Surgical case of partial anomalous pulmonary venous connection to superior vena cava with cor triatriatum: Williams’ modification and excision of diaphragm

Abstract We experienced a successful pediatric surgical case of partial anomalous pulmonary venous connection to the superior vena cava with cor triatriatum. Echocardiography and multidetector-row computed tomography showed partial anomalous pulmonary venous connection (right upper pulmonary vein connected to the high superior vena cava) and atypical cor triatriatum (analogue to type III-A2 of Lucas–Schmidt classification: left upper pulmonary vein had dual connection to the innominate vein via vertical vein and the accessory chamber). At 8 years of age, the male patient underwent extracardiac right atrial pedicle repair of partial anomalous pulmonary venous connection to the superior vena cava (Williams’ modification) and excision of the diaphragm between the accessory chamber and the left atrium simultaneously. The postoperative course was uneventful in normal sinus rhythm and there was no stenosis of newer drainage root from right upper pulmonary vein.

Key words Partial anomalous pulmonary venous connection · Cor triatriatum · Williams’ modification

Introduction

The number of variants of cor triatriatum has been described previously,¹ but the pattern in which only left upper pulmonary vein (PV) returns to the proximal chamber and also connects to the innominate vein via the vertical vein (analogue to type III-A2 of the Lucas–Schmidt classification), is unusual. We experienced a combined case of the right upper PV returning to the higher side of the superior vena cava (high SVC) with this atypical cor triatriatum, and a radical repair for this combined cardiac anomaly.

Case report

The male patient was diagnosed as having atrial septal defect (ASD) at birth; he was asymptomatic and was followed at the outpatient clinic. At 7 years of age, echocardiography showed anomalous septum in the left atrium (Fig. 1), and multidetector-row computed tomography showed that the right upper PV connected to the high SVC and the left upper PV had dual connection to the innominate vein via vertical vein and the accessory chamber (Fig. 2). Subsequent catheterization showed a pulmonary to systemic blood flow ratio (Qp/Qs) of 2.2, 18 mmHg mean pulmonary arterial pressure, and normal ejection fraction and end-diastolic pressure of both ventricles. The diagnosis was partial anomalous pulmonary venous connection (PAPVC) to the high SVC and atypical cor triatriatum (analogue to type III-A2 of the Lucas–Schmidt classification). A radical operation was indicated.

The patient was 8 years of age at the time of operation and his body weight was 22 kg. Standard cardiopulmonary bypass was utilized and venous cannulae were placed in the innominate vein and the inferior vena cava. The anomalous right upper PV drained into the SVC far above the SVC–right atrial (RA) junction and direct anastomosis of the divided SVC to RA did not seem feasible because of the likelihood of excessive tension on the anastomosis. After the SVC was divided just distal to the anomalous PV orifice, the lower end of the divided SVC was closed directly. Then a pedicle flap of RA appendage and RA lateral wall was created. The sinus venosus ASD was small, 3 mm in diameter, and enlarged caudally. The left upper PV had dual connection into the accessory chamber and the innominate vein via a vertical vein. Trans-septal excision of the diaphragm between the accessory chamber and the left atrium was performed. The interatrial septum was repositioned by...
suture to the right-sided RA wall through the atriotomy directing the pulmonary venous blood from the caval orifice across the ASD into the left atrium. The tip of the pedicle was sutured to the posterior half of the divided end of the cephalad SVC. The anterior wall of the SVC–RA conduit was completed using an autologous pericardial patch (Williams’ modification, Fig. 3a,b). Finally, the vertical vein from the left upper PV to the innominate vein was simply ligated.

The postoperative course was uneventful in normal sinus rhythm. Postoperative echocardiography demonstrated that there was no stenosis of newer drainage root from the right upper PV. The patient is doing well 1 year after his operation.