The impact of extracardiac conduit-total cavopulmonary connection on apicocaval juxtaposition

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Abstract

Objectives: Modifications of the Fontan procedure are applied to a wide range of complex congenital heart defects with single ventricle physiology. We examined the pathway and the clinical results of extracardiac conduit-total cavopulmonary connection (EC-TCPC) for a malpositioned heart with apicocaval juxtaposition. Methods: Of the 365 patients who underwent EC-TCPC since 1994, 56 patients with a malpositioned heart with apicocaval juxtaposition were included in this retrospective study (group 1). The pathway for the EC was selected after careful consideration of the results of preoperative angiography and computed tomography, as well as intra-operative findings. A concurrent malpositioned heart with apicocaval juxtaposition were included in this retrospective study (group 1). The pathway for the EC behind the ventricle or crossing the vertebra can be used without conduit stenosis or pulmonary venous obstruction, and no patient required re-operation. Three patients developed late complications, including arrhythmias requiring medication in two and subdural haematoma in one. The incidence of death or late complications did not differ among the pathways of conduits. In group 2, there were eight late deaths, and 27 patients developed late complications. The Kaplan–Meier survival rate was 93.5% at 5 and 10 years in group 1, and 97.3% at 5 years and 96.1% at 10 years in group 2 (log-rank test, \( P = 0.29 \)). The haemodynamics in groups 1 and 2 during the intermediate term were identical with respect to IVC pressure (8.9 ± 2.5 in group 1 and 9.6 ± 2.6 mmHg in group 2), left ventricular end-diastolic pressure (4.4 and 4.3 mmHg, respectively), cardiac index (3.3 and 3.4 l min\(^{-1} \) m\(^{-2} \)), respectively) and arterial oxygen saturation (94 and 94.2%, respectively). No patient in either group had a pressure gradient >2 mmHg between the IVC and central PA. Postoperative catheterisation data showed no significant differences in haemodynamics between the conduit pathways in group 1. Conclusions: EC-TCPC can be performed in children with apicocaval juxtaposition with excellent mid-term outcomes compared with other Fontan candidates. Based on individual cardiac anatomy, the pathway of the EC behind the ventricle or crossing the vertebra can be used without conduit stenosis or pulmonary venous obstruction. © 2010 European Association for Cardio-Thoracic Surgery. Published by Elsevier B.V. All rights reserved.

Keywords: Apicocaval juxtaposition; Extracardiac total cavopulmonary connection; Fontan operation

1. Introduction

The surgical procedure for single ventricle physiology has evolved for decades, and modifications of the Fontan procedure are applied to a wide variety of complex congenital heart defects with single ventricle physiology [1–7]. The current modifications in common use are intra-atrial lateral tunnel-total cavopulmonary connection (LT-TCPC) and extracardiac conduit-total cavopulmonary connection (EC-TCPC). In Fukuoka Children’s Hospital, EC-TCPC has been the procedure of choice for single ventricle physiology even with complex cardiac anatomy since the mid-1990s because of its feasibility and potential advantages, such as optimal laminar flow in the systemic venous pathway, minimal atrial surgery and cardiac ischaemic time and avoidance of pressure-related atrial stretch [8,9].

Apicocaval juxtaposition is a morphological feature of the cardiac apex pointing towards the ipsilateral side of the inferior venacava (IVC). Apicocaval juxtaposition makes it difficult to construct a lateral tunnel or a long, curved EC...
route. If the heart is malpositioned with apicocaval juxtaposition, several issues should be considered in selecting the best course of the IVC and pulmonary artery (PA) pathway. When the conduit is placed between the PA and the contralateral side of the IVC, the conduit has to cross the spinal column (Fig. 1). Crossing the spine may cause a kink and collapse of the conduit at the crossing point. In contrast, if the conduit is positioned straight between the IVC and the same side as the PA, the conduit should run behind the ventricle (Fig. 1). With this pathway, potential stenosis of the conduit due to compression by the vertebrae is of concern.

Little information is available on outcome of the Fontan procedure for apicocaval juxtaposition, mainly because of the relatively rare morphological features of this cardiac anomaly [10,11]. Therefore, we examined the pathway and the clinical results of EC-TCPC for a malpositioned heart with apicocaval juxtaposition.

2. Material and methods

2.1. Patient population

Between January 1994 and October 2008, 365 patients underwent EC-TCPC at the Fukuoka Children's Hospital in Fukuoka, Japan. Of these, a cohort of 56 patients with malpositioned heart with apicocaval juxtaposition was included in this retrospective study (group 1). The pathway of the EC was selected after careful consideration of the results of preoperative angiography and computed tomography (CT) as well as intra-operative findings. A concurrent group of 299 patients undergoing EC-TCPC without apicocaval juxtaposition was used as a control group (group 2). The mean follow-up was 5.5 years (range: 0–12 years) for group 1 and 5.5 years (range: 0–14 years) for group 2.

2.2. Surgical procedures

After median sternotomy, the pulmonary artery, atrium, ventricle, IVC and hepatic vein were dissected carefully and extensively without tension. After establishing cardiopulmonary bypass, an extracardiac graft was anastomosed with the caudal surface of the PA. A GORE-TEX® tube graft was used as an EC for TCPC. Seven patients in group 1 underwent primary TCPC without a staged bidirectional Glenn operation, and 21 patients in group 2 also underwent primary TCPC during this period. Since 1999, we have employed a staged Fontan operation.

For the first three cases in this series, ringed Gore-Tex was used as a conduit; however, in one of these three patients, the ring was removed to avoid pulmonary venous obstruction. The size of the GORE-TEX® tube graft is shown in Table 1. The IVC was then fully mobilised, transected and anastomosed without cardiac arrest. The extracardiac graft was anastomosed with the IVC. The patients were then weaned off cardiopulmonary bypass. Fenestration was not routinely used for the TCPC operation; it was performed only when high CVP or haemodynamic instability occurred during the surgery. Concomitant procedures were performed if necessary, and these procedures are shown in Table 1.

The pathway for the EC was selected after careful consideration of the results of preoperative angiography and CT as well as the intra-operative findings. The main factors considered in pathway selection were avoidance of pulmonary venous obstruction, avoidance of severe adhesions, balancing of pulmonary flow, placement of the graft on the ipsilateral superior venacava (SVC) and avoidance of a previously stented pulmonary artery. The main factors that were used in each patient for pathway selection are shown in Table 2. The pathway selection was also based on individual anatomy and haemodynamic function. The avoidance of pulmonary venous obstruction was the most important factor for pathway selection. However, placing the conduit on the ipsilateral side of the SVC was a relatively weak factor for pathway selection. If the ipsilateral pulmonary artery was very small, we preferred to connect the EC-TCPC graft to the contralateral pulmonary artery.

2.3. Postoperative anticoagulation

Acetylsalicylic acid (2 mg kg⁻¹ per day) and warfarin sodium (target prothrombin time: 1.5–2.0 international normalised ratio) were provided. When the prothrombin time was 2.5 or more, intravenous heparin was administered. The anticoagulation was maintained for at least 6 months.

Table 1
Operative procedures.

<table>
<thead>
<tr>
<th>Size of conduit</th>
<th>16 mm</th>
<th>18 mm</th>
<th>20 mm</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>21</td>
<td>30</td>
<td>5</td>
</tr>
</tbody>
</table>

Table 2
Main factors to consider in pathway selection.

<table>
<thead>
<tr>
<th>Factor</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Avoiding PVO</td>
<td>9</td>
</tr>
<tr>
<td>Placing the conduit on the PA and ipsilateral side of the SVC</td>
<td>25</td>
</tr>
<tr>
<td>Avoiding severe adhesions</td>
<td>7</td>
</tr>
<tr>
<td>Avoiding a contralateral small pulmonary artery</td>
<td>3</td>
</tr>
<tr>
<td>Avoiding a stented pulmonary artery</td>
<td>2</td>
</tr>
</tbody>
</table>

PVO: pulmonary venous obstruction; PA: pulmonary artery; SVC: superior venacava.
normalised ratio (INR)) were routinely administered in the long term to all patients after the Fontan procedure.

2.4. Data analysis

Surgical, haemodynamic and laboratory data for all of the patients who underwent EC-TCPC were reviewed retrospectively from the database of the Fukuoka Children’s Hospital. The diagnosis, operative and additional procedures, age and weight at the time of operation, status of the patient, cause of late death and postoperative clinical parameters were analysed. Data were sampled at the latest catheterisation following the EC-TCPC procedure. Catheter analysis was routinely planned at 6 months postoperatively. Additional evaluation was planned at ~5-year intervals, if needed, as these were the regular diagnostic procedures performed in our hospital for such procedures. Early death was defined as death within 30 days of surgery, and late death was defined as death occurring beyond 30 days after surgery. All data are expressed as mean ± standard deviation (SD). Comparisons between the two groups were performed with an unpaired, two-tailed t-test for normally distributed variables. The χ² or Fisher’s exact test was used to analyse differences between groups for categorical variables. A P-value <0.05 was considered statistically significant.

3. Results

3.1. Patients and operation

The patient characteristics are listed in Table 3. The mean age at operation was 4.2 ± 3.2 years and the median conduit size was 18 mm (range: 16–20 mm). The most common diagnosis was single right ventricle and 31 patients with heterotaxia were included in group 1 (Table 3). In 30 patients, the conduit was placed between the IVC and the opposite side of the PA crossing the vertebra. In 25 patients, the conduit was positioned behind the ventricle between the IVC and on the same side as the PA. There was one patient with a Y-style conduit placed between the IVC and bilateral PA behind the ventricle. There were no patients in group 1 who required fenestration, whereas four patients in group 2 required fenestration. Three of these four patients had impaired ventricle function and was referred from another hospital.

3.2. Mortality

In group 1, there were no early deaths but three patients died in the intermediate term. The first patient with single right ventricle died of sudden haemoptysis 37 months after the operation. The second patient with unbalanced atrioventricular septal defect (AVSD) had sudden death at 12 months postoperatively. The third patient with a single right ventricle died of gastric bleeding 5 months after the operation. In group 2, there were no early deaths but there were eight late deaths. The Kaplan–Meier survival rates (Fig. 2) were 93.5% at 5 years and 10 years in group 1, and 97.3% at 5 years and 96.1% at 10 years in group 2 (log-rank test, P = 0.29).

3.3. Morbidity

In group 1, none of the patients developed conduit stenosis or pulmonary venous obstruction. No patient required re-operation. Three patients developed late complications during the follow-up period. Two patients had new-onset arrhythmia including paroxysmal ventricular contractions and atrial tachycardia that required medication soon after the operation. Another patient had a minor subdural haematoma 3 months after the surgery without any neurological complications.

In group 2, 27 patients developed late complications. Fourteen patients had new-onset arrhythmia including sick sinus syndrome in three, paroxysmal supraventricular tachycardia in seven, second-degree atrioventricular block in two and paroxysmal ventricular contractions in two patients. In addition, six patients developed protein-losing enteropathy and seven patients had bleeding or thromboembolic events. Freedom from adverse events, including deaths and complications are shown in Fig. 3.

3.4. Catheterisation study

A postoperative cardiac catheterisation study was performed at 2.3 ± 2.7 years in patients who survived the Fontan operation. The data are summarised in Table 4. There was no

![Fig. 2. Kaplan–Meier survival curve.](https://academic.oup.com/ejcts/article-abstract/38/4/439/408692/The-impact-of-extracardiac-conduit-total by guest on 05 October 2017)
significant pressure gradient between the IVC and the PA. A non-obstructive systemic venous pathway to the PA was created with each method.

Haemodynamics in group 1 during the intermediate term were identical with those in group 2 with respect to central venous pressure (8.9 ± 2.5 in group 1 and 9.6 ± 2.6 mmHg in group 2), left ventricular end-diastolic pressure (4.4 and 4.3 mmHg, respectively), cardiac index (3.3 and 3.4 l min⁻¹ m⁻², respectively) and arterial oxygen saturation (94% and 94.2%, respectively).

3.5. Comparison among the pathways

In 30 patients, the conduit was placed between the IVC and the contralateral side of the PA crossing the vertebra (group CV: group crossing the vertebra). In 25 patients, the conduit was positioned behind the ventricle between the IVC and the same side as the PA (group BV: group behind the ventricle). There was one patient with a Y-style conduit placed between the IVC and right and left PA behind the ventricle. Three patients died in the intermediate term as previously described. One patient died in group CV, and two patients died in group BV. Three patients developed late complications during the follow-up period, one in group CV and two in group BV.

The haemodynamics in groups CV and BV during the intermediate term were identical with respect to IVC pressure (9.1 ± 2.9 in group CV and 8.5 ± 1.7 mmHg in group BV), left ventricular end-diastolic pressure (4.2 ± 3.2 and 4.3 ± 3.5 mmHg, respectively), cardiac index (3.3 ± 0.8 and 3.4 ± 0.8 l min⁻¹ m⁻², respectively) and arterial oxygen saturation (94.2% and 94.3%, respectively). The data are summarised in Table 5.

Table 4
Cardiac catheterisation data.

<table>
<thead>
<tr>
<th></th>
<th>Group 1</th>
<th>Group 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number</td>
<td>47</td>
<td>271</td>
</tr>
<tr>
<td>CVP (mmHg)</td>
<td>8.9 ± 2.5</td>
<td>9.6 ± 2.6</td>
</tr>
<tr>
<td>SVEDP (mmHg)</td>
<td>4.4 ± 3.5</td>
<td>4.3 ± 2.9</td>
</tr>
<tr>
<td>CI (l min⁻¹ m⁻²)</td>
<td>3.3 ± 0.8</td>
<td>3.4 ± 0.8</td>
</tr>
<tr>
<td>SaO₂ (%)</td>
<td>94.1 ± 2.3</td>
<td>94.2 ± 2.2</td>
</tr>
</tbody>
</table>

CVP: central venous pressure; SVEDP: systemic ventricular end-diastolic pressure; CI: cardiac index; SaO₂: arterial oxygen saturation.

Table 5
Cardiac catheterisation data.

<table>
<thead>
<tr>
<th></th>
<th>Group CV</th>
<th>Group BV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number</td>
<td>27</td>
<td>20</td>
</tr>
<tr>
<td>CVP (mmHg)</td>
<td>9.1 ± 2.9</td>
<td>8.5 ± 1.7</td>
</tr>
<tr>
<td>SVEDP (mmHg)</td>
<td>4.2 ± 3.2</td>
<td>4.3 ± 3.5</td>
</tr>
<tr>
<td>CI (l min⁻¹ m⁻²)</td>
<td>3.3 ± 0.8</td>
<td>3.4 ± 0.8</td>
</tr>
<tr>
<td>SaO₂ (%)</td>
<td>94.2 ± 2.3</td>
<td>93.4 ± 2.2</td>
</tr>
</tbody>
</table>

CVP: central venous pressure; SVEDP: systemic ventricular end-diastolic pressure; CI: cardiac index; SaO₂: arterial oxygen saturation.

4. Discussion

The present study showed excellent clinical outcomes of the EC-TCPC operation in patients with apicocaval juxtaposition. All 56 patients underwent the Fontan operation without early mortality, and the pathway of EC behind the ventricle or crossing the vertebra was employed without conduit stenosis or pulmonary venous obstruction. No patient required re-operation. However, there were three late deaths and three patients developed minor late complications.

It is very difficult to construct the LT-TCPC in the Fontan procedure for apicocaval juxtaposition. Compared to the LT-TCPC, the EC-TCPC is very versatile and can be performed for all anatomical situations, including a malpositioned heart with apicocaval juxtaposition. When EC-TCPC is used for a malpositioned heart with apicocaval juxtaposition, the graft must bridge the ventricle from the IVC to the contralateral PA. The graft may be compressed by the vertebra, which is a possible cause of a stenotic venous pathway after EC-TCPC. Another possible pathway from the IVC to the pulmonary artery is behind the ventricle; the graft could run behind the ventricle from the IVC to the ipsilateral pulmonary artery. In this situation, the pulsating left ventricle may compress the graft, which is a possible cause of low cardiac output postoperatively. Surgeons should decide the side on which the extracardiac graft should run according to the morphology of each heart. Another option is the Fontan procedure with intra-atrial artificial grafting from the IVC to the contralateral PA [10]. This procedure requires a prolonged cardiac arrest time to anastomose the intra-atrial graft from the IVC to the contralateral PA and residual right-to-left shunt. Although the anastomotic site is often confirmed postoperatively, it may cause pulmonary venous obstruction inside the atrium because the large conduit may disturb the pulmonary venous flow at the orifice of the pulmonary vein.

Some reports recommended the pathway crossing the vertebra or intra-atrial graft [11]. Kawahira and co-workers [12] reported a series of five patients who underwent EC-TCPC using an artificial graft bridging the vertebra. They suggested that this modification might be advantageous for a univentricular heart associated with apicocaval juxtaposition to prevent a postoperative stenotic venous pathway. Lim and co-workers suggested use of the in situ retrograde coronary sinus and the left SVC conduit behind the ventricle as the conduit material [13]. In addition, Adachi and co-workers presented a pedicled pericardial conduit placed behind the ventricle to make a straight pathway between the IVC and PA [14]. In some cases, either the pathway crossing the vertebra or behind the ventricle can be selected, but in other cases, such as those with severe adhesions, the choice of pathway is limited.
In our series, there was no obvious superiority of the pathway behind the ventricle compared with the pathway crossing the vertebra by itself. There are many factors to consider in selecting the pathway. In some cases, such as with a stented PA, the anastomotic site should be contralateral and an alternate pathway should not be used. However, it is possible to place the conduit on the ipsilateral side of the SVC. Other factors should also be considered, such as the dimension of the apex and the need for pulmonary augmentation. It is most important to select a pathway that avoids pulmonary venous obstruction and conduit stenosis.

The EC Fontan operation also has potential disadvantages, such as thrombo-embolism, conduit stenosis and lack of growth potential [9]. The optimal anticoagulant prophylaxis after EC-TCPC is unknown [15]. There is little agreement regarding the efficacy of various forms of prophylactic anticoagulant therapy in reducing the morbidity and mortality from thrombo-embolic events. However, it has been well known that post-Fontan patients have abnormalities in both procoagulant and anticoagulant factors and the imbalance between these factors [16]. We previously presented excellent mid-term results of EC-TCPC with the combination of afterload reduction therapy and lifelong anticoagulant prophylaxis [17], and have consequently continued this policy after EC-TCPC operation [16]. Our policy for anticoagulant prophylaxis is lifelong treatment with warfarin in combination with aspirin. The results of the present study showed no thrombus formation in the conduit. In addition, the low central venous pressure (CVP) observed during late cardiac catheterisation in our patients may have been due to the low risk of clinically silent small pulmonary emboli with our anticoagulant strategy. In 2003, a study evaluating the prevalence of clinically silent pulmonary emboli in adults after Fontan operation was reported, and 17% of the patients had an intermediate or high probability for pulmonary embolism on ventilation perfusion [18]. However, three late deaths were associated with anticoagulant-related complications. The other concern for the conduit is the potential for longitudinal growth, and the diameter of the conduit should be selected based on the adult body size [19,20]. In children aged 2—4 years, it has been reported that the distance between the IVC and the undersurface of the right PA is ~60% of that in adults [20]. Therefore, it is possible that the conduit may cause distortion of the right PA, compression of the pulmonary vein or flow disturbance in the conduit as the patient grows, especially for a malpositioned heart with apicocaval position. Although we and other investigators have not observed such problems or longitudinal torsion of the conduit in mid-term follow-up thus far [19], further investigations are necessary for long-term outcome of the conduit and the circumference, especially for the apicocaval position because of the length of the conduit.

5. Limitation

While the patient data were collected retrospectively, a small, non-randomised patient population and an institutional bias for the EC-TCPC approach are limitations of this study.

6. Conclusions

In conclusion, EC-TCPC can be performed in children with apicocaval juxtaposition with excellent mid-term outcomes compared with other anatomical Fontan candidates. The pathway of the EC behind the ventricle or crossing the vertebra can be employed without conduit stenosis or pulmonary venous obstruction.

References


Appendix A. Conference Discussion

Dr T. Jones (Birmingham, UK): First of all, your group is to be commended on reporting a large Fontan series with a 96% 10-year survival. That’s obviously a great achievement in itself.

The first observation is that you’re reporting quite a high incidence of 16% of the need to do this type of procedure in the juxta-ventricular apical position. Now, that’s not something that we see in our own practice. And what is quite interesting is there are very few people who have published on this, and they all seem to come from Japan. I know there is a higher incidence of heterotaxy in Japan, but I wonder if this is something where you see a higher incidence of this alignment needing this type of corrective surgery at the Fontan than we see in our own population.

Secondly, you mention the need to bring the conduit across the midline. We usually find that if we use a bypass strategy to completely decompress the heart, including cardiopulmonary arrest, we can work down low on the IVC, and take off the IVC with a large RA cuff as an open technique and mobilize the...
pulmonary veins right out of the periphery, we can create the space to go on the same side as the IVC.

By doing this, we don’t find the need to cross the midline. But there again, it might be that we have a different patient population.

You mentioned one of the reasons to cross the midline is to avoid a small pulmonary artery. Looking at the ‘ten commandments’ of the Fontan, and the more recently published ‘two commandments’ of a good Fontan, one is maintenance of good size PAs. So a small PA to us is something to go after to patch into. And again, that can sometimes create a good sump to put a Fontan in. So those are just a few comments.

But the questions I’ve got relate to, first of all, the Gore-Tex tube size. You’ve told us you’re using a 16 to 18 mm tube. I think that’s on the smaller side, and certainly I would be wanting to be using a 20 to 22 mm conduit. Is there a difference in your groups in this size Gore-Tex tube? And particularly if you’re going to need to cross the mediastinum over the spine, are you downsizing those tubes?

Dr Sakurai: The question is about the size of the Gore-Tex conduit. Actually, I didn’t show the results of the conduit size in comparison between an apicocaval juxtaposition and the extracardiac tissue, but actually, there are no differences between apicocaval juxtaposition and the EC-TCPC.

But I think that this is a study of about 15 years, and in this time we changed the indication of the Fontan pressure, I mean, especially for age. We used to perform the EC-TCPC around 3 or 4 years of age, but now much more often we do the EC-TCPC between 1 to 2 years of age.

Therefore the size of the conduit is getting smaller and smaller. So now we use mostly 16 or 18 in both anatomy, and we never use a conduit size less than 16. And recently a paper showed that 16 mm of the Gore-Tex size is enough to grow up.

Dr Jones: My other comment is regarding the beautiful angiograms you’ve shown, and in the paper, you demonstrate there is no significant hold-up in the pathways. But we know from the work by Bove and Marc de Leval, the computational flow dynamic studies that have been done in Fontan circuits, that the best way to avoid loss of energy in the circuit is to have as short and straight a tube as possible not meeting any competitive flow.

You’re, of course, going away from those principles, and you’re putting in a long curved tube, whereas the angiogram and the pressure data at rest under anaesthetic seems to be comparable between the groups. I wonder if when you start exercising these patients who have got the longer curved tube, you might find their functional ability is less. Do you have any information on that?

Dr Sakurai: Well, actually, we have very much curve, but if we use a longer bending tube, it maybe happen.

But following this result, I think we can do either way, and I don’t think that that’s a very big problem if we choose a longer bent pathway or short compressed pathway so far.