Case report - Congenital

Anatomical repair of a persistent left superior vena cava into the left atrium

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Abstract

The anatomy of a persistent left superior vena cava (SVC) to the left atrium (LA) without the innominate vein can make it challenging to complete intracardiac repair. We reviewed our five cases of the direct end-to-side anastomosis of SVCs to facilitate anatomical repair of SVC-right atrial connection for biventricular repair. Diagnoses were two partial atrioventricular septal defect with left isomerism, one complete atrioventricular septal defect (CAVSD) with left isomerism, one CAVSD without isomerism and one atrioventricular discordance and double outlet right ventricle with right isomerism. Mean age at the operation was 20 ± 23 months (4–58 months) and body weight was 7.8 ± 3.4 kg (4.8–12.7 kg). After completion of intracardiac repair, the SVC to LA was divided and end-to-side anastomosed to the SVC to the right atrium during cardiopulmonary bypass. No early or late death occurred during follow-up of 14.4 ± 6.9 months (7–23 months). None of the patients developed an obstruction at the anastomosis site of the SVCs. The direct end-to-side anastomosis of SVCs achieved an excellent anatomical SVC–right atrium connection in complex congenital heart diseases.

Keywords: Congenital heart disease; Pediatric

1. Introduction

Persistent left superior vena cava (SVC) that connects directly with the left atrium (LA) can result in a significant intracardiac shunt. If there is no connecting vein between the two SVCs, surgical repair is generally performed by intracardiac rerouting using a baffle to divert flow from the left SVC to the right atrium [1]. In intra-atrial rerouting, the anatomy between the orifices of systemic veins and pulmonary veins can render the procedure to correct the lesion more complex, thus causing a disturbed venous flow [2–4]. There have been few reports describing extracardiac correction for directly connecting SVCs. We herein review our cases involving direct end-to-side anastomosis of SVCs to repair a SVC–left atrial connection for biventricular repair.

2. Case report

From July 2007 to December 2009, we performed the direct end-to-side anastomosis of SVCs in five patients (Table 1). The diagnoses were two partial atrioventricular septal defect with left isomerism, one complete atrioventricular septal defect (CAVSD) with left isomerism, one CAVSD without isomerism and one atrioventricular discordance and double outlet right ventricle with right isomerism. Mean age at the operation was 20 ± 23 months (4–58 months) and body weight was 7.8 ± 3.4 kg (4.8–12.7 kg). Computed tomography revealed that the mean distance between the SVCs at the level of the orifice of the brachiocephalic artery was 32.3 ± 5.5 mm (25.8–41 mm) and the mean size of right SVCs was 6.9 ± 2.3 mm (5.2–10.9 mm) and that of left SVCs was 6.5 ± 2.0 mm (4.9–10.0 mm).

Before cardiopulmonary bypass, the persistent left SVC to the LA was mobilized well and its length was measured to assess the possibility of extracardiac redirection. Cardiopulmonary bypass was established with cannulation to the aortic root, the SVC to the right atrium and the inferior vena cava. After the correction of intracardiac defects, the occlusion of the left SVC showed a significant rise in the venous pressure (> 30 mmHg) and we abandoned the option of the simple ligation of the persistent left SVC. The left SVC was divided at the SVC–atrial junction and was end-to-side anastomosed to the right SVC superior to the aortic arch (akin to a new innominate vein) during cardiopulmonary bypass. Postoperative anticoagulant therapy was administered for eight weeks after surgery.

No early and late death occurred during mean follow-up of 14.4 ± 6.9 months (7–23 months). No children experienced disturbed flow or obstruction at anastomosis sites by transthoracic echocardiogram. A catheter examination in patient 1 one year after surgery demonstrated no pressure gradient and a smooth venous flow in the anastomosis site of SVCs (Fig. 1).
Table 1. Patients’ demographics

<table>
<thead>
<tr>
<th>No.</th>
<th>Diagnosis</th>
<th>Age (months)</th>
<th>Body weight (kg)</th>
<th>Distance between SVCs (mm)</th>
<th>Right SVC size (mm)</th>
<th>Left SVC size (mm)</th>
<th>Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>CAVSD(A), PLSVC to LA</td>
<td>8</td>
<td>5.4</td>
<td>25.8</td>
<td>5.5</td>
<td>4.9</td>
<td>Modified one patch repair, direct anastomosis of SVCs</td>
</tr>
<tr>
<td>2</td>
<td>Dextrocardia, partial AVSD, PRSVC to LA, interrupted IVC and hemiazygos</td>
<td>24</td>
<td>10.2</td>
<td>41.0</td>
<td>6.8</td>
<td>6.0</td>
<td>Patch closure of ASD, cleft closure of mitral valve, direct anastomosis of SVCs</td>
</tr>
<tr>
<td>3</td>
<td>CAVSD(A), PLSVC to LA, interrupted IVC and azygos connection, left isomerism</td>
<td>4</td>
<td>6.1</td>
<td>31.2</td>
<td>5.9</td>
<td>6.4</td>
<td>Modified one patch repair, direct anastomosis of SVCs</td>
</tr>
<tr>
<td>4</td>
<td>DORV, PS, PLSVC to LA, right isomerism, s/p Bil.mBTS</td>
<td>58</td>
<td>12.7</td>
<td>33.1</td>
<td>10.9</td>
<td>10.0</td>
<td>VSD patch closure, atrial septation, RVOTR, direct anastomosis of SVCs</td>
</tr>
<tr>
<td>5</td>
<td>Partial AVSD, PLSV to LA, MR, left isomerism</td>
<td>6</td>
<td>4.8</td>
<td>30.5</td>
<td>5.2</td>
<td>5.4</td>
<td>Patch closure of ASD, cleft closure of mitral valve, direct anastomosis of SVCs</td>
</tr>
</tbody>
</table>

SVC, superior vena cava; CAVSD, complete atrioventricular septal defect; PLSVC, persistent left superior vena cava; LA, left atrium; AVSD, atrioventricular septal defect; PRSVC, persistent right superior vena cava; IVC, inferior vena cava; ASD, atrial septal defect; DORV, double outlet right ventricle; PS, pulmonary stenosis; Bil.mBTS, bilateral modified Blalock–Taussig shunt; VSD, ventricular septal defect; RVOTR, right ventricular outflow tract reconstruction; MR, mitral regurgitation.

3. Discussion

Persistent left SVC into the pulmonary venous atrium creates a right–left shunt and could cause a brain abscess or cerebral infarction [3]. Several surgical procedures to correct this anomaly have been reported, including ligation of the left SVC, intra-atrial redirection of flow from the left SVC to the right atrium, and reimplantation of the left SVC into the right atrium, pulmonary artery or SVC. Ligation of the vein obliterates the intracardiac shunt, but this procedure is risky unless there are large collateral links in the head that allow unobstructed head and neck venous return into the heart [5]. Reimplantation of the persistent left SVC is preferable, especially when there is a possibility that an intra-atrial baffle may obstruct systemic or pulmonary venous return due to the location of the veins’ orifices [2, 3]. In addition, extracardiac procedures can render intracardiac repair easier and shorten cardiac arrest time. The cavopulmonary connection is advantageous in terms of the proximity between the SVC and pulmonary artery [2]; however, high pulmonary artery pressure may well reverse the flow into the left SVC and elevated pulmonary resistance may not permit this connection [3, 4]. The use of a polytetrafluoroethylene graft to connect the SVCs has been successful [3], but this technique is not suitable for small children. Shumacker et al. reported a direct connection into the right atrium [5], but they are commonly located remotely and this anastomosis may have technical difficulties [2]. Reddy et al. also reported a direct reimplantation of the left SVC to the right SVC through a tunnel that had been created between the aortic arch and the pulmonary artery [4]. They thought the persistent left SVC runs through the mediastinum more posterior than the right SVC in most cases and this approach can create a more natural connection. However, creating sufficient space to pass the SVC under the aortic arch may be difficult in some cases due to a dilated aorta or pulmonary artery. Furthermore, this approach may restrict the anastomosis site between the SVCs and cause distortion of the left SVC. Therefore, we chose a route superior to the aortic arch to redirect the flow of the left SVC. We have utilized this procedure since 2007 as the initial choice to repair anomalous venous return to the LA. We have been able to perform this procedure in all cases without technical difficulties. Moreover, all patients experienced good outcomes with no obstruction during follow-up.

In conclusion, the long-term results of this technique are still unknown, but our findings suggest that the anatomical repair of bilateral SVCs can be an effective treatment strategy for the correction of complex intracardiac anomalies with a persistent left SVC to the LA.

Fig. 1. The catheter study revealed no stenosis at the anastomosis site between the left SVC and the right SVC in the first patient with atrioventricular septal defect. SVC, superior vena cava.
References


