Intraatrial Conduit Fontan Procedure: Indications, Operative Techniques, and Clinical Outcomes

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Background. The intraatrial conduit (IAC) Fontan procedure is one of the Fontan modifications and is usually not considered the first choice. In this large series, we report our experience of the IAC Fontan procedure for the treatment of a functional single ventricle and review its indications, techniques, and clinical outcomes.

Methods. Between 2009 and 2013, 101 patients with a functional single ventricle underwent an IAC Fontan procedure. The median surgical age was 44 months and weight was 15 kg. The cardiac malformations included tricuspid atresia in 10, double-inlet left ventricle in 8, double-inlet right ventricle in 15, double-outlet right ventricle in 18, congenitally corrected transposition of the great arteries in 11, complete atrioventricular septal defect in 8, criss-cross in 1, pulmonary atresia with intact ventricular septum in 3, Ebstein anomaly in 3, and others in 3. Heterotaxy syndrome was found in 21 patients. An intraatrial polytetrafluoroethylene conduit was implanted to construct the Fontan pathway. In 75 patients, a 2.7-mm to 4.5-mm fenestration was made to reduce the intraconduit pressure. Eighty-three patients had previously undergone a Glenn operation. Median follow-up was 29 months (range, 2 to 60 months). Doppler echocardiography, electrocardiography, and Holter monitoring were used to evaluate hemodynamic performance and arrhythmias.

Results. There were 2 hospital deaths, 1 Fontan take-down, and 3 midterm deaths. Conduit thrombosis developed in 1 patient. Atrial flutter developed in 2 patients, who underwent electrical cardioversion. Junctional bradycardia developed in 2 patients, but they did not require permanent pacemakers. Overall survival was 97.0% at 1 year and 94.1% at 5 years.

Conclusions. The IAC modification provides excellent operative and midterm outcomes in most patients with a functional single ventricle. However, a longer follow-up time is required to demonstrate its real advantages.
2013. All operations were performed by 2 surgeons (Z.Q.Z. and H.B.Z.). There were 60 male and 51 female patients, with a median age of 44 months (range, 29 to 177; mean 59.3 ± 37.5 months) and a median weight of 15 kg (range, 10 to 60; mean 17.8 ± 8.5 kg).

The underlying diagnoses and associated anomalies are reported in Table 1. Sinus rhythm was normal in 87 patients (86%). One patient with complete heart block had a permanent pacemaker. The patients’ surgical histories are summarized in Table 2. All patients were evaluated with preoperative echocardiography and computed tomography or magnetic resonance imaging. Catheterizations were performed in patients suspected to have high pulmonary vascular resistance or aortopulmonary collateral vessels. Five patients (all with heterotaxy syndrome and a common atrioventricular valve) had moderate (n = 3) or severe (n = 2) regurgitation of the atrioventricular valve. Two patients had subvalvular systemic outflow obstruction. Coil embolization was successfully performed to obliterate the aortopulmonary collateral vessels in 5 patients.

Eighteen patients (17.8%) with relatively old age met the “ten commandments” criteria of the Fontan [9] and underwent single-stage Fontan procedures, 74 (72.3%) underwent two-stage Fontan procedures, and the remaining 9 patients (8.9%) underwent three-stage Fontan procedures. One patient had a single-lung Fontan procedure. Patients were monitored regularly in our outpatient clinic at 3 months, 6 months, and yearly thereafter with the necessary electrocardiographic or echocardiographic studies.

Table 1. Diagnosis and Associated Anomalies

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Patients (No.)</th>
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<tbody>
<tr>
<td>Primary diagnosis</td>
<td></td>
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<tr>
<td>Tricuspid atresia</td>
<td>10</td>
</tr>
<tr>
<td>Double-inlet left ventricle</td>
<td>8</td>
</tr>
<tr>
<td>Double-inlet right ventricle</td>
<td>15</td>
</tr>
<tr>
<td>Double-outlet right ventricle</td>
<td>18</td>
</tr>
<tr>
<td>Heterotaxia</td>
<td>21</td>
</tr>
<tr>
<td>Congenitally corrected TGA</td>
<td>11</td>
</tr>
<tr>
<td>CAVSD</td>
<td>8</td>
</tr>
<tr>
<td>Criss-cross</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary atresia with IVS</td>
<td>3</td>
</tr>
<tr>
<td>Ebstein anomaly</td>
<td>3</td>
</tr>
<tr>
<td>Other</td>
<td>3</td>
</tr>
<tr>
<td>Associated anomaly</td>
<td></td>
</tr>
<tr>
<td>Coarctation</td>
<td>3</td>
</tr>
<tr>
<td>TAPVC</td>
<td>5</td>
</tr>
<tr>
<td>AVVR</td>
<td>11</td>
</tr>
<tr>
<td>Apicocaval juxtaposition</td>
<td>6</td>
</tr>
<tr>
<td>Separate hepatic vein</td>
<td>8</td>
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</tbody>
</table>

AVVR = atrioventricular valve regurgitation; CAVSD = complete atrioventricular septal defect; IVS = intact ventricular septum; TAPVC = total anomaly of pulmonary vein connection; TGA = transposition of the great arteries.

Operative Technique

All procedures were performed through a median sternotomy using cardiopulmonary bypass (CPB) and moderate hypothermia. Cannulation of the aorta was performed first, followed by cannulation of the superior vena cava (SVC) and inferior vena cava (IVC). The heart was arrested with antegrade cold blood cardioplegia. More recently, cold histidine-tryptophan-ketoglutarate cardioplegia was infused as a single dose (30 mL/kg) over 6 minutes.

Tailoring to the patients’ different age, weight, height, and IVC diameter, an expanded polytetrafluoroethylene (PTFE) conduit (Gore-Tex; W.L. Gore & Associates Inc, Flagstaff, AZ), sized at 16 mm in 3 patients, 18 mm in 13, 19 mm in 25, 20 mm in 52, and 22 mm in the remaining 8 patients, was interposed intraatrially between the IVC and the pulmonary artery. An anastomosis between the PTFE conduit and the inner orifice of the IVC was then completed. The graft was routed intraatrially and brought out from the orifice of the SVC or the roof of the right atrium in staged Fontan patients. The orifice of the SVC or the incision at the roof of the right atrium was enlarged in most patients to avoid “bottle neck” stenosis of the conduit.

A longitudinal incision was made at the underside of the right pulmonary artery, which was carried centrally in the presence of a proximal right or left pulmonary artery stenosis or laterally in the presence of a distal right pulmonary artery stenosis. The conduit was pulled straight and trimmed with an appropriate angle to create a large anastomosis with the inferior aspect of the right branch pulmonary artery. The posterior wall of the conduit and the posterior right atrium wall were Anastomosed end-to-side to the incision at the right pulmonary artery in a sandwich fashion. The anterior wall of the conduit was extended with an elliptical autologous or bovine pericardium to construct a flared junction (Fig 1). If a small conduit was used, a relatively larger bevel angle was trimmed to make up the flared opening, and if necessary, this pericardial flap also allowed concomitant enlargement of the proximal or distal pulmonary artery stenosis in moderate-risk or high-risk patients. Finally, the incision of the right atrium was closed with a running suture.

During the operation, a vent was placed in the atrium to decompress the ventricle and allow for measurement
of the collateral blood flow. The CPB flow rates were then increased to account for the measured collateral flow. A period of modified ultrafiltration was performed essentially on all patients. Associated procedures at the time of the Fontan operation included atrial septectomy in 15, branch pulmonary arterioplasty in 6, relief of subaortic obstruction in 3, total anomalous pulmonary vein connection repair in 1, atrioventricular valvuloplasty in 7, and replacement in 2.

After weaning from CPB, the hemodynamics were assessed by directly measuring pressures in the IVC, SVC, right pulmonary artery, and left atrium. Intraoperative transesophageal echocardiography was used to evaluate myocardial and valve functions and confirm laminar flow in the conduit. Fontan takedown was performed if the SVC pressures were elevated (>22 mm Hg) or if the hemodynamics were suboptimal after weaning from CPB. In these cases, the entire conduit was completely taken down and all incisions were closed under CPB and cross-clamp with cardiopulmonary arrest.

Low-dose dopamine (5 to 7.5 µg/kg/min) and milrinone (0.5 to 0.75 µg/kg/min) were used postoperatively. If patients had no complication, early extubation, within 4 to 6 hours, was performed whenever possible. Chest tubes were removed when the drainage was less than 2 mL/kg/d. Fraxiparine (0.01 mL/kg, once every 12 hours, hypodermic injection; GlaxoSmithKline, Research Triangle Park, NC) was routinely used for approximately 48 to 72 hours after the operation, and then Bamyl (LIF, Stockholm, Sweden) was used instead. If the patients were prone to thrombosis, warfarin was used. Bamyl (5 mg/kg/d) anticoagulation was arbitrarily maintained for 1 year in all patients who survived.

**Data Analysis**

Data were collected by retrospective review of the patients’ medical records. Statistical analyses were performed with SPSS 18.0 software (SPSS Inc, Chicago, IL). Results are expressed as mean ± standard deviation and median (range). Continuous variables before and after the Fontan operation or between two groups of patients were compared with the use of the paired or independent samples t test, respectively. Survival was analyzed by the Kaplan-Meier method. Multivariable analyses were performed using Cox proportional hazard models.

**Results**

Early deaths occurred in 2 patients (2%), and the IAC was taken down in another patient, for a total early failure rate of 3%. The causes of death included myocardial failure in a patient with subaortic stenosis for which a subaortic myectomy was performed and liver failure in a patient with separate hepatic venous drainage and atrioventricular valve regurgitation. The causes of IAC takedown included junctional bradycardia and low cardiac output at the conclusion of the procedure in 1 patient.

The median CPB time was 100 minutes (range, 47 to 210 min) and clamp time was 53 minutes (range, 27 to 99 min). Fenestrations were required in 78 patients (78%).

The central venous pressure and transpulmonary gradient after the operation were low, at 14 ± 4 and 8 ± 4 mm Hg, respectively. Mean arterial oxygen saturation on room air increased from 78% ± 6% before IAC to 90% ± 5% at the time of discharge (p = 0.001) and continued to improve at follow-up (Table 3). The median duration of inotropic support was 3 days (range, 1 to 15 days). Median mechanical ventilation was 20 hours (range, 6.5 to 95.5 hours), excluding 6 patients in whom ventilatory support was prolonged (100 hours). The median chest tube duration was 5 days (range, 3 to 28 days). Prolonged chest tube drainage (>14 days) occurred in 10 patients. The median intensive care unit and hospital lengths of stay were 5 days (range, 2 to 45 days) and 12 days (range, 7 to 55 days), respectively.

Major postoperative complications included low cardiac output and transient renal failure with peritoneal dialysis in 16 patients (16%), unilateral diaphragmatic paralysis with the plication procedure in 3 (3%), and

<table>
<thead>
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<th>Table 3. Perioperative and Follow-Up Hemodynamic Results</th>
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<tr>
<td><strong>Variable</strong></td>
</tr>
<tr>
<td>CVP, mean ± SD mm Hg</td>
</tr>
<tr>
<td>TPG, mean ± SD mm Hg</td>
</tr>
<tr>
<td>SpO2, mean ± SD %</td>
</tr>
<tr>
<td>Sinus rhythm, No.</td>
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CVP = central venous pressure; SD = standard deviation; SpO2 = oxygen saturation measured by pulse oximetry; TPG = transpulmonary gradient.
conduit thrombosis with conduit replacement in 1 (1%). Multivariable analyses showed that the risk of dialysis was greater with increasing CPB time (>120 minutes; \( p = 0.013 \)) and heterotaxy syndrome (\( p = 0.028 \)). The single-stage Fontan procedure was not a risk factor for dialysis (\( p = 0.117 \)). The other 2 patients also had a sinus rhythm by electrocardiogram or Holter monitoring before death (Table 3). During the first 48 postoperative hours, 3 patients had a transient junctional rhythm with no hemodynamic consequences. Two patients had junctional ectopic tachycardia that responded to antiarrhythmic drugs or hypothermia. Finally, 1 patient with congenital complete atrioventricular block had dual permanent pacemaker placement at the time of the operation.

Mean follow-up was 29 ± 26 months and complete in all patients. There were 3 deaths during midterm follow-up. The first patient, with Ebstein anomaly, who had undergone staged IAC, had progressive ventricular dysfunction and died 1 year after the operation. The second patient, with right single ventricle, complete atrioventricular defect, and separate hepatic venous drainage, died of liver failure 1.5 years after the operation after thrombosis developed in the hepatic vein. The last patient, also with right single ventricle, had a sudden death after sports in school 2.1 years after the operation. The overall survival rate for these patients was 97.0% at 1 year and 94.1% at 5 years (Fig 2).

Atrial flutter developed in 2 patients, who underwent electrical cardioversion at 40 months and 48 months. Junctional bradycardia developed in another 2 patients at 24 and 38 months but did not require permanent pacemakers. No patients have required reoperation for late complications or intervention for the IAC conduit thus far. Ninety-six percent of the patients are free of arrhythmias, and 99% of the patients have been free from any thromboembolic events. No pulmonary vein pathway obstruction was found in any patient by follow-up (Fig 3). Long-term complications, such as pulmonary arteriovenous malformation, protein-losing enteropathy, and plastics trachitis, were not reported in this series. According to the New York Heart Association Functional Classification, 6 patients were class II and the remaining 89 survivors (93.6%) were class I.

**Comment**

In the present series of IAC Fontan procedures, we have achieved a favorable survival rate of 97.0% at 1 year and 94.1% at 5 years. We have demonstrated excellent midterm functional status, with 93.6% of patients achieving New York Heart Association class I. The incidence of arrhythmias and thrombus formation was low. Our results are in agreement with other recent series of Fontan procedures, with reported operative mortalities of 0% to 13% and 5-year survival rates of 89% to 98% [10–14].

Despite some limitations, our experience suggests that the IAC Fontan procedure has some notable advantages over the LT and EC [5–8, 14]:

1. This technique is simple and reproducible. It avoids extensive IVC dissection, which is difficult in patients with a short IVC, separate hepatic vein, or severe adhesions from a staged Fontan. Creating fenestrations on the conduit is also very easy.

2. This technique offers a shorter and straighter pathway, resulting in a significantly more uniform blood streamline and less stagnant region. This reduces the Fontan circulation power loss and the risk of thromboembolic complications.

We have also made technical modifications to improve the safety and efficiency of the IAC Fontan procedure. First, the sandwich anastomosis between the conduit and the posterior pulmonary artery wall or pericardium (anterior wall), the atrial tissue, like a mattress, can help to decrease

![Fig 2. Kaplan-Meier survival curve for the entire cohort.](image1)

![Fig 3. The pulmonary venous pathway around the intraatrial conduit without obstruction.](image2)
anastomotic tension and the risk of bleeding. Bleeding at
the anastomosis between the prosthesis and autologous
tissue is common, especially in cyanotic patients with thin
and fragile pulmonary artery walls. Second, a flared junc-
tion design between the IAC and the pulmonary artery
creates more efficient hemodynamics [15].

In recent years, computational fluid dynamics tech-
niques have been applied to the design, refinement, and
assessment of the Fontan procedure by calculating flow
patterns and pressure changes within a virtual model of
the Fontan pathway. Our computational fluid dynamics
study also demonstrated [14] that a flared junction of the
IAC Fontan has a cycle-averaged power loss of 4.1%,
which is significantly lower than the 5.23% power loss
with the LT Fontan procedure. The IAC Fontan procedure
also generates even hepatic flow distribution at 33.6% and
66.4% in the left and right pulmonary arteries, respec-
tively, compared with the EC Fontan, which generates an
unequal blood flow at 84.5% and 15.5%, respectively.
Taken together, these results suggest that the IAC Fontan
may be an ideal total cavopulmonary connection with
superior hemodynamics.

The IAC Fontan procedure has traditionally
been considered an alternative to the Fontan procedure
[5-8, 14]. We have been using the IAC Fontan as the
procedure of choice in our institution since 2009. On the
basis of our excellent outcomes, we have extended the
indications to almost all patients with a functional single
ventricle and to those with apicocaval juxtaposition and
separate hepatic vein, which are challenging for the LT or
EC Fontan procedures [16-19]. Apicocaval juxtaposition is
a morphologic feature of the cardiac apex pointing toward
the ipsilateral side of the IVC. Dextrocardia is associated
with the right-sided IVC, and levocardia is associated
with the left-sided IVC. This feature makes it difficult to
construct the LT and EC routes, whereas the IAC Fontan
provides an easy choice.

Placing the conduit between the pulmonary artery and
the IVC at the ipsilateral side can create a straight pas-
sage. The conduit is connected to the intraatrial inner
orifice and hidden inside the atrium. Therefore, potential
stenosis of the conduit due to compression by the
ventricle, which can be of concern in selected cases where
EC may be used, is not of concern in IAC procedures. It
also avoids the long, curved crossing conduit used during
the EC Fontan where the conduit connects the IVC to the
contralateral pulmonary artery. Excellent early and
midterm results were achieved in the 6 patients with
apicocaval juxtaposition in this series.

For the patients with a separate hepatic vein, the conduit
was trimmed obliquely to cover the hepatic vein and IVC
orifices. Results were good in 6 of the 8 patients in this
series. The other 2 patients with the 2 veins widely sepa-
rated by the vertebral died of liver failure within 1.5 years
after the operation, possibly caused by hepatic blood flow
obstruction. In this condition, an intraatrial separation or
two separate pathways for the IVC and hepatic vein might
be an effective alternative approach [20].

On the basis of our experience, the following conditions
may be considered relative contraindications for the IAC
procedure:

1. The morphologically systemic atrial cavity is not large
enough to accommodate an adequate conduit. The conduit
selection in our center was based on the patient’s age, weight, and the diameter of IVC. Generally, a Gore-Tex conduit with a diameter of 18 to 20 mm was implanted in children aged between 2 and 4 years with a weight of 12 to 18 kg, and 22-mm conduits were implanted in the adolescents. The diameter of IVC is also an important reference standard. If the diameter of atrial cavity were less than the diameter of the IVC, as measured by imaging before the operation, this means the atrial cavity was not large enough to accommodate an expected size conduit. Five patients underwent an alternative Fontan in the same period because of relatively small atrial cavity.

2. If the patient’s pulmonary vein drainage is located at
the ipsilateral IVC, or very close with IVC, the IAC
may hinder pulmonary venous return, and the IAC
pathway should be abandoned. Meanwhile, oversized
conduit should be also avoided in patients with
normal pulmonary vein connections, which may
create “right of domain” issues and possibly pulmo-
nary venous obstruction. In our series, no pulmonary
vein obstruction was found in any patients during
follow-up (Fig 3).

3. The IVC and the hepatic vein are widely separated by
the column. The beveled conduit is unable to cover
the two orifices of the hepatic vein and IVC
completely.

4. In patients with azygous continuation of the IVC, the
IAC carrying only the hepatic venous flow may have a
higher risk of thrombosis [21]. In fact, the final deci-
sion to choose IAC is only made after direct inspec-
tion of the atrial morphology and the relationship
between the systemic and pulmonary vein.

When IAC is not a good choice for the patient, patient-
specific EC or IL Fontan procedures can be considered.
Furthermore, the IAC Fontan has some inherent disad-
vantages [5-8], which include (1) the lack of growth
potential of the conduit and the risk of thromboembolic
complications, (2) atrial incision and suture line related to
postoperative atrial arrhythmias, (3) the need for CPB and
aortic clamping; and (4) the theoretical need for
reoperation.

This study has some limitations. First, this is a retro-
spective analysis, not a randomized prospective study. In
particular, this study was not designed to compare the
outcomes of LT, EC, and IAC. Second, due to lack of
health insurance, some patients could not afford the
cardiac catheterization, and the hemodynamic evaluation
could not be made before and after Fontan procedure,
which could have affected postoperative morbidity and
mortality.
In conclusion, our experience suggests that construction of an IAC connection is a simple, safe, and reproducible technique. As an alternative Fontan modification, this technique yields a reliable Fontan circulation and is associated with excellent operative and midterm survival, with a low incidence of arrhythmias and thrombosis for most patients with a functional single ventricle. However, this procedure is not a “one fits all” technique. Some anatomic factors, including systemic atrial cavity volume, systemic vein, and pulmonary vein location need to be considered when constructing IAC pathway, and meanwhile, long-term follow-up results are required to further demonstrate its advantages and disadvantages.

Funding for this study was supported by Shanghai Hygiene Science Research (20114121), Innovation Program of Shanghai Municipal Education Commission (13YZ027), and the grant of Shanghai Science and Technology Commission (1441196490).

References


INVITED COMMENTARY

Rationales supporting one form of Fontan over another have been extensively articulated, supported in the main by single-center clinical reports. The technical issues in play are well known: atrial suture burden, myocardial preservation requirements, geometric maintenance of optimal fluid dynamics, ease of fenestration, and concern regarding the presence or absence of Fontan pathway growth capability. What is now a mature and expansive literature is decidedly mixed, and conclusive evidence establishing superiority of one technique over another is lacking. The report by Zhu and colleagues [1] present an opportunity to place the intraatrial conduit Fontan into the broader context of Fontan palliation.

There are certainly anatomic arrangements in which the use of an intraatrial conduit is technically advantageous. This would include anatomies with abnormalities of pulmonary or systemic venous drainage, or both, requiring complex baffle configurations, additional extracardiac connections (hepatic vein to inferior vena cava, for instance), or that place an extracardiac conduit at risk for compression by the cardiac mass. The intraatrial conduit Fontan has also been used with success in the setting of Fontan revision [2]. In addition to its geometric simplicity, ease and reliability of fenestration is an attractive attribute of the intraatrial conduit Fontan. Conversely, lack of conduit growth, right-of-domain issues causing pulmonary venous obstruction, atrioventricular inflow disturbance, and creation of areas of stasis within the systemic atrium are potential issues when an intraatrial conduit Fontan is entertained. The authors...