Medical Expertise*

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

Title: An adult patient with a single ventricle RCD code: IV-2A.3

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Date of the CRCD meeting: 09.09.2013r.

EXPERT: Prof. Vladimir Alexi-Meskishvili, pediatric cardiac surgeon

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[* The expertise should be written in English]

Medical Expertise

CASE SUMMARY

This is a high-risk patient with single ventricle, low mean pulmonary arterial pressure, severe ventricular hypertrophy, subnormal EF. Endocarditis of tricuspid valve (?) is described, but valve function (incompetence) is not given in detail

DISCUSSION

There are several treatment options:

1. Primary.Fontan operation bears very high risk, because ventricular hypertrophy, possible hepato-renal insufficiency after operation, usually seen in an older patients
2. Cavo-pulmonary shunt is insufficient in increasing arterial saturation in an adult patient, because of insufficient flow from upper body, contrary to small children
3. Cavo-pulmonary shunt with additional blood flow source (systemic-pulmonary shunt) seems to optimal option before Fontan operation
4. Cardiac transplantation on this stage to my opinion is not indicated

EXPERT'S OPINION

Cavo-pulmonary shunt with additional blood flow source (systemic-pulmonary shunt) seems to optimal option before Fontan operation

CONCLUSION

I recommend cavo-pulmonary shunt combined with systemic (central) aorto-pulmonary anastomosis. Operation can be performed without use of extracorporeal circulation, to reduce risk of operation. If TV incompetence is significant it must be corrected using ECC

REFERENCES

See below

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PEDIATRIC CARDIAC

Midterm to Long-Term Outcome of Total Cavopulmonary Connection in High-Risk Adult Candidates

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Background. Adult patients who do not fulfill the classical Fontan criteria now undergo total cavopulmonary connection (TCPC). However, limited information is available on the results for high-risk adult TCPC.

Methods. Twenty-five consecutive adult patients (aged 16 years or more) who underwent TCPC were retrospectively reviewed. The mean age at operation was 27 ± 9 years (range, 16 to 52). The following items were considered as the potential risk factors according to previous reports: (1) aged more than 30 years (7 of 25); (2) heterotaxy (9 of 25); (3) systemic ventricular ejection fraction less than 50% (6 of 25); (4) atrioventricular valve regurgitation moderate or greater (6 of 25); (5) pulmonary arterial index less than 200 (7 of 25); (6) mean pulmonary arterial pressure 15 mm Hg or greater (3 of 25); (7) pulmonary arterial resistance 2.0 wood units or greater (11 of 25); (8) arrhythmias (13 of 25); (9) protein-losing enteropathy (3 of 25); (10) New York Heart Association (NYHA) functional class III or greater (9 of 25); (11) previous Fontan procedure (10 of 25); (12) systemic ventricular outflow obstruction (1 of 25); and (13) end-diastolic pressure of the systemic ventricle 11 mm Hg or higher (4 of 25).

Results. The mean follow-up period was 57 ± 45 months (range, 0 to 154). All patients had at least 2 risk factors (range, 2 to 8). There was 1 early death and 2 late deaths. Comparing the late survivors and nonsurvivors, no statistical significance was identified in the above risk factors. However, the patients with 6 or more risk factors had a significantly higher mortality rate than patients with fewer than 6 risk risk factors ($p < 0.01$). Age ($p = 0.08$), NYHA class ($p = 0.13$), and protein-losing enteropathy ($p = 0.08$) may be risk factors for late death.

Conclusions. The majority of the adult TCPC candidates tolerated the TCPC procedure in the early postoperative period. However, the accumulation of risk factors influences late mortality.

(Ann Thorac Surg 2009;87:562–70)

Since Fontan and Baudet first described their procedure for the correction of tricuspid atresia in 1971, the principles of the Fontan procedure have been applied to all forms of functional univentricular heart defects [1]. After several modifications of this operation, the total cavopulmonary connection (TCPC), which was first reported by de Leval and colleagues [2], has become a standard method for the Fontan procedure because it provides better venous hemodynamics [3] and is less arrhythmogenic [4] than the other Fontan modifications. In addition, several management strategies have been incorporated to achieve an improved mortality: universal risk factors that have resulted in better patient selection [5]; a "staged" approach to the Fontan procedure with an interim superior cavopulmonary connection [6]; the use of fenestration [7]; and the use of modified ultrafiltration at the end of cardiopulmonary bypass [8].

In Japan, more than 300 patients have undergone the Fontan procedure annually with a hospital mortality of 3% to 4%, and approximately 10% of these patients are aged 18 years or older [9]. The TCPC procedure carries a greater risk for the adult patient than for children because the adult functional ventricle usually presents with complications caused by long-term chronic hypoxia, ventricular volume overload, and increased venous pressure, such as arrhythmia, protein-losing enteropathy (PLE), pleural effusion, ventricular dysfunction, and limited exercise capacity [10, 11]. Because heart transplantation is not a realistic surgical option in Japan owing to a very strict organ transplantation law, much more complicated adult patients with a functional single ventricle have presented to our institute. However, unlike the management of children with a functional single ventricle, management guidelines, including the preoperative predictors for mortality, have yet to be established for the adult patient who is a candidate for the Fontan procedure because few studies have so far addressed this issue. The

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Early to Midterm Results of Total Cavopulmonary Connection in Adult Patients

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Background. Total cavopulmonary connection (TCPC) has not been studied in adults. We investigated early and midterm morbidity and mortality in adults undergoing TCPC and assessed risk factors for mortality.

Methods. Between June 1994 and October 2010, 30 adults (21.3 ± 5.5 years) underwent TCPC (extracardiac conduit). Twenty-two patients who had palliated single ventricles underwent TCPC completions and 8 patients underwent TCPC conversions. Preoperative and perioperative data were reviewed retrospectively.

Results. Six of 9 patients with preoperative atrial flutter or fibrillation or intraatrial reentry tachycardia were treated in the catheterization room. An aortic cross-clamp was necessary in 12 patients, and 16 TCPCs were fenestrated. Mean follow-up was 51 months (range, 4–198 months). Early mortality was 10%: 2 of 8 conversions and 1 of 22 completions. There was 1 late conversion death (at 56 months postoperatively). Postoperatively, 4 patients required pacemakers and 1 patient required long-term antiarrhythmic medication, but no heart transplantations were necessary. Risk factors for early mortality were arrhythmia ($p = 0.02$), aortic cross-clamp ($p = 0.054$), and extracorporeal circulation in hypothermia ($p = 0.03$). Risk factors for overall mortality were conversion ($p = 0.047$), absence of fenestration ($p = 0.036$), surgery before January 2006 ($p = 0.036$), aortic cross-clamp ($p = 0.018$), extracorporeal circulation in hypothermia ($p = 0.008$), and arrhythmia ($p = 0.005$). New York Heart Association functional class had improved at the last follow-up: preoperatively, 17 patients were in class II and 12 patients were in class III versus 18 patients in class I and 9 patients in class II postoperatively ($p < 0.001$). At the last clinical visit, systemic ventricular function was maintained, and no late supraventricular arrhythmia was found.

Conclusions. Early and midterm TCPC results for adults are encouraging for completion but are disappointing for conversion. Identified risk factors for mortality should improve patient selection for TCPC.

(Ann Thorac Surg 2013;95:941–7)

In 1971 Fontan and Baudet [1] reported the first physiologic repair of tricuspid atresia: this atrial–pulmonary connection became known as a Fontan procedure. Fontan surgery was then applied to different forms of single-ventricle circulation and resulted in definitive palliation. Since 1971, numerous modifications have been introduced to this operation: the ideal age at which to undergo surgery has been defined, and the technical procedure itself has evolved [2, 3]. The main technical evolution has been a total cavopulmonary connection (TCPC) using an intraatrial tunnel [2] or an extracardiac conduit [3] instead of an atrial–pulmonary connection to improve hemodynamics and, consequently, the Fontan circulation.

Concerning age, the original selection criteria included those older than 4 years but younger than 15 years [4]. Currently, TCPC is generally performed between 2 and 6 years of age [5]: risk factors for mortality have been studied and defined for this age group [6]. In contrast, TCPC has been rarely studied in adult patients (>16 years old) [7–9], and no risk factors have been defined. However, some adult patients (with a single ventricle), with or without surgical palliation, are candidates for TCPC surgery [10]. Adults who have a failed "classic" Fontan surgery (chronic arrhythmia, protein-losing enteropathy, pleural effusions, ventricular dysfunction, limited exercise capacity) are also possible candidates for TCPC [11].

This single-center study assessed early and late morbidity and mortality after TCPC performed in adults to either complete interim palliation (TCPC-comp) or to convert a failing atrial-pulmonary connection (TCPC-conv). The risk factors for mortality were also assessed.

Patients and Methods

Study Population

Approval for this study was granted by the Institutional Ethics Committee, and the need for individual consent

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The Fontan procedure in adults

G R Veldtman, A Nishimoto, S Siu, M Freeman, P M Fredriksen, M A Gatzoulis, W G Williams, G D Webb

Abstract
Setting—Tertiary adult congenital cardiac referral centre.
Design—Retrospective cross sectional analysis.
Objectives—To report our 20 year experience with adult Fontan operations, and to compare late outcome in patients with single ventricle with definitive aortopulmonary or cavopulmonary shunt palliation.
Patients and main outcome measures—Patients older than 18 years undergoing Fontan operation between 1 January 1982 and 31 December 1998 were identified. Mortality and late outcome were derived from hospital records. These patients were compared with a cohort of 50 adults with single ventricle who had not undergone a Fontan operation.
Results—61 adults, median age 36 years (range 18–47 years), with a median follow up of 10 years (range 0–21 years) were identified. Actuarial survival was 80% at one year, 76% at five years, 72% at 10 years, and 67% at 15 years. Compared with before the Fontan operation, more patients were in New York Heart Association (NYHA) functional class I or II at the latest follow up (80% *v* 38%, $p < 0.001$). Systolic ventricular function deteriorated during follow up such that 34% had moderate to severe ventricular dysfunction at the latest follow up compared with 5% before Fontan ($p < 0.001$). Arrhythmia increased with time (10% before Fontan *v* 57% after 10 years, $p < 0.001$). Fontan patients had improved NYHA functional class, ventricular function, atrioventricular regurgitation, and fewer arrhythmias than the non-Fontan group at the latest follow up.
Conclusion—The Fontan operation in adults has acceptable early and late mortality. Functional class, systolic ventricular function, atrioventricular regurgitation, and arrhythmia deteriorate late after surgery but to a lesser degree than in non-Fontan patients with a single ventricle.
(*Heart* 2001;86:330–335)

Keywords: Fontan operation; univentricular heart; cavopulmonary shunt; aortopulmonary shunt

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Accepted 17 April 2001

The Fontan operation allows total bypass of the right heart.[1] It was first described in 1971 in three patients with tricuspid atresia.[2] The ideal age to undergo the Fontan operation is thought to be between 18 months and 6 years.[3–4] Not infrequently, however, adult patients with or without surgical palliation are encountered who are suitable for the operation.[4] Under these circumstances volume unloading the single ventricle and simultaneously improving its oxygen delivery may permit better ventricular preservation and may result in less atrioventricular (AV) valve regurgitation, better functional capacity, and potentially improved longevity. Late outcome is, however, not well understood and comparative data with more conservative management are not well documented.[4] We report our experience with the Fontan operation in adults over a 20 year period. We compared the study patients with a cohort of patients who had had either cavopulmonary or aortopulmonary shunts as their definitive palliation.

Patients and methods

PATIENTS

Fontan group

Consecutive patients older than 18 years at the time of surgery who underwent a modified Fontan procedure at the University of Toronto Congenital Cardiac Centre for Adults (UTCCCA), Toronto, Canada, between 1 January 1982 and 31 December 1998 were identified from the UTCCCA database. Hospital records, and reports of cardiac catheterisation and echocardiography were reviewed for patient characteristics. The cardiac morphology was reviewed and categorised according to operative notes, and echocardiographic and cineangiographic descriptions of the anatomy. Patients were assigned one or more of the following diagnostic categories based on their underlying anatomy: tricuspid atresia, double inlet left ventricle, pulmonary atresia with intact ventricular septum, double outlet right ventricle, atrial isomerism, transposition of the great arteries complex, or other hypoplastic ventricle.

Aortic saturations, mean pulmonary artery pressures, systolic ventricular function, the presence of pulmonary arterial stenoses (any focal or diffuse stenosis evident on either cardiac catheterisation or echocardiogram), dominant ventricular anatomy (left or other), and the severity of AV valve regurgitation were recorded from cardiac catheterisation and two dimensional (cross sectional) echocardiography reports before Fontan operation. Surgical history was obtained from operative notes. Recent follow up status was documented from hospital records and contact with the patient and referring physicians. Thirty seven study patients also formed part of a concurrent study looking at outcomes of late atrial arrhythmias after the Fontan operation.[5] Ethics approval was obtained from the Toronto General Hospital research ethics board.

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Early and medium-term results after modified Fontan operation in adults

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Abstract

Objective: Single ventricle palliation is rarely performed in adults and the results are less optimal than in children. In this article we analyze our experience with the modified Fontan operation in this age group. **Methods**: Data of 15 consecutive patients with single ventricle with a mean age of 26 (range 16–38) years, who underwent Fontan operation between 3/92 and 1/2000 were retrospectively analyzed. Five patients had previously had an aortopulmonary shunt in childhood and two patients had previously received a bi-directional cavopulmonary shunt as adults. Eleven patients were preoperatively in NYHA class III and four in class II. The main factors for the selection of the patients before surgery were well-developed pulmonary arteries with lower lobe index 120 + 30 mm/m^2, pulmonary artery pressure <18 mmHg, good cardiac function and enddiastolic systemic ventricular pressure <12 mmHg. The lateral tunnel Fontan operation (LTFO) was performed in ten patients and extracardiac Fontan operation (ECFO) in five. A fenestration 4–5 mm in size was constructed in all patients with LTFO and in three of five patients with ECFO. **Results**: There was one intraoperative and one late death (total mortality 13%). The mean extubation time and hospital stay were 24 h and 21 days, respectively. Severe postoperative complications were observed in three patients (20%). Two LTFO patients out of a total of eight patients (53%) with perioperative arrhythmias received a permanent pacemaker due to bradyarrhythmia. During the median follow-up of 5.0 (range 2.3–10.1) years, four patients developed arrhythmias; one of them had new onset bradyarrhythmia after LTFO and required permanent pacemaker implantation. The median postoperative oxygen saturation was 93% (range 90–98%). NYHA class improved significantly in 12 survivors. Cardiac catheterization (0.5–4 years postoperatively, $n = 12$) showed excellent Fontan hemodynamics in all patients. **Conclusions**: The modified Fontan operation can be performed in adults with acceptable early and midterm mortality and morbidity and leads to either complete or marked relief of cyanosis and enhanced exercise tolerance in all survivors. Postoperative arrhythmias are one of the main drawbacks but the incidence of arrhythmias after ECFO seems to be lower. The long-term follow-up has yet to be established.

Keywords: Fontan operation in adult; Extracardiac; Lateral tunnel

1. Introduction

In patients with single ventricle, the Fontan operation relieves cyanosis and ventricular volume overload. It improves exercise tolerance and preserves ventricular function. The ideal time for the operation is in early childhood to avoid long-term volume overload and cyanosis, which may lead to valve regurgitation and myocardial fibrosis. In adult patients the Fontan operation bears these special risk factors for an adverse outcome [1–3]. To establish whether late surgical Fontan palliation in adults is beneficial, we analyzed our results of the last 8 years.

2. Patients and methods

Between March 1992 and January 2000, 15 consecutive adults (23%) of the total of 64 patients with single ventricle physiology underwent a Fontan operation at our institution. Their mean age was 26 years (range 16–38 years) and mean body weight 60 kg (range 42–90 kg). Eight patients were older than 25 years. Previous surgical procedures during childhood were establishment of an aortopulmonary shunt in five patients and of pulmonary artery banding in two. Two patients had received a bi-directional cavopulmonary shunt prior to the definitive Fontan operation. Preoperatively four patients were in NYHA class II and 11 in class III (Fig. 1). Preoperative arterial oxygen saturation was 82% (range 64–85%, Fig. 2). The anatomical diagnoses are presented in Table 1. All patients were seen for the first time in our outpatient department as adults.

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REVIEW ARTICLE

Role of Systemic to Pulmonary Artery Shunt after Cavopulmonary Anastomosis

Pankaj Garg, M.Ch., Sachin Talwar, M.Ch., Shyam Sunder Kothari, D.M., Palleti Rajashekar, M.Ch., Rachit Saxena, M.Ch., and Balram Airan, M.Ch.

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ABSTRACT Superior cavopulmonary anastomosis and total cavopulmonary anastomosis are the procedures of choice for the management of patients with a functionally univentricular heart. We review the various indications, sites, advantages, and complications of a systemic to pulmonary artery shunt after the creation of superior cavopulmonary anastomosis. Systemic pulmonary artery shunt may be useful as a palliative strategy in patients who have hypoxemia and completion of total cavopulmonary anastomosis is not feasible. doi: 10.1111/jocs.12154 *(J Card Surg 2013;28:599–603)*

Since its introduction in the 1940s, a systemic to pulmonary shunt has played an important role in the management of patients with congenital cyanotic heart disease. Currently it is performed as an initial palliation in neonates and small infants with a structurally or functionally univentricular heart as the older children commonly undergo a bidirectional superior cavopulmonary anastomosis (BDG) or one of the types of total cavopulmonary connection (TCPC).[1] Both BDG and TCPC have good short- and mid-term results as both these procedures improve functional capacity of the patient, decrease cyanosis, and decrease the volume overload of the single ventricle.[2,3] Advantages of BDG over systemic pulmonary artery shunt are that it decreases the volume loading of the heart leading to a decrease in ventricular end diastolic pressure and ventricular work. In addition, the mean pulmonary artery pressure and pulmonary vascular resistance remains low compared to patients with a systemic pulmonary artery shunt.[4] The Fontan operation further underloads the ventricle, increases the pulmonary blood flow, and prevents the formation of pulmonary arteriovenous collaterals by delivering the hepatic factor into the pulmonary circulation.[5] However, in the long term both procedures are fraught with complications such as cerebrovascular accidents, arrhythmias, progressive cyanosis and dyspnea on exertion due to the development of systemic to pulmonary artery collaterals, pulmonary arteriovenous fistulae and systemic venous collaterals[1] protein-losing enteropathy, and progressive Fontan failure that may necessitate a cardiac transplantation.[6] This may be due to the development of systemic venous hypertension, pulmonary arterial hypotension, or ventricular dysfunction.[7]

Encouraged by the early results of both these procedures, it was thought that the right ventricular pump is not mandatory for the pulmonary blood flow and energy present in the vena-caval blood flow is sufficient for blood to pass through the pulmonary circulation.[8] But the development of complications after both of these procedures in the long-term has led to a renewed interest in the role of the right ventricle in pumping blood into the pulmonary circulation.

It is well known that the essential functions of the right ventricle are to pump blood into the pulmonary circulation at an adequate pressure to keep the pulmonary capillary bed open.[8,9]

A low systemic venous pressure is essential for prevention of peripheral edema and hepatic congestion. At the same time, a pulsatile pulmonary blood flow provides better transfer of pressure energy from the main pulmonary artery to the pulmonary capillary bed compared to linear flow, which leads to recruitment of additional capillary bed requiring increased pressure to open.[10] Recruitment of additional capillary bed leads to a decrease in the pulmonary vascular resistance and improvement in gaseous exchange.[11] Also there is improvement in pulmonary lymphatic drainage by transmitting the arterial contractions to the lymphatic vessels wrapped around the pulmonary arteries.[10] Finally it also provides growth stimulus to the pulmonary arteries.[12]

Patients who have undergone a BDG or a TCPC are known to gradually develop an increase in systemic

Conflict of interest: The authors acknowledge no conflict of interest in the submission.

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The Effect of Additional Pulmona
Timing of the Total Cavopulmona

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Joost P. van Melle, MD, PhD, Rolf M.F. Berger, MD, PhD,

Departments of Cardiothoracic Surgery, Cardiology, and Pediatric Cardiology, Cente
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PEDIATRIC CARDIAC

Background. The staged Fontan procedure is used to palliate functionally univentricular hearts. The effect of additional pulmonary blood flow combined with a bidirectional cavopulmonary shunt in these patients remains a controversial subject.

Methods. This retrospective study included all 82 patients with a unilateral or bilateral bidirectional cavopulmonary shunt at our institution between April 1990 and July 2010. Patients with hypoplastic left heart syndrome were excluded. Two groups, based on the presence (n = 57) or absence (n = 25) of additional pulmonary blood flow after the bidirectional cavopulmonary shunt, were compared.

Results. Patients with a bidirectional cavopulmonary shunt combined with additional pulmonary blood flow had higher arterial oxygen saturations postoperatively (86% [interquartile range, 85% to 90%] vs 82% [80% to 85%]; $p = 0.001$) and had a longer median interval before the total cavog
years vs 2.90 [2
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Conclusions.
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The staged Fontan procedure has been proven to reduce death and provide early volume unloading in patients with functionally univentricular hearts [1–3]. The bidirectional cavopulmonary shunt (BCPS) combined with additional pulmonary blood flow (APBF) on track to the total cavopulmonary connection (TCPC) remains a controversial subject.

Theoretical advantages of APBF may include higher oxygen saturations, increased pulmonary artery growth, and implantation of a larger extracardiac conduit at completion of the TCPC. Disadvantages may include elevated central venous pressure in the upper body, increased ventricular volume load, a negative effect on final Fontan candidacy, and increased interval morbidity [4–7]. In this study we analyzed some of the effects of APBF on the timing of the TCPC in patients with functionally univentricular hearts.

Patients and Methods

This study is a retrospective, single-center cohort study of patients with single-ventricle physiology. We included all 82 patients
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Accepted for publication Feb 20, 2012.

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[** Signing the Expertise will mean an agreement for its publication on www.crcd.eu]

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