CASE REPORT

Successful correction of unroofed coronary sinus with pulmonary vein stenosis

Yang Li, Qi An* and Eryong Zhang

Department of Thoracic and Cardiovascular Surgery, West China Hospital, Sichuan University, Chengdu, Sichuan, China

* Corresponding author. Department of Thoracic and Cardiovascular Surgery, West China Hospital, Sichuan University, Chengdu, Sichuan, China
e-mail: docli2002@hotmail.com (Q. An).

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Abstract

We present a case of an infant with an unroofed coronary sinus associated with a persistent left superior vena cava draining into the left atrium, right superior pulmonary vein stenosis, an atretic left superior pulmonary vein and a double-outlet right ventricle. For pulmonary vein stenosis and atresia, we used a sutureless technique with an autologous pericardial patch to create a neoatrium.

Keywords: Pulmonary veins • Unroofed coronary sinus • Autograft • Surgery

CASE REPORT

An 8-month-old male infant (weight 4 kg) was referred to our institution for surgical repair. Preoperative transthoracic echocardiography showed an unroofed coronary sinus, a coronary sinus atrial septal defect (diameter = 12 mm), a persistent left superior vena cava (SVC) that drained into the left atrium, a double-outlet right ventricle with subaortic ventricular septal defect (diameter = 11 mm) and a patent ductus arteriosis (Fig. 1A). The pulmonary valve was bicuspid and mildly regurgitant but without stenosis. The right ventricle measured 17 mm and the main pulmonary artery 19 mm. The left ventricular ejection fraction was 70%. Intraoperatively, we discovered the right superior pulmonary vein stenosis, with a diameter of 1.5 mm at the narrowest but with a normal distal segment. The left superior pulmonary vein connected to the left SVC but was atretic (Fig. 1B). The bilateral lower pulmonary veins were normal. The patient also had hypospadias.

The operation was performed with cardiopulmonary bypass. Prior to the initiation of cardiopulmonary bypass, a pericardial patch was harvested and the patent ductus arteriosus ligated. We divided the right superior pulmonary vein and left SVC at the inlet portion of the left atrium. The left superior pulmonary vein was then transected at the entrance into the left SVC. Next we included part of the left atrial tissue along with our incision to enlarge the ostia and incised the superior pulmonary veins longitudinally to enlarge the opening. The proximal segments of the superior pulmonary veins were left in the pericardium. The neoatrium was created by suturing the posterior wall of the left atrium and the autologous pericardial patch to the pericardium around the opening of the superior pulmonary veins, so that the effluent of the pulmonary veins was contained by the left atrium and pericardium (Fig. 2A). We then proceeded to repair the double-outlet right ventricle, the subaortic ventricular septal defect and the atrial septal defect. Finally, the posterior wall of the proximal left SVC was anastomosed directly to the right atrial appendage, while the anterior part was patched with autologous pericardium (Fig. 2B). The patient was rewarmed and weaned from cardiopulmonary bypass in normal sinus rhythm, and primary closure of the sternum was carried out.

The patient was discharged from hospital without signs of congestive heart failure on postoperative day 21. At 9 month follow-up, the infant’s clinical status was excellent, without signs of pulmonary venous restenosis or any obstruction at the site of the anastomosis of the left SVC to the right atrium. Transthoracic echocardiography showed that the right and left ventricular outflow tracts were unobstructed. There was no residual shunt. The right ventricle measured 12 mm, the main pulmonary artery measured 20 mm, and the left ventricular ejection fraction was 60%.

DISCUSSION

In the patient with an unroofed coronary sinus and a left SVC, intracardiac or extracardiac correction is required. We chose the latter technique because we were worried that an intracardiac repair would leave this patient with a left atrium that would be too small, because part of it would be used to repair the stenotic and atretic pulmonary veins. In addition, the extracardiac technique is much quicker and reduces the expected long duration of cardiopulmonary bypass.

The left SVC was too short to anastomose directly to the right atrial appendage or the right SVC. Although the usage of a graft has been previously described in the literature [1], the long-term patency of a graft is unclear. To prevent graft stenosis and occlusion, we anastomosed the posterior part of the proximal left SVC directly to the right atrial appendage, while the anterior part was patched with autologous pericardium, a technique we have previously published [2].

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Congenital pulmonary vein stenosis is a rare disease with high mortality. One or multiple veins may be affected, and the median number is two [3]. Using transthoracic echocardiography, it can be challenging to identify the pulmonary vein stenosis because this technique is limited in its ability to visualize the posterior cardiac structures, such as the pulmonary vein [4]. In our case, although the patient had two preoperative echocardiographic examinations, the pulmonary vein stenosis was not identified until during the operation. Computed tomography and/or cardiac catheterization might help the diagnosis to be made preoperatively.

The sutureless repair of pulmonary vein stenosis has been proposed as the ideal approach, because the technique avoids anastomosis and reduces trauma to the pulmonary veins, which might result in postoperative obstruction [5]. Other potential surgical interventions include balloon dilatation, stent insertion and an expanding repair using a patch or autologous atrial tissues for right pulmonary vein stenosis and/or the left atrial appendage for left pulmonary vein stenosis by direct suturing. However, all these approaches have the potential to injure the pulmonary vein intima, leading to early restenosis.

Figure 1: (A) Persistent left SVC draining into the left atrium, double-outlet right ventricle with subaortic VSD and PDA. (B) The unroofed coronary sinus, the coronary sinus ASD, the right superior pulmonary vein stenosis and the left superior pulmonary vein, which connected with the root of the left SVC but was atretic. ASD: atrial septal defect; CS: coronary sinus; LPVs: left pulmonary veins; PDA: patent ductus arteriosus; RPVs: right pulmonary veins; SVC: superior vena cava; VSD: ventricular septal defect.

Figure 2: (A) The neoatrium was created by suturing the left atrium and the autologous pericardial patch to the pericardium around the opening of the superior pulmonary veins. (B) The left SVC was anastomosed to the right atrial appendage by using an autologous pericardial patch. LA: left atrium; LPVs: left pulmonary veins; RAA: right atrial appendage; RPVs: right pulmonary veins; SVC: superior vena cava.

For our patient, we were able to achieve an excellent short-term outcome. We will continue to monitor the patient carefully in the long term.

Conflict of interest: none declared.

REFERENCES


