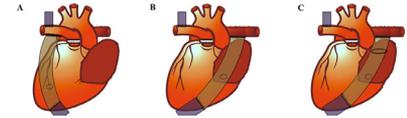


Conduit Route Selection for Total Cavopulmonary Connection in Patients With Apicocaval Juxtaposition



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Apicocaval juxtaposition (ACJ) may complicate the selection of conduit route in patients with single ventricles when total cavopulmonary connection (TCPC) is performed. We reviewed our experience of pathway selection and evaluated the clinical results. Of 128 patients who underwent TCPC at our hospital between January 2009 and April 2016, 31 with ACJ were included in this study. In 24 patients, the conduit was placed between the inferior vena cava (IVC) and the ipsilateral pulmonary artery. To avoid compression of the conduit and pulmonary veins in 5 patients, the conduit was placed between the IVC and the contralateral pulmonary artery. In 2 patients, the tube graft was anastomosed with the IVC orifice within the atrium, then guided through the atrial free wall and anastomosed with the contralateral pulmonary artery outside the heart (intra/extracardiac Fontan). Patient demographics were compared with those of patients without ACJ. The mean age and body weight at surgery were 58.5 ± 32.4 months and 16.2 ± 6.0 kg, respectively. The mean postoperative pulmonary artery pressure was 15 ± 3 mm Hg. The postoperative data did not differ significantly from that of patients without ACJ who underwent extracardiac TCPC. One patient died of overwhelming infection. The mean follow-up was 17.5 ± 15.4 months (range, 1–65 months). There were no conduit-related early or late complications. TCPC in patients with ACJ can be performed with excellent early and midterm results. The route between the IVC and the ipsilateral pulmonary artery is our preference.



Modified Fontan procedure for ACJ.

Central Message

Total cavopulmonary connection in patients with ACJ has excellent results. The route from the inferior vena cava to the ipsilateral pulmonary artery is our preference.

Perspective Statement

ACJ may complicate the conduit route selection for patients with single ventricles when TCPC is performed. From our experience of 31 patients with ACJ, we conclude that TCPC can be performed with excellent early and midterm results. The route between the inferior vena cava and the ipsilateral pulmonary artery is our preference. Sometimes, contralateral pulmonary artery was chose to avoid compression.

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INTRODUCTION

Modifications of the Fontan procedure are also applied to a wide variety of complex congenital heart defects. Extracardiac conduit total cavopulmonary connection (TCPC) has been widely performed and has become the preferred operation. It avoids aortic cross-clamping and atriotomy, allowing for minimal atrial surgery and cardiac ischemic time.

Furthermore, near-optimal laminar flow in the systemic venous pathway precludes atrial stretch or arrhythmia in the long term.^{1–4}

One challenging problem is the pathway of the extracardiac conduit for patients with apicocaval juxtaposition (ACJ). ACJ is a morphologic feature of the cardiac apex pointing toward the ipsilateral side of the IVC. When the heart is malpositioned

Abbreviations: ACJ, apicocaval juxtaposition; CI, confidence interval; IVC, inferior vena cava; LCOS, low cardiac output syndrome; MRI, magnetic resonance imaging; TCPC, total cavopulmonary connection

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with ACJ, several issues should be considered in choosing the best pathway for the extracardiac conduit from the inferior vena cava (IVC) to the pulmonary artery. If the conduit is placed between the IVC and the pulmonary artery on the contralateral side, the long curved conduit crosses the spinal column, which may cause a kink or serious energy loss. In contrast, if the conduit is positioned directly between the IVC and the ipsilateral pulmonary artery, it is confined to the narrow space behind the ventricle. Thus, obstruction of the conduit by compression from the ventricle and pressure exerted by the conduit itself on the pulmonary veins could both be matters for potential concern.

However, available data regarding this pathway selection are limited and few studies have addressed this issue specifically.^{5–7} The present study was undertaken to evaluate the impact of ACJ on the early and midterm results of TCPC and to examine our experience of route selection in patients with ACJ.

MATERIALS AND METHODS

The study protocol was reviewed and approved by the Institutional Review Board at Guangzhou Women and Children's Medical Center; individual consent for the study was waived owing to its retrospective medical record review design. The databases of the Department of Cardiac Surgery were searched for patients with a functional single ventricle and ACJ who had undergone a modified Fontan procedure at our center between January 2011 and March 2016; 31 consecutive patients met the criteria. Patients with mesocardia were excluded in this study. Medical records, operative notes, and all available cardiac magnetic resonance imaging (MRI) reports, echocardiography reports, and cardiac catheterization reports were reviewed. Cardiac MRI was performed to identify compression from the heart or vertebrae and any kinking of a curved conduit due to excessive curvature. Echocardiography was performed to access the pressure gradient between IVC and the conduit. All survivors were closely followed-up to provide sufficient information. These results were compared with the corresponding data from 97 patients without ACJ who also underwent a TCPC procedure during the same period. Prolonged pleural effusions were defined as effusions requiring drainage for more than 2 weeks.

Surgical Technique

After median sternotomy, hypothermic cardiopulmonary bypass was used in all patients. The ascending aorta was cannulated, and up to 4 venous cannulas were inserted in the superior vena cava, IVC, and hepatic veins when these did not drain into the IVC. Aortic cross clamping and cardioplegic arrest were performed in 8 patients. In patients without a prior bidirectional Glenn shunt, a unilateral or bilateral superior cavopulmonary anastomosis was constructed; the pulmonary artery was divided, any systemic-to-pulmonary arterial shunts were dismantled, and a pulmonary arterioplasty was performed when necessary. The pathway of the extracardiac conduit was chosen to obtain unobstructed flow of IVC and hepatic vein blood to the pulmonary artery without compromising pulmonary venous blood flow. The conduit was anastomosed to the undersurface of the pulmonary artery with a bevel extending toward one of the pulmonary arteries, avoiding impingement of superior vena cava flow on IVC flow. Generally, after extensive dissection of aorta, pulmonary artery, atrium and ventricle, we first accessed the route between the IVC and the ipsilateral pulmonary artery behind the ventricle. If the space was narrow, and the ventricle compressed the conduit or the conduit compressed the pulmonary veins, we changed it to the route between the IVC and the contralateral pulmonary artery. If the vertebra compressed the conduit or a long curved pathway caused a kink, it was changed to intra/extracardiac route finally. In 24 patients (77.4%), the GoreTex tube graft was anastomosed to the IVC and the ipsilateral pulmonary artery (Fig. 1A). In 5 patients, the ventricular and venous anatomy did not allow for a pathway between the IVC and the ipsilateral pulmonary artery, and a total extracardiac Fontan was constructed using an 18–22 mm GoreTex tube graft anastomosed to the IVC and the contralateral pulmonary artery (Fig. 1B). In another 2 patients, the tube graft was anastomosed with the IVC orifice within the atrium, then guided through the atrial free wall and anastomosed with the contralateral pulmonary artery outside the heart (intra/extracardiac Fontan, Fig. 1C). In patients with partial or total anomalous hepatic venous drainage, where the hepatic veins entered the atrium separately, the separate entrance sites of the IVC and hepatic veins were incorporated and anastomosed with the conduit. A 3–5 mm fenestration was placed routinely.

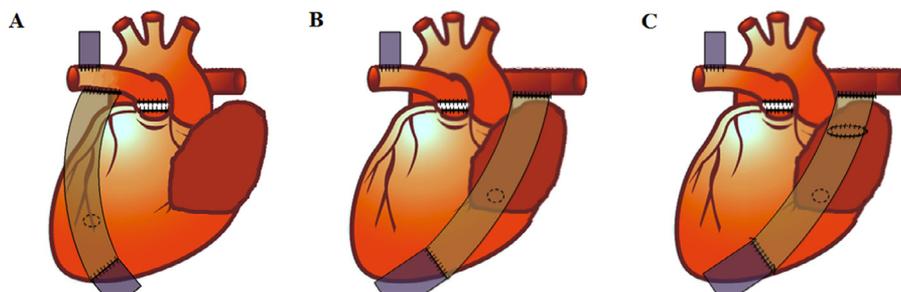


Figure 1. Modified Fontan procedure for ACJ. (A) The conduit is positioned straight between the IVC and the ipsilateral pulmonary artery. (B) The conduit is placed between the IVC and the contralateral pulmonary artery. (C) Intra/extracardiac Fontan.

CONGENITAL – CONDUIT ROUTE SELECTION FOR TOTAL CAVOPULMONARY CONNECTION

Together with the TCPC operation, a surgical pulmonary arterioplasty was performed in 2 patients. In another 4 patients, a significantly regurgitant atrioventricular valve was repaired at the time of the TCPC. Mean cardiopulmonary bypass time was 121 ± 37 minutes, and mean aortic cross-clamp time was 53 ± 24 minutes (8 patients).

Anticoagulation Therapy

Anticoagulation therapy with oral aspirin (5 mg/kg daily) was initiated 1 day after surgery and maintained for 6 months. However, for patients with favism, oral dipyridamole (5 mg/kg daily) was used instead of aspirin.

Statistical Analysis

Data are presented as mean \pm standard deviation or median and range, as appropriate. Comparisons between the two groups were performed using an unpaired two-tailed *t* test for normally distributed variables. Estimated survival was determined by the Kaplan-Meier method, based on the product-limit estimator, and 95% confidence intervals (CI) were constructed around the curves according to Greenwood's formula. All reported *P* values are two-tailed. Differences were considered statistically significant if the *P* value was <0.05 .

RESULTS

The characteristics of the patients with ACJ are listed in Table 1. Twenty-five patients (80.6%) were male and 6 (19.4%) were female. The mean age was 58.5 ± 32.4 months (range, 16–172 months), and the mean body weight was 16.2 ± 6.0 kg (range, 9.1–40.3 kg). Total anomalous pulmonary venous connection was found to coexist in 7 patients. Seven patients had right atrial isomerism. In 9 patients, there was a complete atrioventricular canal, and in most cases, there was an unbalanced

opening into the morphologic right ventricle. Echocardiography identified more than moderate atrioventricular valve regurgitation in 4 patients, tricuspid atresia in 3 patients, and mitral atresia in 2 patients. Anomalous hepatic venous drainage was present in 4 patients; in these patients, the hepatic veins entered the atrium separately. Ten patients had bilateral superior vena cava, usually without a connecting innominate vein.

Mortality and Morbidity

One patient died of overwhelming infection 7 days after operation. The mortality was 3.2%. The mean follow-up duration was 17.5 ± 15.4 months (range, 1–65 months). There were no late deaths. The Kaplan–Meier survival rate was 96.8% at 5 years in patients with ACJ and 92.3% at 5 years in patients without (log-rank test, *P* = 0.431; Fig. 2). None of the patients developed conduit stenosis or pulmonary venous obstruction requiring reoperation. MRI was performed 6 months to 1 year after Fontan procedure in 13 patients (8.7 ± 2.6 months, range, 6–12 months). It showed no conduit obstruction or compression by the ventricle, nor any undue pressure on the pulmonary veins (Fig. 3). One patient developed new-onset supraventricular tachycardia and required medication. During the follow-up period, no patient exhibited bleeding or thromboembolic events. One developed protein-losing enteropathy, while one was readmitted for severe valve insufficiency and cardiac functional failure.

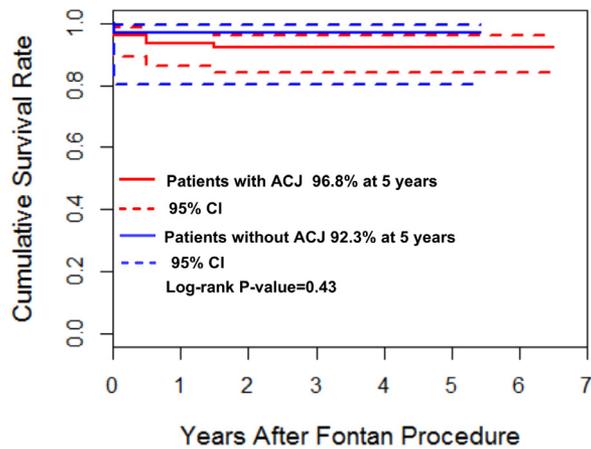
Comparison Among the Different Routes

In 24 patients, the conduit was placed behind the ventricle, between the IVC and the ipsilateral pulmonary artery (group A). In 5 patients, the conduit was placed between the IVC and the contralateral pulmonary artery (group B). There were 2 patients with an intra/extracardiac conduit placed

Table 1. Patient Characteristics, and Preoperative, Intraoperative, and Postoperative Data

Variables	Patients With ACJ (<i>n</i> = 31)	Patients Without ACJ (<i>n</i> = 97)	<i>P</i> Value
Age, mo	58.5 ± 32.4	56.0 ± 33.6	0.460
Weight, kg	16.2 ± 6.0	16.0 ± 7.0	0.386
Preoperative mPAP, mm Hg	14.5 ± 2.3	14.0 ± 1.9	0.271
CPB time, min	121 ± 37	127 ± 43	0.311
Mechanical ventilation time, min	8.1 ± 2.7	7.8 ± 4.3	0.298
ICU stay, d	4 ± 1.6	5 ± 2.1	0.408
Conduit size, mm	19.4 ± 1.28	19.1 ± 1.6	0.421
Follow-up, mo	27.5 ± 15.4	34.5 ± 23.3	<0.001
Pressure gradient (IVC to conduit), mm Hg	1.27 ± 0.5	1.32 ± 0.7	0.394
Readmission	2	9	
Adverse event			
LCOS	1	5	
Arrhythmia	3	8	
Death	1	7	
Pleural effusion (>14 d)	8	17	

ACJ, apicocaval juxtaposition; CPB, cardiopulmonary bypass; IVC, inferior vena cava; LCOS, low cardiac output syndrome; mPAP, mean pulmonary artery pressure.



NO. at risk

With ACJ	31	30	30	30	30	30	
Without ACJ	97	91	90	90	90	90	90

Figure 2. Kaplan-Meier survival curve. The number in parentheses indicates the patients at risk, and the dashed lines enclose the 95% confidence limits.

between the IVC and the contralateral pulmonary artery (group C). There was no significant difference in the preoperative, intraoperative, and postoperative data between groups A and B (Table 2).

DISCUSSION

In a malpositioned heart with ACJ, the completion of TCPC may be compromised by difficulty in selecting an appropriate conduit route. Our experience shows excellent clinical outcomes in patients with a functional single ventricle and ACJ. All 31 patients underwent TCPC without heart-related mortality, and the alternative routes for the conduit—behind the ventricle, crossing the spine, or intra/extracardiac—were all employed successfully, with no patient developing conduit stenosis or pulmonary venous obstruction requiring reoperation.

When the graft runs behind the ventricle from the IVC to the ipsilateral pulmonary artery, the ventricle may compress the graft, possibly causing obstruction of the conduit, and the conduit may compress the pulmonary veins, leading to pulmonary venous obstruction, especially in patients with total anomalous pulmonary venous connection. Sakurai et al reported 56 patients with ACJ who underwent TCPC.⁵ In 30 patients, the conduit was placed between the IVC and the contralateral pulmonary artery, crossing the vertebra. In the other 25 patients, the conduit was placed behind the ventricle, between the IVC and the ipsilateral pulmonary artery. No patient developed conduit obstruction or pulmonary venous obstruction, and none required reoperation. The incidence of death or late complications did not differ between the conduit routes. Even with the risk of compression from the ventricle, based on individual cardiac anatomy, the conduit route behind the ventricle can be used without conduit obstruction or pulmonary venous

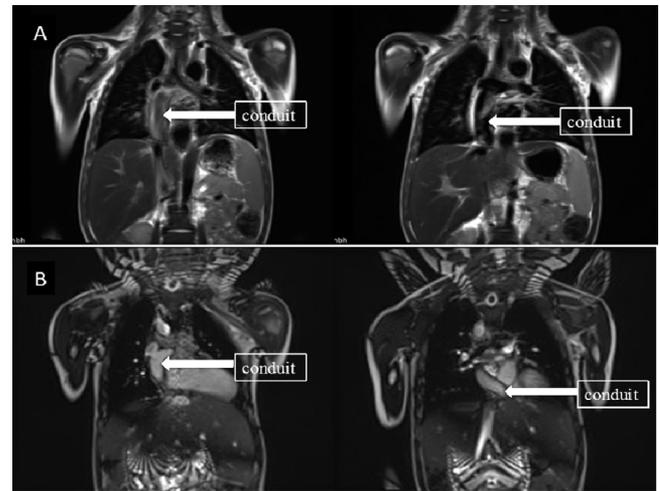


Figure 3. MRI image of the extracardiac conduit. (A) The conduit runs behind the ventricle but is not compressed. (B) The conduit crosses the spinal column but is not compressed by the spine and shows no kinking.

obstruction. Furthermore, Yoshida et al reported that the conduit length between the IVC and the ipsilateral pulmonary artery was significantly shorter,⁷ while computational fluid dynamics studies revealed that a straighter and shorter conduit between the IVC and the ipsilateral pulmonary artery provided better laminar blood flow with less energy loss. In this study, we chose the route between the IVC and the ipsilateral pulmonary artery in most of our patients (77.4%, 24/31). Postoperative MRI showed no conduit obstruction or pulmonary venous obstruction. The route behind the ventricle appears to be a valid surgical option for patients with ACJ.

When the conduit bridges the vertebra from the IVC to the contralateral pulmonary artery, the vertebra may compress the conduit, potentially causing obstruction. Morizumi et al reported 10 patients with ACJ who underwent TCPC.⁶ In most of the patients (80%, 8/10), the conduit was positioned between the IVC and the contralateral pulmonary artery, taking a slightly roundabout route. In 2 patients, the extracardiac conduit was positioned behind the ventricle, between the IVC and the ipsilateral pulmonary artery, to create the shortest route. There was no conduit-related reoperation. However, in 1 patient with a conduit placed between the IVC and the contralateral pulmonary artery, a postoperative cineangiogram and computed tomography showed a curved pathway and acute conduit occlusion developed. In another patient who had a large and poorly functioning single ventricle, a conduit placed between the IVC and the contralateral pulmonary artery developed acute thrombotic occlusion; the patient died of bleeding and multiple organ dysfunction despite surgical thrombectomy and thrombolysis. The authors advocated that the route between the IVC and the contralateral pulmonary artery should be the primary choice when the relevant pulmonary artery was in good condition, as the conduit placement could

Table 2. Comparison Among the Routes

Variables	Group A (n = 24)	Group B (n = 5)	Group C (n = 2)	P Value (A and B)
Age, mo	61.8 ± 36.0	44.7 ± 9.6	52.2	0.11
Body weight, kg	16.9 ± 6.6	13.4 ± 1.6	14.6	0.165
Preoperative mPAP, mm Hg	14.7 ± 1.4	14.2 ± 3.2	14.4	0.521
CPB time, min	114.7 ± 34.6	152.6 ± 37.2	123	0.868
Diameter of conduit, mm	19.4 ± 1.2	20 ± 1.4	18	0.457
Postoperative mPAP, mm Hg	17.2 ± 2.7	18.8 ± 1.9	19	0.309
ICU stay, d	4.2 ± 1.7	3.8 ± 2.2	4.4	0.542
Follow up, mo	30.9 ± 15.0	21.1 ± 9.0	4	0.145
Pressure gradient (IVC to conduit), mm Hg	1.04 ± 0.3	1.39 ± 0.2	1.22	0.431
Complication				
Arrhythmia	2	0	1	
Pleural effusion (>14 d)	7	1	0	
Death	1	0	0	

ICU, intensive care unit. Other abbreviations as in Table 1.

be easily secured without flipping over the ventricle or performing extensive dissection. Based on individual anatomy and hemodynamic function, to avoid compression from the ventricle and pulmonary venous obstruction, we chose the route between the IVC and the contralateral pulmonary artery in only 5 patients (16.1%, 5/31). The longer route may result in compression by the vertebra and is likely to be a curved pathway, which is a risk for thrombotic occlusion.

Although the ability to perform a satisfactory cephalad anastomosis and provide better fluid dynamics is very important in choosing the conduit route for Fontan circulation, some issues should not be ignored. When the ventricle is large and poorly functioning, the available space is limited, or severe adhesions cause extensive dissection, the intra/extracardiac conduit may be more suitable. This requires cardiac arrest to anastomose the intra-atrial graft from the IVC to the contralateral or ipsilateral pulmonary artery. The conduit inside the atrium may disturb the pulmonary venous flow and cause pulmonary venous obstruction. Menon et al reported that the net energy loss across the anastomosis was significantly higher with lateral tunnel TCPC in comparison with extracardiac TCPC, because of flow impingement.⁸ Compared with the intra/extracardiac conduit, a relatively longer and curved route between the IVC and the contralateral pulmonary artery has been demonstrated to be a suitable connection option, regardless of the anatomical situation. Therefore, the intra/extracardiac route between the IVC and the contralateral pulmonary artery should not take precedence over the other two routes. Additionally, it is difficult to construct the lateral tunnel TCPC with a graft from the IVC to the ipsilateral pulmonary artery. In our series, only 2 patients received an intra/extracardiac conduit between the IVC and contralateral pulmonary artery because the available space was narrow. For the lateral tunnel conduit, Menon et al placed it between the IVC and the ipsilateral pulmonary artery.⁸ It is

more convenient to anastomose to the contralateral rather than to the ipsilateral pulmonary artery.

Thromboembolism is one of the major postoperative complications after the Fontan operation, with a prevalence that varies from 0% to 28%.^{9–12} The pathophysiology of thromboembolic events in Fontan patients may be explained in the context of Virchow's triad: that is, abnormal blood flow, blood constituents, and vessel wall.¹³ A curved and compressed conduit, early postoperative hypoxia, and low cardiac output in conjunction with the unique features of the Fontan circulation immediately after the operation, could also predispose patients to thromboembolic events.^{12,14,15} There is little agreement regarding the efficacy of various forms of prophylactic anticoagulant therapy for thromboembolism after a Fontan operation. Our policy for anticoagulant prophylaxis is treatment with aspirin or dipyridamole for 6 months. The results of our study showed no thrombus formation in the conduit.^{16,17}

LIMITATIONS

The major limitations of the present study are its retrospective nature and the relatively small number of patients included. The patient cohort was limited to one center, which might have resulted in selection bias as a result of clinical management practices. Computational fluid dynamics were not available for a quantitative investigation of the effects of different Fontan conduit routes.

CONCLUSIONS

In patients with a functional single ventricle associated with ACJ, TCPC can be performed with excellent results. Based on individual anatomy and hemodynamic function, a conduit positioned in any of the alternative routes can provide satisfactory results, as long as it is neither compressed nor kinked. The ideal strategy for minimizing energy loss is to create a straight route between the IVC and the ipsilateral pulmonary artery using an extracardiac conduit. Therefore, the route

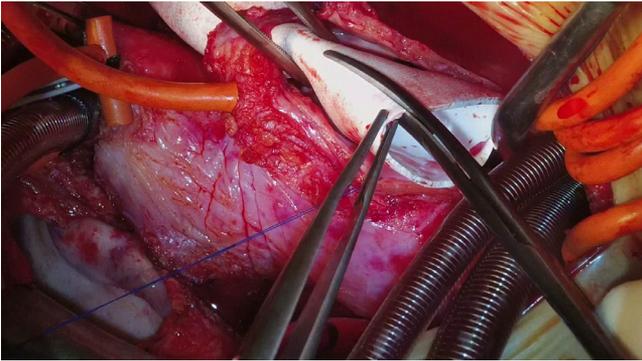
between the IVC and the ipsilateral pulmonary artery is our preference. Sometimes, contralateral pulmonary artery was also chose to avoid compression. Close follow-up and further investigations are required.

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SUPPLEMENTARY MATERIAL

The following is the supplementary data to this article:



Video 1. Intra/extracardiac Fontan.

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