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Sugato Nawa* Makoto Yamada† Koichi Kino‡
Shigeru Teramoto** Kiyoshi Morita††

*Okayama University,
†Okayama University,
‡Okayama University,
**Okayama University,
††Okayama University,

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Abstract

Changes in the hemodynamics of six patients having received Fontan-like operations were closely observed during the first 48 h after the operation. Catheterization studies and simultaneous angiocardiography were also performed before and after the operation. Hemodynamic derangement was particularly severe during the first 24 h postoperatively as indicated by a low cardiac output of less than 2.01/min/m2, which persisted in spite of very high central venous pressure. Furthermore, the central venous pressure needed to re-establish the circulation soon after the Fontan procedure significantly correlated with the angiocardiographically assessed preoperative size of distal pulmonary arteries. Accordingly, the preoperative evaluation of the distal pulmonary arterial size is very important, that provides a good guide-line for the degree of circulatory volume expansion necessary to elevate the central venous pressure and to sustain the circulation in the early postoperative period.

KEYWORDS: Fontan's operation, central venous pressure, pulmonary arterial size, hemodynamic characteristics, post-Fontan care

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Post-Fontan Care Based on Hemodynamic Characteristics, with Special Reference to the Central Venous Pressure

Sugato Nawa*, Makoto Yamada, Koichi Kino, Shigeru Teramoto and Kiyoshi Morita[a]

Departments of Surgery and [a]Anesthesiology, Okayama University Medical School, Okayama 700, Japan

Changes in the hemodynamics of six patients having received Fontan-like operations were closely observed during the first 48 h after the operation. Catheterization studies and simultaneous angiocardiography were also performed before and after the operation. Hemodynamic derangement was particularly severe during the first 24 h postoperatively as indicated by a low cardiac output of less than 2.0 l/min/m^2, which persisted in spite of very high central venous pressure. Furthermore, the central venous pressure needed to re-establish the circulation soon after the Fontan procedure significantly correlated with the angiocardiographically assessed preoperative size of distal pulmonary arteries. Accordingly, the preoperative evaluation of the distal pulmonary arterial size is very important, that provides a good guide-line for the degree of circulatory volume expansion necessary to elevate the central venous pressure and to sustain the circulation in the early postoperative period.

Key words : Fontan's operation, central venous pressure, pulmonary arterial size, hemodynamic characteristics, post-Fontan care

There have been many reports that describe clinical and hemodynamic characteristics of Fontan circulation (1-3). It is well known that high central venous pressure (CVP) is needed to sustain Fontan circulation. Many reports have documented that the CVP needed in the early post-Fontan period was lower than 20 mmHg in the majority of the cases, and it was recommended that the CVP should be kept lower than this value to avoid body fluid accumulation. However, some cases require a CVP higher than this value, while other cases survive with a CVP lower than 20 mmHg. In this study, changes in hemodynamics before and soon after Fontan-like operations were observed in conjunction with angiocardiographic evaluation of the size of distal pulmonary arteries. A search was made for a parameter to predict preoperatively the degree of volume expansion, that is, the CVP, necessary to re-establish and sustain Fontan circulation in the early postoperative period.

*To whom correspondence should be addressed.

Subjects and Methods

Six patients, aged 7 to 21 years (mean age of 12.8 years), were adopted for this study. The cases were restricted to those without pulmonary hypertension or systemic ventricular dysfunction, such as reduced contractility. The patients' characteristics including associated cardiac anomalies are presented in Table 1. The pulmonary circulation was re-established by using the right atrium with or without a conduit. Whole venous blood was diverted via the right atrium in cases 1, 2, 3, and 4, while in cases 5 and 6 venous blood from the superior vena cava (SVC) was directly drained into the pulmonary artery (SVC-PA anastomosis). No valves were inserted at the cavo-atrial junctions or between the right atrium and the pulmonary artery. In all cases cardiac surgery was performed using a cardiopulmonary bypass (CPB) with moderate hypothermia and myocardial protection with the St. Thomas Hospital cardioplegic solution and topical cooling with ice slush.

Serial changes in hemodynamics during the first 48 h after surgery were closely monitored in the ICU (intensive care unit). A hemodynamic study was also performed by means of pre- and post-operative catheterizations and perioperative needle punctures. Postoperative catheterization, including repeated angiocardiography, was performed after 30 to 45 days (mean interval of 40.6 days). The hemodynamic indicators were: heart rate (HR, beats/min), central venous pressure (CVP, mmHg) represented by the mean right atrial pressure (mean RAP, mmHg), mean pulmonary arterial pressure (mean PAP, mmHg), cardiac index (CI, l/min/m^2), and stroke volume index (SVI, ml/beat/m^2). CI was measured by a thermodilution method, and systemic vascular resistance (SVR, dynes · sec · cm^{-5} · m^2) and total pulmonary resistance (TPR, dynes · sec · cm^{-5} · m^2) were calculated.

The size of distal pulmonary arteries was angiocardiographically estimated before and after the operation. The diameters of the right and left pulmonary arteries were measured according to Blackstone *et al.* (4). The cross-sectional area of the right pulmonary artery (RPAa) and the mean value of the cross-sectional areas of the right and left pulmonary arteries ($\overline{PAa}$) were compared with

Table 1 Patients' characteristics, operative procedures and results

Case	Age[a] / Sex	Diseases[b]	Associated anomalies	Previous operations	Modifications of Fontan's procedure	Results
1	19 M	TA (IIb)	ASD, VSD, Juxta-RAA	None	Direct RA-PA anast.	Alive
2	10 F	DORV (SDD) Non-com. VSD	ASD, LSVC, TS, PS, Juxta-RAA	B-T shunt	RA-PA anast. with a patch	Dead
3	7 F	Pulmonary atresia	PDA, ASD, TS, Hypoplastic RV	B-T shunt	Conduit between RA and PA	Alive
4	7 F	Pulmonary atresia	PDA, ASD, TS, Hypoplastic RV	None	Conduit between RA and PA	Alive
5	21 M	TGA (III), SA, SV, C-ECD	Bi-SVC, Hepatocard. V., Absent coronary sinus	B-T shunt	RA-PA anast. using proximal part of SVC, AVVR	Dead
6	13 M	TA (Ib)	ASD, VSD, Hypoplastic RV	B-T shunt, Glenn's op.	Conduit between RA and PA	Alive

a: Ages are presented in years.
b: Type of disease is presented in parentheses.
Abbreviations: TA, tricuspid atresia; ASD, atrial septal defect; VSD, ventricular septal defect; RA-PA anast., right atrium to pulmonary artery anastomosis; M, male; Juxta-RAA, juxta-position of right atrial appendage; DORV, double-outlet right ventricle; SDD, situs solitus, D-loop, D-position; LSVC, left superior vena cava; TS, tricuspid stenosis; B-T, Blalock-Taussig; F, female; Non-com., non-committed; PS, pulmonary stenosis; PDA, patent ductus arteriosus; RV, right ventricle; TGA, transposed great arteries; SA, single atrium; Bi-SVC, bi-sided superior vena cavae; Hepatocard. V., hepatocardiac vein; SV, single ventricle; C-ECD, complete endocardial cushion defect; AVVR, atrioventricular valve replacement.

the prospective area of the right pulmonary artery of the normal heart (N-RPAa), which was calculated by the equation of Castellanos *et al.* (5). These results are presented as ratios RPAa to N-RPAa (RPAa/N-RPAa) and $\overline{PAa}$ to N-RPAa ($\overline{PAa}$/N-RPAa).

Variables are expressed as means±standard deviation (SD) except in Figs. 1 and 2, where only the mean values of the variables of three cases are presented. Student's *t*-test was used to compare the values, and an exponential regression curve and the correlation coefficient (r) were calculated using the Hewlett-Packard microcomputer system (HP-97) with Standard Pac (curve fitting). A $p<0.05$ was considered to be statistically significant.

Results

The operative procedures and the results are presented in Table 1. The overall mortality rate was 33%. Case 2, having presented a persistent low cardiac output state soon after the Fontan operation, succumbed to a second operation, which was done for residual interatrial shunt on the day after the first surgery. Case 5 also succumbed 25 days postoperatively to multiorgan failure following low cardiac output.

The serial changes in hemodynamics during the first 48 h after the operation of three surviving patients, except case 6, are shown in Figs. 1 and 2. A high CVP, represented by mean RAP, in the range of 15.3 to 19.3 mmHg was needed to maintain Fontan circulation. There was no significant difference between CVP and mean RAP. CI was observed to increase gradually as time passed, and it exceeded 2.0 l/min/m² 24 h postoperatively. HR decreased and became stable at about 100 beats/min after 24 h. Consequently, SVI increased (Fig. 1). The serial changes in TPR and SVR during the same period are illustrated with the changes in CI (Fig. 2). TPR showed a very high value of 557 dynes·sec·cm^{-5}·m^2 even 48 h postoperatively, although it decreased gradually as time passed. SVR showed extremely high peaks of 3,120 and 2,830 dynes·sec·cm^{-5}·

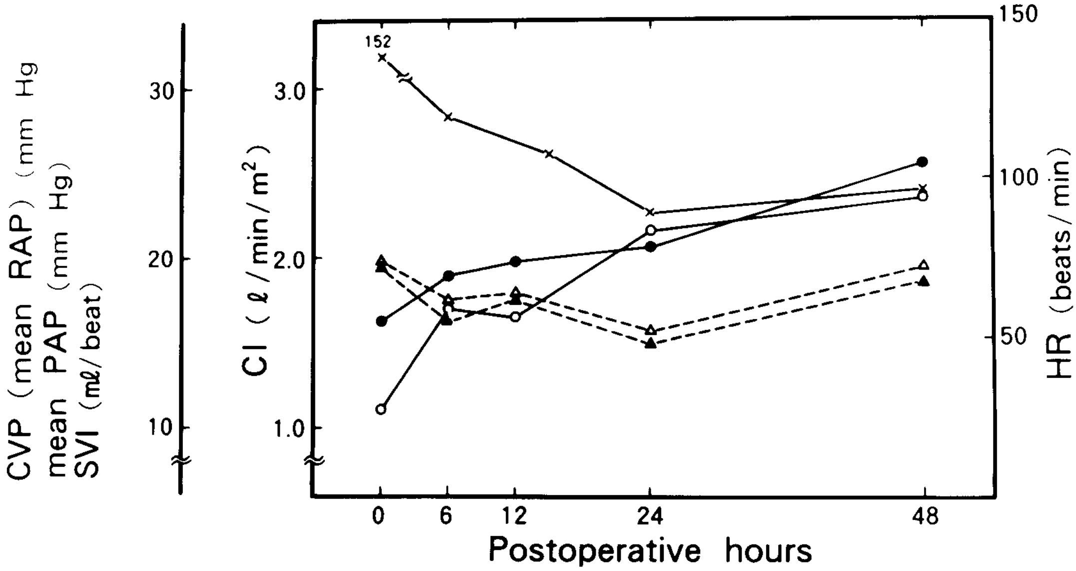

Fig. 1 Serial changes in hemodynamics during the first 48 h after the operation. The variables are presented as means. Abbreviations and symbols: HR, heart rate, ×——×; CI, cardiac index, ●—●; SVI, stroke volume index, ○——○; CVP (mean RAP), central venous pressure (mean right atrial pressure), △·······△; mean PAP, mean pulmonary arterial pressure, ▲·······▲.

m², 6 and 12 h postoperatively, respectively. After that, SVR also gradually decreased.

It was revealed that the mean RAP needed perioperatively to re-establish and to sustain the circulation was significantly higher than the preoperative value (19.0 ± 3.0 mmHg versus 6.2 ± 3.0 mmHg, $n=6$, $p<0.01$), and decreased significantly from 19.0 ± 3.0 mmHg ($n=6$) to 12.1 ± 3.2 mmHg ($n=4$) at the time of postoperative catheterization ($p<0.05$), though it still remained at a higher level than the preoperative value ($p<0.05$) (Table 2). The serial change in mean PAP showed a similar course to that in mean

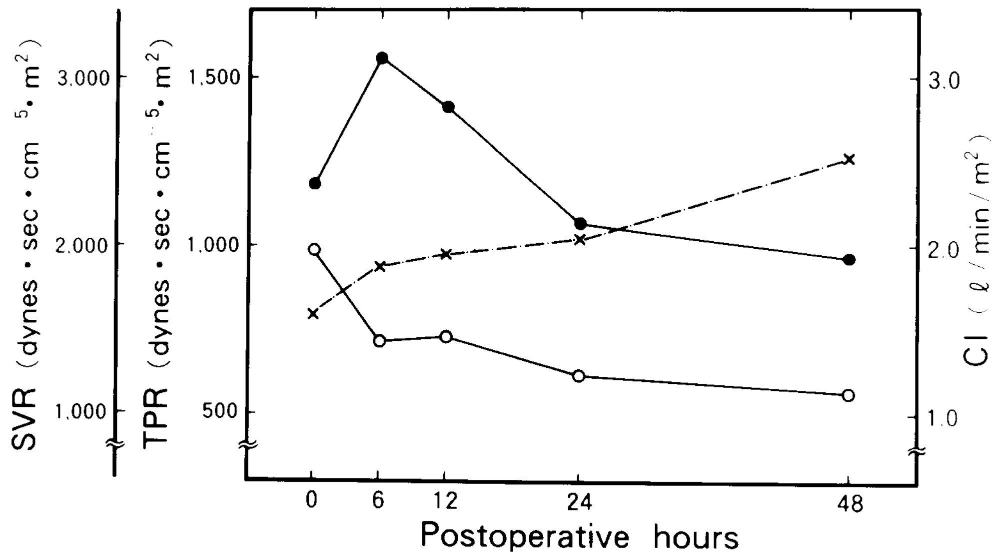

Fig. 2 Serial changes in vascular resistance during the first 48 h after the operation. The variables are presented as means. Abbreviations and symbols: SVR, systemic vascular resistance, ●——●; TPR, total pulmonary resistance, ○——○; CI, cardiac index, ×—·—×.

Table 2 Serial changes in mean right atrial pressure (mean RAP) and mean pulmonary arterial pressure (mean PAP)

Case	Preoperation		Early postoperation		Postop. cathe.	
	Mean RAP[a]	Mean PAP[a]	Mean RAP[a]	Mean PAP[a]	Mean RAP[a]	Mean PAP[a]
1	4	8	14	12	13	12
2[b]	10	15	21	20	—	—
3	5	22	18	20	8.5	7
4	4	10	19	18	11	11
5[b]	10	16	23	22	—	—
6[c]	4	8	18	18	16	14
Mean± SD	6.2 ± 3.0	13.2 ± 5.5	19.0 ± 3.0*, **	18.3 ± 3.4	12.1 ± 3.2***	11.0 ± 2.9

a : Pressures are presented in mmHg.
b : Died postoperatively.
c : Glenn's operation (superior vena cava-pulmonary artery anastomosis) had been done.
Abbreviations and symbols: Postop. cathe., postoperative catheterization. *, $p<0.01$ and ***, $p<0.05$ compared to the preoperative value; **, $p<0.05$ compared to the value at postoperative catheterization.

RAP. No significant difference was observed between these two variables throughout the postoperative course. There were no remarkable differences in the postoperative pressure in the vena cavae and pulmonary arteries either.

The results of angiocardiographic assessment of the size of distal pulmonary arteries before and after the operation are summarized in Table 3. No statistical difference was observed between the pre- and postoperative values. An exponential correlation was observed between the preoperative $\overline{\text{PAa}}$/N-RPAa (x) and the perioperative mean RAP (*i. e.*, CVP) (y) as indicated by the equation: $y = 23.45 \cdot e^{-0.40x}$ ($r = 0.83$, $p < 0.05$) (Fig. 3). The preoperative $\overline{\text{PAa}}$/N-RPAa was 0.36 and 0.29, and perioperative mean RAP needed was 21 mmHg and 23 mmHg in the dead cases, 2 and 5, respectively. Accordingly, two out of three cases having a preoperative $\overline{\text{PAa}}$/N-RPAa value of less than 0.50 succumbed to the operation even though all cases could wean from CPB and the circulation seemed to be tentatively sustained with a very high CVP.

Table 3 Angiocardiographic assessment of the size of distal pulmonary arteries[a]

Case	RPAa/N-RPAa		$\overline{\text{PAa}}$/N-RPAa	
	Preop.	Postop.	Preop.	Postop.
1	1.32	1.04	1.14	0.80
2[b]	0.39	—	0.36	—
3	0.72	0.49	0.90	0.83
4	0.55	0.47	0.51	0.71
5[b]	0.33	—	0.29	—
6[c]	0.41	0.51	0.30	0.39
Mean ± SD	0.62 ± 0.32	0.63 ± 0.24	0.58 ± 0.32	0.68 ± 0.18

a: All data are presented as ratios. No statistically significant difference in either ratio was noted between the pre- and postoperative values.
b: Died postoperatively.
c: Glenn's operation (superior vena cava-pulmonary artery anastomosis) had been done.
Abbreviations: RPAa, cross-sectional area of the right pulmonary artery; N-RPAa, the prospective value of RPAa of the normal heart (5); $\overline{\text{PAa}}$, the mean value of the cross-sectional areas of the right and left pulmonary arteries; Preop., preoperation; Postop., postoperation.

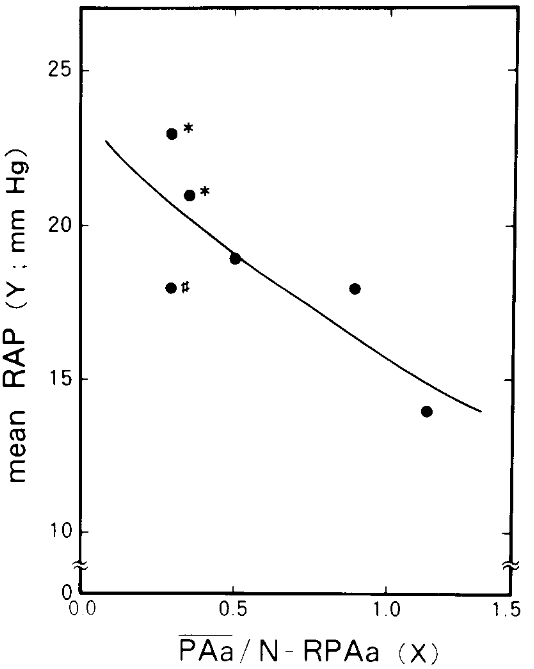

Fig. 3 Correlation between the early postoperative mean RAP and preoperative $\overline{\text{PAa}}$/N-RPAa. There was a significant correlation as indicated by the equation: $y = 23.45 \cdot e^{-0.40x}$ ($r = 0.83$, $p < 0.05$). e: the natural number; other abbreviations are the same as in Tables 2 and 3. *: Died postoperatively. #: Glenn's operation (superior vena cava-pulmonary artery anastomosis) had been done.

Discussion

The Fontan procedure produces non-pulsatile perfusion of the lungs and non-pulsatile filling of the left atrium. Consequently, the pulmonary circulation and the systemic ventricle have to accommodate this unphysiological, non-pulsatile flow. A certain amount of time may be required for this physiological adaptation to be completed. Serial hemodynamic changes were evaluated in three surviving cases. Case 6, however, had received a SVC-PA anastomosis, so that CI could not be estimated by a thermodilution method. It was revealed that CI was less than 2.0 $l/min/m^2$ during the first 24 h after the operation and TPR and SVR were extremely high during the first 12 h after the operation. Accordingly, the problem is how to correct the early postoperative hemodynamic derangement during the first 24 h, while cardiac function recovers following surgical invasion (6) and the pulmonary circulation and systemic ventricle adapt physiologically to the non-pulsatile flow. In fact, at the time of catheterization restudy, Fontan circulation could be sustained with a significantly lower CVP than that needed soon after the operation.

An important problem concerning the measurement of the cardiac output may be the use of a thermodilution method. In Fontan circulation, the venous ventricle as a mixing chamber was totally excluded. It may be thought, therefore, that the data obtained by this method will be unreliable. Repeated measurements, however, were similar and little variance. Furthermore, other clinical signs, such as arteriovenous oxygen content difference, urinary output, and peripheral circulation, were observed to concomitantly improve as the thermodilution-determined CI increased. Accordingly, the thermodilution method is both convenient and useful for the evaluation of hemodynamic changes in cases with Fontan circulation.

It is generally considered that a high CVP is necessary to sustain Fontan circulation, particularly in the early postoperative period. However, as the criteria indicative of a Fontan operation (7) have expanded (8, 9), it has been observed that in some cases, an unusually high CVP was needed to sustain the circulation (10), while in other cases the circulation could easily be sustained with a relatively low CVP, as after corrective surgery for atrial septal defect. The atrial contribution to Fontan circulation is still controversial. Therefore, the major impediment to the pulmonary circulation after the Fontan procedure must be the pulmonary vascular resistance when the pump function of the systemic ventricle is sufficiently preserved. It is not always possible, however, to preoperatively determine the pulmonary vascular resistance in all patients requiring the Fontan operation since such patients usually have very complicated cardiac anomalies so that the direct measurement of the pulmonary arterial pressure and pulmonary blood flow is difficult.

On the other hand, it is generally accepted that the pulmonary arterial size influences the postoperative right heart circulation in tetralogy of Fallot (4, 11, 12). Accordingly, it is considered that the pulmonary arterial size may influence the pulmonary circulation after the Fontan operation in a similar way as in cases with tetralogy of Fallot. The catheterization restudy performed an average of 40.6 days postoperatively revealed that the distal pulmonary arteries did not enlarge after the Fontan operation. Therefore, the preoperative size is very important, and it could be one of the definitive factors impeding the pulmonary circulation after the Fontan procedure. On the other hand, the size of the main pulmonary artery, even if it may be hypoplastic, can be enlarged by the operative methods with modifications of the Fontan procedure (13).

This study revealed that the smaller was the size of distal pulmonary arteries, the higher was the CVP needed perioperatively. However, this does not mean that the high CVP appropriate to the size of distal pulmonary arteries is always sufficient to sustain the pulmonary circulation as observed in cases 2 and 5, in which the distal pulmonary arteries were very small. On the other hand, all cases with a preoperative $\overline{PAa}$/N-RPAa value of more than 0.50 survived the operation. Accordingly, considering the size of pulmonary arteries as a selection criterion for the Fontan operation, the expansion of the criterion should be restricted to a $\overline{PAa}$/N-RPAa value of 0.50. Case 6 survived the operation in spite of severely hypoplastic distal pulmonary arteries. This case, however, received a SVC-PA anastomosis on the right side, and the right lung had already accomodated to the non-pulsatile perfusion via the anastomosis. Therefore, cases having received an anastomosis which produced a non-pulsatile flow might be considered separately.

One can not discuss post-Fontan care only by referring to CVP, *i.e.*, circulatory volume expansion. Medical support with catecholamines will be mandatory, and in some cases mechanical circulatory assists, such as intraaortic balloon pumping and/or ventricular assist devices, may be required to re-establish the falling Fontan circulation (14, 15). Nevertheless, the regression curve obtained will be beneficial to preoperatively predict the CVP necessary to wean from CPB and to sustain Fontan circulation in the early postoperative period. Uniform and/or unreasonable volume expansion may cause massive body fluid accumulation followed by lethal complications.

It was concluded that sophisticated and meticulous postoperative care was particularly important during the first 24 h to re-establish and sustain Fontan circulation, keeping CVP at a high level appropriate to the size of distal pulmonary arteries, and reducing SVR and TPR. Our series, however, was too small to draw a definitive conclusion, and further evaluations in a larger series will be needed to support our observation.

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