmonary Fontan, 24 after a lateral tunnel and 38 after an extracardiac conduit. A total of 39 patients of the 236 who had an atriopulmonary connection (17%) were recorded to have undergone a surgical conversion in 6 surgical units from January 1990 to May 2011. Patient characteristics are given in Table 1.

Predominant surgical indication was intractable cardiac arrhythmia (26/39; 66.7%), followed by pathway obstruction (6/39; 15.4%), of which 2 were secondary to thrombus formation. The remaining were due to obstruction of pathway by dilated right atrium and cardiac failure (2/39; 5.1%). Two patients underwent conversion due to need for concomitant surgery—one with severe aortic stenosis and another with restrictive ventricular septal defect obstructing the systemic outflow tract. Only 1 patient (from the early conversion group) received surgery as the management of refractory protein-losing enteropathy. One patient in the early conversion group had also surgical conversion due to amiodarone therapy-induced pulmonary fibrosis. Another patient received surgery after suffering an intracranial haemorrhage, due to inability to continue warfarin anticoagulation for his underlying atrial dilatation.

Eighteen patients were operated in one centre with a lower threshold for intervention (early Fontan conversion), and 21 in the remaining centres (late Fontan conversion). Comparative characteristics are displayed in Table 1. Patients operated in the centre practising early Fontan conversion had shorter interval since first documented arrhythmia, lower mean NYHA class, and were on less antiarrhythmic medications at the time of surgery. Patients in the early conversion group had 1 ± 0.9 cardioversions while those of the late group had 2.0 ± 3.1 cardioversions ($P = 0.13$).

Two patients underwent conversion to an extracardiac conduit without arrhythmia procedure. Thirty-seven patients had concomitant right atrial cryoablation, of which 16 also had pacemaker implantation. Comparison between the surgeries performed in the centre practising early conversion and those practising late conversion is displayed in Table 2. Patients operated in the centre that had a lower threshold for conversion had less concomitant insertion of a pacemaker.

Table 1: Patient characteristics

Characteristics	Total (n = 39)	Early conversion (n = 18)	Late conversion (n = 21)
Male	19	6	13
Cardiac morphology			
Dextrocardia	3	1	2
Dominant ventricle			
Left	31	16	15
Right	8	2	6
Dominant diagnosis			
Tricuspid atresia	15	11	4
Double-inlet left ventricle	9	3	6
Double-outlet right ventricle	9	2	7
Pulmonary atresia/stenosis	6	2	4
Dextrocardia	3	1	2
Common AV valve	2	0	2
Heterotaxia	1	1	0
Age at Fontan surgery	4.3 ± 5.0	5.6 ± 6.5	3.4 ± 3.1
Preconversion status			
Average NYHA ± SD	2 ± 0.7	2 ± 0.5	3 ± 0.7
AV valve incompetence			
Mild	9	4	5
Moderate	4	2	2
Severe	1	0	1
Atrial arrhythmia	32	15	17
Atrial flutter	18	8	10
Atrial fibrillation	5	4	1
Both	9	3	6
No. of antiarrhythmics (mean ± SD)	1 ± 0.6	1 ± 0.4	2 ± 0.6
No. of cardioversions (mean ± SD)	2 ± 2.4	1 ± 0.9	2 ± 3.2
Previous radiofrequency ablation	4	2	2
Pacemaker	16	9	7
Right atrial dilatation	38	18	20
Mild	1	1	0
Moderate	1	0	1
Severe	29	14	15
Unknown	7	3	4
Fontan conversion			
Age at conversion	23.9 ± 7.3	25.1 ± 8.6	23.7 ± 5.7
Indication			
Arrhythmia	24	11	13
Pathway obstruction	8	2	6
Cardiac failure	2	1	1
Other	5	4	1
Duration since arrhythmia onset	3.0 ± 4.2	2.9 ± 3.8	4.5 ± 4.4
Aortic cross-clamp time	66.5 ± 57.4	63 ± 45.9	79 ± 73.9
Cardiopulmonary bypass time	283 ± 90.3	270 ± 66.7	303 ± 112.5

AV: atrioventricular; NYHA: New York Heart Association class; SD: standard deviation.

In the early intervention group, all patients were converted into extracardiac conduit. In the late intervention group, 21 of 22 patients were converted to an extracardiac total cavopulmonary circulation. One patient received a lateral tunnel connection.

Early postoperative outcomes

There were 4 hospital deaths. Nine of the 38 patients suffered major cardiac-related complications perioperatively. A total of 7 patients had low cardiac output state postoperatively, of which 3 required extracorporeal membrane oxygenation. Three patients developed acute renal failure and 1 patient had a stroke resulting in permanent hearing impairment.

Table 2: Concomitant surgeries with Fontan conversion

Concomitant procedure	Early group (n = 18)	Late group (n = 21)
Right atrial maze	18	18
Biatrial maze	0	1
Insertion of permanent pacemaker	2	14
Atrial reduction	15	11
Septectomy	3	0
Pulmonary artery plasty	1	0
Outlet resection	1	1
Atrial baffle takedown	1	0
Bilateral atrioventricular valve repair	0	1
Thrombectomy	1	1

Seven patients (all from early intervention group) returned to theatre in same admission for delayed insertion of pacemaker box for postoperative bradycardia or junctional rhythm for atrial pacing. Epicardial leads were prophylactically placed during initial surgery. Four patients had prolonged hospitalization for management of persistent pleural effusion.

Long-term follow-up

Patients were followed up for a median duration of 6.0 years (IR 0.9–9.3) after Fontan conversion. There were 4 late deaths occurring after a median of 0.9 years (IR 0.8–2.4) post conversion. Two patients needed cardiac transplantation at 1 and 8.8 years post conversion. The 10-year freedom from heart transplantation was 86% (95% CI 51–97%) for the entire cohort.

Atrial arrhythmia recurred in 7 (17.9%) patients all of whom had right atrial ablation during surgical conversion. At last follow-up, survivors were taking an average of 1 ± 0.66 antiarrhythmic agents, with 4 patients on amiodarone. Seventeen of 32 patients (54%) on antiarrhythmic therapy before conversion remained in sinus rhythm without treatment at late follow-up. Four patients (3 from early conversion group, 1 from late conversion group) required late insertion of permanent pacemaker.

During the course of follow-up, 10 patients had thromboembolic events: 7 right atrial thrombus and 3 strokes. Seven of the 10 patients were on warfarin anticoagulation during the time of thromboembolic event.

Strategy of early versus late conversion

The early outcomes of the patients operated in the centre practising early conversion were compared with the early outcomes of the remaining centres practising late conversion. There were 4 in-hospital deaths, all occurring in the late intervention group (4/21 vs 0/18, $P = 0.05$). Of the 9 patients that suffered perioperative major cardiac complications, 2 were from the 18 patients of the early intervention cohort and 7 from the 21 in the late intervention group ($P = 0.1$).

There were 3 late deaths in the late conversion group and 1 late death in the early conversion group (3/17 vs 1/18). Two survivors operated in the centre with late conversion were transplanted at 1 and 8.8 years post conversion. Eight-year freedom from death or heart transplantation was 86% (95% CI: 53–96%) in the patients operated in the centre with a low threshold for conversion versus 51% (95% CI: 22–74) in the others ($P = 0.007$) (Fig. 1). There were no differences between rate of recurrence of arrhythmias between the surviving patients of the two groups (early vs late: 3/14 vs 4/17).

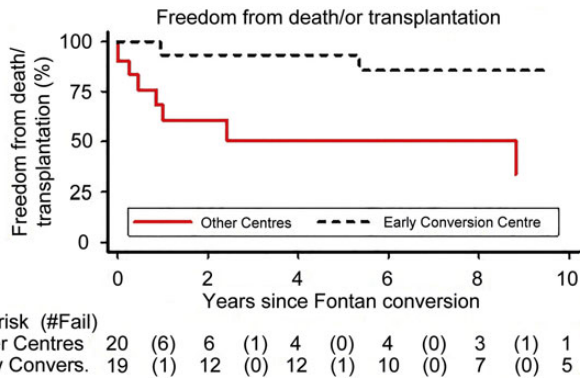

Figure 1: Kaplan–Meier survival curve of freedom of death and transplantation in the centre with a low threshold for Fontan conversion versus other centres.

Adult versus paediatric versus facilities

There were 25 patients who were operated in paediatric facilities (paediatric hospital or paediatric hospital with integrated adult service) and 14 patients in adult hospitals. The number of cases operated varied between 18 for the paediatric hospital with integrated adult service, 7 for the paediatric hospital and 7, 4, 2 and 1 for the adult hospitals. All the 4 early deaths occurred in patients operated in adult hospitals. Four of the 9 patients who suffered major cardiac-related complications postoperatively (4/25, 16%) had been operated in paediatric facilities and 5 (5/14, 35.7%) in general hospital ($P = 0.16$). All 4 patients developing complications in the paediatric facilities survived while only one of the 5 developing complications in the adult hospital survived (100 vs 20%, $P = 0.02$).

DISCUSSION

Since its first description by Laschinger *et al.* in 1993, Fontan conversion has been recognized as a potentially successful strategy for rescuing patients with a Fontan circulation from late failure. While the potential success of this strategy is not disputed, its relative indication has been the topic of heated debate. The main issue precluding the delineation of indications for the procedure is the fact that Fontan conversion is associated in most hands with a high mortality. The team from Chicago who has promoted this strategy has reported a perioperative mortality of 0.9% in 111 patients, but it is likely that they performed the procedure at a lower threshold than other teams [14]. The mortality rates of Fontan conversion in other centres have been described between 0 and 15%, and one would anticipate that mortality may even be higher in individual centres performing only a few of these procedures [9–13, 17–22]. There is growing evidence suggesting that Fontan conversion will be associated with a lower mortality if offered early [23].

Contraindications for the procedure are slowly being delineated. Severe ventricular dysfunction with elevated end-diastolic pressures of the systemic ventricle [24], severe atrioventricular valve regurgitation [12, 14] and poor functional status such as that encountered in protein-losing enteropathy [20, 24] are likely significant obstacles to the success of the procedure.

Offering the procedure in a timely fashion to patients with atriopulmonary connection with progressively enlarging atria remains difficult, mainly because of the slow and insidious progression of their symptoms. Most of the patients with atriopulmonary Fontan connections and dilated atria attending their annual clinic reviews are typically young active adults with a good quality of life, remaining mostly asymptomatic with daily activities. There is therefore often great reluctance in referring them for surgery.

While in some hands Fontan conversion has been a successful strategy, it is unclear whether it is always a sustainable strategy as it is not yet clear how many of those surviving a Fontan conversion will ultimately die or require a heart transplantation.

The most striking finding of our study is that patients who have undergone an early Fontan conversion maintain good cardiac function and NYHA scores for up to 8 years. Those undergoing a late conversion not only had at a higher risk of hospital mortality, but also had an earlier decline of their status. Half of them were dead or needed transplantation by 2 years after the procedure. The reason why patients with early conversion seemed to have more durable results cannot be demonstrated in the frame of this study. One can speculate that patients with delayed conversion would have a worse ventricular function at the time of conversion. The team who had a strategy for earlier conversion managed to have shorter procedures. They also adopted a strategy of reduced line of ablation to preserve atrioventricular conduction. It is possible that the better late outcomes observed in their patients were related to the preservation of the atrial function. A later right atrial maze procedure may achieve control of arrhythmia but some of these atria may no longer contract as a result of the procedure.

The perioperative management of adult patients after Fontan conversion is a challenging task of adult congenital heart disease. The Fontan patient while anaesthetized and under positive pressure ventilation has a significantly reduced cardiac output, making them a major challenge for the team to get them safely through the early postoperative period after this surgery, especially with the dangers of bleeding and the associated impact of blood products transfusion on pulmonary vascular resistance. It requires attention to detail and a careful multidisciplinary approach. This topic has been the focus of many recent studies. In a nationwide audit of close to 8000 adult congenital heart surgeries in 2010, Karamlou identified that best survival was achieved in patients operated by a paediatric surgeon in paediatric hospitals [25]. We found a 4-fold increase in risk of failure to rescue when patients experienced perioperative complications in an adult instead of paediatric facility.

Limitations and strengths

The main criticism to be raised is the lack of cohesion in our separation between early and late Fontan conversion. While the attitude of the centre adopting early conversion was found to be different from the other centres when examining the individual files, the distinction between early and late conversion was not as definite as this separation implied. Their experience developed over the course of two decades during which time their indication for surgery varied. Additionally, patients presenting late for surgery were not denied Fontan conversion. It is therefore difficult to demonstrate clear difference between their experience and one of the other centres. Because of the retrospective nature of the study, there was great variation in the evaluation of preoperative status of the patients. More precise delineation using cardiac MRI or exercise studies was inconsistently performed. There was therefore inadequate data in the small patient cohort to demonstrate significant differences in outcomes using variables other than that used in the current analysis. Clearly, the indications for successful Fontan conversion would be best delineated by large randomized multicentric studies, but they do not seem to be feasible. Without the comparative assessment of the impact of a strategy, Fontan conversion on an entire population of patients with atriopulmonary Fontans, it remains difficult to ascertain whether earlier conversion effectively prolongs the life of these patients. Patients operated in adult facilities seemed to have more complications and mortality but this finding may be related to the fact that these centres were also operating on less patients per centre.

In conclusion, Fontan conversion is associated with lasting survival outcomes up to 8 years after conversion. A strategy of surgical conversion at earlier stage of failure may be associated with better survival free from transplantation.

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APPENDIX. CONFERENCE DISCUSSION

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Dr T. Ebels *(Groningen, Netherlands):* There is a lot that can be said about your data and your presentation, but first I would like to remark that cardiac failure and NYHA class in Fontan patients have to be seen in the context of their Fontan circulation, and all Fontan patients have a lower output than people without a Fontan circulation.

So my question is, when do you consider a Fontan patient to be in cardiac failure? You could also state that all Fontan patients are in cardiac failure from the time of operation.

Dr Poh: This was actually a question that my supervisor Professor Yves d'Udekem and I were discussing in preparation for this session. I think that there is a big gap in the existing literature about the evaluation of the Fontan circulation and the definition of Fontan failure. As you mentioned, they all have a low cardiac output state immediately after surgery.

At present, the best marker we have, besides imaging and invasive monitoring of their haemodynamics, is their NYHA score and the severity of symptoms, and I think that's what we have used to define heart failure in this population.

Dr Ebels: Yes. I think if we really want to define cardiac failure in the Fontan population, then we need to clearly define what measure we use to say that.

The other thing is you had this wonderful Kaplan-Meier curve indicating a difference between two curves, but I think it is somewhat of a self-fulfilling prophesy. If you do the conversion very early, then obviously the longevity is going to be longer. But that doesn't really show that, as you state, this means that you rescue patients from late failure. I think that the conclusion is not really warranted.

Dr Poh: The population we're looking at are patients who have all ultimately undergone Fontan conversion, the only difference is the timing. What we're trying to possibly explore is that doing the surgery at an earlier stage may result in possibly better preservation of the cardiac or atrial function and therefore resulting in better long-term outcomes. That is something I think our results suggest at this point.

Dr Ebels: Well, you have clearly shown that it's a good treatment for the arrhythmias, but do the patients actually live longer? That is the question that is really unsolved. I think it is very difficult to conclude that from your or anybody else's data because the attrition of the Fontan circulation keeps on going whatever we do.

Dr Poh: I believe my supervisor; Yves d'Udekem is hoping to make a few statements.

Dr Y. d'Udekem *(Victoria, Australia):* So the whole point is to know whether or not you should do a Fontan conversion and at what stage. I was stricken by the fact that they seem to have lasting results. I mean, some of these patients up to eight, ten years seem to have lasting results from the Fontan conversions which was longer than I expected because there is no sign of failure up to this stage when you look at these patients. There are things that are transpiring from the direct reading of the data that are difficult to translate in statistical terms.

They seem to be all right for eight to ten years, which is really not the case of those you're doing when they're starting to become really symptomatic and very limited.

Dr Poh: There have been a lot of intermediate, mid-term results published with regards to Fontan conversion. The Mavroudis group, the biggest cohort study till date, and multiple smaller studies have found good survival rates of about 70 to 80% at three to four year after a Fontan conversion.

Recently, there was a study published by a Korean group, Jung *et al.*, which had a median duration follow-up of 6.6 years, similar to our study, and found a freedom from death rate of about 87%. That is similar to the rates that we have demonstrated in our early intervention group as well.

So, I think there is quite convincing evidence that these patients seem to be doing quite well up to eight to ten years.

As to how they'd progress subsequently, I think there's a lot more work to be done and we hope our research will shed some light on this in the near future.