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Pulmonary blood flow distribution after the total cavopulmonary connection for complex cardiac anomalies

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Summary. In total cavopulmonary connection (TCPC), the anastomotic portion of the caval veins to the pulmonary artery (PA) is decided empirically based on personal experience. To compare the pulmonary flow distribution from both caval veins in various types of cavopulmonary anastomosis, intrapulmonary ventilation-perfusion distribution after TCPC was studied using lung scanning. We studied 11 patients, 2 to 37 years old, at 30–84 months after TCPC. Lung scanning was performed by administering 185 MBq of xenon-133 saline solution from their upper extremities and, after xenon-133 was washed out, from their lower extremities. Radionuclide counts on both lungs were obtained and intrapulmonary ventilation-perfusion distribution was assessed. In 4 patients whose superior vena cava (SVC)-PA anastomosis was on the right side of the inferior vena cava (IVC)-PA anastomosis, the blood flow distribution of the right and left lungs was 57.4%: 42.6%. In 3 patients whose SVC-PA anastomosis was on the left side of the IVC-PA anastomosis, the blood flow distribution of the right and left lungs was equal in both lungs (right, 53.1%; left, 46.9%). Systemic arterial oxygen saturation increased after TCPC (before TCPC, 85.3% ± 2.7% and after TCPC, 89.8% ± 2.3% ($P < 0.05$) in group R; before TCPC, 86.1% ± 2.8% and after TCPC, 93.6% ± 0.6% ($P < 0.02$) in group L). After TCPC, the value in group L had a tendency to be greater than that in group R ($P < 0.04$), in spite of the same values of systemic arterial oxygen saturation before TCPC and cardiac index (group R, 2.9 ± 0.96; group L, 3.4 ± 0.37). Lung scanning with xenon-133 revealed the distribution of pulmonary blood flow in the patients after TCPC quantitatively, and in the patients whose SVC-PA anastomosis was on the left side of the IVC-PA anastomosis, the right and left balance of the pulmonary blood flow distribution appeared to be more balanced compared with patients whose connection was done the opposite way.

Key words: Total cavopulmonary connection – Xenon-133 – Pulmonary perfusion lung scanning

Introduction

The total cavopulmonary connection (TCPC) in the Fontan-type procedure has become popular as an alternative to atriopulmonary connection for treatment of tricuspid atresia, single ventricle, and several other complex malformations of the heart. It has been reported that this modification reduces postoperative arrhythmia and minimizes the risk of atrial thrombosis from the standpoint of blood flow dynamics and less blood flow turbulence [1–3]. In this procedure, there is almost no mixing chamber for systemic venous return, and the anastomotic portions of the superior vena cava (SVC)-pulmonary artery (PA) and the inferior vena cava (IVC)-PA may provide some sort of obligatory or abnormal distribution of pulmonary blood flow to the right or left lung and, consequently, influence lung or cardiac function over the long term after these procedures.

De Leval et al. investigated the competition of flows from the SVC and IVC in TCPC using computational fluid dynamic simulating methods, and revealed that a minimal energy loss with optimal pulmonary blood flow distribution between the two lungs was obtained by enlarging the inferior vena caval anastomosis toward the right PA [4]. However, there are few reports of

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investigations of this matter in patients late after Fontan operation. The present study attempted to clarify the characteristics of pulmonary blood flow distribution from the SVC and from the IVC separately in the patients after TCPC by means of pulmonary ventilation-perfusion scanning using xenon-133 as the radionuclide tracer.

Patients and methods

Patients

Eleven patients who underwent the TCPC operation as the procedure for Fontan-type repair were included in this study. The cardiac anatomy and surgical procedure for each patient is summarized in Table 1. There were 5 patients with single ventricle, 3 with tricuspid atresia, 2 with mitral atresia, and 1 with double-outlet right ventricle. Age at TCPC ranged from 2 to 37 years (mean 8.1 ± 10.1). Ten of the 11 patients had previously undergone palliative surgery: 6 had a Blalock-Taussig shunt, 1 had a bidirectional Glenn shunt, and 5 had pulmonary artery banding. All of them were released at the time of the TCPC.

We divided the patients into three groups according to the difference in the anastomotic portion of the caval veins to PA (Fig. 1). The SVC-PA anastomosis was on the right side of the IVC-PA anastomosis in 4 patients (group R), and the SVC-PA anastomosis was on the left side of the IVC-PA anastomosis in 3 patients (group L). The IVC-PA anastomosis was between bilateral SVC-PA anastomoses in 4 patients (group B).

Radionuclide assessment

Lung scanning was performed 36–90 months (mean 74 ± 15) after TCPC, using a Hitachi (Tokyo, Japan) gamma camera (2600I) interfaced to a Hitachi nuclear medical data processor (RW-3000). The method utilized was according to that of Ohno et al. and Matsushita et al. [5, 6]. For the ventilation scans, the patient was in the sitting position and inhaled a mixture of 925 MBq xenon-133 gas and room air in a xenon-133 gas control system (ANZAI, AZ-701-NTS) from the maximal expiratory level to the maximal inspiratory level (total lung capacity level). Radionuclide counts on both sides were obtained while the patient held his breath for several seconds ($\dot{V}$). The counts at the equilibrium state were also obtained after rebreathing in a closed circuit ($\dot{V}$). Thereafter, xenon-133 was washed out of the airway in an open circuit with steady breathing. Finally, at the background level in perfusion scans, 185 MBq of xenon-133 saline solution was rapidly injected intravenously in an arm while the patient held his breath at total lung capacity level until the radionuclide count rate reached a plateau on the recorder. After xenon-133 was washed out of the airway in an open circuit with steady breathing, 185 MBq of xenon-133

Table 1. Diagnosis and operative procedures in 11 patients

Group	Patient no.	Diagnosis	Surgical procedure	Age (years) at TCPC procedure	Previous palliative surgery
R	1	SV, l-TGA, 2AVV	TCPC	13	PAB, Rt B-T
R	2	SV, PDA, SAS	TCPC	5	PAB, Lt B-T
R	3	SV, PA, RAA, ASD	TCPC	5	LT B-T, BDG
R	4	MA, l-TGA, TR(3/4)	f TCPC + AVVP + VSD enlar., closure of fenestration	37	PAB, PMI, ASD creation
L	5	SV, l-TGA, 2AVV, SAS	TCPC	3	PAB
L	6	SV, l-TGA, PFO, AVVR, SAS	TCPC + AVVP, VSD enlar., D-K-S	4	CoA repair, PAB
L	7	TA(1b)	TCPC	4	RT B-T
B	8	MA, TGA, PS, BiSVC, PDA, PFO	TCPC	4	ASD creation
B	9	DORV, CAVC, PS, BiSVC, Rt.isomerism	TCPC	3	Rt B-T
B	10	TA(1b), PS, PDA, BiSVC, ASD	TCPC	4	Lt B-T
B	11	TA(1b), BiSVC, ASD	TCPC	2	None

TCPC, total cavopulmonary connection; SV, single ventricle; TGA, transposition of the great arteries; AVV, atrioventricular valve; PAB, pulmonary artery banding; Rt, right; B-T, Blalock-Taussig shunt; PDA, patent ductus arteriosus; SAS, subaortic stenosis; Lt, left; PA, pulmonary atresia; RAA, right aortic arch; ASD, atrial septal defect; BDG, bidirectional Grenn shunt; MA, mitral atresia; TR, tricuspid regurgitation; f TCPC, fenestrated TCPC; AVVP, atrioventricular valve annuloplasty; VSD enlar., enlargement of ventricular septal defect; PMI, pacemaker implantation; PFO, patent foramen ovale; AVVR, atrioventricular valve regurgitation; D-K-S, Damus-Kay-Stansel's operation; CoA, coarctation of the aorta; TA, tricuspid atresia; PS, pulmonary stenosis; Bi, bilateral; SVC, superior vena cava; DORV, double-outlet right ventricle; CAVC, common atrioventricular canal defects

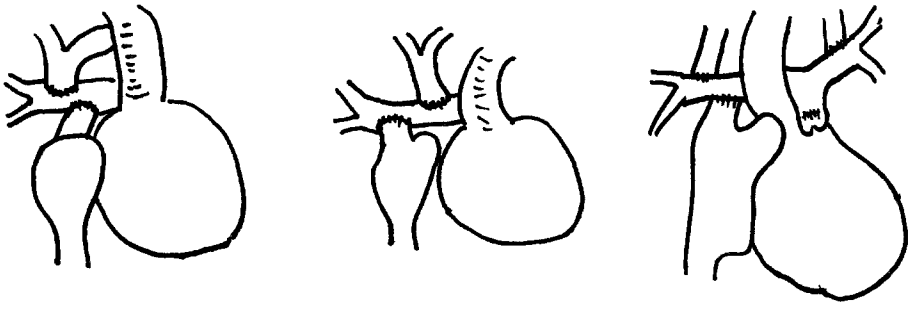

Fig. 1. The three options for total cavopulmonary connection in the Fontan-type operation. The superior vena cava-pulmonary artery anastomosis is on the right side of the inferior vena cava-pulmonary artery anastomosis (group R, 4 patients). The superior vena cava-pulmonary artery anastomosis is on the left side of the inferior vena cava-pulmonary artery anastomosis (group L, 3 patients). The inferior vena cava-pulmonary artery anastomosis is between the bilateral superior vena cava-pulmonary artery anastomoses (group B, 4 patients)

saline solution was rapidly injected, intravenously, in the leg. A count rate of lung analogous to the perfusion of each lung was thus obtained ($\dot{Q}$) when xenon-133 saline solution was injected intravenously into either the arm or the leg. The data determined for the ventilation distribution and perfusion distribution of each lung were corrected for each lung capacity measurement ($\dot{V}/V$, $\dot{Q}/V$). Considering the SVC to IVC flow ratio to be 3:5 [7], the distribution of the right lung is calculated as:

$$\frac{3 \times \text{R from SVC} + 5 \times \text{R from IVC}}{3 \times (\text{R from SVC} + \text{L from SVC}) + 5 \times (\text{R from IVC} \times \text{L from IVC})}$$

The distribution of the left lung is calculated as:

$$\frac{3 \times \text{L from SVC} + 5 \times \text{L from IVC}}{3 \times (\text{R from SVC} + \text{L from SVC}) + 5 \times (\text{R from IVC} + \text{L from IVC})}$$

R from SVC: count rate of the right lung when xenon-133 was perfused from the SVC; R from IVC: count rate of the right lung when xenon-133 was perfused from the IVC; L from SVC: count rate of the left lung when xenon-133 was perfused from the SVC; L from IVC: count rate of the left lung when xenon-133 was perfused from the IVC.

The risk of radiation exposure and the necessity of this examination were sufficiently explained, and informed consent to permit the radionuclide investigation was obtained from each patient and his parent.

Cardiac catheterization and angiography was performed 1–77 (mean 41 ± 25) months after the TCPC operation. Cardiac output was measured by the dye dilution method. Systemic arterial oxygen saturation was measured. On the posteroanterior projection of the pulmonary arteriogram, the diameter of the right and left PA proximal to the origin of the first lobar branch was measured [8].

Statistical analysis

Values were expressed as mean ± standard deviation using the paired *t*-test. Differences were considered to be significant when $P < 0.05$.

Results

Intrapulmonary ventilation-perfusion distribution

Group R (the SVC-PA anastomosis is on the right side of the IVC-PA anastomosis)

In the ventilation scan, the radioactive images of both lung fields were homogeneous in all patients (Table 2). The percentage of the radionuclide count in the right and left lungs was equal (right, 48.5% ± 2.6%; left, 51.5% ± 2.6%). Pulmonary blood flow from the SVC perfused to the right lung (77.9% ± 11.6%) more predominantly than to the left lung (22.1% ± 11.6%). Pulmonary blood flow from the IVC perfused to the right lung (45.1% ± 6.3%) less predominantly than to the left lung (54.9% ± 6.3%) (Fig. 2). Considering the SVC to IVC flow ratio to be 3:5, the distribution of the right and left lungs was 57.4%:42.6%.

Group L (the SVC-PA anastomosis is on the left side of the IVC-PA anastomosis)

In the ventilation scan, the radioactive images of both lung fields were homogeneous in all patients (Table 3). The percentage of the radionuclide count in the right and left lungs was equal (right, 49.5% ± 0.1%; left,

Table 2. Size and mean pressure of the pulmonary artery, and intrapulmonary ventilation-perfusion distribution in group R

		Right	Left
Diameter of PA (mm)		13.5 ± 1.9	14.6 ± 3.5
V	(counts)	44.1 ± 3.8	55.6 ± 3.9
V̇/V		48.5% ± 2.6%	51.5% ± 2.6%
Q̇/V	from SVC	77.9% ± 11.6%	22.1% ± 11.6%
Q̇/V	from IVC	45.1% ± 6.3%	54.9% ± 6.3%
Q̇/V	total	57.4%	42.6%

Table 3. Size and mean pressure of the pulmonary artery, and intrapulmonary ventilation-perfusion distribution in group L

		Right	Left
Diameter of PA (mm)		9.2 ± 1.4	9.2 ± 2.1
V	(counts)	51.0 ± 4.3	48.9 ± 4.3
V̇/V		49.5% ± 0.1%	50.5% ± 0.1%
Q̇/V	from SVC	23.6% ± 11.2%	76.4% ± 11.2%
Q̇/V	from IVC	70.8% ± 11.6%	29.2% ± 11.6%
Q̇/V	total	53.1%	46.9%

Table 4. Size and mean pressure of the pulmonary artery, and intrapulmonary ventilation-perfusion distribution in group B

	Right	Left
V (counts)	46.3 ± 6.7	53.3 ± 6.4
V̇/V	48.1% ± 2.7%	51.9% ± 2.7%
Q̇/V from right SVC	75.6% ± 1.5%	24.4% ± 1.5%
Q̇/V from left SVC	33.8% ± 28.4%	66.2% ± 28.4%
Q̇/V from IVC	54.4% ± 10.3%	45.6% ± 10.3%

50.5% ± 0.1%). Pulmonary blood flow from the SVC perfused to the left lung (76.4% ± 11.2%) more predominantly than to the right lung (23.6% ± 11.2%). Pulmonary blood flow from the IVC perfused to the left lung (29.2% ± 11.6%) less predominantly than to the right lung (70.8% ± 11.6%) (Fig. 3). Considering the SVC to IVC flow ratio to be 3:5, the distribution of the right and left lungs was 53.1%:46.9%.

This right and left balance of the pulmonary blood flow distribution is superior to that observed in group R.

Group B (IVC-PA anastomosis is between bilateral SVC)

In the ventilation scan, the radioactive images of the both lung fields were homogeneous in all patients (Table 4). The percentage of the radionuclide count in the right and the left lung was equal (right, 48.1% ± 2.7%; left, 51.9% ± 2.7%). Pulmonary blood flow from the right SVC perfused to the right lung (right vs left, 75.6% ± 1.5% vs 24.4% ± 1.5%) predominantly, and that from the left SVC perfused to the left lung (right vs left, 33.8% ± 28.4% vs 66.2% ± 28.4%) predominantly. The percentage of pulmonary blood flow from the IVC perfused to the right lung was 54.4% ± 10.3% and the percentage to the left lung was 45.6% ± 10.3% (Fig. 4).

Angiographic and hemodynamic findings (the PA size and pulmonary hemodynamics) (Tables 2–5)

In group R, the size of the right and left PA was the same (right, 13.5 ± 1.9 mm; left, 14.6 ± 3.5 mm). Also, in group L, the size of the right and left PA was the same (right, 9.2 ± 1.4 mm; left, 9.2 ± 2.1 mm). Pulmonary arteriography revealed that none of the patients had stenosis in the PA.

Systemic arterial oxygen saturation increased after TCPC (before TCPC, 85.3% ± 2.7%; after TCPC, 89.8% ± 2.3% ($P = 0.049$) in group R; before TCPC, 86.1% ± 2.8%; after TCPC, 93.6% ± 0.6% ($P = 0.011$) in group L). After TCPC, the value in group L had a tendency to be greater than that in group R ($P = 0.041$), in spite of the same values of systemic arterial oxygen saturation before TCPC and cardiac index (group R, 2.9 ± 0.96; group L, 3.4 ± 0.37).

Discussion

This study attempted to discover the hemodynamic characteristics after the Fontan circulation particular to TCPC. It has been predicted, from some in vitro studies and venography of the patients, that the pulmonary blood flow from a vena cava may distribute to the lung of the anastomotic side. However, there has been no previous clinical demonstration of such a characteristic. In the present study, we demonstrated pulmonary blood flow distribution in the patients after TCPC quantitatively. Moreover, in the patients whose SVC-PA anastomosis was on the left side of the IVC-PA anastomosis (group L), the right and left balance of the pulmonary blood flow distribution was more even in both lungs compared with the connection of the opposite situation. Systemic arterial oxygen saturation after TCPC in group L was greater than that in group R ($P < 0.05$).

In basic flow dynamics, TCPC represents two T-junctions with blood flow in opposite directions from the SVC and IVC into the PA. These abrupt changes in geometry and velocity inevitably generate energy losses. Our results appeared to coincide with the report of de Leval et al., who used computational fluid simulation methods [4]. They mentioned that minimal energy loss with optimal pulmonary blood flow distribution between the two lungs was obtained by enlarging the inferior vena caval anastomosis toward the right PA.

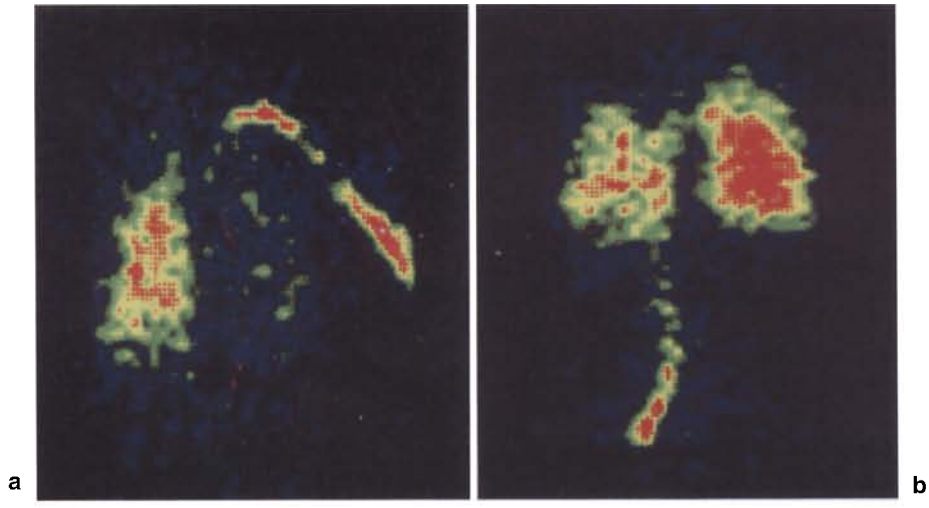

Fig. 2. Pulmonary perfusion from the superior vena cava (**a**) and from the inferior vena cava (**b**) in a patient whose superior vena cava-pulmonary artery anastomosis is on the right side of the inferior vena cava-pulmonary artery anastomosis (group R)

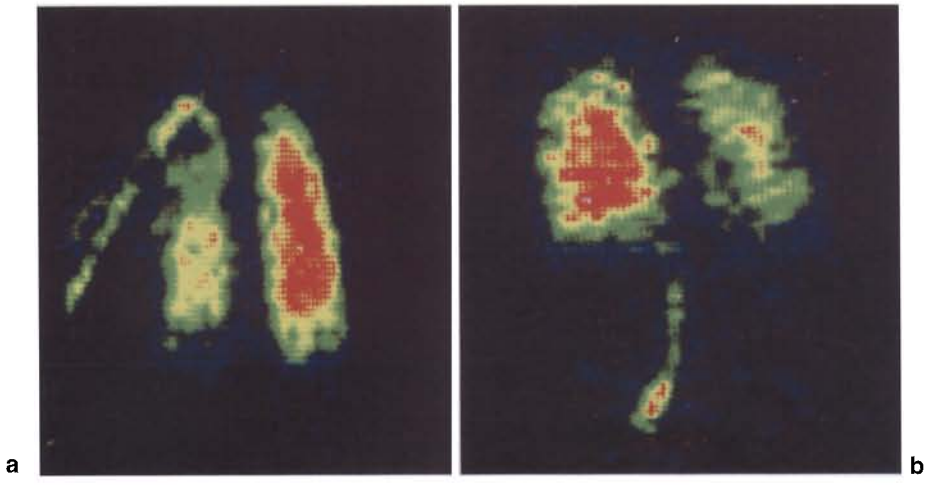

Fig. 3. Pulmonary perfusion from the superior vena cava (**a**) and from the inferior vena cava (**b**) in a patient whose superior vena cava-pulmonary artery anastomosis is on the left side of the inferior vena cava-pulmonary artery anastomosis (group L)

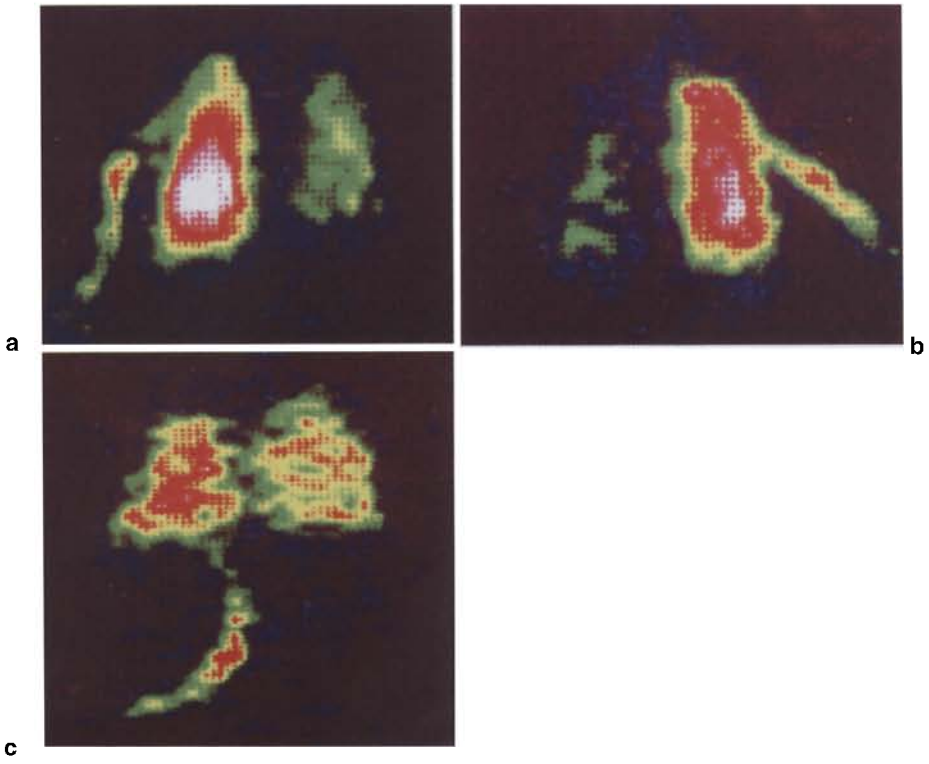

Fig. 4. Pulmonary perfusions from the right superior vena cava (**a**), from the left superior vena cava (**b**), and from the inferior vena cava (**c**) in a patient whose inferior vena cava-pulmonary artery anastomosis is between bilateral superior vena cava-pulmonary artery anastomoses (group B)

Table 5. Pre- and postoperative hemodynamic data in 11 patients

Patient number	Preoperative data			Postoperative data			
	mPAP [mPVWP] (mmHg)	PVR (units/m²)	SaO_2 (%)	mPAP (mmHg)	PVR (units/m²)	SaO_2 (%)	CI (l/min/m²)
Group R							
1	15	2.1	88.2	10	2.4	92.2	3.4
2	8	1.0	81.7	13	3.0	88.5	2.0
3	10	2.0	85.0	13	2.0	91.1	4.0
4	[20]	2.8	86.4	13	2.3	87.2	2.2
Mean ± SD	13.3 ± 5.4	2.0 ± 0.8	85.3 ± 2.7	12.3 ± 1.5	2.4 ± 0.42	89.8 ± 2.3*	2.9 ± 0.96
Group L							
5	12	0.7	88.3	11	1.4	93.7	2.9
6	[16]	1.5	87.0	19	1.4	92.9	3.6
7	[12]	2.9	82.9	9	2.2	94.1	3.6
Mean ± SD	13.3 ± 2.3	1.7 ± 1.1	86.1 ± 2.3	13 ± 5.3	1.7 ± 0.46	93.6 ± 0.5**,†	3.4 ± 0.37
Group B							
8	13	1.6	75.7	8	1.0	90.6	3.8
9	10	3.0	80.9	15	2.1	88.8	1.9
10	13	4.0	76.8	13	1.5	92.0	3.2
11	[11]	1.9	80.2	12	1.6	84.5	3.2
Mean ± SD	11.8 ± 1.5	2.7 ± 1.1	78.4 ± 2.5	12.0 ± 2.9	1.6 ± 0.45	89.0 ± 3.3	3.0 ± 0.80

mPAP, mean pulmonary artery pressure; mPVWP, mean pulmonary venous wedge pressure; PVR, pulmonary vascular resistance; SaO_2, systemic arterial oxygen saturation; [20] means 20 mmHg of the mean pulmonary venous wedge pressure
* $P < 0.05$ vs before TCPC in group R; ** $P < 0.02$ vs before TCPC in group L; † $P < 0.04$ vs after TCPC in group R

The size of the PA and pulmonary hemodynamics may be significant factors responsible for the distribution of pulmonary blood flow. The patients in this study were balanced in terms of right and left pulmonary arterial architecture, right and left pulmonary vascular resistance, and right and left lung ventilation distribution. However, if pulmonary artery branch stenosis exists, the balance of the distribution of pulmonary blood flow can be readily changed [9]. This it is a more logical solution for those patients who are not optimal candidates or whose anatomy will not be suitable for TCPC.

Regarding the limitations of this study, the lung scanning image we obtained was of two-dimensional data. The information from the radioisotope decreased with the decrease in the distance from the lung surface. In a further geometric study, we may need to explore the three-dimensional display of the distribution of pulmonary blood.

The clinical implications of the present study for the decision of the appropriate anastomotic portion of the SVC or IVC to the PA may be still preliminary, due to small number of patients in the study. However, we believe that these results may assist in making a decision in TCPC. In this issue, it has to be kept in mind that there are differences in the blood flow between the SVC and the IVC according to body size and age. Also, we have to be concerned about the long-term results and effects of the flow characteristics with the patient's growth.

In summary, a xenon-133 perfusion scan revealed the distribution of pulmonary blood flow in patients given TCPC after the Fontan procedure, and the pulmonary blood flow from the SVC and the IVC perfused predominantly to the anastomotic side. When the site of SVC-PA anastomosis was left of the IVC-PA anastomosis, the right and left balance of the pulmonary blood flow distribution seemed more balanced compared with connections of the opposite type, and this connection might provide a better pulmonary perfusion status in TCPC.

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