Fontan completion in patients with atrial isomerism and separate hepatic venous drainage

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Abstract

Objective: Fontan completion in patients with atrial isomerism, in which the inferior vena cava (IVC) and the hepatic vein (HV) drain separately, is technically challenging. Herein, we review our surgical approach to these patients.

Methods: The medical records of 50 consecutive patients with atrial isomerism who underwent Fontan completion between 1998 and 2008 were reviewed retrospectively.

Results: Separate HV drainage was present in 17 patients. Patients with interrupted IVC were excluded. Patient characteristics were as follows: median age, 26 months (range 15—149); median weight, 9.6 kg (range 8.1—47.2); right atrial isomerism, 16 patients; and left atrial isomerism, one. The IVC and the separate HV at the level of diaphragm were contralateral in 16 patients, and ipsilateral in one. The surgical procedures for directing blood flow from the IVC and the separate HV to the pulmonary arteries were as follows: en bloc resection of the IVC and the HV and anastomosing these veins to an extracardiac conduit in 10 patients; connecting the IVC to the HV in a side-to-side fashion before anastomosing them to an extracardiac conduit in one; and lateral tunnel in another. When the IVC and the HV were widely separated by the vertebrae, we chose an intra-extracardiac conduit (intra-atrial septation) in four patients and an extracardiac conduit for the IVC and the right HV and lateral tunnel for the separate left HV in one. There was no mortality. Five re-operations were performed (pacemaker in two patients; one each of fenestration, release of outflow obstruction and ligation of collateral arteries).

Conclusions: The mid-term results of the Fontan completion in patients with atrial isomerism and separate HV drainage were excellent. The distance between the IVC and the separate HV and the position of the vertebrae should be considered when choosing a surgical technique.

Keywords: Functional single ventricle; Separate hepatic venous drainage; Atrial isomerism; Fontan completion

1. Introduction

The modified Fontan procedure has been established as a definitive surgical treatment for patients with functional single ventricle (FSV), and with improvements in treatment strategies, surgery and management, the outcomes of Fontan completion for FSV patients, even those with atrial isomerism, have improved dramatically [1—4]. However, owing to complex anatomical anomalies in patients with atrial isomerism [5—7], the technical aspects of constructing the pathway for Fontan completion are complicated. Among these anomalies, Fontan completion in patients in whom the inferior vena cava (IVC) and the hepatic vein (HV) drain separately remains technically challenging. Moreover, not much information is available about such patients, because most relevant reports have focused only on successful cases [8—12], and to the best of our knowledge, there have been no reports focussing on patients with separate HV drainage. This study examines the technical variations and modifications associated with separate HV drainage and evaluates the early and mid-term outcomes for total cavopulmonary connection (including HV) in FSV patients with atrial isomerism who had separate HV drainage.

2. Patients and methods

2.1. Study design

We retrospectively reviewed the clinical data of 50 consecutive FSV patients with atrial isomerism who underwent Fontan completion between January 1998 and December 2008 at the Shizuoka Children’s Hospital. This study was...
approved by the Institutional Review Board of the hospital and the need for patient consent was waived. Among the patients reviewed, we identified 17 (34%) patients who had separate HV drainage. In this study, we defined 'separate HV drainage' as the existence of one or more HVs that drain into the atrium separately from the IVC. Therefore, patients with interrupted IVC withazygos or hemiazygous continuation were excluded. The medical, operative, angiographic and echocardiographic data for all the patients were reviewed. Information regarding their current clinical status was obtained from the medical records and the referring cardiologist was contacted. The mean follow-up period was 66.0 ± 34.8 months and was completed for all patients.

2.2. Patients population

There were 10 male and seven female patients. Sixteen patients had right atrial isomerism (RAI) and one had left atrial isomerism (LAI). The median age at Fontan completion was 26 months (range: 15—149), and the median body weight was 9.6 kg (range: 8.1—47.2).

Separate HV drainage was diagnosed by preoperative cardiac catheterisation and enhanced or multidetector-row computed tomography, and all cases were diagnosed preoperatively. The IVC and the separate HV at the level of diaphragm were contralateral in 16 (94%) patients and ipsilateral in one (6%). The separate HV was multiple in two (12%) patients. Other systemic and pulmonary venous anomalies included bilateral SVC in seven (41%) patients, and extracardiac total anomalous pulmonary venous connection (TAPVC) in 12 (71%) (supracardiac TAPVC, eight; infracardiac TAPVC, two; and mixed TAPVC, two).

Concomitant diagnoses included apicocaval juxtaposition (the cardiac apex pointing at the same side of the IVC) in 10 (59%) patients, dextrocardia in four (24%), right aortic arch in five (29%), pulmonary atresia in seven (41%) and pulmonary stenosis in four (24%).

All patients had undergone at least one operation before Fontan completion. The Fontan completion was preceded by an interim cavopulmonary anastomosis in 15 (88%) patients. Patient profiles are shown in Table 1.

2.3. Surgical technique and management

Fontan completion was performed through a median sternotomy, under cardiopulmonary bypass using a vacuum-assisted venous drainage system, with mild-to-moderate hypothermia. The arterial cannula was inserted in the ascending aorta in all but one patient (in whom the femoral artery was used), and the venous cannula was inserted in the SVC or the innominate vein. The decision about whether to cannulate the IVC and the separate HV was based on the need to maintain a reasonable surgical field and to prevent HV stenosis at the cannulation site. For patients weighing more than 15 kg (n = 3), the venous cannula was inserted into the femoral vein, and either the IVC or the HV was left uncannulated. For patients weighing less than 15 kg (n = 14), cannula placement was decided according to the position and size of the IVC and the separate HV: both the IVC and the HV were cannulated in four patients, only the IVC was cannulated in seven and neither in three (in these 10 patients without the HV cannulation, pump suction and intermittent simple clamp were used).

Aortic cross clamping and cardiopulmonary bypass were performed in all but one patient, in order to avoid clamping the atrium close to the atroventricular groove and to prevent injury or deformity of the atroventricular valve or coronary system. The IVC and the separate HV were fully dissected and mobilised before cross clamping. The atrium was opened and the ostium of the IVC and the separate HV were inspected after cross clamping. The type of Fontan completion was chosen such that an unobstructed pathway was obtained for both the IVC and the separate HV to the pulmonary artery without disturbing the pulmonary venous flow. The Fontan completion was performed by a single surgeon.

### Table 1

Patient profiles.

<table>
<thead>
<tr>
<th>Case</th>
<th>Primary diagnosis</th>
<th>Pulmonary obstruction</th>
<th>Extracardiac connection</th>
<th>SVC</th>
<th>IVC</th>
<th>Apex</th>
<th>Separate HV</th>
<th>IVC and HV widely separated?</th>
<th>Arch</th>
<th>Surgical procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>RAI</td>
<td>—</td>
<td>—</td>
<td>Rt.</td>
<td>Rt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>2</td>
<td>RAI</td>
<td>—</td>
<td>—</td>
<td>Rt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Rt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC</td>
</tr>
<tr>
<td>3</td>
<td>RAI</td>
<td>Atresia</td>
<td>Infra</td>
<td>Lt.</td>
<td>Rt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Y</td>
<td>Lt.</td>
<td>EC for IVC + HV, LT for Lt. HV</td>
</tr>
<tr>
<td>4</td>
<td>RAI</td>
<td>—</td>
<td>—</td>
<td>Rt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Rt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>5</td>
<td>RAI</td>
<td>Stenosis</td>
<td>Supra (1b)</td>
<td>Rt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>6</td>
<td>LAI</td>
<td>Atresia</td>
<td>—</td>
<td>Bilateral</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>7</td>
<td>RAI</td>
<td>—</td>
<td>Supra (1b)</td>
<td>Bilateral</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>(multiple)</td>
<td>Y</td>
<td>Lt.</td>
</tr>
<tr>
<td>8</td>
<td>RAI</td>
<td>Atresia</td>
<td>Supra (1a)</td>
<td>Bilateral</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>(multiple)</td>
<td>Y</td>
<td>Lt.</td>
</tr>
<tr>
<td>9</td>
<td>RAI</td>
<td>Stenosis</td>
<td>—</td>
<td>Bilateral</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>10</td>
<td>RAI</td>
<td>Atresia</td>
<td>Supra (1b)</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>11</td>
<td>RAI</td>
<td>Atresia</td>
<td>Supra (1b)</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>12</td>
<td>RAI</td>
<td>—</td>
<td>—</td>
<td>Rt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Y</td>
<td>Lt.</td>
<td>Intra-extra conduit</td>
</tr>
<tr>
<td>13</td>
<td>RAI</td>
<td>—</td>
<td>Infra</td>
<td>Bilateral</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (connecting IVC to HV)</td>
</tr>
<tr>
<td>14</td>
<td>RAI</td>
<td>—</td>
<td>Supra (1b)</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>15</td>
<td>RAI</td>
<td>Stenosis</td>
<td>Supra (1b)</td>
<td>Bilateral</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>16</td>
<td>RAI</td>
<td>—</td>
<td>Supra (1b)</td>
<td>Bilateral</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>N</td>
<td>Lt.</td>
<td>EC (en bloc resection)</td>
</tr>
<tr>
<td>17</td>
<td>RAI</td>
<td>Stenosis</td>
<td>Mixed</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Lt.</td>
<td>Y</td>
<td>Lt.</td>
<td>Intra-extra conduit</td>
</tr>
</tbody>
</table>

Abbreviations — EC: extracardiac conduit; HV: hepatic vein; IVC: inferior vena cava; LAI: left atrial isomerism; Lt.: left; LT: lateral tunnel; N: No; RAI: right atrial isomerism; Rt.: right; SVC: superior vena cava; TAPVC: total anomalous pulmonary venous connection; Y: yes.
The surgical procedures performed concomitant to Fontan completion included atrioventricular valve repair in three patients, repair of extracardiac TAPVC in one and atrial septectomy in one.

Fenestrations were placed in two patients, and in one of these, single-lung Fontan procedure was performed (because of obstruction of the unilateral pulmonary veins).

The mean cardiopulmonary bypass time was 174±55 min and the mean aortic cross-clamping time \( (n=16) \) was 70±21 min.

2.4. Data analysis

The Dr SPSS II statistical software for Windows (SPSS, Inc., Chicago, IL) was used for data analysis. Data are expressed as the median value and range or the mean ± standard deviation as appropriate. Data from groups were compared with the chi-square \( (\chi^2) \) test. Freedom from re-operation was estimated by the Kaplan–Meier method, and 95% confidence intervals (CIs) were constructed around the curves according to Greenwood’s formula.

3. Results

3.1. Surgical procedures

We performed five types of the surgical procedures for directing blood flow from the IVC and the separate HV to the pulmonary artery. When the IVC and the HV were ipsilateral \( (n=1) \) or contralateral, but not widely separated by the vertebrae \( (n=11) \), we chose en bloc resection of the IVC and the HV and anastomosing these veins to an extracardiac conduit (Fig. 1) in 10 patients and lateral tunnel in one. We took special care in one patient after the repair of infracardiac TAPVC (in whom the right lower pulmonary venous branch was occluded totally before cavopulmonary anastomosis) to avoid obstructing or deforming the previous common chamber and the ostium of the pulmonary veins located inferiorly. In this patient, we chose to connect the IVC to the HV in a side-to-side fashion before anastomosing them to an extracardiac conduit.

When the IVC and the HV were widely separated by the vertebrae \( (n=5, \text{ including two patients with multiple separate HVs}) \), we chose an intra-extracardiac conduit (intra-atrial septation; Fig. 2) in four patients. The degree of separation was assessed by both preoperative imaging and intra-operative impressions after the IVC and the HV were fully dissected and mobilised. We took special care in one patient after the repair of infracardiac TAPVC as well. In this patient, we chose an extracardiac conduit for the IVC and the right HV and lateral tunnel for the separate left HV (Fig. 3).

As a conduit, we used an expanded polytetrafluoroethylene tube conduit (Gore-Tex; W.L. Gore & Associates, Inc., Flagstaff, AZ, USA), without a reinforced ring. The size of the extracardiac conduit used was 16 mm in six patients, 18 mm in two and 20 mm in four, while the size of the intra-extracardiac conduit used was 16 mm in two patients and 20 mm in two.

We routinely administered 5 mg kg\(^{-1}\) ticlopidine as an anticoagulant. Warfarin was used for several months only in patients with an intra-extracardiac conduit.

Fig. 1. Preoperative (A) and postoperative (B) multidetector-row computed tomography and postoperative angiography (C) of case 16. The IVC was right-sided, and the separate HV and apex were left-sided. The IVC and the HV were not widely separated. We performed en bloc resection of the IVC and HV, and anastomosed these veins to an extracardiac conduit.

3.2. Survival and post-Fontan re-operations

There was no mortality after Fontan completion. One patient exhibited acute liver damage during the early
postoperative period, as revealed by serum levels of aspartate aminotransferase and alanine aminotransferase of 500 U l\(^{-1}\) or greater. This patient quickly recovered and was discharged with normal levels of liver enzymes.

Early postoperative tachyarrhythmias (e.g., atrioventricular reciprocating tachycardia, junctional ectopic tachycardia or paroxysmal atrial tachycardia) were documented in nine patients with RAI. Five of these nine patients also had a tachyarrhythmic event prior to these surgical procedures (e.g., after the initial palliation or Glenn procedure). All of them were treated successfully with pharmacologic intervention. The incidence of early postoperative tachyarrhythmias in patients with RAI did not differ significantly between patients undergoing these special techniques and the usual Fontan completion (9 of 16, 56% vs 9 of 25, 36%). Sinus node dysfunction developed in two patients (one with RAI and one with LAI); both were treated with implantation of a permanent pacemaker in the same hospitalization.

One patient had respiratory dysfunction and elevation of Fontan pressure on postoperative day 2. The patient was
supported with extracorporeal membrane oxygenation for 5 days and successfully weaned after adding a fenestration.

Besides these three early re-operations, two late re-operations (release of ventricular outflow obstruction and ligation of collateral arteries) were performed. Kaplan–Meier method-estimated freedom from re-operation was 82% (95% CI: 64–100%) and 76% (95% CI: 55–96%) at 1 and 5 years, respectively.

Postoperatively, no thrombo-embolic events occurred, and no patient required conduit replacement. Finally, no protein-losing enteropathy or plastic bronchitis was reported.

3.3. Follow-up cardiac catheterisation

Follow-up cardiac catheterisations were performed in 16 patients after Fontan completion. We evaluated the latest data for patients who underwent follow-up catheterisations several times (n = 11). The interval between Fontan completion and the latest follow-up cardiac catheterisation was 50.3 ± 34.1 months. The haemodynamic variables were as follows: central venous pressure, 12.0 ± 2.0 mmHg; cardiac index, 2.9 ± 0.6 l (min m²)⁻¹; ejection fraction, 53.1 ± 5.7%; and saturation, 92.3 ± 6.4%. Stenosis of the IVC, the separate HV or the pathway of Fontan circuit was not observed in any patient. There were no instances of external compression of the pulmonary vein by the extracardiac conduit or obstruction of the pulmonary vein by the intra-atrial tunnel.

At the latest follow-up, one patient who underwent single-lung Fontan procedure was in New York Heart Association class II, and the remaining 16 patients were in class I.

4. Discussion

The outcomes of Fontan completion for FSV patients with atrial isomerism have improved dramatically in the past decade [1—4]. However, owing to complex anatomical anomalies in patients with atrial isomerism [5—7], the technical aspects of constructing the pathway for Fontan completion are complicated. To the best of our knowledge, there have been no reports focussing on patients with separate HV drainage. In the present study, we defined ‘separate HV drainage’ as the existence of one or more HVs that drain into the atrium separately from the IVC. Interestingly, there is not even a commonly accepted term to describe this condition. In the previous reports, it has been called ‘separate HV’ [4,5,10,12], ‘independent HV’ [6], ‘partial anomalous HV’ [7], ‘isolated HV’ [8], ‘direct return of the HV’ [9], ‘nonconfluency between the IVC and the HV’ [11] and so on. Small, separate HV, which could be simply ligated, have been termed ‘accessory HV’ [13].

The surgical technique to be applied for separate HV drainage is a matter of controversy. In the detailed successful case reports, various methods for directing blood flow from the IVC and the separate HV to the pulmonary arteries have been presented. These include intra-extracardiac conduit Fontan procedure [9,10], connecting the IVC to the HV in a side-to-side fashion before anastomosing them to an extracardiac conduit [8], en bloc resection of the IVC and the HV and anastomosing these veins to an extracardiac conduit [11] and other technique [12]. However, these case reports did not make clear recommendations as to which technique is indicated by which situation or anatomical presentation. Therefore, in the present series, we tried to establish a coherent, anatomically based strategy for Fontan completion in patients with separate HV drainage. Similarly to these other reports on successfully treated cases, we managed to direct blood flow from the IVC and the HV (including the separate HV) to the pulmonary arteries in all patients, and we believe it is very important to create an unobstructed pathway for both the IVC and the HV to the pulmonary artery without disturbing the pulmonary venous flow. For this purpose, we believe there are three important anatomical issues, and one technical issue that should be considered when surgically treating patients with separate HV drainage. These are as follows:

1) The distance between the IVC and the separate HV and the position of the vertebrae are the most important anatomical issue to consider. When the IVC and the HV are ipsilateral or contralateral and not widely separated by the vertebrae, we can perform en bloc resection of the IVC and the HV, and anastomose these veins to an extracardiac conduit (Fig. 1). Kawahira and colleagues [11] reported on a handmade shoetree conduit for a relatively large orifice of en bloc resection of the IVC and the HV. However, we obtained good results by only cutting the conduit obliquely, and making a suitable size for anastomosis. We performed lateral tunnel in one patient (case 2) with this situation, and it was our policy in the early study period to perform lateral tunnel Fontan procedure for patients with low body weight. However, because of the longer cardiac arrest time and late atrial baffle enlargement after lateral tunnel, we changed our policy and adopted extracardiac conduit Fontan procedure even in FSV patients with low body weight [14], and we believed we could install an extracardiac conduit after en bloc resection of the IVC and the HV in this patient.

When the IVC and the HV are contralateral and widely separated by the vertebrae, we did not choose en bloc resection of the IVC and the HV, because a large resection of the atrial wall may lead to kinking of the conduit, the IVC or the separate HV [15], or may deform the atrioventricular valve or the pulmonary vein(s). Therefore, we chose an intra-extracardiac conduit (Fig. 2) at the expense of a longer cardiac arrest time and postoperative warfarinisation.

2) The relationship of the IVC and the apex (apicocaval juxtaposition) is another anatomical issue to consider. In patients with apicocaval juxtaposition, the ideal pathway of Fontan circuit should be created with careful consideration. It is our policy to perform Fontan completion with a conduit for such patients; therefore, possible pathways are as follows: an extracardiac conduit bridging the vertebrae to the contralateral pulmonary artery [11]; an extracardiac conduit behind the ventricle to the ipsilateral pulmonary artery [16]; or an intracardiac conduit. We mainly chose an extracardiac conduit bridging the vertebrae to the contralateral pulmonary
artery in patients with atrial isomerism and apicocaval juxtaposition, because an intracardiac or an extracardiac conduit behind the ventricle may lead to pulmonary venous obstruction [17].

In patients with separate HV drainage, the technical aspects of constructing the pathway with apicocaval juxtaposition are more complicated. When the IVC is on the opposite side of the apex, and the separate HV is on the same side of the apex (Fig. 1), it is relatively easy to move these veins and construct the pathway with en bloc resection of the IVC and the HV. However, in the reverse case (Fig. 2), owing to the difference in calibre, it is more difficult. In fact, all patients we judged to have widely separated HV had apicocaval juxtaposition.

3) The status of pulmonary veins in patients following the repair of infracardiac TAPVC is another anatomical issue to consider. In two detailed reports on the Fontan procedure in patients with heterotaxy [2,4], among the three types of extracardiac TAPVC, Fontan completion was successfully achieved in only supracardiac TAPVC. Furthermore, few reports [18] have been published about patients with infracardiac TAPVC who underwent Fontan completion. Therefore, it is difficult to recognize the pitfalls of Fontan completion after the repair of infracardiac TAPVC. We believe that en bloc resection of the IVC and the HV can deform the previously common chamber, and that an intra-extracardiac conduit can obstruct the ostium of the pulmonary veins located more inferiorly than usual. In case 3 (Fig. 3), if we had tried to create one pathway to direct blood flow from the IVC and the widely separated HV, it would have resulted in compression of one of the pulmonary veins. Therefore, we chose lateral tunnel for only the left side widely separated HV and extracardiac conduit for the IVC and the right HV. We had good results of two cases in which we took special care not to obstruct the pulmonary veins. Thus, we concluded that patients with infracardiac TAPVC and separate HV drainage warrant specially tailored surgical techniques.

4) The placement of venous cannula is a technical issue to consider. In case of separate HV, achieving a reasonable surgical field and preventing HV stenosis must be considered. We believe that connecting the IVC and the HV in a side-to-side fashion prior to anastomosing them to an extracardiac conduit may cause acute liver damage, if performed without HV cannulation (with only a simple clamp), because of the longer HV clamp time required. Therefore, we undertook mainly en bloc resection because it can be performed with a simple clamp. Furthermore, direct HV cannulation should be avoided if there are multiple separate HVs. In the two patients we encountered with multiple separate HVs (Fig. 3), we did not cannulate the separate HVs and avoided deforming the HVs. Therefore, we chose an intra-extracardiac conduit and preserved the multiple separate HVs as they were.

Undoubtedly, the characteristics of our cohort with atrial isomerism and separate HV drainage (extracardiac TAPVC, 71%; apicocaval juxtaposition, 59%) represented a significantly more challenged patient population than that of other reports [1—4]. After Fontan completion with various modifications in this series, the central venous pressure of 12.0 ± 2.0 mmHg and cardiac index of 2.9 ± 0.6 l (min m²)⁻¹ were excellent considering the severity of our patient population. Furthermore, our patients had maintained good physical status at the time of their latest follow-up.

Finally, we should discuss the issue of the Fontan procedure paired with fenestration and partial HV exclusion [19]. Different institutions have different strategies for fenestration in the Fontan circuit [20,21], and we believe that fenestration is not necessary in most FSV patients [14,18]. It has been observed that the performance of a small fenestration in the Fontan circuit results in decreased systemic venous pressure and reduced morbidity from serious effusions, at the expense of lower arterial oxygen saturation, balanced by improved cardiac output and oxygen delivery. Among various modifications in fenestration, partial HV exclusion [19] may be indicated in patients with separate HV drainage, partially because Fontan procedures can lead to liver function disturbances that are presumably caused by a high central venous pressure [22]. It seems beneficial to leave separate HV draining into the lower-pressure atrium. However, increasing right-to-left shunt can develop over time because of the pressure gradient, and there have been reports of massive intrahepatic shunting after Fontan completion with partial HV exclusion, requiring surgical intervention [23—25]. Because there have been no detailed report on the long-term follow-up of intrahepatic shunting after Fontan completion with partial HV exclusion, and no large studies on late liver cirrhosis after Fontan completion without HV exclusion, we cannot conclude whether our strategy for separate HV drainage or partial HV exclusion is better. Further long-term studies are needed to resolve this issue.

5. Study limitations

The present study is retrospective and non-randomised with a relatively small number of patients. Furthermore, the mean follow-up period was only 66.0 ± 34.8 months. Therefore, a larger number of patients and longer follow-up are required.

6. Conclusions

The mid-term results of the Fontan completion in patients with atrial isomerism and separate HV drainage were excellent. The distance between the IVC and the HV, the position of the vertebrae, the relationship of the IVC and apex (apicocaval juxtaposition) and the site of pulmonary venous return (especially after the repair of infracardiac TAPVC) should be considered when choosing a surgical technique.

References


