

Two-stage approach for mixed total anomalous pulmonary venous connection

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Abstract

A 4-day-old boy underwent an urgent operation for mixed total anomalous pulmonary venous connection with the left upper pulmonary vein draining into the innominate vein and the other pulmonary veins draining into the coronary sinus. The left upper pulmonary vein was left uncorrected at that time. After periodical follow-up for 5 years, repair of the uncorrected anomalous pulmonary vein was performed. This two-stage operation is a viable option in cases of mixed type total anomalous pulmonary venous connection, leaving the isolated left upper vein uncorrected in the neonatal period, instead of an aggressive full repair.

Keywords

Cardiac surgical procedures, heart atria, heart defects, congenital, pulmonary veins, time factors

Introduction

Although the surgical outcome of total anomalous pulmonary venous connection (TAPVC) has improved, repair of mixed-type TAPVC in neonates is still technically challenging. The surgical management of mixed TAPVC remains controversial.^{1–4} We report a successful two-stage operation in a case of mixed TAPVC. As far as we know, this is the first report of a two-stage operation for mixed TAPVC.

Case report

The patient was a 5-year-old boy who had undergone urgent intervention at the age of 4 days for mixed TAPVC with the left upper pulmonary vein draining into the innominate vein and the other pulmonary veins draining into the coronary sinus (1 + 3 arrangement). In the initial operation, the collecting chamber was sutured to the left atrium and the atrial septal defect was closed using deep hypothermic arrest; the left upper pulmonary vein was left uncorrected. Postoperative echocardiography showed no pulmonary vein stenosis. The right ventricle was slightly dilated, and the grade of tricuspid insufficiency was mild. After the operation, the boy was followed up periodically. Five years after the operation, echocardiography showed good biventricular function and no pulmonary

hypertension; however, the tricuspid insufficiency had progressed to moderate and the right ventricle was moderately dilated because of the residual anomalous connection of the left upper pulmonary vein (Figure 1). The pulmonary-to-systemic flow ratio was 2.2 on cardiac catheterization. To prevent progressive dilation of the right heart and the risk of arrhythmias, repair of the residual anomalous pulmonary venous connection was scheduled. After re sternotomy and dissection of the heart, cardiopulmonary bypass was established using bicaval cannulation with aortic crossclamping. Cardiac arrest was initiated and the heart was enucleated. The vertical vein was transected just below the insertion of the vertical vein into the innominate vein, and anastomosed to the left atrial appendage. The crossclamp time was 53 min. Echocardiography showed no pulmonary vein stenosis. The postoperative course was uneventful, and the child was discharged from hospital on the 5th postoperative day.

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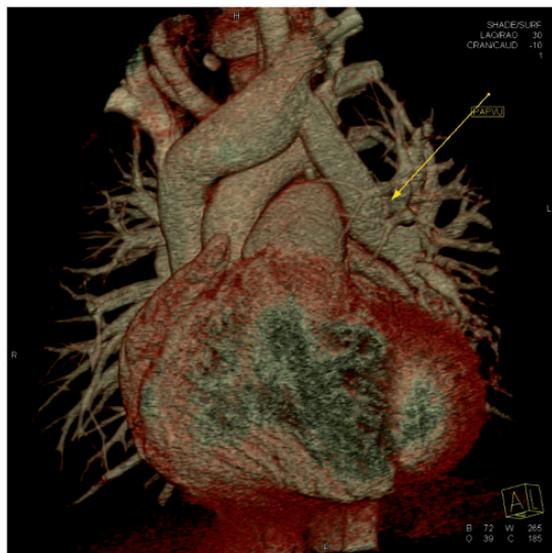


Figure 1. Three-dimensional computed tomography showing the residual anomalous pulmonary venous connection of the left upper pulmonary vein (arrows).

There was no stenosis at the site of anastomosis of the left upper pulmonary vein, and the size of right ventricle became normal one year after the operation.

Discussion

Although the surgical outcome of TAPVC correction has improved, repair of mixed-type TAPVC in neonates is still technically challenging, and the mortality remains high. In this case of mixed type TAPVC, the left upper pulmonary vein was left unrepaired at the initial operation. After periodical and careful follow-up for 5 years, repair of the uncorrected partial anomalous pulmonary vein was performed. Imoto and colleagues² reported good long-term outcomes of patients who had unrepaired isolated anomalous pulmonary veins. Honjo and colleagues³ showed excellent results of primary sutureless repair of mixed TAPVC, in which isolated small anomalous pulmonary veins were unrepaired, especially an isolated right or left upper pulmonary vein (1 + 3 arrangement), with the majority

of the pulmonary veins anastomosed without residual obstruction. Delius and colleagues⁴ stated that if there is evidence that the isolated anomalous pulmonary vein of mixed TAPVC is obstructed, anastomosis to the left atrium is mandatory, but the vein may be left uncorrected if it is not obstructed. On the other hand, conventional repair of mixed TAPVC has been reported, but the patency of the vertical vein-to-left atrial appendage anastomosis is uncertain when it is performed in the neonatal period. Although in adults, the results of left-sided partial anomalous pulmonary venous connection is excellent,⁵ the small size of the isolated pulmonary vein in neonates poses the risk of pulmonary vein stenosis. This case suggests that in mixed-type TAPVC, it is a viable option to leave the isolated left upper vein uncorrected in the neonatal period, instead of the aggressive full repair.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Conflict of interest statement

None declared.

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