Case report - Congenital

Total cavo-pulmonary connection without foreign material for asplenic heart associated with partial anomalous pulmonary venous connection

Kouta Agematsu*, Yuji Naito, Mitsuru Aoki, Tadashi Fujiwara

Department of Cardiovascular Surgery, Chiba Children’s Hospital, 579-1, Heta-cho, Midori-ku, Chiba City, Chiba, 266-0007, Japan

Received 11 June 2007; received in revised form 4 November 2007; accepted 5 November 2007

Abstract

The presented case was a 3-year-old boy diagnosed with asplenia (SLL), double outlet right ventricle, pulmonary stenosis, atrioventricular septal defect, hypoplastic left ventricle and partial anomalous pulmonary venous connection to the superior vena cava. Partial anomalous pulmonary venous connection was repaired by translocation of pulmonary artery to avoid pulmonary venous obstruction when Glenn anastomosis was performed. Total cavo-pulmonary connection was established by re-routing the inferior vena cava to pulmonary artery using the atrial septal remnant and the left atrium free wall flap.

Keywords: Total cavo-pulmonary connection; Partial anomalous pulmonary venous connection

1. Introduction

Many procedures have been reported for the repair of partial anomalous pulmonary venous connection (PAPVC) with return to the high superior vena cava (SVC), and total cavo-pulmonary connection (TCPC) has been a common procedure but anatomical complexity of pulmonary veins and systemic venous return in asplenic heart makes the definitive repair more difficult. The PAPVC repair concomitant with Glenn anastomosis and shift of the inferior vena cava (IVC) orifice has been rarely reported. We present a novel technique, in which the pulmonary artery (PA) translocation was performed to avoid obstruction of the pulmonary veins when Glenn anastomosis was established and TCPC was completed by re-routing the IVC connection without foreign material. The postoperative course was uneventful and the obstructions of the PA, pulmonary veins and systemic venous pathway were not observed.

2. Case report

A 3-year-old boy weighing 13.7 kg was diagnosed with asplenia (SLL), double outlet right ventricle (DORV), pulmonary stenosis (PS), atrioventricular septal defect (AVSD), hypoplastic left ventricle and PAPVC returned to the right SVC. Cardiac catheterization examination demonstrated a ventricular volume of 284% of normal value, a ventricular ejection fraction of 51%, Nakata index of 414 mm²/m², pulmonary artery resistance of 1.38 wood/unit. The angiography revealed second degree atroventricular valve regurgitation, bilateral SVC and IVC returning to the right side atrium. The left pulmonary veins returned to the right side atrium and the right upper and lower pulmonary veins took a course behind the right PA and were connected to the right SVC at high level (Fig. 1a). No preparatory cardiac operation had been performed.

The median full sternotomy was made. The gross appearance was consistent with the preoperative diagnosis. The right upper and lower pulmonary veins, which took a course behind the right PA, joined the right SVC at a high level. The IVC returned to the right side atrium slightly on the left side of the supine. The cardiopulmonary bypass was established and the PA trunk was divided at the origin and the proximal end was closed directly. A direct right SVC-PA anastomosis was then performed. The intra-cardiac anatomy was compatible with the preoperative diagnosis. The right upper and lower pulmonary veins, which took a course behind the right PA, joined the right SVC at a high level. The IVC returned to the right side atrium slightly on the left side of the supine. The cardiopulmonary bypass was established and the PA trunk was divided at the origin and the proximal end was closed directly. A direct right SVC-PA anastomosis was then performed. The intra-cardiac anatomy was compatible with the preoperative diagnosis. The IVC orifice was on the right side of the atrial septal remnant and the left pulmonary veins returned to the right side atrium on the right side of the septal remnant. The septal remnant of the left side of the IVC orifice was resected and transected remnant muscle bar was sutured inferiorly on the right side of the orifice of the IVC to create a ridge, and the free wall of the left side atrium was then sutured to the ridge (Fig. 2). The left SVC was divided from the left side atrium and sutured to the right PA and the proximal
stump of the left SVC was sutured to the divided distal end of the PA trunk (Fig. 1b). The common atrioventricular valve was repaired by valvulo and annuloplasty.

Postoperative clinical course was uneventful. Postoperative catheterization findings demonstrated the unobstructed TCPC pathway, PA and the right pulmonary veins, and postoperative Holter electrocardiogram performed when the patient was discharged did not reveal any kind of arrhythmia, and catheterization performed seven-months after the operation findings demonstrated smooth TCPC pathway without obstruction, mean SVC pressure 9 mmHg, mean IVC pressure 10 mmHg and remarkable intra-atrial shunting was not detected.

3. Comment

Repair of the PAPVC to high SVC has significant morbidity, including obstruction of the pulmonary veins, SVC stenosis or obstruction and atrial arrhythmia [1–3]. Many surgical techniques have been reported for the repair of the PAPVC with atrial septal defect [4, 5]. However, the PAPVC repair concomitant with Glenn anastomosis has rarely been reported. Also, lateral tunnel established by converting the right side IVC to the left side atrium without foreign material has been rarely reported.

In our case, the right pulmonary vein obstruction might have been caused by lifting up the right PA which was sutured to the right SVC when Glenn anastomosis was performed because the right pulmonary vein rose up to the right SVC behind the right PA. The right pulmonary venous obstruction was avoided by translocating the PA in front of the right side atrium (Fig. 1). The translocation was easily performed by dividing the right and left PA at free PA trunk. The risk of PA stenosis could be avoided by wide reconstitution of the PA continuity by direct anastomosis.

Due to complex systemic and pulmonary venous returns and situs anomaly, the mortality for patients with heterotaxy who had Fontan operation remains substantial [6], and heterotaxy was one variable that predicted poor outcome [7] in spite of the improvement of outcome after Fontan operation.

The design of the inferior-caval connection for asplenic heart with complex anatomy is difficult. The extracardiac conduit operation and the lateral tunnel are used to complete Fontan circulation. In our case, IVC was returning to the right side atrium and it was difficult to create the inferior-caval connection by using extra cardiac conduit and the free wall of the left side atrium was relatively large which could be used as intra-atrial baffle without any augmentation. In addition, the lateral tunnel procedure has the growth potential and decreased risk of thromboembolism, although the extracardiac conduit operation has potential disadvantages related to using the extracardiac tube, including the lack of growth potential, conduit stenosis and increased risk of thromboembolism and the need for reoperation [8]. Despite of the complex inferior venous return to the atrium, TCPC could be completed by re-routing the IVC to the PA on the left side using autologous tissue. With this technique the benefit of the growth potential of TCPC route and avoidance of anticoagulation are expected.

We performed a successful repair for asplenic heart with complex anatomy.

References


