Intra- or extracardiac Fontan operation? A simple strategy when to do what

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Abstract

Introduction: The complete Fontan circulation is the definite palliation for many complex congenital cardiac lesions. After bi-directional Glenn anastomosis (BDG), two well-established techniques – intracardiac tunneling and extracardiac prosthesis – are available for completion, although the choice of technique is still a matter of debate.
Material and methods: We retrospectively reviewed the surgical and clinical records of patients with single ventricle physiology, who underwent intracardiac (group I) or extracardiac (group II) Fontan palliation after BDG.
Results: Complete data were available in 72 patients. Thirty-eight patients received intracardiac (median weight: 12.6 kg) and 34 patients extracardiac repair (median weight: 15.6 kg). Patients with intracardiac tunneling had longer cardiopulmonary bypass (CBP) time (170 min vs. 104 min; $p < 0.001$), longer ventilatory (39 h vs. 21 h; $p = 0.009$) and longer inotropic support (48 h vs. 10 h; $p < 0.001$). Ventilatory and inotropic support were dependent on CPB ($r = 0.69$ and $r = 0.637$) and on aortic cross-clamping ($r = 0.785$ and $r = 0.705$ only group I), but not dependent on age, weight or pulmonary artery pressure (PAP).
Conclusions: Both techniques are feasible without perioperative mortality. Normally developed children with good hemodynamics after BDG received an elective extracardiac procedure without fenestration later. Patients with developmental retardation, severe progressive cyanosis, myocardial dysfunction, or moderate to severe atrio-ventricular valve insufficiency are scheduled for an earlier intracardiac baffle repair with routine fenestration, as they are at higher risk. Prolonged CPB and aortic cross-clamping times adversely impact the early postoperative course. Further strategies must be developed to avoid these effects, particularly in the patient group at higher imminent risk.

Key words: Fontan, univentricular heart.

Introduction

The concept of the Fontan circulation was first clinically introduced in 1971 [1]. Since the first description of this operation for single ventricle pathology, numerous modifications have been developed [2, 3]. Understanding and improving the Fontan circulation is still evolving with numerous *in vitro* and *in vivo* studies [4–7]. Bidirectional Glenn anastomosis (BDG) has become a standard intermediate step towards a Fontan circulation [8, 9]. In 1988 de Leval *et al*. introduced the concept of total cavopulmonary connection [4]. In this, a BDG anastomosis is performed while the inferior caval blood flow is directed towards the pulmonary arteries through

an intraatrial tunnel. At a later date an extracardiac polytetrafluoroethylene (PTFE) conduit to divert inferior vena cava flow to the pulmonary arteries was advocated and has become the procedure of choice for most single ventricle variants in many centers [9–11]. The advantages described to date are: 1. improved blood flow dynamics with less stasis and turbulence, and less thrombus formation, 2. fewer atrial arrhythmias in the early and long-term follow-up, 3. easier technique without aortic clamping in normothermia or mild hypothermia with preserved pulmonary and myocardial function and drainage of the coronary sinus to the low pressure atrial chamber [7, 12–16].

Despite continuing improvements, the Fontan procedure (FP) still carries significant early mortality and morbidity. To optimize the early results, it is now firmly established that a staging strategy with early relief of volume load on the single ventricle and concomitant performance of auxiliary intracardiac procedures plays a major role [11].

In this report, we present our experience both with intracardiac baffle Fontan and with the extracardiac conduit modification. The perioperative influence of age, weight, pulmonary artery pressure (PAP), duration of cardio-pulmonary bypass and aortic cross-clamping time on the duration of intensive care therapy, duration of ventilation and need for catecholamines was compared in both groups. From our observed results we propose a strategy for an appropriate choice between both techniques.

Material and methods

We retrospectively reviewed the surgical and the clinical records of 72 patients (48 males and 24 females) with single ventricle physiology, who underwent an intracardiac or extracardiac Fontan procedure between January 1995 und February 2008. Patients were divided into two groups: group I: a BDG and total cavo-pulmonary connection (TCPC) with an intracardiac lateral PTFE tunnel (n = 38, 24 males, 14 females, median age: 38 months, range: 19 to 99 months; median weight: 12.6 kg, range: 7.9 to 36.1 kg) and group II: a BDG with an extracardiac PTFE conduit (n = 34, 24 males, 10 females, median age: 44 months, range: 26 to 115 months, median weight: 15.6 kg, range: 7.9 to 37.5 kg). In all patients cardiopulmonary bypass (CPB) was used: in group I with aortic clamping and cardioplegic solution, in group II without aortic clamping. Fenestrations (2.7 mm to 4.0 mm) were performed in all patients of group I, and in two patients of group II with a preoperative mean pulmonary artery pressure of 20 mm Hg and 22 mm Hg. Gender and preoperative hemodynamic data matched between the groups. All patients were subjected to preoperative echocardiography and catheterization. The anatomical diagnoses are listed in Table I. No patient had significant atrioventricular valve regurgitation, and 4 patients had subvalvular systemic outflow tract obstruction. All except 2 patients had previously undergone several palliative procedures.

Surgical technique

All procedures were performed through a median sternotomy with standard aortic and bicaval cannulation.

In group I to construct a lateral intra-atrial channel draining the inferior vena cava (IVC), a PTFE baffle was implanted during aortic cross-clamping with cardioplegic arrest using CPB with moderate hypothermia. The coronary sinus was left on the "left" side of the baffle. The fenestrations were created with a 2.7 mm to 4.0 mm aortic punch.

In group II extracardiac PTFE conduits were implanted under CPB without aortic cross-clamping on a warm, beating heart. The diameter of the conduits ranged from 18 mm to 22 mm.

Statistical analysis

Statistical analysis was performed using descriptive statistics such as median, minimum, maximum and mean. For graphical demonstration box plots were used to describe the distributions of continuous variables in the two OP groups. For the evaluation of monotonic relationships between two continuous variables Spearman correlation coefficients were calculated. For the comparison of continuous variables in the two OP groups Mann-Whitney U tests were performed and the values of p are presented. These values of p are explorative only and were calculated for the purpose of description.

Table I. Cardiac anatomy, diagnoses

Diagnosis	Group I (lateral tunnel)	Group II (extracardiac conduit)
Tricuspid atresia	9	10
Hypoplastic left heart syndrome (HLHS)	11	5
Pulmonary atresia with VSD and hypoplastic right ventricle	8	3
Double-inlet left ventricle	4	3
Double-outlet right ventricle/hypoplastic left ventricle	5	7
Various univentricular situations	1	6

Results

There was no perioperative mortality. The duration of CPB ranged in group I from 50 min to 399 min (median: 170 min) and in group II from 53 min to 247 min (median: 104 min). This difference in duration was statistically significant ($p = 0.001$). No patient was subjected to more than one period of CPB during the completion FP.

Median respiratory support (ventilation time) in group I was 39 h (range: 0–504 h) and in group II was 21 h (range: 4–736 h). This difference was statistically significant ($p = 0.009$).

The duration of stay on the intensive care unit ranged in group I from 0 to 40 days (median 19.5 days) and in group II from 4 to 108 days (median: 14.0 days). Again, this difference showed statistical significance despite the wide ranges ($p = 0.002$).

The duration of inotropic support with catecholamines ranged in group I from 0 to 278 h with a median of 48 h and in group II from 0 to 104 h with a median of 10 h. The value of p was < 0.001, indicating significance.

Prolonged CPB time correlated well with longer mechanical ventilatory support ($r = 0.687$), with days on the intensive care unit ($r = 0.671$) and with inotropic support (> 0.1 µg/kg/min) ($r = 0.637$).

Aortic cross-clamping correlated well with longer mechanical ventilatory support ($r = 0.785$), stay on the intensive care unit ($r = 0.562$) and inotropic support (> 0.1 µg/kg/min) ($r = 0.705$).

Age was not correlated with mechanical ventilatory support ($r = 0.005$), with stay on the intensive care unit ($r = 0.017$) or with inotropic support ($r = –0.078$). Group II was definitely older than group I.

The same effect was observed for weight at FP completion, which was not correlated with ventilatory support ($r = –0.009$), with stay on the intensive care unit ($r = 0.038$) or inotropic support ($r = –0.256$).

Mean preoperative pulmonary artery pressure at completion of FP was 13.38 mm Hg (6–23 mm Hg), showing no differences between groups, and had no effect on the postoperative mechanical ventilatory support ($r = 0.266$), inotropic support ($r = 0.240$) or stay on the intensive care unit ($r = 0.403$).

Discussion

Patients with unrepaired univentricular hearts have a poor prognosis. The most common causes of death are dysrhythmia, congestive heart failure and sudden death [17]. The Fontan procedure has long challenged cardiologists and cardiac surgeons. The completion of the FP is the final phase of a staged procedure. Conceptually, at the level of the heart, systemic and pulmonary venous flows are separated and the single ventricle provides the pumping function for the systemic circulation only [1, 4, 17]. Today the short-term and medium-term results of the Fontan palliation are excellent, particularly considering the spectrum of severe congenital cardiac malformations for which it is now performed. Operative mortality of the FP has steadily decreased and in the best centers is no higher than for many biventricular repairs [18, 19]. This improvement over the past decade is a result of a number of factors, including better patient selection and improved surgical technique [4, 10, 12, 14, 16].

The effect of age and weight on outcomes after the FP is unclear. Some have reported satisfactory outcome after FP performed in patients as young as 1 year, and older age at Fontan completion has been associated with worse hemodynamics [20]. Others have reported that age at FP had no effect on operative mortality [18]. In multivariable analysis, age and weight were not significantly associated with outcome, whereas lower weight-for-age z-score was associated with significantly increased in-hospital morbidity and mortality [21]. There are also controversial reports.

The median age of 3.2 years in the whole study group reflects our general preference to complete the FP at about 3 years of age in most patients, and slightly later (in relation to patient size) in those for whom an extracardiac conduit technique is chosen. We generally try to follow the advice of Lardo that for hemodynamic reasons the diameter of an extracardiac conduit should not exceed that of the inferior vena cava by a factor of 1.5, meaning that in order to avoid conduit changes the children have to be older [7]. Generally, there is a tendency to perform the operation relatively early in life, at 2 to 4 years, before serious ventricular impairment has occurred [18]. Interestingly, age and weight in both our groups at operation had no effect on the postoperative need for mechanical ventilatory support, inotropic support or length of stay on the intensive care unit (ICU). Both artificial ventilation and prolonged administration of catecholamines have a negative effect on the vulnerable Fontan circulation, which requires the lowest pulmonary vascular resistances possible. This was already recognized by Fontan in his original publications [1] and was again confirmed in our study, with the extracardiac group doing better on the ICU (Figures 1–4).

Mean pulmonary artery pressure in our selected groups at completion of FP was 13.38 mm Hg and ranged from 6 mm Hg to 23 mm Hg. It had no detectable effect on the postoperative course.

Many of the early postoperative problems can result from perioperative factors such as the CPB and/or aortic cross-clamping and cardioplegic arrest times [14]. Despite widespread acceptance of the FP for the treatment of functional single ventricle, significant controversy remains over the optimal operative strategy [11, 19].

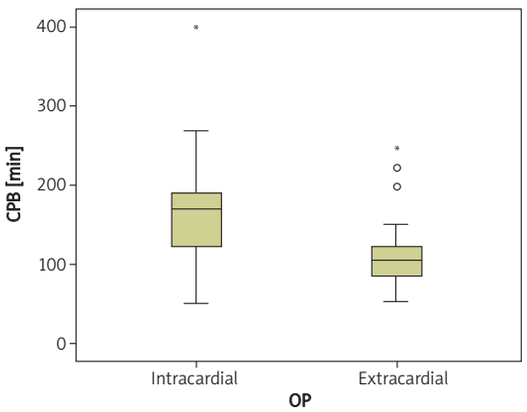

Figure 1. Cardiopulmonary bypass in intracardiac and extracardiac groups

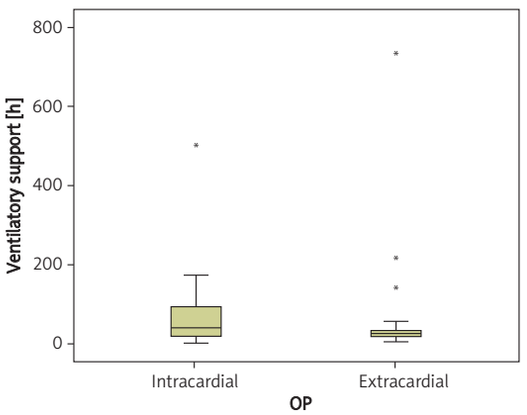

Figure 2. Time of postoperative ventilation in intracardiac and extracardiac groups

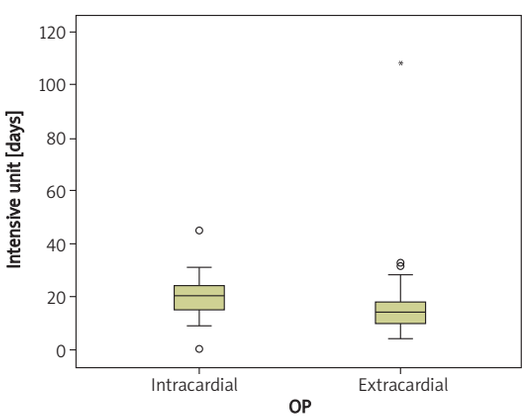

Figure 3. Hospitalization in intensive care in days in intracardiac and extracardiac groups

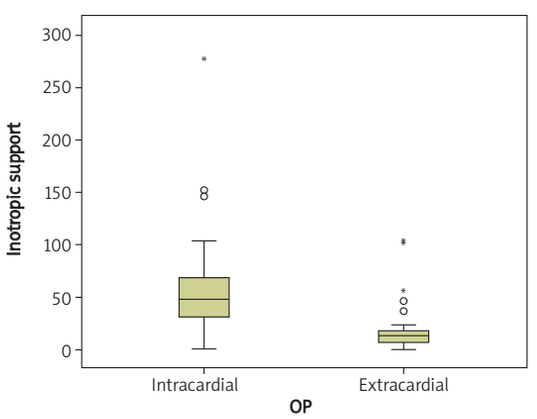

Figure 4. Hours of inotropic support in intracardiac and extracardiac groups

Extracardiac and lateral tunnel reconstructions are widely used, and each technique has its advocates [14, 16, 22]. Recent reports advocate avoidance of CPB and aortic cross-clamping to ameliorate systemic inflammation and potentially to achieve improved myocardial function, hemodynamics and outcome [23]. Certainly there are cases where these strategies are applicable, and excellent results have been reported [23]. Others have published reports of large series of completion FP using deep hypothermia with circulatory arrest. Surprisingly, mortality was significantly lower in the deep hypothermia with circulatory arrest group, vs. a group with continuous CPB with moderate hypothermia [24].

The intracardiac tunnel procedure necessitates aortic cross-clamping and cardioplegic arrest. We and other authors have observed a longer time of mechanical ventilatory support, inotropic support and stay on the intensive care unit in cases of prolonged aortic cross-clamping time [25]. This influence was seen in both of our groups.

Regarding a technical detail of another controversy [18, 22], we currently fenestrate all intracardiac baffles at our institution. In the extracardiac group, where it is technically more difficult to perform, we deemed it necessary only in two high-risk cases with preoperative PA pressures of 20 mm Hg and 22 mm Hg.

Long-term evaluation has shown that for patients with this physiology, life-long follow-up by experts in pediatric cardiology is necessary [26].

In conclusion, from our experience as described above we have developed the following strategy in our unit: children who show normal development and good hemodynamics during the preparatory steps undergo an elective extracardiac conduit procedure. For this we try to achieve at least a diameter of 18 mm to avoid growth-related conduit changes later in life. A fenestration procedure is done in cases of markedly elevated preoperative PA pressure (20 mm Hg and higher).

Patients who have retarded development, show signs of myocardial dysfunction or AV valve insufficiency, or who suffer from ongoing cyanosis are scheduled for an earlier intracardiac baffle repair with routine fenestration, as they are higher-risk patients anyway.

Our data have shown that the need for prolonged CPB and aortic cross-clamping times has a negative influence on the early postoperative course. Ideally,

a prospectively performed matched-pair analysis would strengthen this evidence. Further strategies must be developed to avoid these effects, particularly in the patient group at higher imminent risk.

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References

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax 1971; 26: 240-8.
2. Kreutzer G, Galindez E, Bono H, de Palma C, Laura J. An operation for the correction of tricuspid atresia. J Thorac Cardiovasc Surg 1973; 66: 613-21.
3. Björk VO, Olin CL, Bjarke BB, Thoren CA. Right atrial-right ventricular anastomosis for correction of tricuspid atresia. J Thorac Cardiovasc Surg 1979; 77: 452-8.
4. de Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. J Thorac Cardiovasc Surg 1988; 96: 682-95.
5. Voss B, Sack FU, Saggau W, Hagl S, Lange R. Vorhofkardiomyoplastik in einer experimentellen "Fontan-Zirkulation". Z Herz- Thorax- Gefäßchir 2003; 17: 115-22.
6. Konstantinov IE, Alexi-Meskishvili W. Cavo-pulmonary shunt: from the first experiments to clinical practice. Ann Thorac Surg 1999; 68: 1100-6.
7. Lardo AC, Webber SA, Friehs I, del Nido PJ, Cape EG. Fluid dynamic comparison of intra-atrial and extracardiac total cavopulmonary connections. J Thorac Cardiovasc Surg 1999; 117: 697-704.
8. Hopkins RA, Armstrong BE, Serwer GA, Peterson RJ, Oldham HN Jr. Physiological rationale for a bidirectional cavopulmonary shunt. A versatile complement to the Fontan principle. J Thorac Cardiovasc Surg 1985; 90: 391-8.
9. Berdat PA, Belli E, Lacour-Gayet F, Planche C, Serraf A. Additional pulmonary blood flow has no adverse effect on outcome after TCPC. Thorac Cardiov Surg 2004; 52: 280-6.
10. Marcelletti C, Corno A, Giannico S, Marino B. Inferior vena cava-pulmonary artery extracardiac conduit. A new form of right heart bypass. J Thorac Cardiovasc Surg 1990; 100: 228-32.
11. Vouhe PR. Fontan completion: Intracardiac tunnel or extracardiac conduit? Thorac Cardiov Surg 2001; 49: 27-9.
12. Tokunaga S, Kado H, Imoto Y, et al. Total cavopulmonary connection with an extracardiac conduit: experience with 100 patients. Ann Thorac Surg 2002; 73: 76-80.
13. Petrossian E, Reddy VM, McElhinney DB, et al. Early results of the extracardiac Fontan operation. J Thorac Cardiovasc Surg 1999; 117: 688-96.
14. Kuroczynski W, Kampmann C, Choi YH, et al. The Fontan-operation: from intra- to extracardiac procedure. Cardiovasc Surg 2003; 11: 70-4.
15. Klimes K, Abdul-Khaliq H, Ovroutski S, et al. Pulmonary and caval blood flow patterns in patients with intracardiac and extracardiac Fontan: a magnetic resonance study. Clin Res Cardiol 2007; 96: 160-7.
16. Kirklin JK, Brown RN, Bryant AS, et al. Is the "Perfect Fontan" operation routinely achievable in the modern era? Cardiology in the Young 2008; 18: 328-36.
17. Khairy P, Poirier N, Mercier LA. Univentricular heart. Circulation 2007; 115: 800-12.
18. Mair DD, Puga FJ, Danielson GK. The Fontan procedure for tricuspid atresia: early and late results of a 25-year experience with 216 patients. JACC 2001; 37: 933-9.
19. de Leval MR, Deanfield JE. Four decades of Fontan palliation. Nat Rev Cardiol 2010; 7: 520-7.
20. Shiraishi S, Yagihara T, Kagisaki K, et al. Impact of age at Fontan completion on postoperative hemodynamics and long-term aerobic exercise capacity in patients with dominant left ventricle. Ann Thorac Surg 2009; 87: 555-60.
21. Wallace MC, Jaggers J, Li JS, et al. Center variation in patient age and impact on postoperative outcomes. Ann Thorac Surg 2011; 91: 1445-52.
22. Alphonso N, Baghei M, Sundar P, et al. Intermediate-term outcome following the Fontan operation: a survival, functional, and risk-factor analysis. Eur J Cardiothorac Surg 2005; 28: 529-35.
23. Yetman AT, Drummond-Webb J, Fiser WP, et al. The extracardiac Fontan procedure without cardiopulmonary bypass: technique and intermediate-term results. Ann Thorac Surg 2002; 74: 1416-21.
24. Mosca RS, Kulik TJ, Goldberg CS, et al. Early results of the Fontan procedure in one hundred consecutive patients with hypolastic left heart syndrome. J Thorac Cardiovasc Surg 2000; 119: 1110-8.
25. Haas GS, Hess H, Black M, et al. Extracardiac conduit Fontan procedure: early and intermediate results. Eur J Cardiothorac Surg 2000; 17: 648-54.
26. Trojnarska O, Gwizdala A, Katarzynski S, et al. Evaluation of exercise capacity with cardiopulmonary exercise testing and BNP levels in adult patients with single or systemic right ventricles. Arch Med Sci 2010; 6: 192-7.