In Vivo Evaluation of Fontan Pathway Flow Dynamics by Multidimensional Phase-Velocity Magnetic Resonance Imaging

Eliezer Be’eri, MD; Stephan E. Maier, MD, PhD; Michael J. Landzberg, MD; Taylor Chung, MD; Tal Geva, MD

Background—Hemodynamic efficiency of Fontan circulation is believed to be a major determinant of outcome. Prior research on flow dynamics in different modifications of Fontan circulation used in vitro models and computer-based simulation. This study was designed to compare in vivo flow dynamics in the systemic venous pathway between patients with atriopulmonary anastomosis (APA) and those with total cavopulmonary connection (TCPC).

Methods and Results—Multidimensional phase-velocity magnetic resonance imaging (PV-MRI) studies were performed on 10 patients who had undergone a modified Fontan operation (5 with TCPC and 5 with APA) and were free of symptoms. The groups were comparable in terms of age and body surface area. The interval since surgery was longer for APA than for TCPC subjects. In each subject, the phase-velocity data sets were used to generate dynamic velocity-vector maps and to calculate quantitative flow indices describing the 3-dimensional blood-flow patterns throughout the cardiac cycle at the widest diameter of the Fontan pathway. Mean flow rate was comparable between groups. Velocity-vector maps showed areas of flow reversal, flow stagnation, and circular flow within APA but not TCPC pathways. Analysis of quantitative flow indices showed that compared with the APA group, flow velocities in the TCPC patients were significantly higher (mean velocity, 14±6 cm/s versus 5±3 cm/s; P=0.02), less variable (coefficient of variation, 19±2% versus 37±3.5%; P<0.0001), and more unidirectional (degree of unidirectionality, 89±7% versus 71±12%; P=0.03). APA pathways were significantly more dilated than were TCPC pathways (P<0.01) and showed a trend toward larger diameter with increased interval since surgery (R²=0.6, P=0.09). Fontan pathway dilatation correlated with flow velocity variability (R²=0.57, P=0.01) and inversely with flow unidirectionality (R²=0.75, P=0.001).

Conclusions—Blood flow patterns are more organized and uniform in TCPC than in APA pathways and are significantly influenced by pathway diameter. We speculate that TCPC may result in a more hemodynamically efficient circulation than APA because of differences in pathway dimension and uniformity. (Circulation. 1998;98:2873-2882.)

Key Words: magnetic resonance imaging ■ blood flow ■ hemodynamics ■ Fontan procedure ■ heart defects, congenital

Since its inception in the early 1970s, the Fontan operation and its modifications have been used for a wide spectrum of congenital heart disorders in which a biventricular repair cannot be achieved.1,2 It has been speculated during the past decade that the hydrodynamic characteristics and energy efficiency of the Fontan circulation influence the medium- and long-term outcomes of these patients.3 Several reports indicated that progressive right atrial dilatation may lead to flow stagnation, thrombus formation, supraventricular arrhythmias, exercise intolerance, and low cardiac output.4,5 de Leval et al3 recognized the importance of minimizing energy losses across the Fontan pathway. They hypothesized that in a Fontan circuit with an atriopulmonary anastomosis (APA), disorganized, and thus energy-inefficient, venous flow patterns occur because of the variations in pathway diameter inherent to that surgical technique. Thus, Puga et al6 and de Leval et al7 advocated the creation of a total cavopulmonary connection (TCPC) by means of a prosthetic intra-atrial baffle of uniform diameter, postulating that this would result in a more streamlined, and thus energy-efficient, blood-flow pattern. To date, however, nonrandomized clinical studies comparing outcome for APA versus TCPC have yielded equivocal results.5,6 Recent studies performed on in vitro models7 and computer-based simulations8 demonstrated that energy loss is greater and flow is more disturbed in Fontan circulation with APA compared with TCPC. These in vitro studies, however, may not accurately reflect in vivo conditions, and any differences between APA and TCPC pathways in this regard still remain unknown.
Figure 1. Methodology of PV-MRI velocity-vector mapping. A, Systolic frame of a magnitude image obtained in sagittal plane in a healthy volunteer. This 6-mm–thick image depicts the right atrium (RA), superior vena cava (SVC), inferior vena cava (IVC), and cross-sections of the right pulmonary artery (RPA) and right upper pulmonary vein (RUPV). The azygous vein (Azyg. V.) is seen entering the superior vena cava. B, ROI is then defined (red line). For the purpose of quantitative analysis, another ROI is defined at the widest diameter of the right atrium by 2 horizontal lines separated from each other by a vertical distance of 10 cm. Because vertical (y axis) distance is set at 10 cm and depth (z axis) is standardized at 6 mm, the contained volume of ROI is dictated by the width (x axis) of the right atrium. C, Systolic frame of velocity-vector mapping within ROI. Each vector represents the sum of 3×3 voxels. The origin of the line-vector is indicated by a dot located at the center of the relevant group of voxels. Length of line vector is proportional to magnitude of instantaneous in-plane blood-flow velocity. Background color represents a speed map. D, Flow velocity–vector map with a z-axis velocity map. z-axis (through plane) velocity component for each group of voxels is depicted as a background color scale. Gray represents zero velocity, progressively darker red indicates progressively higher instantaneous velocities out of the plane (ie, toward the viewer) and progressively darker blue indicates progressively higher instantaneous velocities into the plane (ie, away from the viewer). For any data set, 24 images per cardiac cycle are obtained and can be viewed in a cine loop mode.
Phase-velocity magnetic resonance imaging (PV-MRI) has recently been shown to be capable of noninvasively depicting flow direction and measuring flow velocity in 3 dimensions throughout the cardiac cycle.\textsuperscript{9,10,14–16} It has also been shown to allow for accurate site-specific quantification of flow rate.\textsuperscript{9,10,14–16} The present study, therefore, was designed to analyze and compare the in vivo 3-dimensional blood-flow patterns in APA and TCPC Fontan pathways by the use of PV-MRI.

**Methods**

**Patients**

Patients who had previously undergone a Fontan-type operation were recruited from the outpatient clinic of the Boston Adult Congenital Heart program at Children’s Hospital to undergo a voluntary MRI scan. Patients were excluded from the study if they had any contraindication to the performance of an MRI. The study protocol was approved by the institutional review boards of Children’s Hospital and Brigham and Women’s Hospital in Boston. Informed consent was obtained from each patient.

**MRI Protocol**

MRI was performed on a standard SIGNA 1.5-T whole-body unit with software release 5.4 (General Electric). A wrap-around radiofrequency coil was used in all patients. Prospectively ECG-gated TI-weighted spin-echo images in coronal, transverse, sagittal, and, when necessary, oblique planes were obtained so as to image the entire Fontan pathway and to locate appropriate planes for subsequent velocity mapping (echo time = 20 ms, repetition time = R-R interval, slice thickness = 5 to 8 mm, image matrix size = 256 x 128, field of view = 24 to 40 cm, number of excitations = 2 to 4). Retrospectively ECG-gated phase-velocity images, encoded for flow in the x-, y-, and z-axis directions were then acquired in coronal, sagittal, or oblique planes, and were intended to include the caval, atrial, and pulmonary components of the Fontan pathway in the imaging planes (echo time = 13 ms, repetition time = 24 ms, image matrix = 256 x 128, 20° flip angle, 2 excitations per acquisition, 24 images per cardiac cycle). Respiratory compensation was used to minimize motion artifacts. The scan time for a single location was ~2 to 4 minutes, depending on the patient’s heart rate. No sedation or contrast material was used in this study.

**MRI Data Processing**

Flow quantification and analysis were performed off line on a SUN workstation with customized software (XPhase) developed by one of the authors (Dr Maier). The accuracy of the system for measuring flow rate was validated by our group.\textsuperscript{14} This dedicated MRI flow analysis software automatically correlates the x-, y-, and z-axis velocity-vector components of each pixel of a 3-dimensional MRI phase-velocity data set. The instantaneous x- and y-axis velocity vectors are resolved into a single “in-plane” velocity vector, which is depicted as a line vector on a velocity-vector map. The origin of the line vector is indicated by a dot located at the center of the relevant pixel. The length of the line vector is proportional to the magnitude of the instantaneous in-plane blood-flow velocity at that pixel, and the direction of the line vector indicates the instantaneous direction of in-plane flow (Figure 1A through 1D). The z-axis (through plane) velocity component for each pixel is depicted as a background color scale (Figure 1D). Gray represents zero velocity, progressively darker red indicates progressively higher instantaneous velocities out of the plane, and progressively darker blue indicates progressively higher instantaneous velocities into the plane. By superimposing the x-y flow-vector map on a z-axis color scale, a depiction of instantaneous 3-dimensional flow is achieved. For each cardiac cycle, 24 sequential instantaneous flow-vector maps are generated from each phase-velocity data set. Both instantaneous and dynamic flow patterns can be reviewed by either displaying each frame individually or by viewing the 24 frames as a continuous cine loop. This results in the depiction of blood flow in terms of 5 dimensions: x, y, and z spatial axes; velocity; and time.

Because the amount of flow data generated by each vector map was substantial, the mean velocity of each 2x2 or 3x3 group of pixels was represented by a single line vector. This facilitated graphic representation and data interpretation. In addition, we found that when we analyzed flow patterns in which the z-axis component was relatively static, overall flow analysis was facilitated by the use of a background color scale depicting the speed of blood, rather than the z-axis velocity component (Figure 1C). Speed, as opposed to velocity, is a scalar quantity possessing magnitude but not direction. It is derived from the x, y, and z velocity-vector components by the following formula:

$$\text{Speed} = \sqrt{x^2 + y^2 + z^2}$$

**Qualitative Analysis of Flow Patterns**

The qualitative nature of 3-dimensional blood flow through the Fontan pathways was assessed by inspecting the flow-vector maps.

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**TABLE 1. Characteristics of Study Patients**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Diagnosis</th>
<th>Pathway Type</th>
<th>Age, y</th>
<th>Gender</th>
<th>BSA, m²</th>
<th>Interval Since Surgery, y</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>(S,D,D)</td>
<td>Fen TCPC</td>
<td>25</td>
<td>M</td>
<td>1.82</td>
<td>3.6</td>
</tr>
<tr>
<td>2</td>
<td>(S,LL)</td>
<td>Fen TCPC*</td>
<td>39</td>
<td>M</td>
<td>1.81</td>
<td>0.2</td>
</tr>
<tr>
<td>3</td>
<td>(S,LL)</td>
<td>Fen TCPC</td>
<td>43</td>
<td>F</td>
<td>1.64</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>(S,D,D)</td>
<td>TCPC</td>
<td>16</td>
<td>M</td>
<td>2.10</td>
<td>3</td>
</tr>
<tr>
<td>5</td>
<td>(S,LL)</td>
<td>Fen TCPC</td>
<td>12</td>
<td>F</td>
<td>1.44</td>
<td>1</td>
</tr>
</tbody>
</table>

Mean±SD

<table>
<thead>
<tr>
<th>Mean±SD</th>
<th>Age, y</th>
<th>BSA, m²</th>
<th>Interval Since Surgery, y</th>
</tr>
</thead>
<tbody>
<tr>
<td>27±12</td>
<td>1.76±0.22</td>
<td>1.8±1.2</td>
<td></td>
</tr>
</tbody>
</table>

*Extracardiac conduit from inferior vena cava to superior vena cava; †, extracardiac conduit from right atrium to left pulmonary artery. AS, aortic stenosis; BSA, body surface area; DORV, double outlet right ventricle; DI-DORV, double inlet-double outlet right ventricle; DILV, double inlet left ventricle; Fen, fenestrated; PS, pulmonary stenosis; Tri, tricuspid valve; VSD, ventricular septal defect.
The 24 instantaneous maps comprising 1 cardiac cycle were inspected frame by frame to identify the following flow patterns: (1) laminar flow (characterized by parallel orientation of the velocity vectors throughout the vessel or pathway), (2) disorganized flow (characterized by velocity vectors that are simultaneously oriented in different spatial directions), (3) circular flow (characterized by a circular flow pattern within the pathway or vessel as seen by dynamic vector mapping), and (4) stagnant flow (characterized by velocity vectors \( \leq 5 \text{ cm/s} \)). By viewing the 24 frames as a cine loop, the dynamic nature of the blood-flow patterns could be appreciated.

Quantitative Analysis of Flow Patterns

To facilitate meaningful statistical analysis of differences in flow patterns between TCPC and APA pathways, we measured flow through a standardized region of interest (ROI) in the center of each Fontan pathway. The ROI was defined at the widest mid-right atrial diameter, encompassing the entire width of the pathway between 2 parallel horizontal lines separated by a vertical (or superior-inferior) distance of 10 cm (Figure 1B). Because the \( z \)-axis dimension (ie, the imaging slice thickness) of the ROI was standardized at 6 mm in each case and the \( y \)-axis dimension was standardized at 10 cm, the ROI represented a 3-dimensional volume in the mid-Fontan pathway whose volume varied from patient to patient depending on the degree of pathway dilation (ie, variation in the \( x \)-axis dimension of the ROI). In TCPC patients with baffle fenestration, the ROI included the systemic venous aspect of the fenestration. To determine interobserver variability in determining the ROI, it was traced by 2 investigators who were unaware of each other’s measurements. Simple linear regression analysis was used to calculate the correlation of measurements by the 2 observers. The absolute difference between observers’ measurements was divided by the mean value of measurements and expressed as a percentage. Interobserver variability was expressed as the mean \( \pm SD \) of these percentages.

The mean, maximum, and minimum flow velocities through the entire ROI of each Fontan pathway during the course of the cardiac cycle were computed from the relevant \( x \)-, \( y \)-, and \( z \)-axis phase-velocity data sets. Instantaneous flow rates were calculated for each of the 24 phase-velocity maps by multiplying the mean flow velocity by the cross-sectional area of the pathway. Flow volume was then computed by integrating the flow rates throughout the cardiac cycle. Flow within the ROI was then analyzed in terms of 3 parameters that quantify the uniformity of flow velocity and direction.

1. Temporal coefficient of variation (CV) of flow speed: To quantitatively express the degree of variation in flow speed throughout the cardiac cycle, the mean instantaneous flow speed through the ROI for each of the 24 flow maps (comprising 1 cardiac cycle) was determined. Subsequently, the mean and SD for the 24 resulting data points were computed and the CV of flow speed was derived and expressed as a percentage by use of the following formula:

\[
\text{COV} = \frac{\text{standard deviation}}{\text{mean}} \times 100.
\]

2. Percentage unidirectionality of flow (PUF): To evaluate the degree to which blood flow was unidirectional, the 3-dimensional velocity vectors within the ROI for 1 cardiac cycle were combined into a single antegrade velocity vector (representing all antegrade velocity vectors) and a single retrograde velocity vector (representing all retrograde velocity vectors). The relative magnitudes of the antegrade and retrograde velocity vectors were expressed as the PUF, which was defined as follows:

\[
\text{PUF} = \frac{\text{Antegrade Flow} - \text{Retrograde Flow}}{\text{Antegrade Flow}} \times 100
\]

A PUF of 0% would imply that the amount of retrograde flow during 1 cardiac cycle equals the amount of antegrade flow.

3. Vector angle (VA): In addition to the relative magnitudes of the antegrade and retrograde flow vectors, the angle between the 2 vectors also quantifies an element of flow reversal. For an antegrade-retrograde vector pair with a given PUF, a wider angle between the 2 vectors implies a greater degree of flow reversal during the cardiac cycle than does a narrower angle.

The angle between the antegrade (a) and retrograde (r) flow vectors, defining the VA, was therefore calculated for each ROI by use of the following formula:

\[
\text{VA} = \arccos \left( \frac{(x_a x_a + y_a y_a + z_a z_a)}{(\sqrt{x_a^2 + y_a^2 + z_a^2} \times \sqrt{x_r^2 + y_r^2 + z_r^2})} \right)
\]

where \( x \), \( y \), and \( z \) are the spatial components of each vector.

Statistical Analysis

Data are reported as mean \( \pm SD \) for each group of measurements. A 2-tailed paired Student \( t \) test was used to compare continuous variables between APA and TCPC groups. Potential associations between continuous variables were examined by linear regression analysis. Data analysis was performed with a commercially available statistical package (StatView 4.1, Abacus Concepts Inc). For all tests, \( P \leq 0.05 \) was considered statistically significant.

Results

Patients

Ten adult subjects, 5 with APA and 5 with TCPC, volunteered to participate in the study. Their diagnoses and demographic data are summarized in Table 1. All subjects had successfully undergone a Fontan procedure and were being monitored on an outpatient basis. One of the APAs was accomplished via an extracardiac conduit from the anterior free wall of the right atrium to the left pulmonary artery (patient 8), and the other 4 via anastomosis of the right atrial appendage to the main pulmonary artery. In 4 patients, TCPC was performed by the creation of an intra-atrial baffle connecting the inferior vena cava to the cardiac end of the superior vena cava and an end-to-side anastomosis between the superior vena cava and the right pulmonary artery. In 1 TCPC patient, an extracardiac conduit was placed between the inferior vena cava and the pulmonary arteries (patient 2). In 4 of the TCPC patients, a fenestration had been placed in the conduit/baffle. One subject (patient 7) had recently undergone radiofrequency catheter ablation of an intra-atrial reentrant tachycardia and another subject (patient 8) was receiving diuretic therapy. No patient exhibited signs or symptoms that suggested a failure of the Fontan circuit that might indicate the need for reoperation. There were no significant differences between groups with regard to age, gender, weight, or body surface area. The interval between the date of Fontan surgery and the MRI study was significantly longer for the APA group (11.2 \( \pm 2.2 \) years) than for the TCPC group (1.8 \( \pm 1.2 \) years, \( P < 0.01 \)).

Anatomic Findings

The anatomy of the Fontan pathways and pulmonary arteries was clearly imaged by spin-echo MRI in all subjects and were shown to be unobstructed. Whereas TCPC pathways were characterized by a relatively uniform diameter throughout their length (Figure 2), the diameter of APA pathways varied substantially from the cava to
the pulmonary arteries and was maximal at the midright atrium (Figure 3 and 4). Dilatation of systemic venous pathways in APA patients was not limited to the Fontan pathway but was manifest in the superior and inferior venae cavae of those patients with the most dilated right atria (patients 8 and 10). The average midright atrial diameter for APA pathways was 3 times wider than for TCPC pathways (Table 2). Furthermore, in patients with APA pathways, a trend between larger pathway diameter and longer interval since surgery could be discerned but did not reach statistical significance ($R^2=0.61$, $P=0.09$). No such trend was evident in the TCPC group.
Analysis of Flow Patterns

Inspection of the cine-loop flow vector maps revealed that flow through each of the 5 TCPC pathways was characteristically unidirectional and, at any instant in the cardiac cycle, of relatively uniform velocity throughout the pathway (Figure 2). During the course of the cardiac cycle, flow was biphasically pulsatile, but with a relatively small difference between the maximal and minimal velocities. There were no areas of flow stagnation. In patients 1 to 3 and 5, in whom a baffle fenestration was present, there was streaming toward the fenestration without disturbance of the overall laminar flow pattern. In 4 TCPC patients, flow through the cavopulmonary anastomosis itself was laminar and of a similar velocity to flow through the midbaffle. In these pathways, the cardiac and cephalic ends of the cavopulmonary anastomosis were offset from each other. Accordingly, blood from the superior vena cava streamed preferentially to the right pulmonary artery and blood from the inferior vena cava to the left pulmonary artery, with the 2 oppositely directed streams producing an area of circular flow between them. This was the only circular flow phenomenon noted within TCPC pathways. In the patient with an extracardiac TCPC conduit, a small area of turbulence occurred immediately after the proximal anastomosis where the inferior vena cava and the conduit had a 135° angle between them. There were no other occurrences of turbulent flow within the Fontan pathways in either subgroup.

Figure 3. PV-MRI velocity-vector mapping showing flow pattern in nondilated APA-type Fontan (patient 9). A, Magnitude image in an oblique sagittal plane showing right atrium (RA), SVC and IVC, RPA, and RUPV. B, C, and D, Flow velocity-vector mapping with a background speed map. Note parallel orientation of vectors within the center of right atrium and superior vena cava and the nearly stagnant flow along the anterior right atrial wall. There is no retrograde flow in this patient at any time during the cardiac cycle.
Flow patterns in the APA group were far more varied than those found in the TCPC patients. The APA patient with the narrowest right atrial diameter (patient 9) demonstrated flow through the right atrium that was consistently unidirectional throughout the cardiac cycle. Blood velocity was maximal in the center of the pathway, becoming progressively slower toward the periphery and almost stagnant adjacent to the anterior wall of the right atrium (Figure 3). In contrast to the TCPC pathways, flow accelerated markedly through the APA. The flow velocities in this patient remained relatively constant throughout the cardiac cycle and there were no areas of disorganized or circular flow. In contrast to the flow pattern observed in the narrowest APA pathway, the patient with the widest right atrial diameter (patient 8) exhibited a complex and disorganized pattern of very low–velocity flow (Figure 4). Flow within the markedly dilated right atrium was characterized by large, slow, circular flow swirls that gradually changed their location and velocity during the course of the cardiac cycle. Areas of flow stagnation occurred intermittently at multiple locations within the chamber. Flow at the points of Fontan pathway inflow (the superior and inferior venae cavae) and outflow (the APA) was characterized by periods of flow reversal occurring in mid- to late ventricular systole. The remaining 3 APA patients, each with intermediate degrees of right atrial dilation, exhibited flow patterns intermediate to the 2 described above. An area of transient circular flow within the body of the right atrium occurred in all 3 patients. Transient flow reversal was noted within the inferior vena cava in 1 subject, within the right pulmonary artery in a second, and within the body of the right atrium in the third patient. All 3 exhibited areas of flow stagnation toward the periphery of the

Figure 4. PV-MRI velocity-vector mapping showing flow pattern in dilated APA-type Fontan (patient 8). A, Magnitude image obtained in sagittal plane. B, C, and D, Flow velocity-vector mapping with a background speed map. Note the very low velocities at any location in the right atrium (RA). Inspection of cine loop demonstrates multiple regions of very low velocity swirls with near-zero flow at periphery of dilated right atrium. Flow reversal is evident in SVC and IVC in late diastole (D).
right atrium, and 1 subject demonstrated stagnation of flow within the body of the right atrium.

### Quantitative Flow Analysis

The qualitative observations described above were corroborated by the flow measurements performed within the ROI (Table 2). Although the amount of blood flow through the 2 pathway types was similar (mean flow, 28±7 versus 27±7 mL/s), there were marked differences between APA and TCPC pathways with regard to the nature of the flow. Compared with APA patients, flow velocities in TCPC pathways were 2 to 3 times faster, 49% more uniform (CV, 19±2 versus 37±3.5; \( P<0.0001 \)), consistently more antegrade (PUF, 89±7% versus 71±12%; \( P=0.03 \)), and with a smaller angle between antegrade and retrograde velocity vectors (112±6° versus 135±20°, \( P=0.058 \)). The diameter of the Fontan pathway correlated negatively with mean \( (R^2=0.58, P=0.01) \), maximal \( (R^2=0.5, P=0.02) \), and minimal \( (R^2=0.6, P<0.01) \) flow velocities, and with PUF \( (R^2=0.75, P=0.001) \). A positive linear correlation was found between pathway diameter, CV \( (R^2=0.57, P=0.01) \), and vector angle \( (R^2=0.47, P<0.03) \).

### Interobserver Variability

The mean interobserver variability for ROI measurements was 2.3±7.2% (0.8±2 mm). The correlation coefficient by simple linear regression analysis for measurements by the 2 observers was 0.99 (\( P<0.0001 \)).

### Discussion

The noninvasive application of multidimensional flow imaging by PV-MRI flow vector mapping allowed for in vivo visualization and quantitative analysis of flow dynamics in Fontan pathways. The results of this study demonstrate marked differences in flow patterns between APA and TCPC pathways. Flow through TCPC pathways was characteristically unidirectional and streamlined (as evidenced by a high PUF and a low VA, biphasic, and of relatively high velocity. In contrast, flow through APA pathways was significantly slower, more disorganized, and varied. Except for the narrowest APA (patient 9), flow patterns in dilated APA pathways were characterized by multiple circular, reversed, or stagnant flow systems within the right atrium. Stagnation of blood near the periphery of the right atrium was observed in all APA patients, even in the patient without significant chamber dilatation who exhibited a relatively organized flow pattern at the center of the pathway. The study found evidence of increased right atrial diameter as a function of the interval since surgery in the APA group. It also found strong correlations between indices of impaired flow profiles and pathway diameter.

The results of this study provide in vivo corroboration of the mounting evidence from in vitro and computer simulation studies indicating that flow dynamics in APA is inefficient. de Leval and his colleagues first studied hemodynamic and energy factors in an in vitro model of APA-type Fontan circulation.\(^7\) They found that the hydrodynamic design of the APA was relatively poor and that the presence of a large passive chamber between the systemic veins and the pulmonary circulation was a major cause of flow inefficiency. On the basis of the results of those experiments, they advocated the use of TCPC that was designed to produce a more streamlined flow pattern that should lead to less energy loss. Subsequent in vitro\(^17\)\(^–\)\(^21\) and computational\(^18\)\(^–\)\(^19\) studies confirmed those observations. The effect of progressive right atrial dilatation on energy loss and resistance to flow was recently studied by Lardo and associates\(^20\) in an in vitro flow model of explanted sheep hearts with APA-type Fontan pathways. Their experiment showed that progressive right atrial dilatation is associated with increased energy loss and resistance to flow, which were more pronounced at higher flow rates. Other investigators studied various geometries of
TCPC, attempting to find the most efficient connection. 7,8,18,19,21,22 Although these in vitro and computational experiments provided important information regarding isolated aspects of the Fontan circulation, they have not been validated in vivo.

Energy Considerations

The complex flow patterns observed in dilated APA pathways that consisted of multiple circular, reversed, or stagnant flow systems imply the presence of multiple planes of shear between adjacent fluid flow systems. In the clinical context of Fontan circulation, friction between these layers and at the interface with the walls of the pathway may result in a loss of pressure energy. In an attempt to confirm the hemodynamic advantage of TCPC over APA, several researchers studied the magnitude of energy loss in simulated APA and TCPC circuits. 8,17 Their results, however, predict that the losses of pressure energy due to increased friction would be of very small absolute magnitude (<1 mm Hg for an APA pathway). 8 Our experience, too, has been that in the absence of a significant anatomical stenosis even markedly dilated Fontan pathways do not exhibit clinically measurable pressure gradients across the right atrial chamber and APA at cardiac catheterization. 23 Consideration of the principles of hydraulic energetics in light of the preceding observations regarding Fontan pathway dilation, however, would suggest an alternative explanation for the possible advantage of TCPC over APA. Because total blood volume exerts a profound effect on myocardial energetics, the tendency of APA pathways to dilate over time may, therefore, cause a gradual increase in the myocardial energy expenditure required to maintain normal cardiac output and blood pressure. The possible protection against pathway dilatation afforded by TCPC may thus be of significant long-term benefit in terms of myocardial energy expenditure.

Clinical Implications

The findings of this study provide a link between fluid dynamics and several clinical observations. Progressive right atrial dilation in APA has been linked to exercise intolerance, thrombus formation, atrial arrhythmias, and increased systemic venous pressure and its sequelae. 3,5,24 We found that blood velocities within APA pathways were significantly slower than those in TCPC. In addition, blood velocity was less constant over time and space, manifesting as a significantly higher speed CV for the APA group than for the TCPC group. The dynamic 3-dimensional flow maps confirmed this finding, revealing multiple areas of transient or even constant flow stagnation within the most dilated APA pathways, particularly toward the periphery of the chamber. These flow patterns likely contribute to the substrate that may lead to thrombus formation in some of these patients.

Study Limitations

The APA group was monitored for a significantly longer period after the operation than the TCPC group. This limitation is somewhat offset by the fact that the 2 groups showed no significant differences in terms of demographic variables, body surface area, clinical well-being, and Fontan flow rate. In addition, it is important to note that acquisition of PV-MRI data was not gated to the respiratory cycle and was acquired with patients in the supine position and at rest. Each data set represents an average of multiple heart beats and respiratory cycles. The effects of respiration, body position, and exercise on the observed flow patterns require further study. Although the results of this study demonstrate an association between pathway diameter and flow patterns, it does not establish a cause and effect relationship between these variables. A much larger number of participants is required for such analysis.

Conclusions

PV-MRI mapping of Fontan pathways in a small group of patients demonstrates in vivo that TCPC results in venous flow patterns that are significantly more organized and uniform with higher flow velocities than those occurring in APA pathways. The flow dynamics observed in patients with APA connection is thought to be hemodynamically inefficient and more thrombogenic. The disadvantageous flow characteristics increase with pathway dilatation.

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