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 patient selection 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. To clarify whether or not pulmonary artery size is indispensable as one of the selection criteria for the Fontan operation, we considered the physiological importance of pulmonary artery size and investigated how pulmonary artery size 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 ($r=0.71, P=0.001$). Furthermore, Cp influenced postoperative hemodynamics of the Fontan operation by affecting peak central venous pressure and total impedance, which was the afterload to the ventricle. Impedance increased abruptly when PAI was less than $100 \text{ mm}^2/\text{m}^2$.

Conclusions The smaller pulmonary artery size is hemodynamically disadvantageous after the Fontan operation, with resultant rise in peak central venous pressure and increased afterload to the single ventricle. (Circulation. 1994;89:266-271.)

Key Words • hemodynamics • pressure

Since Fontan and Baudet first described a new surgical procedure for the repair of 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. Because this operation is not anatomic but a functional repair in which both pulmonary and systemic circulations must be maintained fundamentally by a single pump, patient selection is important in operative outcome. A number of original criteria for patient selection were suggested by Choussat, Fontan, and colleagues. Among these various criteria, the size of the pulmonary artery has been said to be one of the most important factors influencing the results of the Fontan operation. Several quantitative indicators such as the McGoon ratio or pulmonary artery index (PAI) have been recommended for the evaluation of size and development of the pulmonary artery. There have been several reports, however, which suggest that pulmonary artery size was not related to the outcome of the Fontan operation.

In the light of physiology, pulmonary circulation can be replaced by the electrical circuit shown in Fig 1. Inductance is negligible, as we consider blood flow to be steady. According to this analogy, the primary determinants 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 importance 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 not by Q but only by resistance and compliance. Pulmonary artery size (Q) itself is the product of pulmonary blood flow, compliance, and resistance as shown by the formula $Q=\frac{iR}{Cp}$. It is then apparent that the size of the pulmonary artery never primarily predicts the postoperative Fontan circulation.

When we consider the pulmonary circulation in a living body, however, we must also take into consideration the secondary effect of pulmonary artery size on the pulmonary circulation. 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 patients (age, 5 months to 18 years; mean, 7 years) were examined for the analysis mentioned below. Among the patients were those who had mildly increasing pulmonary...
blood flow secondarily as a result of 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 the pulmonary artery. We examined the relations of PAI to Rp and Cp, which are the physiological properties of the pulmonary artery, to reveal the physiological meaning that PAI comprises secondarily. 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 the PAI.13

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 pul-

**Patient Characteristics**

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<td>DORV, PS</td>
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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 with asplenia syndrome were included.
†Blood flow through the pulmonary valve was disregarded because the stenosis was very severe.

**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 a larger PAI. Some cases showed normal resistance with a small PAI, and others showed high resistance with a normal to larger PAI. Even the case of a PAI of 111 mm²/m² kept the normal resistance of 2.1 resistance units · m². On the other hand, significant correlation (r = .71, P = .001) could be found between PAI and Cp (Fig 3). Cp decreased as PAI became smaller.

**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.

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

**Fig 2.** Plot shows relation between pulmonary artery index (PAI) and pulmonary vascular resistance (Rp). No significant correlation was found (r = .21).

**Fig 3.** Plot shows relation between pulmonary artery index (PAI) and pulmonary vascular compliance (Cp). Significant correlation was found (y = 0.149x - 2.579, r = .71, P = .001).
Discussion

It is widely acknowledged that the selection of patients is a key point in determining the outcome of the Fontan operation. Choussat and Fontan\(^4\) presented the first original criteria for patient selection, and it was shown that patients who met these original criteria had good operation results.\(^8\) On the other hand, the patients who exceeded one or more of the criteria in some areas have undergone the Fontan procedure and have had a good outcome. From this viewpoint, the original criteria have been reevaluated\(^19\)\(^-\)\(^23\) and some of them have been suggested to be dispensable, but among these criteria, the importance of pulmonary artery size remains controversial. Nakata et al\(^13\) 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 mm\(^2/m^2\); however, Girod et al\(^4\) reported that there was no significant difference in mortality rate between patients with a PAI >250 mm\(^2/m^2\) and those with a PAI <250 mm\(^2/m^2\). The 188 mm\(^2/m^2\) PAI was reported to be the lowest value among survivors. Bridges et al\(^2\) also showed that there was no statistical difference in PAI between survivors and nonsurvivors after the Fontan operation. In their series, the lowest PAI value among survivors was as low as 48 mm\(^2/m^2\). On the contrary, Fontan et al\(^24\) 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 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, it is possible that pulmonary artery size secondarily predicts the postoperative Fontan circulation by its relation to pulmonary vascular resistance or compliance, whereas the primary predictors of pulmonary circulation are pulmonary vascular resistance and compliance.

In general, resistance of the vessel can be expressed approximately as 8 kL/πr\(^4\), according to the law of Poiseuille,\(^25\) and compliance of the vessel as 3πLr\(^2\)(t/ h+1)/2'E(2t/h+1)\(^26\) (L is length of the vessel; r, radius of the vessel; k, viscosity of blood; E, Young's modulus of the vessel wall; and h, wall thickness of the vessel). As the size of the vessel is expressed as πr\(^2\)L, the radius of the vessel (r) increases, accompanied with the increase in 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 radius of the pulmonary artery.

As shown in Figs 2 and 3, there was no significant correlation between pulmonary artery size and Rp, whereas a 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 Reuben's method (Cp'). As shown in Fig 4, there was a significant correlation between Cp and Cp' (r=.80, P=.001). This indicates the validity of our method.

Furthermore, the Cp value is 46 mm\(^2/m^2\) per mm Hg when the normal value of 330 mm\(^2/m^2\) is assigned to PAI in the correlation equation in Fig 3, and the Cp value, calculated by using the Q=JR equation, is 41 to 55 mm\(^2\)·m\(^{-2}\)·mm Hg\(^{-1}\) when the normal values of 330 mm\(^2/m^2\), 3 to 4 L·min\(^{-1}\)·m\(^{-2}\), and 2 RU·m\(^2\) are assigned to Q, I, and Rp, respectively. This also confirms the validity of our method for calculating the Cp. Regarding the relation between pulmonary artery size and Rp, further analysis will be required for more precise understanding.

When we apply the law of Poiseuille (R=8kL/πr\(^4\), resistance) to the pulmonary vascular bed, Q (pulmonary artery size) is expressed as πr\(^4\)L, so the relation between R and Q is shown as R=8k\(r^2/L^2\). As Q can be supposed to be equivalent to L(αPAI) if PAI reflects the whole size of the pulmonary artery (α, coefficient between PAI and true cross-sectional area of pulmonary artery), R is represented as 8kL/αPAI\(^2\). Fig 5 shows the relation between Rp and kL/PAI\(^2\) in 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 1 when hematocrit is 40%. There was no statistically significant correlation between Rp and kL/PAI, either. This indicates that the difference in radius of proximal arteries does not reflect the difference in radius of the vessel, 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 by a difference in radius of the “resistance vessel.”

In this way, pulmonary artery size, which is 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 the hemodynamics after the Fontan operation because of its relation to compliance.

How, then, 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:

\[ I = i_0 + \sum i_n \sin(\omega_n t) \]

where \( \omega_n = 2\pi f_n \), \( f_n = n \) HR/60

where HR is heart rate, \( n = 1, 2, 3, \ldots \), and \( t \) is length of sequence. A constant term \( i_0 \) represents mean steady flow going through resistance \( R_p \), and the subsequent term represents the pulsatile components of flow. Because the integral calculus of pulsatile components is zero during one cardiac cycle, cardiac output is determined by the constant term. On the other hand, pulmonary artery pressure corresponding to the pulmonary blood flow represented by Formula 1 can be described by using the pulmonary artery input impedance \( Z_n(\theta_k) \) as the following:

\[ P = p_0 + \sum Z_i \sin(\omega_i t + \theta_k) \]

where \( P \) is pulmonary artery pressure, \( Z_n \) is impedance modulus of the nth harmonic, and \( \theta_k \) is the 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), that is, \( p_0 = i_0 R_p \). The second term represents the pulsatile components of pressure, which are determined by impedance and pulsatile components of flow.

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). As one of the determinant factors of impedance, \( C_p \) can influence 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 is impedance of the single vessel continuing from the aorta to the 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 \( p_0 (i_0 R_p) \) in Formula 2. On the other hand, pCVP is affected by peak PAP, which varies according to pulmonary artery input impedance. As \( C_p \) is 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 disregarded. Thus, in total cavopulmonary connection (TCPC), the influence of \( C_p \) on pCVP is negligible. On the other hand, in the right atrial to pulmonary artery connection (APC) of the Fontan-type repair, \( C_p \) influences pCVP according to Formula 2 because of pulsatility caused by the atrial contraction. By substituting 0.12 mm Hg · sec⁻¹ · m⁻² · mL⁻¹ (2 RU/m²) for \( R_p \) as the normal value, the relation between \( C_p \) and \( Z_n \) at a heart rate of 120 beats per minute is represented by Formula 3:

\[ Z_n = 0.12/(1 + a_n^2 n^2 C_p^2)^{1/2} \]

where \( a = 0.2304 \)

Because the first several harmonics at the lower frequency domain mostly decide the actual pressure waveform, consideration up to the 10th harmonic is sufficient for satisfactory analysis. From the first to 10th harmonics 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 standard case at a heart rate of 120 beats per minute when \( C_p \) showed a normal value of 1.5 mL · m⁻² · mm Hg⁻¹. Because we assumed that
Fig 7. Curves show the change of peak central venous pressure (pCVP) according to the change of pulmonary vascular compliance (Cp). Heart rate (HR) was set at 120 beats per minute. The first harmonic and harmonics up to the fifth were evaluated. Curve 1 represents the change of pCVP when only the first harmonic is taken into consideration and curve 2 when the harmonics up to the fifth are done. pCVP ranges from 21.8 to 22.4 mm Hg when Cp is 0.75 mL·m⁻²·mm Hg⁻¹ and from 23.8 to 25.4 mm Hg when Cp is 0.5 mL·m⁻²·mm Hg⁻¹.

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 Cp decreases from normal to half (0.75) and one third (0.5) of normal, respectively. If the Cp decreases further, pCVP will also increase further. In this way, Cp has an influence on pCVP after the Fontan operation. Because PAI is related to Cp (as shown in Fig 3), PAI also affects pCVP in the same way. Although it is unclear how high pCVP is tolerable, the smaller Cp 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 Cp on postoperative hemodynamics of the Fontan operation. Because after the Fontan operation a single ventricle maintains both systemic and pulmonary circulation, which are located in series without the interposition of a ventricle, resistance and compliance of the pulmonary artery are directly related to total resistance (Rt) and total compliance (Ct) of the single vessel continuing from the aorta to the pulmonary artery. Rt and Ct also play important roles as 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 (Zt). Zt is the afterload to the single ventricle that undertakes both systemic and pulmonary circulation. Rt and Ct can be expressed as Formula 4:

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

where Rs is vascular resistance from the aorta to the central veins, and Cs is vascular compliance from the aorta to the central veins. Zt is expressed as Formula 5 by using Rt and Ct:

\[
Z_{t,n} = R_t/(1 + \omega_n^2 R_t C_t^2)^{1/2}
\]

\[
\theta_n = \tan^{-1} \omega_n R_t C_t
\]

On the basis of Formula 5, Fig 8 shows the change of Ztn at n of 1, 3, 10 when Cp changes variously at a heart rate of 120 beats per minute. Rp, Rs, and Cs were substituted by 0.12 mm Hg·sec⁻¹·m⁻²·mL⁻¹ (2 RU/m²), 1.2 mm Hg·sec⁻¹·m⁻³·mL⁻¹ (20 RU/m²), and 1.0 mL·m⁻²·mm Hg⁻¹ as the normal values, respectively. Furthermore, Fig 9 shows the change of Ztn when PAI replaces Cp, according to the regression analysis in Fig 3. Zt starts to rise sharply when Cp is about 0.2 to 0.4 mL·m⁻²·mm Hg⁻¹ and PAI about 71 to 122 mm²/m². Considering the first three harmonics as the most decisive ones, Zt starts to increase abruptly when Cp is about 0.3 to 0.4 mL·m⁻²·mm Hg⁻¹ and PAI about 97 to 122 mm²/m². These results approximately agree with those presented by Fontan et al, who reported that the risk of death or takedown increased abruptly when the size of the pulmonary artery decreased beyond a certain point. It is understandable that the increase in Ztn resulting from the smaller pulmonary artery size causes unstable hemodynamics after the Fontan operation associated 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.

Although we cannot definitely set the cutoff line, it appears that postoperative hemodynamics of the Fontan operation become less stable when PAI is less than...
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