Case report - Congenital

Tricuspid atresia with common arterial trunk: successful treatment using Fontan procedure

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Received 19 May 2003; received in revised form 1 October 2003; accepted 11 November 2003

Abstract

Tricuspid atresia (TA) coexisting with common arterial trunk (CAT) is extremely rare, and successful surgical treatment has been reported in only one case until now. We have recently performed surgical treatment for TA with CAT on a 6-year-old boy using the staged Fontan procedure.

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1. Case report

A 2802 g male infant was delivered naturally with an Apgar score of 9. On the second postnatal day, cyanosis and tachypnea were noted, and he was admitted to our institution for further evaluation. Chest radiography showed an increased pulmonary vascular shadow. Electrocardiography indicated a normal sinus rhythm. Echocardiography revealed tricuspid atresia (TA), common arterial trunk (CAT), ventricular septal defect, and atrial septal defect in the context of situs solitus. The main trunk of the pulmonary artery originated from the ascending aorta and divided into right and left branches. Neither regurgitation nor stenosis of the truncal valve was found. The left ventricular diastolic diameter was 32 mm, which was 139% of the normal value. The left ventricular systolic diameter was 23 mm. The left ventricle ejection fraction was 64%. Cardiac catheterization revealed that the diameter of the right pulmonary artery was 5.8 mm, which was 77% of the normal value and the diameter of the left pulmonary artery was 5.8 mm, which was 88% of the normal value. The pulmonary arterial pressure was 50 mmHg in systole, 26 mmHg in diastole, with a mean of 34 mmHg. The $Q_p/Q_s$ value was 5.6.

When the infant was 1 month old, we banded the main trunk of the pulmonary artery, resulting in a decrease in mean pulmonary arterial pressure from 29 to 14 mmHg. It was difficult to band the pulmonary artery because of the shortness of its main trunk. After the procedure, the right pulmonary artery was completely occluded because the banding tape slanted.

At the age of 6 months, this patient underwent a right modified Blalock–Taussig (BT) shunt with a 4 mm graft. Since then, follow-up cardiac catheterizations were repeated every 6 months, however, the right pulmonary artery remained small with a moderate degree of pulmonary hypertension. So, we planned to create another shunt and reconstruct continuity between the right and left pulmonary arteries to allow further growth of the pulmonary arteries and improvement of the unequal pulmonary vascular condition in preparation for the Fontan operation. Then at the age of 3 years, he underwent reconstruction of the pulmonary arteries with autologous pericardial patch and central shunting with a 5 mm graft. The previous BT shunt was left open. Cardiac catheterization 4 months after the operation revealed that the diameters of the pulmonary arteries remained the same size. They were 54% of the normal value on the right side and 80% of the normal value on the left side. Pulmonary arterial index (PAI) was 140 and pulmonary resistance (Rp) was 3.7 unit·m². The mean pulmonary arterial pressure was 13 mmHg on the right side and 22 mmHg on the left side. We considered that the Glenn procedure was better than a more systemic pulmonary shunting to allow growth of the pulmonary artery. Consequently, this patient underwent a bidirectional Glenn procedure.
procedure and plasty of the right pulmonary artery at the age of 5 years. After the operation, the diameters of the pulmonary arteries became 86% of normal value on the right side and 63% of the normal value on the left side. PAI was 191 and Rp was 2.3 unit·m². The mean pulmonary arterial pressure was 14 mmHg on the right side and 17 mmHg on the left side.

At the age of 6 years, we performed the Fontan procedure. Only by using a temporary bypass [1] could we successfully place a 22 mm extracardiac graft from the inferior vena cava to the pulmonary artery. Fenestration was not needed. The postoperative course was very good, and extubation was possible on the operative day.

2. Comments

TA is a rare congenital anomaly and constitutes 2.5% of congenital heart diseases. CAT is also rare, and constitutes 0.7% of congenital heart disease. The coexistence of TA and CAT is extremely rare, accounting for only 0.01–0.02% of congenital heart diseases. Of only 14 cases of TA + CAT that have so far been described in the literature [2–8], 12 died within a few days (maximum 2 months) because of low cardiac output. Only one successful case of surgical treatment using the staged Fontan operation was described by Malec et al. [8].

In our patient, we managed to establish Fontan circulation. However, we had great difficulty controlling pulmonary blood flow. This was first caused by pulmonary artery banding. Because the banding tape was slanted, pulmonary perfusion became unbalanced. For this, we needed to repeat systemic pulmonary shunting. Pulmonary artery banding was not a good option. Primary separation of the aorta and the pulmonary branches with construction of a systemic-to-pulmonary shunt would have been a better first stage approach as described by Malec [8].

References