Construction of the right ventricular outflow tract in children with pulmonary atresia associated with ventricular septal defect

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Background: **It is difficult to build up the right ventricular outflow tract for pediatric patients with pulmonary atresia complicated by ventricular septal defect (VSD). This study was undertaken to evaluate the surgical procedures for the reconstruction of the right ventricular outflow tract (RVOT) in infants and young children.**

Methods: **From June 1999 to December 2004, 81 patients with PA associated with VSD were treated by primary repair, palliative repair or staged repair. These patients were divided into 2 groups according to different surgical procedures. In group I (32 patients), 17 underwent biventricular primary repair and transannular patching with autologous pericardial flake. Group II (49 patients) underwent palliative repair including construction of the right ventricular outflow tract according to preoperative conditions. Fifteen patients underwent staged radical repair, during which transannular patching was performed with homovariant pericardial flake in 8 patients.**

Results: **Four patients (12.5%) died after primary radical repair. Three patients died (6.1%) after palliative repair. But no death occurred for stage II radical repair. Follow-up for 3 months to 2 years showed satisfactory results. The oxygen saturation ($SatO_2$) was 79%-87% for the palliative repair group, and the McGoon index was increased from 0.4-0.7 to 1.1-1.6.**

Conclusions: **With the development of pulmonary artery and pulmonary artery confluence, RVOT construction by the palliative method includes transannular autologous pericardial patching, Gortex conduit, and pericardial rolling. By the radical procedure, the back wall of the traverse pulmonary artery trunk is anastomosed to the incision and homograft besides the transannular autologous pericardial patch.**

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Introduction

Pulmonary atresia associated with ventricular septal defect (VSD) is a common cyanotic congenital heart disease. The severity of clinical symptoms is dependent on the quantity of the pulmonary blood flow which is supplied by the systemic arteries. The early upbuilding of the continuity of the right ventricle to the pulmonary artery will provide favorable pulmonary blood flow and promote the growth of the right ventricle and pulmonary artery.[1-3]

Methods

Patients

Eighty-one of 109 infants and young children with pulmonary atresia associated VSD were operated on by stage I or II biventricle repair at our hospital from March 1999 to December 2004. The patients who had received reconstruction of the right ventricular outflow tract (RVOT) were divided into two groups. Group I (32 patients), aged from 3 months to 11 years (average 3.2 ±1.6 years) and weighing from 5 kg to 33 kg (average 13.5±2.1 kg) were given stage I radical operation. Group II (49 patients) underwent palliative operation; 15 of the 49 patients were subjected to the two-stage

radical operation. These patients were aged from 7 days to 8 years (average 1.9±1.1 years) and weighed from 2.4 kg to 17 kg (average 9.4±1.3 kg). The average interval between the two operations was 0.9-2.7 years. Color Doppler interrogation and catheterization before operation showed neither obvious central confluence between the left and right pulmonary artery (PA), which was supplied by collateral arteries, nor the pulmonary artery trunk (PAT) in 6 patients; the confluence but no pulmonary artery trunk (PAT) in 31 patients; both the confluence and pulmonary artery trunk in 44 patients. The smallest diameter was only 2.3 mm for left PA and 3.1 mm for right PA. The McGoon index was 1.1-1.9 and oxygen saturation ($SatO_2$) in the artery was 63%-89%. Eleven patients were complicated by major aortopulmonary collateral arteries (MAPCAs).

Surgical techniques

Thirty-two patients of group I underwent the radical procedure. Because of the continuity of the main pulmonary artery (MPA) to the right ventricle and full growth of either the PAT or the left and right PA (Table), 17 patients received transannular patching from the right ventricular outlet to the main pulmonary artery. Since the length of pulmonary atresia was 1-2 cm, the back wall of the traverse main pulmonary artery was anastomosed to the incision of the right ventricular outlet in 5 patients.

Forty-nine patients of group II underwent palliative operation. Construction of RVOT was carried out because of growth of the pulmonary artery (Table). In 4 patients, the back wall of the traverse PAT was anastomosed to the incision of the RVOT, and in 1 patient the front wall of the PAT was turned down as the back wall to the incision of the RVOT. The front wall was again enlarged by transannular pericardial patching. Because of discontinuation of the left and right PA, the back wall of the left and right PA was anastomosed to the RVOT with a Gortex conduit in 3 patients. In this group, aortopulmonary shunt plus central shunt or Blalock-Taussig shunt was performed in 18 patients because of low $SatO_2$ after operation. The time for cardiopulmonary bypass and aorta clamping was 29-164 seconds and 24-109 seconds respectively. In this group, 15 patients received two-stage bi-ventricle repair during which transannular homovariant pericardial patching was done in most of the patients. The diameter for enlarged RVOT was 10-12 mm for patients weighing below 10 kg and 13-18 mm for those weighing above 10 kg.

Results

The mortality for primary repair, palliative repair and staged repair was 6.1% (5 patients), 3.0% (3) and 0 respectively. Most patients died from right heart dysfunction and low $SatO_2$. The ratio of pRV/pLV (right ventricle pressure / left ventricle pressure) was below 0.75 except in 4 patients. The total duration of ICU stay and ventilatory support was 4-19 days and 28-92 hours respectively, and the right atria pressure was 9-18 mmHg for the patients. The time for treatment of nitric oxide caused by postoperative high pulmonary resistance was 6 days. The $SatO_2$ was 95%-98% for the patients receiving two-stage biventricle repair and 79%-87% for the patients having palliative method two weeks after operation. Follow-up for 3 months to 2 years showed that the McGoon index increased from 0.4-0.7 to 1.1-1.6.

Discussion

It is difficult to build up the right ventricular outflow tract for patients with pulmonary atresia associated with VSD. In this study we evaluated the surgical procedures for construction of the right ventricular outflow tract according to the PAT, right ventricle,

Table. The procedures for right ventricular outflow tract construction

Procedures	Group I (*n*)	Group II (*n*)	
	Biventricular primary repair	Palliative repair	Staged radical repair
Transannular autologous pericardial patch	17	23	3
Transannular homovariant pericardial patch	0	0	8
Traverse PAT back wall anastomosed to incision	5	4	0
Connected with Gortex conduit	1	6	0
Connected with pericardial roll	2	13	2
Connected with homograft	7	0	2
Link between left and right PA	0	3	0
Unifocalization	2	2	0
B-T shunt	0	18	0

PAT: pulmonary artery trunk; PA: pulmonary artery; B-T: Blalock-Taussig.

pulmonary origin and abnormal connection and coarctation of MAPCAs in infants and young children.

Palliative procedure for RVOT construction

Palliative procedure is used to alter the pathophysiological and anatomical changes for final repair of the RVOT.[4] The criteria for the palliative operation include the upgrowth of the PAT but hypogenesis of the left and right PA, the McGoon index <1.2, the pRV/pLV rate between 0.7-1, and poor growth of the left and right PA with or without the confluence. Transannular patching is suitable for the RVOT to make the pulmonary artery endure the pressure from the right ventricle and to stimulate the development of the pulmonary artery.[5-7] In the reconstruction of the RVOT, it is better to use the self-tissue which is beneficial to growth.[8] Only if the length of pulmonary atresia is limited at or less than 2 cm from the level of the pulmonary valve, can we anastomose the back wall of the main pulmonary artery to the incision of the right ventricular outlet or turn down the front wall of the PAT as the back wall to the incision of the RVOT. Otherwise, a valveless nonelastic conduit is used to connect the right ventricle and the pulmonary arteries.[9] The use of distensible materials such as pericardium or homograft tissue can result in aneurismal dilation of these materials, which absorb the energy that should be transmitted to the distal pulmonary arteries.[10] But the downside of this method is potential coarctation in the future.[11] Though it is beneficial to use the homograft such as valve, the shortcoming is that it is easy to calcify. Because of non-limited pulmonary blood flow for congestive heart failure, it is necessary to reduce the amount of pulmonary blood flow by palliative operation and unifocalization of collaterals including patent ductus arteriosis and MAPCAs as early as possible and build up suitable pulmonary blood flow.[12] In our study, unifocalization of collaterals was performed in 2 patients, of whom 1 died.

Radical operation for RVOT construction

The indications for radical operation include well-development of the main pulmonary artery (MPA) or the left and right PA, normal distribution of pulmonary blood flow to one lung, the existence of the left and right PA and central confluence despite the disappearance of the MPA, the left and right PA at the divarication level of more than 1/2 of the descending aorta at the level of the diaphragm and a McGoon index of more than 1.2.[13] In the stage I radical operation, pericardial patching was done for the construction of the RVOT in most patients of this series, of whom 4 patients were given valved pericardiac patch because of higher pulmonary artery resistance.[14] In the longer length of pulmonary atresia or poorer growth of pulmonary arteries, we used homograft in most patients to avoid the regurgitation of pulmonary blood flow.[15]

In patients with MAPCAs, it is favorable to perform one stage unifocalization to avoid the disadvantage of operation. Moreover, the tiny vessels can be brought into unifocalization of pulmonary blood circulation.[16] The operation should be performed before 1.5 years old[3,4] when the tiny vessels are not severely affected. In our study, 4 patients were subjected to unifocalization of collateral arteries to pulmonary blood circulation as well as RVOT construction by stage I and II operations. The small collateral arteries should be ligated to avoid nerve complications caused by the lower pump pressure. Radical operation showed that the mortality after stage II operation is lower than that after stage I operation.[5] The ratio of pRV/pLV should be kept below 0.75 after cardiopulmonary bypass (CPB) to avoid the low cardiac output in VSD patch. The VSD can be closed operatively if continuous pulmonary perfusion is performed at a rate of more than 2.5 L/min/m^2 and the pulmonary artery pressure is less than 30 mmHg.[17] Otherwise, the VSD should not be closed at that time.[7] We think that the most important factor after CPB is the ratio of pRV/pLV. The pRV/pLV ratio less than 0.45 after operation indicates a well-growing pulmonary artery and better prognosis in a CPB patient.

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