Extracardiac Fontan with direct cavopulmonary connections: midterm results

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Abstract

OBJECTIVES: The aim of this study was to analyse the midterm results following superior and inferior direct cavopulmonary connections (DCPC) to create a Fontan circulation in patients with functionally univentricular hearts.

METHODS: A retrospective review of patients operated on between January 2005 and December 2011 was carried out.

RESULTS: Thirty consecutive patients who underwent this type of operation were retrospectively reviewed. There were 18 (60%) males and 12 (40%) females, with a median age of 69 months (range 16–150 months) and median weight of 23 kg (range 11–46 kg). Aortic cross-clamping was used in 10 patients, with a median cross-clamp time of 40 min (range 23–99) and a median cardiopulmonary bypass (CPB) time of 135 min (range 76–179 min). The remaining 20 patients were operated on without aortic cross-clamping. Their median CPB time was 104 min (range 78–139 min). Fenestration was performed in 16 patients. The associated intracardiac procedures were performed in 10 patients. The follow-up period ranged from 2 months to 6 years. Operative mortality and late mortality after discharge was zero. The major postoperative complications included supraventricular tachycardia in one patient, oliguria and peritoneal dialysis in one and chest drainage (>30 ml/day) persisting >7 days in five (20%). One patient developed sinus bradycardia in association with sinus pauses 2 months after discharge. One patient developed pericardial effusion 1 month after discharge. A computational fluid dynamic study was performed in one patient. The computational fluid dynamic study showed that DCPC may have a better power efficiency.

CONCLUSIONS: Superior and inferior DCPCs to create a Fontan circulation in appropriately selected patients with functionally univentricular hearts can be performed with a low risk and a low rate of reinterventions. The midterm results are favourable.

Keywords: Total cavopulmonary connection • Functionally univentricular heart • Fontan • Congenital heart surgery

INTRODUCTION

Since first being introduced in 1971 by Dr Francis Fontan [1], the Fontan palliation for patients with functionally univentricular hearts has undergone significant evolution. From the atropulmonary connection procedures independently pioneered by Fontan and Kreutzer for repair of tricuspid atresia, surgical approaches have transitioned to a total cavopulmonary connection with either an intracardiac lateral tunnel (LT) or, most recently, the extracardiac conduit (EC) technique [2, 3].

With advances in anaesthetic techniques, perfusion strategies and postoperative care, the EC type of reconstruction is often accomplished with the avoidance of systemic cooling, cardioplegic cardiac arrest and extensive atrial manipulation, all of which may have a negative effect on survival [4]. However, compared with the LT circuit, the EC pathway cannot grow and, as such, future reoperations may be required with the potential for associated significant morbidity and mortality. In addition, the prosthetic material may predispose the patient with a single ventricle to an already increased risk of thromboembolic complications.

More than 600 Fontan operations of all types have been performed at the Shanghai Children’s Medical Center (SCMC), University of Shanghai Jiaotong University (SJTU). The EC technique has been used in parallel with LT connections since 2000. More recently, we have adopted the alternative extracardiac Fontan with DCPC since 2005. We reviewed our total experience and report here the midterm results with the first 30 patients.

MATERIALS AND METHODS

The present study was approved by our Institutional Review Board of SCMC, and the need for written informed consent from the patients was waived.
Table 1: Preoperative patient demographics

<table>
<thead>
<tr>
<th>Preoperative catheterization data</th>
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<tbody>
<tr>
<td>PA pressure (mmHg)</td>
<td>10.7 ± 4.6</td>
</tr>
<tr>
<td>( R_p ) (Wood units)</td>
<td>1.7 ± 0.7</td>
</tr>
<tr>
<td>( Q_p/Q_s )</td>
<td>1.4 ± 0.48</td>
</tr>
<tr>
<td>Nakata index</td>
<td>356 ± 137</td>
</tr>
<tr>
<td>Ejection fraction (%)</td>
<td>49 ± 8</td>
</tr>
<tr>
<td>Room air arterial saturation (%)</td>
<td>77 ± 9</td>
</tr>
</tbody>
</table>

PA: pulmonary artery; \( R_p \): pulmonary vascular resistance index; \( Q_p/Q_s \): pulmonary to systemic flow ratio.

Table 2: Morphological features and previous procedures

<table>
<thead>
<tr>
<th>Morphological features</th>
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<tbody>
<tr>
<td>Univentricular heart</td>
<td>11</td>
</tr>
<tr>
<td>TA</td>
<td>6</td>
</tr>
<tr>
<td>DILV with small LV</td>
<td>3</td>
</tr>
<tr>
<td>PAIVS</td>
<td>2</td>
</tr>
<tr>
<td>d-TGA VSD PS</td>
<td>6</td>
</tr>
<tr>
<td>ccTGA VSD PS</td>
<td>6</td>
</tr>
<tr>
<td>DORV CAVSD PS</td>
<td>4</td>
</tr>
<tr>
<td>Isomerism/heterotaxy syndrome</td>
<td>3</td>
</tr>
<tr>
<td>Previous procedures</td>
<td></td>
</tr>
<tr>
<td>Right bidirectional Glenn</td>
<td>1</td>
</tr>
<tr>
<td>Bilateral bidirectional Glenn</td>
<td>1</td>
</tr>
</tbody>
</table>

TA: tricuspid atresia; DILV: double inlet left ventricle; LV: left ventricle; PAIVS: pulmonary atresia intact ventricular septum; d-TGA: d-transposition of great arteries; VSD: ventricular septal defect; PS: pulmonary stenosis; ccTGA: congenitally corrected transposition of great arteries; DORV: double outlet right ventricle; CAVSD: complete common atrioventricular septal defect.

Patients

A total of 321 Fontan operations were performed between January 2005 and December 2011. Thirty (9%) patients were operated on using the DCPC extracardiac Fontan procedure. All patients underwent a preoperative evaluation with transthoracic echocardiography, with cardiac catheterization used in all patients.

Patients were identified by a review of the institutional cardiothoracic surgical database. The medical record of each subject was reviewed. The study cohort included 18 (60%) males and 12 (40%) females. The mean age was 49 ± 35 months (range 16–150 months) and mean weight was 19 ± 10 kg (range 11–46 kg). The preoperative patient demographics are summarized in Table 1. The morphological features and previous procedures are summarized in Table 2.

The sizes of the main pulmonary artery (MPA) and both right and left pulmonary arteries (RPA and LPA) were assessed by cardiac catheterization. A direct connection was performed without other procedures if the PAI was >250 mm²/m² and the MPA–aorta ratio was >0.75. The direct connection between the posterior wall of the MPA and inferior vena cava (IVC) with an autologous pericardial patch enlargement or branch pulmonary arteries reimplantation if 200 mm²/m² < pulmonary artery index (PAI) < 250 mm²/m² and 0.5 < MPA–aorta ratio < 0.75.

Surgical technique

After systemic heparinization, conventional cardiopulmonary bypass (CPB) support was established with cannulation of the ascending aorta, the superior vena cava (SVC) close to the brachiocephalic vein (innominate vein) and the IVC close to the suprahepatic veins. CPB was started with mild hypothermia (32°C).

The arterial duct was divided if it existed. The LPA and RPA were mobilized out to the lobar branches. The MPA was sectioned close to the pulmonary valve and a suture closure of the proximal stump was accomplished with 5-0 and 6-0 polypropylene sutures using the pericardium after the pulmonary valve was cut. The SVC was divided above the sinus node and the cardiac end was closed.

A downward retraction of the diaphragm allowed a careful incision of the pericardial attachments to the IVC and hepatic veins, increasing their effective intrapericardial length by several centimetres. Continuing this dissection along the right pericardial reflection allowed the pulmonary veins to fall away from the atrium and increased the space for creation of the cavopulmonary pathway. The mobilized MPA was then brought to the side of the IVC, confirming that a direct connection would be possible.

If the atrial septum was intact, or the septal defect was a restrictive defect, or additional intracardiac procedures were needed, a cardiopulmonary arrest was used. After cross-clamping the aorta, the IVC was divided from the atrium leaving as much length with the IVC as possible. Indicated intracardiac procedures were then performed. The atrial stump at the site of the division of the IVC was then closed using a 5-0 polypropylene suture.

Five manoeuvres were used, singly or in combination, to facilitate the connection between the caval veins and the pulmonary trunk with a 5-0 or 6-0 polypropylene suture: (i) direct connection between the MPA and IVC (15 patients); (ii) direct connection between the posterior wall of the MPA and IVC with an autologous pericardial patch enlargement of the anterior wall of the MPA (four patients); (iii) direct connection between the posterior wall of the MPA and IVC with an autologous pericardial patch enlargement of the LPA (seven patients); (iv) direct connection between the posterior wall of the MPA and IVC with an autologous pericardial patch enlargement of the SVC (three patients). These techniques, used in conjunction with superior cavopulmonary anastomosis, are illustrated in Fig. 1. In each instance, the azygos vein was divided and the SVC was anastomosed to the RPA using a 6-0 polydioxanone suture (bidirectional Glenn operation). The site of the superior cavopulmonary connection was selected with a goal of minimizing the tension on the anastomosis.

Fenestration was required if the pressure of the SVC was >15 mmHg after CPB. A 4-mm Gore-Tex tube graft (W.L. Gore & Associates, Flagstaff, AZ, USA) was interposed between the IVC cannulation site and the lateral wall of the atrium using partial occlusion clamps during post bypass ultrafiltration.

Computational fluid dynamic study

A 3-year old boy was diagnosed with univentricular heart/complete common atroventricular canal/pulmonary stenosis.
A one-stage DCPC Fontan procedure had been performed at the age of 2 years. The direct connection between the posterior wall of the MPA and IVC was performed with an autologous pericardial patch enlargement of the LPA. A 4 mm fenestration was made between the pericardial patch and the left atrium. Magnetic resonance imaging (MRI) was performed. A series of continuous 5-mm-thick MRI images were acquired on a 1.5 Tesla Signa Hispeed scanner (GE Healthcare, USA) with a 256 × 192-pixel field of view. The MRI showed that the fenestrations had closed spontaneously. In addition to the anatomical slices, magnetic resonance phase-contrast velocity measurements were performed to acquire the mean flow rate: left innominate vein (LIV) 0.48 l/min, right innominate vein (RIV) 0.5 l/min, IVC 0.67 l/min, LPA 0.71 l/min and RPA 1.02 l/min.

A three-dimensional reconstruction of the DCPC connection area was performed in Mimics12.0®. A reconstructed model is shown in Fig. 2. The vessels were cut orthogonally to their axes near their distal ends and then extruded over a distance sufficient for computational purposes.

Grid generation was performed in ANSYS-ICEM®. An unstructured grid was generated in the central connection area and a structured grid was generated in the extended section of the vessels. CFD simulations were carried out at a different grid number of the central connection area to perform a grid sensitivity analysis. We chose grid number 500,983 for the following CFD simulations, as shown in Fig. 3.

CFD simulations were performed in ANSYS-CFX®. We assumed that the vessel walls were rigid and impermeable. We also assumed no-slip boundary conditions at the vessel walls (velocity = 0). The assumed fluid was homogeneous and Newtonian with a constant density \( q = 1060 \text{ kg/m}^3 \) and viscosity \( \mu = 3.5 \times 10^{-3} \text{ Pa s} \).

The mass flow rate of the innominate veins and IVC was set as inflow boundary conditions in the CFD models. The different levels of the LPA/RPA flow ratios were simulated by changing the mass flow rate of the LPA artificially. Thus, we also performed another five groups of simulations in different kinds of the LPA/RPA flow ratios (30:70, 40:60, 50:50, 60:40 and 70:30). Controlled energy loss under these conditions was calculated respectively. Furthermore, in order to simulate moderate and severe exercises, we increased the mass flow rate of the IVC to two and three times of that measured and calculated the controlled power loss in these conditions too. Inflow boundary conditions were as follows: LIV (kg/m³) 0.00848, RIV (kg/m³) 0.01025, IVC (kg/m³) 0.01184, ×2 IVC (kg/m³) 0.02367 and ×3 IVC (kg/m³) 0.03551.

Control volume power loss was calculated as follows:

\[
\dot{E}_{\text{loss}} = \sum_{\text{inlet}} P_{\text{total}} Q_{i} - \sum_{\text{outlet}} P_{\text{total}} Q_{i}
\]

where \( P_{\text{total}} \) and \( Q_{i} \) are the mean pressure and volume flow rate on the cross sections where the vessels were cut.
Follow-up

The early postoperative period was defined as the first 30 days after the operation. Operative mortality was defined as any death within 30 days after surgery or after 30 days but during the same hospitalization subsequent to the operation. The mean follow-up period after the operation was 28 ± 17 months (range 2–72 months). Chest X-ray examinations, electrocardiogram and echocardiographical examination were performed every 6 months during the follow-up.

Statistical analysis

The data were presented as the mean ± standard deviation and/or median and range for continuous variables. All analyses were performed using SPSS version 11.0 (SPSS Inc., Chicago, IL, USA).

RESULTS

Morbidity and mortality

Aortic cross-clamping was used in 10 patients. Their mean CPB time was 114 ± 32 min (range 76–179 min); the mean aortic cross-clamp time was 57 ± 22 min (range 23–99). Twenty patients underwent DCPC operations without aortic cross-clamping. Their mean CPB time was 99 ± 20 min (range 78–139 min). Associated intracardiac procedures were performed in 10 patients, including atrial septectomy in nine, common atioventricular valve repair in one, patent arterial duct ligation in 10 and ligation of aortopulmonary collaterals in two. Fenestration was utilized in 16 patients.

Inotropic support with milrinone was administered to all patients postoperatively. Twenty-five (83%) patients also received infusions of dopamine hydrochloride in the cardiac ICU. The mean duration of the ventilatory support time was 14 ± 7 h (range 4–24 h). The mean length of the intensive care unit stay was 3 ± 1 days (range 2–6 days) and the mean length of hospital stay was 12 ± 5 days (range 7–28 days). At the time of discharge, the mean oxygen saturation was 93 ± 5% (range 80–100%). Operative mortality and late mortality after discharge was zero. The major postoperative complications included supraventricular tachycardia in one patient, oliguria and peritoneal dialysis in one and chest drainage (>30 ml/day) persisting >7 days in six (20%).

A 10-year-old boy with heterotaxy syndrome with the dominant right ventricle underwent DCPC Fontan with concomitant valvuloplasty of the common atioventricular valve. The 12-lead electrocardiographic records were performed before and shortly after the operation, at the time of hospital discharge and during the period of the follow-up. He had regular sinus rhythm at the time of hospital discharge, but was found to have developed sinus bradycardia in association with sinus pauses 2 months after discharge. He recovered regular sinus rhythm after 2 weeks. A 4-year-old boy with the single left ventricle was found to have developed a significant pericardial effusion 1 month after discharge. He had not taken the medications (digoxin, furosemide and acetylsalicylic acid) prescribed at the time of discharge from the hospital. The effusion resolved with medical therapy.

None of the patients had evidence of thromboembolic events during hospitalization or during the period of follow-up. No echocardiographic signs of caval flow obstruction were detected.

Computational fluid dynamic study results

Controlled power loss at different levels of the pulmonary flow ratio was calculated when the mass flow rate of the IVC was that measured and had increased to two and three times, as shown in Fig. 4.

At the resting state, the mass flow rate of the RPA proportioned 58.96% of the total inflow volume, and the control volume power loss was 3.518 mW. As the flow rate of the IVC increased, power loss increased prominently. Control volume power loss reached its lowest level when the mass flow rate of the RPA proportioned 70% of the total flow rate and was 3.119, 5.272 and 8.619 mW, respectively, when the flow rate of the IVC was 1, 2 and 3 times of that measured. On the contrary, the highest power loss was 6.302, 11.515 and 18.828 mW, respectively in these three states when the mass flow rate of the RPA proportioned 30% of the total inflow rate.

The streamlines when the mass flow rate of the IVC was set as measured were demonstrated by particle path plots, as shown in Fig. 5.

DISCUSSION

Since its introduction in 1971 as definitive repair for tricuspid atresia, the Fontan procedure has undergone multiple modifications, encompassing several forms of palliative surgery that divert systemic venous return to the PA, usually without the interposition of a subpulmonary ventricle. The outcomes after the Fontan procedure have improved significantly over the past four decades [5–7]. The optimization of the outcomes in part requires the construction of a Fontan circuit with optimal flow dynamics and the preservation of ventricular and pulmonary
vascular functions, factors that can be influenced by operative techniques.

Experimental or computerized fluid dynamic studies suggest that all types of cavopulmonary connection are more efficient than atrioventricular connection for minimizing energy losses. Some studies show that the EC conduit seems to provide the least fluid energy dissipation at all flow rates and, especially, at high flows [8–10]. Moreover, the spatial arrangement of the EC conduit that minimizes the energy loss is with the cephalad end of the conduit oriented towards the LPA and the SVC anastomosis towards the RPA. This spatial arrangement produces a beneficial vortex that facilitates forward cavopulmonary flow, especially at high flow rates [11].

As the fundamental physiology of the Fontan circulation is the venous return dissociated from a ventricular power source, respiration and gravity have a more significant influence on the venous return. Gravity and respiration have important influences on the infradiaphragmatic venous return in Fontan patients [12–14]. There is extensive clinical experience supporting the use of the EC Fontan connection [15–18]. Those who prefer EC Fontan connections over the LT technique cite good haemodynamics, low incidence of arrhythmias, minimal requirement for cardiopulmonary bypass, possible avoidance of aortic cross-clamping, low operative morbidity and mortality and possibly reduced cost.

Our DCPC Fontan procedure is viewed as an alternative to an EC type of the Fontan connection in appropriately selected patients with functionally univentricular hearts and has the advantage of the balanced PA and the IVC flow distribution. The directly bowed anastomosis with the SVC and IVC favours smooth laminar mixing and low SVC and IVC pressures. Our DCPC Fontan experience has included zero operative mortality, a lower incidence of early postoperative and late arrhythmias, improved haemodynamics, and shorter cardiopulmonary and cross-clamp times. It also may be performed without cross-clamping if intracardiac procedures are not required.

Fenestration is difficult to perform in DCPC Fontan, as in the EC Fontan procedure. To address this challenge, Jonas [19] advocated the intra/extracardiac fenestrated conduit Fontan. In our series, a 4-mm Gore-Tex tube graft was easily interposed between the IVC cannulation site and the lateral wall of the atrium using partial occlusion clamps during the post bypass ultrafiltration if needed.

We are aware of four previous reports describing experiences with the DCPC Fontan [20–23]. Those reports suggested that the DCPC Fontan might be feasible in patients with an elongated SVC and a well-developed main PA located to the right or anteroposteriorly relative to the aorta, and facilitated the accomplishment of a single-stage Fontan procedure in small patients. However, it is possible to construct this route by using an autologous pericardium and the atrial wall, and replanted pulmonary branches or a reanastomosed Glenn connection. The aortopulmonary relationship did not influence this technique using our criteria. In this series, 12 of 30 operations were performed with the use of an autologous pericardium to augment the anterior wall of the MPA, LPA and/or RPA. Reimplantation of the LPA was accomplished in one patient and a previous bidirectional Glenn connection was reanastomosed into the RPA in one patient. The pericardial patch has no growth potential, but the posterior wall was of self-tissue and could grow with age. The postoperative and follow-up echocardiogram showed a normal drainage of both the SVC and IVC. Previous bidirectional Glenn does not seem to be a relative contraindication to this technique. For many two-stage patients with a small PA with a PAI <250 mm$^2$/m$^2$ and a MPA–aorta ratio <0.5, however, a DCPC Fontan may be more suitable than a one-stage Fontan. We recently performed this procedure using a portion of aortic tissue to augment the IVC to the MPA connection in a patient with a double outlet right ventricle, unbalanced common atroventricular canal defect and diminutive PA.

Our computational fluid dynamic study showed that the increase in controlled power loss was only 2.86 times that in the resting state. From this point of view, the DCPC may have a better power efficiency because of avoiding the small angle between the IVC and the PAs. In the measured data, the flow rate of the RPA is larger than that in the LPA. The mass flow rate of the RPA proportioned 58.96% of the total inflow rate. In our opinion, the main cause is the diameter of the RPA being larger than the LPA which leads to the power loss decrease when more blood flows into the RPA. We find another interesting phenomenon in that the majority of the streamlines from the IVC go directly to the LPA, which may result in an unbalanced pulmonary flow distribution in the DCPC.

LIMITATIONS

The validity of our study is clearly limited by its retrospective, nonrandomized nature. An additional limitation is the relatively small study size, including the follow-up MRI and computational model data. Extrapolating the results to the whole cohort is wrong. A larger series with a randomized design would probably address the differences between DCPC Fontan and other Fontan procedures.

CONCLUSIONS

The direct cavopulmonary connection (DCPC) type of Fontan operation can be performed on some patients with functionally univentricular hearts using an autologous pericardium and the atrial wall to facilitate a one- or two-stage extracardiac Fontan, particularly for patients with a good PA. The use of prosthetic material in the Fontan pathway is avoided. Our midterm results are favourable. We have demonstrated a low risk of mortality and a low rate of reinterventions. However, there is concern about the long-term patient survival and late mortality and this will be the subject of further investigations.
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