Reconsideration of Criteria for the Fontan Operation

Influence of Pulmonary Artery Size on Postoperative Hemodynamics of the Fontan Operation

Hideaki Senzaki, MD; Takayoshi Isoda, MD; Akira Ishizawa, MD; Toshio Hishi, MD

Background The outcome of the Fontan operation largely depends on the selection of patients because this procedure is a physiological correction. Among the several selection criteria for the Fontan operation, the importance of adequate size of the pulmonary artery remains controversial. In this series, in order to clarify whether the pulmonary artery size is indispensable or not as one of the selection criteria for the Fontan operation, we considered the physiological meaning of pulmonary artery size and investigated how it influenced postoperative hemodynamics of the Fontan operation.

Methods and Results In congenital heart disease of decreasing pulmonary blood flow, 40 patients were examined for this analysis. Pulmonary artery indexes (cross-sectional area of the right and left pulmonary arteries divided by body surface area) were measured as the expression of pulmonary artery size, and the relations of pulmonary artery index (PAI) to pulmonary vascular resistance (Rp) and compliance (Cp) were studied. There was no significant correlation between PAI and Rp, whereas a significant correlation was found between PAI and Cp (r=.71, P=.001). Furthermore, Cp influenced postoperative hemodynamics of the Fontan operation by affecting the peak central venous pressure (pCVP) and total impedance, which was the afterload to the ventricle. Impedance increased abruptly when PAI was <=100 mm²/m².

Conclusions The smaller pulmonary artery size causes more disadvantageous hemodynamics after the Fontan operation, with resultant effects of the rise in pCVP and the increase in afterload to the single ventricle. (Circulation, 1994;89:1196-1202.)

Key Words • Fontan procedure • hemodynamics

Since Fontan and Baudet first described a new surgical procedure for tricuspid atresia in 1971, this procedure and its modification have been applied to many other types of congenital heart disease to which biventricular repair is inapplicable.3,9

Because this operation is not anatomic but functional repair in which both pulmonary and systemic circulation must be maintained fundamentally by a single pump, selection of patients occupies a very important part in influencing the outcome of the operation. A number of original criteria for selection of patients were suggested by Choussat, Fontan, and colleagues.10 Among these various criteria, the size of the pulmonary artery has been said to be one of the most important factors to influence the results of the Fontan operation, and several quantitative indicators such as the McGoon ratio11,12 or pulmonary artery index (PAI)13 have been recommended for the evaluation of size and development of the pulmonary artery. However, there have been several reports that have suggested that pulmonary artery size was not related to the outcome of the Fontan operation.14,15

In the light of physiology, pulmonary circulation can be replaced by the electrical circuit shown in Fig 1. Inductance is negligible, as we consider the blood flow to be the steady flow.16,17 According to this analogy, the primary determinant factors of pulmonary circulation are pulmonary vascular resistance (Rp), pulmonary vascular compliance (Cp), and the function of the ventricle as a flow generator. The size of the pulmonary artery does not have any physiological meaning as the primary predictor of pulmonary circulation. In this analogy, the size of the pulmonary artery is represented by Q and the physiological property of the pulmonary vascular bed is decided by Q but by resistance and compliance. Pulmonary artery size (Q) itself is the product of pulmonary blood flow, compliance, and resistance, as shown by the formula Q = iRC. It is apparent then that the size of the pulmonary artery never primarily predicts the postoperative Fontan circulation.

When we consider the pulmonary circulation in a living body, we must also take into consideration the secondary effect of pulmonary artery size on the pulmonary circulation. This is because the difference in vascular radius resulting from the difference in vascular size may secondarily influence pulmonary vascular resistance and pulmonary vascular compliance.

Based on this fundamental consideration, we examined the secondary meaning of pulmonary artery size, that is, the relation of pulmonary artery size to pulmonary vascular resistance and pulmonary vascular compliance, and reevaluated the influence that pulmonary artery size has on postoperative hemodynamics of the Fontan operation.

Methods

In congenital heart disease of decreasing pulmonary blood flow, 40 consecutive patients (aged 5 months to 18 years; mean,
Hemodynamic data were obtained by routine catheter examination. Pulmonary blood flow was determined by the Fick method. Rp was calculated by using mean pulmonary artery pressure, mean pulmonary venous atrial pressure, and pulmonary blood flow. Pulmonary artery wedge pressure and pulmonary venous wedge pressure were used to approximate mean pulmonary venous atrial pressure and mean pulmonary artery pressure, respectively, when they could not be measured. Calculation of pulmonary blood flow and resistance was made only when the source of pulmonary blood flow was a single source. On the other hand, Cp was calculated according to the formula Q=IRC. PAI, pulmonary blood flow, and Rp were substituted into Q, I, and R, respectively.

Statistical Analysis

Association between variables was determined by linear regression analysis, and a correlation coefficient was calculated between these variables. A value of \( P<.05 \) was considered statistically significant.

Results

As shown in Fig 2, there was no statistically significant correlation between PAI and Rp, although there was a trend toward low resistance in patients with larger PAI. Some cases showed the normal resistance with small PAI, and others showed the high resistance with normal to larger PAI. Even the case of a PAI of 111 mm²/m² kept the normal resistance of 2.1 R.U. · m². On the other hand, significant correlation \( (r=.71, P=.001) \) could be found between PAI and Cp (Fig 3). Cp decreased as PAI decreased.

Discussion

The selection of patients is a key point in determining the outcome of the Fontan operation. Choussat and Fontan presented the first criteria for patient selection, and it has been shown that the patients who met these original criteria had good operation results. However, the patients who exceeded one or more of criteria in some area have undergone the Fontan procedure and have had a good outcome. From this viewpoint, the original criteria have been reevaluated, and some of them have been suggested to be dispensable, but among these criteria, the importance of the adequate size of the pulmonary artery remains controversial. Nakata et al quantified the pulmonary artery size by PAI (the cross-sectional area of left and right pulmonary arteries immediately after the first lobe branch divided by body surface area) and examined the relations between mortality and PAI in several types of operations. In the Fontan operation, they concluded that the Fontan procedure should be reserved for those with a PAI >250 mm²/m². However, Girod et al reported that there was no significant difference in mortality rate between patients with a PAI >250 mm²/m² and those with a PAI <250 mm²/m², and the PAI of 188 mm²/m² was reported to be the lowest value among the survivors. Bridges et al also showed that there was no statistical difference in PAI between survivors and nonsurvivors after the Fontan operation. In their series, the lowest value of PAI among survivors was as low as 48 mm²/m². On the contrary, Fontan et al examined the relation between the outcome of the operation and the pulmonary artery size expressed as a McGoon ratio, and they inferred that the probability of

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Patient Characteristics

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*BTS indicates Blalock-Taussig shunt; WS, Waterston shunt; RVOTR, right ventricular outflow tract reconstruction; SLV, single left ventricle; PS, pulmonary stenosis; SRV, single right ventricle; PA, pulmonary atresia; TOF, tetralogy of Fallot; VSD, ventricular septal defect; PDA, patent ductus arteriosus; TGA, transposition of the great arteries; cTGA, corrected TGA; and DORV, double-outlet right ventricle.

*Four patients of asplenia syndrome were included.
†Blood flow through the pulmonary valve was neglected because the stenosis was very severe.

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Fig 1. Analogy of pulmonary circulation by electrical circuit. mPA indicates main pulmonary artery; LA, left atrium; R, resistance; C, capacitance (compliance); Q, electrical charge; i, direct current through resistance; V, potential difference between mPA and LA. Q is equivalent to IRC and represents pulmonary artery size in pulmonary circulation.

7 years) who underwent catheter examination between January 1990 and May 1992 were examined for the analysis mentioned below. Among the patients were those who had mildly increasing pulmonary blood flow that was secondarily due to palliative surgery; yet, the ratio of pulmonary to systemic flow (Qp/Qs) of such patients was to be within 1.5 to avoid the embroidery of Rp and Cp caused by pulmonary vascular obstructive disease. Patient characteristics are given in the Table.

In this series, we used PAI (cross-sectional area of the right and left pulmonary arteries divided by body surface area) as the expression of the size of pulmonary artery. We examined the relations of PAI to Rp and Cp, which are the physiological properties of the pulmonary artery, to reveal the secondary physiological meaning of PAI.

The measurement of the diameter of the pulmonary artery just proximal to the origin of the first lobe branch was expressed as cross-sectional area (mm²) and was normalized by dividing by body surface area. This is referred to as PAI.13
death or takedown increased as the pulmonary artery size decreased.

Why does such controversy exist? To explain this, it is necessary to reconsider the postoperative Fontan circulation from the viewpoint of circulatory physiology.

As mentioned at the beginning, pulmonary artery size has the possibility that it secondarily predicts postoperative Fontan circulation by its relation to pulmonary vascular resistance or compliance, although the primary predictors of pulmonary circulation are pulmonary vascular resistance and compliance.

In general, resistance of the vessel can be expressed as $8\,\text{kL} / \pi r^4$ according to the law of Poiseuille and compliance of the vessel as $3\,\pi r^3L(r/h+1)^2/E(2r/h+1)$ (L is length of the vessel; r, radius of the vessel; k, viscosity of blood; E, Young's modulus of vessel wall; h, wall thickness of vessel). As the size of vessel is expressed as $\pi r^2L$, radius of the vessel (r) increases with the increase in the vessel size. Therefore, the size of the pulmonary artery can influence the Fontan circulation because of its relation to resistance and compliance through the radius of the vessel. The question is how the difference in pulmonary artery size influences resistance and compliance through the difference in the radius of the pulmonary artery.

As shown in Figs 2 and 3, there was no significant correlation between pulmonary artery size and Rp, whereas significant correlation was found between pulmonary artery size and Cp ($r = .71, P = .001$).

**Validity of the Calculation of Compliance**

To evaluate the validity of our method for the calculation of Cp, we compared the compliance calculated by our method (Cp) with compliance calculated by the Reuben’s method (Cp’). As shown in Fig 4, there was
a significant correlation between \( \text{Cp} \) and \( \text{Cp}' \) \((r=.80, P=.001)\), and this indicates the validity of our method.

Further, the \( \text{Cp} \) value is 46 \( \text{mm}^2/\text{m}^2 \text{mm} \text{Hg} \) when the normal value of 330 \( \text{mm}^2/\text{m}^2 \) is assigned to \( \text{PAI} \) in the correlation equation in Fig 3, and the \( \text{Cp} \) value, calculated by using the \( \text{Q}=i \text{RC} \) equation, is 41 to 55 \( \text{mm}^2/\text{m}^2 \text{mm Hg} \) when the normal values of 330 \( \text{mm}^2/\text{m}^2 \), 3 to 4 \( L \cdot \text{min}^{-1} \cdot \text{m}^{-2} \), and 2 \( \text{RU} \cdot \text{m}^2 \) are assigned to \( \text{Q}, i, \) and \( \text{Rp} \), respectively. This also confirms the validity of our method for calculating \( \text{Cp} \).

As for the relation between pulmonary artery size and \( \text{Rp} \), further analysis will be required for more precise understanding.

When we apply the law of Poiseuille \((R=8kL/\pi r^4; R, \) resistance) to the pulmonary vascular bed, \( Q \) (pulmonary artery size) is expressed as \( \pi Lr^2 \), so the relation between \( R \) and \( Q \) is shown as \( R=8\pi k L^2/Q^2 \). As \( Q \) can be supposed to be equivalent to \( L(\alpha \text{PAI}) \) if \( \text{PAI} \) reflects the whole size of the pulmonary artery \((\alpha, \) coefficient between \( \text{PAI} \) and true cross-sectional area of the pulmonary artery), \( R \) is represented as \( 8\pi k L/\alpha^2 \text{PAI} \). Fig 5 shows the relation between \( \text{Rp} \) and \( kL/\text{PAI}^2 \) in the patients listed in the Table. \( L \) was estimated to be proportional to body surface area, and \( k \) was substituted by the value that was corrected according to the relation between hematocrit and relative viscosity, the value of which is one when hematocrit is 40%\(^2\). There was no statistically significant correlation between \( \text{Rp} \) and \( kL/\text{PAI}^2 \), either. This indicates that the difference in radius of proximal arteries does not reflect the difference in radius of vessels, which mainly determines the total resistance of the pulmonary artery, that is, the difference in radius of the "resistance vessel." The radius of proximal arteries reflects the total pulmonary blood volume, which is related to pulmonary vascular compliance, but its difference is not necessarily accompanied with a difference in radius of the "resistance vessel."
In this way, pulmonary artery size represented by PAI is significantly related to pulmonary vascular compliance and has no significant relation to pulmonary vascular resistance, although there is a trend toward low resistance in patients with larger pulmonary artery size. Accordingly, pulmonary artery size can secondarily influence hemodynamics after the Fontan operation because of its relation to compliance.

How does compliance itself play a role in determining the postoperative Fontan circulation? When we consider the stable state in pulmonary circulation represented by Fig 1, pulmonary blood flow (I) can be expressed by the Fourier series as below:26

\[ I = I_0 + \sum_{n=1}^{\infty} I_n \sin(\omega_n t + \phi_n) \]

where \( I_0 \) is the mean steady flow, \( \omega_n = \frac{2\pi}{T} \) and \( T = n \) is the period of the nth harmonic and \( t \) is the time.

A constant term \( I_0 \) represents the mean steady flow. The pulsatile components of flow can be calculated by the Fourier series as below:

\[ Z_n = \frac{P_0 + \sum_{n=1}^{\infty} I_n \sin(\omega_n t + \phi_n)}{R_p + \sum_{n=1}^{\infty} \omega_n^2 \sin(\omega_n t + \phi_n)} \]

where \( P_0 \) is the mean pressure, \( R_p \) is the impedance modulus of the nth harmonic, and \( \phi_n \) is phase angle of the nth harmonic. \( P_0 \) represents mean pulmonary artery pressure (mPAP), which is determined by the relation between resistance and mean flow (cardiac output).

Impedance is a measure of the opposition to pulsatile flow. It determines the pressure waveform and furthermore acts as the afterload to the flow generator (ventricle).26,29 As one of the determinant factors of impedance, \( C_p \) has the possibility to influence the hemodynamics after the Fontan operation in two ways: (1) by influencing peak central venous pressure (pCVP) and (2) by influencing total impedance, which is the afterload to the single ventricle. Total impedance means the impedance of the single vessel continuing from aorta to pulmonary artery.

**Influence on pCVP**

Because in Fontan circulation there is no right ventricle or tricuspid valve for pulmonary circulation, pulmonary artery pressure (PAP) almost directly influences central venous pressure (CVP). Therefore, mean CVP (mCVP) is represented approximately by \( P_0 + \frac{I_0 R_p}{2} \) in Formula 2. On the other hand, pCVP is affected by peak PAP, which varies according to pulmonary artery input impedance.

As one of the determinant factors of impedance as shown in Formula 2, impedance modulus increases and phase angle decreases when \( C_p \) decreases. The increase in impedance modulus and the decrease in phase angle lead to the increase in pulsatility of PAP and the subsequent rise in pCVP.

When the central veins are directly connected to the pulmonary artery (total cavopulmonary connection), pulsatility of pulmonary blood flow is found to be so little that pulsatile components in Formulas 1 and 2 can be neglected. Thus, in total cavo pulmonary connection (TCPC), the influence of \( C_p \) on pCVP is negligible. On the other hand, in right atrial to pulmonary artery connection (APC) of the Fontan-type repair, \( C_p \) influences pCVP according to Formula 2 because of the pulsatility caused by atrial contraction. By substituting 0.12 mmHg sec m²/mL (2 RU · m²) for \( R_p \) as the normal value, the relation between \( C_p \) and \( Z_n \) at heart rate of 120 beats per minute is represented by Formula 3:

\[ Z_n = 0.12 \left( 1 + a \pi^2 n^2 C_p^2 \right)^{1/2} \]

As the first several harmonics at lower frequency domain mostly decide the actual pressure waveform, consideration up to the 10th harmonic is sufficient for satisfactory analysis.31 From the first to 10th harmonic in Formula 3, \( Z_n \) changes according to \( C_p \) as shown in Fig 6, and pCVP subsequently changes as shown in Fig 7 when the first harmonic and the harmonics up to the fifth were evaluated as the most decisive ones. The pCVP was set at 18 mm Hg and mCVP at 12 mm Hg as the normal case at heart rate of 120 beats per minute when \( C_p \) showed normal value of 1.5 mL/m² mm Hg.27 As we assumed that the amplitude of the nth harmonic was equivalent to that of the first harmonic, pCVP must vary approximately between the two curves shown in Fig 7. Accordingly, pCVP rises up to about 22 mm Hg and 24 to 25 mm Hg from 18 mm Hg when \( C_p \) decreases from normal to half (0.75) and one third (0.5) of normal, respectively. If the \( C_p \) decreases further, pCVP will also increase further. In this way, \( C_p \) has an influence on pCVP after the Fontan operation. As PAI is related to \( C_p \) as shown in Fig 3, PAI also affects pCVP in the same way. Although it is unclear how high pCVP is tolerable, the smaller \( C_p \) becomes, the more backflow to central veins in atrial systole and the higher pCVP will be. It is certain that such a circulatory condition after the Fontan operation is not desirable.
Influence on Total Impedance

There is another possible effect of \( C_p \) on postoperative hemodynamics of the Fontan operation. Because after the Fontan operation a single ventricle maintains both systemic and pulmonary circulations, which are located in series without the interposition of a ventricle, both resistance and compliance of the pulmonary artery are directly related to total resistance \( (R_t) \) and total compliance \( (C_t) \) of the single vessel continuing from the aorta to the pulmonary artery. \( R_t \) and \( C_t \) also play important roles as the determinant factors of input impedance of the single vessel continuing from the aorta to the pulmonary artery. This impedance is referred to as total impedance \( (Z_t) \). \( Z_t \) is the afterload to the single ventricle that undertakes both systemic and pulmonary circulation.

\( R_t \) and \( C_t \) can be expressed as Formula 4:

\[
R_t = R_s + R_p, \quad 1/C_t = 1/C_s + 1/C_p
\]

where \( R_s \) is vascular resistance from the aorta to the central veins and \( C_s \) is vascular compliance from the aorta to the central veins.

\( Z_t \) is expressed as Formula 5 by using \( R_t \) and \( C_t \):

\[
Z_t = \frac{R_t}{(1 + \omega^2 R_t^2 C_t)^{1/2}}
\]

\[
\omega = \tan^{-1} \frac{R_p}{R_t}
\]

On the basis of Formula 5, Fig 8 shows the change of \( Z_t \), at \( n = 0, 1, 10 \) when \( C_p \) changes variously at heart rate of 120 beats per minute. \( R_p, R_s, \) and \( C_s \) were substituted by 0.12 mm Hg sec \( m^2/\text{mL} \) \((2 \text{ RU} \cdot \text{m}^2)\), 1.2 mm Hg sec \( m^2/\text{mL} \) \((20 \text{ RU} \cdot \text{m}^2)\), and 1.0 mL/sec mm Hg as the normal values, respectively.32 Furthermore, Fig 9 shows the change of \( Z_t \), when \( PAI \) replaces \( C_p \) according to the regression analysis in Fig 3. \( Z_t \), starts to rise sharply when \( C_p \) is about 0.2 to 0.4 mL/sec mm Hg and \( PAI \) about 71 to 122 mm\(^2/\text{m}^2\). Considering the first three harmonics as the most decisive ones, \( Z_t \) starts to increase abruptly when \( C_p \) is about 0.3 to 0.4 mL/sec mm Hg and \( PAI \) about 97 to 122 mm\(^2/\text{m}^2\). These results approximately agree with those presented by Fontan et al.24 They reported that the risk of death or takedown increased abruptly when the size of pulmonary artery decreased beyond a certain point. It is easy to understand that the increase in \( Z_t \) results from smaller pulmonary artery size should cause the unstable hemodynamics after the Fontan operation, associating with the increase in the afterload to the single ventricle.

As mentioned above, pulmonary artery size is closely related to pulmonary vascular compliance, which is one of the physiological properties of the pulmonary vascular bed, and it postoperatively influences both pCVP and total input impedance, which is the afterload to the single ventricle. The smaller the pulmonary artery size becomes, the greater the increase both in peak CVP and total input impedance and the greater the disadvantages to hemodynamics will be.

Although we cannot definitively set the cut-off line, it seems that postoperative hemodynamics of the Fontan operation become less stable when \( PAI \) is \(<100 \text{ mm}^2/\text{m}^2\) in TCPC. In APC, the margin will be somewhat higher because of the additional effect of high pCVP. On the contrary, the influence of \( PAI \) cannot be found when it is around 250 mm\(^2/\text{m}^2\), which was proposed as the cut-off line by Nakata et al.13

The controversy over the significance of pulmonary artery size in the Fontan operation appears to be an argument about where the cut-off line should be set.

We confirm the necessity of adequate size of the pulmonary artery. However, we should not exclude patients as candidates only because the \( PAI \) of each is \(<250 \text{ mm}^2/\text{m}^2\). We must select patients for the Fontan operation on the grounds that a smaller pulmonary artery causes more disadvantageous hemodynamics after the operation (associated with high pCVP and increase in the afterload to the single ventricle) and with
consideration of other conditions, especially ventricular function and pulmonary vascular resistance.

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References

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