Work in progress report - Congenital
Sutureless pericardial repair of total anomalous pulmonary venous connection in patients with right atrial isomerism

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Abstract

Surgical repair of total anomalous pulmonary venous connection (TAPVC) in patients with right atrial isomerism is associated with a significant risk of recurrent pulmonary venous obstruction (PVO). We evaluate the effect of sutureless repair to reduce the risk of recurrent PVO. Since November 2007, five patients, including three neonates, with right atrial isomerism underwent sutureless repair of TAPVC. The sutureless repair was used in three neonates as an initial procedure and in two infants as a procedure for postrepair PVO. Under deep hypothermic circulatory arrest or low flow cardiopulmonary bypass, pulmonary vein (PV) was incised as long as possible. The atrial wall was partially resected and anastomosed to the pericardial wall around the incised PV. There were no early deaths. No patients showed recurrence of PVO. There was one late death. Two patients underwent a bidirectional Glenn shunt after the sutureless repair. The pulmonary venous confluence was confirmed to be left open at the time of the Glenn surgery. The sutureless technique may be useful not only for postrepair PVO but also for non-operated TAPVC in neonates with right atrial isomerism.

Keywords: Right atrial isomerism; Total anomalous pulmonary venous connection; Sutureless pericardial repair

1. Introduction

Despite improvements in the treatment of complex congenital heart disease, right atrial isomerism associated with total anomalous pulmonary venous connection (TAPVC) has remained a clinical challenge [1–3]. Surgical mortality following palliative procedures is still high. More than half of the patients who needed neonatal repair due to the presence of a pulmonary venous obstruction (PVO) died within a few months [2, 3]. Repair of TAPVC in patients with right atrial isomerism is associated with a significant risk of recurrent PVO, which makes it difficult to achieve right heart bypass operation. The prevention of this problem remains elusive. This article describes a sutureless pericardial repair of TAPVC in patients with right atrial isomerism.

2. Patients and methods

2.1. Patients

A sutureless technique was indicated in neonates with right atrial isomerism or patients who suffered from post-repair PVO. Since November 2007, five patients including three neonates with right atrial isomerism underwent sutureless pericardial repair of TAPVC at Toyama University Hospital (Table 1). The parents of the patients gave written informed consent before the operation. The anatomical subtypes of TAPVC included supracardiac type in three patients and infracardiac type in two patients. The sutureless repair was used in three neonates as an initial procedure and in two infants as a procedure for postrepair PVO. Three neonates demonstrated symptoms consistent with PVO, including pulmonary congestion and tachypnea. One patient required preoperative mechanical ventilation. These neonates underwent primary sutureless repair of TAPVC as a matter of urgency. Age at operation ranged from seven to 19 days and body weight ranged from 1.9 to 2.7 kg. One patient underwent pulmonary artery banding and another patient underwent a commissurotomy of the pulmonary valve at the time of the repair of TAPVC.

Another two infants underwent a standard anastomosis between the pulmonary venous confluence and common atrium at the age of one and seven months, respectively. One patient showed right side branch PVO and pulmonary hypertension at the age of six months. This patient underwent a sutureless repair to release the right side branch PVO at the age of seven months. Another patient showed right side branch PVO at 23 months of age. At the age of 26 months, this patient successfully underwent a Fontan-type operation without the repair of PVO. However, protein losing enteropathy developed six months after the Fontan-type operation. This patient underwent a sutureless repair...
Table 1
Patient clinical characteristics

<table>
<thead>
<tr>
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<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
<th>Case 5</th>
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<tbody>
<tr>
<td>Age</td>
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<td>19 days</td>
<td>2 years</td>
<td>7 months</td>
<td>7 days</td>
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<tr>
<td>Body weight (kg)</td>
<td>1.9</td>
<td>2.5</td>
<td>9.5</td>
<td>6.4</td>
<td>2.7</td>
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<tr>
<td>Type of TAPVC Indications</td>
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<td>Infracardiac</td>
<td>Supracardiac</td>
<td>Branch PVO after Fontan</td>
<td>Supracardiac</td>
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<tr>
<td></td>
<td>Primary</td>
<td>Primary</td>
<td>Branch PVO</td>
<td>after Fontan</td>
<td>Primary</td>
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<tr>
<td>Follow-up (months)</td>
<td>21</td>
<td>21</td>
<td>18</td>
<td>18</td>
<td>3</td>
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<tr>
<td>Echocardiography (m/s)</td>
<td>PVO (–)</td>
<td>PVO (–)</td>
<td>PVO (–)</td>
<td>PVO (–)</td>
<td>PVO (–)</td>
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<tr>
<td>Results</td>
<td>Alive</td>
<td>Alive</td>
<td>Alive</td>
<td>Alive</td>
<td>Late death</td>
</tr>
</tbody>
</table>

BDG (1 year) | BDG (11 months) |

TAPVC, total anomalous pulmonary venous connection; PVO, pulmonary venous obstruction; BDG, bidirectional Glenn shunt.

to release the right side branch PVO at the age of 31 months.

Follow-up was obtained for all five patients. Mean duration of follow-up after sutureless repair was 16.2 ± 7.5 (range: 3–21) months.

2.2. Surgical technique for sutureless repair of TAPVC

Patients were placed on cardiopulmonary bypass. The superior and inferior caval veins were not dissected. Core cooling was continued until a rectal temperature of 23 °C was obtained. The aorta was cross-clamped and myocardial protection was achieved with cold blood cardioplegia [4, 5]. In the three neonates, the apex of the heart was elevated to facilitate exposure of the vertical pulmonary venous confluence located in the posterior mediastinum. The pulmonary venous confluence was dissected minimally. Posterior mediastinal pleura and the dilated pulmonary vein (PV) due to PVO were opened simultaneously with a fine surgical knife. Under hypothermic circulatory arrest or low flow cardiopulmonary bypass, incision of the pulmonary venous confluence was enlarged as long as possible. Effluent blood from the PV was blown out with a carbon dioxide blower. The incised pulmonary venous wall was left open with the aid of two or three 7-0 polypropylene stay sutures (Fig. 1). The atrial wall was partially resected and anastomosed to the posterior mediastinal pleura that surrounded the PV with a fine 7-0 polypropylene running suture. This suture line was placed on the pericardium, away from the pulmonary venous ostium (Fig. 2). Then cardiopulmonary bypass was re-established, and rewarming was done. When the rectal temperature reached 35.5 °C, the patients were weaned from cardiopulmonary bypass. Thereafter, delayed sternal closure was performed in neonates to avoid the compression by the edematous mediastinal tissue.

3. Results

3.1. Primary sutureless repair in neonates

There were no early deaths after the initial operation. No patients suffered from neurological complications. PVO was successfully released in all patients. Intraoperative massive bleeding into the left pleural cavity occurred in one patient. Cardiopulmonary bypass was re-established, the heart and the left sided pericardium were retracted toward the midline, exposing the anterior aspect of the pulmonary hilum. Massive bleeding from the incised left inferior PV was found and was repaired successfully with a fine 7-0 polypropylene running suture. This patient suffered from right sided chylothorax and underwent the ligation of the thoracic duct one month after the first operation. Although

Fig. 1. Effluent blood from the pulmonary vein was blown out with a carbon dioxide blower. The incised pulmonary venous wall was left open with the aid of two or three 7-0 polypropylene stay sutures. PV, pulmonary vein; CO₂, carbon dioxide.

Fig. 2. Operative schema of primary sutureless repair of TAPVC in neonates. SVC, superior vena cava; IVC, inferior vena cava.
the chylothorax was successfully treated, the patient died from congestive heart failure due to the progression of the common atrioventricular valve insufficiency. The patient underwent valvuloplasty at the age of four months, but did not survive the operation. The pulmonary venous confluence was confirmed to be left open at the time of valvuloplasty. One patient underwent a bilateral bidirectional Glenn (BDG) shunt and common atrioventricular valvuloplasty at the age of 12 months. The pulmonary venous confluence was confirmed to be left open at the time of Glenn surgery (Fig. 3).

Latest Doppler echocardiography showed that no detectable PVO was present in the three patients who underwent primary sutureless repair.

3.2. Sutureless repair for postrepair PVO

Both infants successfully underwent the release of right side branch PVO. One patient showed marked improvement in pulmonary hypertension, and underwent bilateral BDG shunt four months after the sutureless repair. The patient is now doing well and scheduled for a Fontan-type operation. Another patient recovered from protein losing enteropathy after the sutureless repair.

4. Discussion

Patients with right atrial isomerism have poor outcomes because of a complex combination of cardiac anomalies [1, 2]. After becoming discouraged at poor outcomes, some institutes have been tempted to withdraw aggressive treatment from patients with right atrial isomerism and extra-cardiac TAPVC particularly when it is associated with PVO [2]. In such patients, abnormal positioning of the descending aorta and the PVs cause abnormal stacking of structures in the confined space which leads to compression of the PVs between the common atrium and the descending aorta or the spine. We have experienced two infants who suffered from right branch PVO after the repair of TAPVC. Although anastomotic stenosis did not occur, right PVs were compressed between the common atrium and the descending aorta and/or the spine in both cases. One patient showed pulmonary hypertension before Glenn surgery, and another patient suffered from protein losing enteropathy after a Fontan-type operation. Sutureless pericardial repair solved these difficult problems.

Neonatal repair of TAPVC in patients with right atrial isomerism is associated with a significant risk of recurrent PVO or death, requiring a reoperation for stenosis and pulmonary hypertension [1–3]. The atrial wall of the right atrial isomerism is thick and the pectinate muscles are extensive all round the vestibule [6, 7]. Direct anastomosis between the atrial wall of the right atrial isomerism and the thin-walled pulmonary venous confluence is unreasonable especially during the neonatal period.

The sutureless technique was originally developed for the surgical management of PVO after TAPVC repair [8–10]. The main principle of a sutureless repair is to avoid trauma to the pulmonary venous wall and the endothelium, so as to reduce the risk of subsequent intimal hypertrophy. In addition, distortion of the pulmonary venous structures is prevented by this technique because the atrial wall is sutured to the less pliable posterior mediastinal pleura rather than to the thin pulmonary venous confluence. The prevention of geometric distortion of the suture line reduces local turbulence which may cause anastomotic stenosis [11].

Yun and associates modified the sutureless repair technique as a primary procedure for TAPVC associated with right atrial isomerism [1, 2]. They reported that the sutureless repair could be extended safely to the native PVO conducted into the adhesion-free pericardial cavity. However, the applicability of the sutureless repair to patients without previous cardiac surgery is not established. The efficacy of the primary sutureless technique in the prevention of PVO in patients with right atrial isomerism also remains to be established. Although the extension of the sutureless repair technique to the primary procedure for TAPVC requires adequate evaluation, there is no report of primary sutureless repair for TAPVC in patients with right atrial isomerism without previous repair. We have adopted the sutureless repair technique as a primary procedure for TAPVC in neonates with right atrial isomerism. Since 2007, three neonates with right atrial isomerism underwent primary sutureless repair of TAPVC. All patients survived the first operation. No patient suffered from a recurrence of PVO. It was confirmed that the reported pulmonary venous confluence remained open into the atrial cavity at the time of a second operation in two patients. The primary sutureless repair of TAPVC might have the potential to improve the early and late outcome of patients with right atrial isomerism in terms of the prevention of recurrent PVO.

One patient suffered from massive bleeding into the left pleural cavity in this series. Yun and associates reported four cases in which such bleeding occurred [1]. They reported that the longitudinal division of the PVs into the pulmonary hilum might be associated with the violation of the thin pleura at the junction between the parietal and the visceral pleura overlying the pulmonary hilum in patients without intrapericardial adhesion. An incision of the pulmonary venous confluence should not extend to the portion of each PV.
5. Conclusion

The sutureless pericardial repair of TAPVC could be performed safely not only for postrepair PVO but also for non-operated neonates with right atrial isomerism. All patients survived the operation and no patient suffered from recurrence of PVO. The sutureless pericardial repair technique is therefore considered to improve the early and late outcome of patients with right atrial isomerism.

References


