Angiography findings after late bidirectional cavopulmonary shunt operation at mid-term follow-up

İki yönlü geç kavopulmoner şant ameliyatından sonra orta süreli izlemde anjiyografi bulguları

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Bidirectional cavopulmonary shunt is performed mostly as a bridge to the modified Fontan operation or as final palliation in high-risk patients with univentricular physiology (1, 2). There have been numerous studies investigating the timing of the bidirectional cavopulmonary shunt, with many showing the potential benefits of performing an early bidirectional cavopulmonary shunt procedure (3, 4). Nevertheless, age limits to perform bidirectional cavopulmonary shunt remain uncertain. Our patients were relatively older than those in series because they presented at the hospital at a later stage and were diagnosed at an older age. The purpose of this report is to evaluate the hemodynamic data of the patients after late bidirectional cavopulmonary shunt at mid-term follow-up and investigate suitability of these older patients for modified Fontan operation.

A retrospective cohort study was performed in 23 patients after late bidirectional cavopulmonary shunt that underwent angiography between February 1999 and September 2009. Data were expressed as either mean±standard deviation or median and range.

Median age at operation was 3.6 years (range, 11 months to 11 years). Median age at catheterization was 6.4 years (range, 23 months to 16 years). Median interval between the bidirectional cavopulmonary shunt operation and catheterization was 3.9 years (range, 12 months to 17 years). Primary diagnoses of the patients are listed in Table 1. Previous surgical procedures were right modified-Blalock-Taussig shunt in four patients, left modified-Blalock-Taussig shunt in four patients and central shunt in four patients. Additionally, four patients without pulmonary stenosis underwent pulmonary banding operation before the age of 6 months.

Demographic characteristics and hemodynamic data of the patients are shown in Table 2. Mean pulmonary artery pressure was 11.4±4 mmHg (range, 6 to 22 mmHg) and it was lower than 15 mmHg in 20 of 23 patients (86.9%). Three patients' pulmonary artery pressures were higher than 15 mmHg. In one of these patients, mean pulmonary artery pressure was 22 mmHg and pulmonary vascular resistance was 5 Wood units/m^2. In the other two cases, pulmonary artery pressures were higher than 15 mmHg (16 and 18 mmHg), but their pulmonary vascular resistance was lower than 3 Wood units/m^2 (2.03 and 0.51 Wood units/m^2, respectively). Mean pulmonary vascular resistance was 1.38±1.31 Wood units/m^2 and pulmonary vascular resistance was lower than 3 Wood units/m^2' in 21 of 23 patients (91.3%). Mean systemic vascular resistance was 20±9.5 Wood units/m^2, oxygen saturation was 80.2±7.4%, pulmonary artery index (Mc Goon ratio) was 2.2±0.54 (range, 1.4 to 3.6) and ventricular end-diastolic pressure was 9±1.95 mmHg. Systemic venous collaterals were detected in 5 patients (21.7%). Systemic venous collaterals were closed with plug in 2 of 5 patients. Aorta-pulmonary collateral artery and pulmonary arteriovenous fistula were not identified.

Seventeen of 23 patients (73.9%) were suitable for modified Fontan operation. Modified Fontan operation was not suitable in six patients (26, 1%): in four patients due to systemic venous collaterals, in one patient due to systemic venous collateral and small pulmonary arteries and in one patient due to high pulmonary artery pressure and pulmonary vascular resistance.

Theoretically, bidirectional cavopulmonary shunt should be performed between 3 and 6 months of age to achieve maximum benefits (4). Nevertheless, age limits to perform bidirectional

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Table 1. Diagnosis in 23 patients undergoing late bidirectional cavopulmonary shunting

Diagnosis	No of patients	Percentage (%)
Tricuspid atresia	9	39.1
Double-outlet right ventricle	5	21.7
Heterotaxy syndrome	4	17.3
Hypoplastic tricuspid valve and right ventricle	1	4.3
Mitral valve atresia and left ventricle hypoplasia	1	4.3
Pulmonary atresia/intact ventricular septum	1	4.3
Other functional univentricular heart	2	8.6
Total	23	100

Table 2. Demographic characteristics and hemodynamic data in 23 patients undergoing late bidirectional cavopulmonary shunting

Variables	Median (range)* or mean±SD (range)**
Age at operation	3.6 years (11 months to 11 years)*
Age at catheterization	6.4 years (23 months to 16 years)*
The interval between bidirectional cavopulmonary shunt and catheterization	3.9 years (12 months to 17 years)*
Males-Females, n	13 males-11 females
Weight, kg	24.6±15.2 (13 to 67)**
Oxygen saturation, %	80.2±7.4 (58.4 to 91)**
Hemoglobin, g/dl	16.5±2 (13 to 22)**
Pulmonary artery pressure, mmHg	11.4±4 (6-22)**
Pulmonary vascular resistance, Wood units/m^2	1.38±1.31**
Systemic vascular resistance, Wood units/m^2	20±9.5**
Pulmonary artery index, McGoon unit	2.2±0.54 (1.4-3.6)**
Ventricular end-diastolic pressure, mmHg	9±1.95 (5-15)**

SD - standard deviation

cavopulmonary shunt remain uncertain. Our patients presented at the hospital at a later stage and were diagnosed at an older age. Of our 23 patients after late bidirectional cavopulmonary shunt, 17 (73.9%) had good clinical and hemodynamic findings at mid-term follow-up and were suitable for modified Fontan operation. Modified Fontan operation was not suitable in 6 patients (26, 1%): in 4 patients due to systemic venous collaterals, in one patient due to systemic venous collateral and small pulmonary arteries and in one patient due to high pulmonary artery pressure and pulmonary vascular resistance. The most common problem after late bidirectional cavopulmonary shunt at mid-term follow-up was systemic venous collateral development.

Although the bidirectional cavopulmonary shunt is widely used for a variety of complex congenital heart diseases, there are several concerns with this procedure as a long-term palliation (5-8). These include progressive desaturation, pulmonary vascular changes such as systemic venous collaterals and arteriovenous fistulas. In some patients, gradual enlargement of collateral venous channels between the superior vena cava and the inferior vena cava occurs with diminished flow thorough the anastomosis and an increase in pressure in the superior vena cava; these decompress the superior vena cava system into the inferior cava system and contribute to progressive cyanosis. Older age may be a risk factor for systemic venous collaterals because of the lower proportion of caval return from the superior vena cava relative to the inferior vena cava (5). In this study, systemic venous collaterals were detected in 5 of 23 patients (21.7%). One of these patients had small pulmonary arteries; therefore she had a tendency to systemic venous collaterals and was not suitable for modified Fontan operation. But the other three patients might not have developed systemic venous collaterals, if they had undergone a bidirectional cavopulmonary shunt operation at the right time.

In conclusion, the most common problem after late bidirectional Glenn anastomosis at mid-term follow-up was systemic venous collateral development.

Conflict of interest: None declared.

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