Case report

Successful total correction of complete atrioventricular canal, total anomalous pulmonary venous drainage and unroofed coronary sinus in an infant

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Abstract

An infant with complete atrioventricular canal (CAVC), total anomalous pulmonary venous drainage into the left superior vena cava and an unroofed coronary sinus successfully underwent total correction. A homograft vein was used to connect the proximal left superior vena cava to the right atrium and can be recommended when other methods of correction of unroofed coronary sinus cannot be used. © 1999 Elsevier Science B.V. All rights reserved.

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1. Case report

The combination of complete atrioventricular canal (CAVC), total anomalous pulmonary venous drainage (TAPVD) into the left superior vena cava draining to the left atrium and unroofed coronary sinus is extremely rare. Only five cases of combination of CAVC and TAPVD were reported, and only one was successfully repaired [1]. We describe one-stage complete repair of the above mentioned anomalies in an infant.

A 6-month-old male infant (weight 6 kg) was referred to our institution for surgical repair. Cardiac catheterization and angiography were performed in another hospital and showed a CAVC – Rastelli Type I, supracardiac type of TAPVD into the left superior vena cava, draining to the left atrium, and absence of the innominate vein (Fig. 1). Additional to his cardiac anomalies, the patient had annal atresia, which was corrected at age 2 days and bilateral Hexadactilia. These was no chromosomal anomaly.

Surgical correction was performed using bicaval and aortic cannulation, continuous cardiopulmonary bypass at 20°C, and crystalloid cardioplegia. CAVC was corrected with the two-patch technique and the mitral cleft was completely closed. The interatrial septum and coronary sinus were absent. The left superior vena cava was divided and sutured above the connection to the pulmonary venous collector, thus allowing its drainage into the left atrium via the distal part of the left superior vena cava. The proximal end of the left superior vena cava was clamped. After rewarming and uncomplicated weaning from cardiopulmonary bypass, the mean pressure in the clamped left superior vena cava was 28 mmHg. Therefore a cryopreserved 5 mm homograft vein was anastomosed to the transected end of the left superior vena cava with interrupted 7.0 monofilament sutures. The homograft was placed through the transverse sinus and anastomosed to the transected end of the left superior vena cava with interrupted 7.0 monofilament sutures. Thereafter pressure in the left superior vena cava dropped to 10 mmHg and was equal to the pressure in the right atrium. The sternum was closed primarily.

The total cardiopulmonary bypass time was 195 min and the aortic clamp time was 79 min. The postoperative period was complicated by a hypertensive pulmonary vascular crisis that was treated by sedation, relaxation and NO inhalation. Child was extubated on 9th postoperative day and discharged from hospital on the 22nd day without signs of congestive heart failure. Echocardiography before dis-
charge demonstrated grade I mitral incompetence, non-obstructed flow in the pulmonary veins and homograft. Six months after operation the patient’s clinical status is excellent without echocardiographic signs of atrioventricular valves incompetence, pulmonary venous stenosis or venous hypertension.

Surgical correction of isolated CAVC or TAPVD can be performed nowadays with low mortality and morbidity. Coexistence of CAVC, TAPVD and unroofed coronary sinus is extremely rare. We were unable to find any such cases in the medical literature. The management of an unroofed coronary sinus in our patient presented a technical challenge. Proposed surgical methods of intracardiac [3,4] or extracardiac [3,4] correction of unroofed coronary sinus could not be used in this case for the following reasons: the distal end of the transected left superior vena cava was used as a connecting channel of the pulmonary venous collector to the left atrium; the proximal end of the left superior vena cava was to short, thus not allowing direct connection to the right superior vena cava or right atrium; simple ligation of the left superior vena cava in our patient was dangerous because of severe venous hypertension. It has been suggested that venous pressure more than 25 mmHg will not allow ligation of an anomalous left superior vena cava [5]. We used a cryopreserved homograft vein as an alternative method to connect the left superior vena cava to the right atrium. The fate of a homograft vein in infants is unclear, but even if homograft stenosis and occlusion develops later, venous collaterals are expected to develop, thus preventing venous hypertension.

In conclusion this case demonstrates the feasibility of one-stage repair of a very rare association of CAVC, TAPVD, and unroofed coronary sinus. The use of a homograft vein can be recommended to connect the left superior vena cava to the right atrium when other methods of correction of unroofed coronary sinus can not be used.

References