Magnetic Resonance and Echocardiographic Imaging of Pulmonary Artery Size Throughout Stages of Fontan Reconstruction

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Background  Because pulmonary artery size is considered by most investigators to be a major prognosticator of outcome in patients undergoing staged Fontan reconstruction, the objective of the present study was to determine the efficacy of noninvasive measures in determining pulmonary artery size.

Methods and Results  This study analyzed the T1-weighted, spin-echo magnetic resonance and echocardiographic images of 36 functional single-ventricle patients throughout stages of Fontan reconstruction (prebidirectional and postbidirectional cavopulmonary anastomosis and after Fontan) and compared them with angiography images at cardiac catheterization. Magnetic resonance imaging had a high degree of agreement with angiography, with the McGoon index agreeing better than the Nakata index and absolute diameters. Although echocardiography had fair agreement with angiography, it agreed less well and had a wider standard deviation than magnetic resonance imaging for all indexes and measurements and, based on the prediction interval, would be a poorer prospective measure of pulmonary artery size in this population. In addition, echocardiography was a poorer measure of pulmonary artery size as the size of the vessel increases. Magnetic resonance imaging correctly detected five of five patients with nonconflu ent branch pulmonary arteries and six of six patients with stenoses, whereas echocardiography was unable to visualize any of the patients with nonconfluent branch pulmonary arteries with certainty and only two of six (33%) with stenoses.

Conclusions  Magnetic resonance imaging is a useful, noninvasive tool to determine pulmonary artery size in patients undergoing Fontan reconstruction and is superior to echocardiography. Echocardiography was a fair predictor of pulmonary artery size, but magnetic resonance imaging agreed with angiography better than echocardiography and outperformed echocardiography in diagnosing branch pulmonary artery discontinuity and stenoses. Magnetic resonance imaging may avoid unnecessary cardiac catheterization, especially in older patients, and may obviate the need for jugular or subclavian catheterization in those who have undergone bidirectional cavopulmonary anastomosis. (Circulation. 1994;90:2927-2936.)

Key Words  Fontan  •  magnetic resonance imaging  •  echocardiography  •  angiography

Since 1971, when Fontan pioneered reconstructive surgery for patients with single-ventricle physiology,1 numerous investigators have sought predictors of successful outcome2-9 through subsequent modifications and application to various congenital heart defects.10-14 Pulmonary artery size, although controversial, appears to be a prognostic indicator.4-8 It may also be used by the surgeon as preoperative information and may ultimately affect the conduct of the surgery if the pulmonary arteries need to be augmented.

The noninvasive assessment of pulmonary artery size would therefore be useful and has been attempted in the past with two-dimensional echocardiography.2,15,16 A rigorous study comparing this modality and magnetic resonance imaging with angiography, however, has not been attempted in this group of patients. Furthermore, since magnetic resonance imaging has found application in a wide variety of congenital heart defects17-20 and in evaluating pulmonary artery size in specific,19,21-29 it may be the preferred method in determining pulmonary artery size with a high degree of accuracy.

The purpose of this study, therefore, was to evaluate the efficacy of magnetic resonance and echocardiographic imaging in the determination of pulmonary artery size in patients with single-ventricle physiology throughout stages of Fontan reconstruction.

Methods

Patients

We prospectively studied 36 consecutive patients with a functional single ventricle who had all of the following—cardiac catheterization, echocardiography, and cardiac magnetic resonance imaging—who presented to The Children’s Hospital of Philadelphia between April 1, 1991, and February 28, 1992. The patients’ ages ranged from 0.4 to 237 months (average, 32.9±49.1 months; median, 14.3 months), median body surface area was 0.44 m² (range, 0.17 to 1.5 m²), and median weight was 9.7 kg (range, 2.5 to 30.1 kg). Informed consent was obtained from all participants. The human investigations committee at The Children’s Hospital of Philadelphia approved the study protocol on December 20, 1990. The exclusion criteria for the study was not being stable enough to undergo a 1-hour magnetic resonance scan under sedation, the
TABLE 1. Types of Functional Single Ventricles and Breakdown by Surgical Subgroups

<table>
<thead>
<tr>
<th>Congenital Heart Lesion</th>
<th>Total</th>
<th>Before Hemi-Fontan</th>
<th>After Hemi-Fontan</th>
<th>After Fontan</th>
</tr>
</thead>
<tbody>
<tr>
<td>HLHS</td>
<td>23</td>
<td>11</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>DORV</td>
<td>5</td>
<td>3</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>1. (S,D,D), MA, subPS, Dbl Ao arch, LSVC→CS, S/P bilat hemi-Fontan</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. (S,D,L), dextro, S-I vent, CC-AVW, hypoplastic RAVV, subPS, S/P Fontan</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. (S,D,D), MA, subPS, unroofed CS, unoperated</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. (S,D,D), valvar and subvalvar PS, hypoplastic RV, unoperated</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. (S,D,D), subpulmonary VSD, muscular VSDs, straddling mitral valve, unoperated</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Tricuspid atresia
1. (S,D,D), TGA, LJAA, multiple muscular VSDs, ASD, S/P hemi-Fontan
2. (S,D,S), S/P Fontan
3. (S,D,S), PS, aneurysm of septum primum, S/P BTS
4. (S,D,S), tricuspid stenosis, LAD aneurysm, LAD→RV fistula, S/P hemi-Fontan
5. (S,D,S), S/P Fontan

PA/IVS
1. (S,D,S), isolated ventricular inversion

TGA
1. (S,D,D), mitral hypoplasia, subPS, valvar PS, LSVC→CS, multiple muscular VSDs, ASD, S/P Fontan

Isolated ventricular inversion
1. (S,L,S), subPS, left atrioventricular valve atresia, S/P BTS×2

Single left ventricle
1. (S,LX), RV aorta with pulmonary atresia, S/P Fontan

ASD indicates ostium secundum atrial septal defect; BTS, modified Blalock-Taussig shunt; CC-AVW, criss-cross atrioventricular relations; CS, coronary sinus; Dbl Ao arch, double aorta arch; dextro, dextrocardia; DORV, double-outlet right ventricle; HLHS, hypoplastic left heart syndrome; LAD, left anterior descending coronary artery; LJAA, left juxtaposition of the atrial appendages; LSVC→CS, left superior vena cava to coronary sinus; MA, mitral atresia; PA/IVS, pulmonary atresia with intact ventricular septum; PS, pulmonic stenosis; RAVV, right atrioventricular valve; RV, right ventricle; {S,D,S}, situs solitus of the viscera and atria, ventricular D loop, solitus normally aligned great arteries; {S,D,D}, situs solitus of the viscera and atria, ventricular D loop, D transposed or malposed great arteries (ie, aortic valve to the right of the pulmonic valve); {S,D,L}, situs solitus of the viscera and atria, ventricular D loop, L transposed or malposed great arteries (ie, aortic valve to the left of the pulmonic valve); S/P, status post; subPS, subvalvar pulmonic stenosis; S-I vent, supraventricular aorta, TGA, transposition of the great arteries; and VSD, ventricular septal defect.

The presence of a pacemaker, or arrhythmias. No exclusion criteria were encountered.

The anatomic features of the 36 study patients are summarized in Table 1. Twenty-three (64%) had hypoplastic left heart syndrome, 5 (14%) had double-outlet right ventricle (with an atrioventricular valve either markedly hypoplastic or atretic and pulmonic stenosis), 3 (8%) had tricuspid atresia, 2 (6%) had pulmonary atresia with intact ventricular septum, 1 (3%) had isolated ventricular inversion with left atrioventricular valve atresia, 1 (3%) had single left ventricle with right ventricular outlet chamber and pulmonary atresia, and 1 (3%) had transposition of the great arteries with mitral and pulmonic hypoplasia. The breakdown by surgical subgroup consisted of 16 (44%) prior to having bilateral cavopulmonary anastomosis (ie, hemi-Fontan procedure—superior vena cava–to–pulmonary artery anastomosis with right atrial exclusion), 11 (31%) who had undergone a hemi-Fontan procedure but had not undergone Fontan completion (baffling inferior vena cava blood flow to the pulmonary artery), and 9 (25%) who had completed the modified Fontan reconstruction. Table 2 gives study patients by surgical subgroup tabulating age, body surface area, height, and weight.

TABLE 2. Study Patients With Single Ventricles Divided by Surgical Subgroup, Age, Weight, Height, and Body Surface Area

<table>
<thead>
<tr>
<th>Surgical Subgroup</th>
<th>No. of Patients</th>
<th>Age, Mean±SD mo (Median)</th>
<th>Weight, kg</th>
<th>Height, cm</th>
<th>Body Surface Area, m²</th>
</tr>
</thead>
<tbody>
<tr>
<td>Before hemi-Fontan</td>
<td>16</td>
<td>5.7±2.0 (6)</td>
<td>6.1±1.7</td>
<td>66.3±8.0</td>
<td>0.33±0.07</td>
</tr>
<tr>
<td>After hemi-Fontan</td>
<td>11</td>
<td>38.2±42.4 (22)</td>
<td>12.6±4.9</td>
<td>89.1±19.4</td>
<td>0.55±0.17</td>
</tr>
<tr>
<td>Fontan</td>
<td>9</td>
<td>74.6±69.4 (37)</td>
<td>20.9±15.5</td>
<td>104.9±24.6</td>
<td>0.76±0.35</td>
</tr>
<tr>
<td>Total</td>
<td>36</td>
<td>32.9±49.1 (14)</td>
<td>11.8±10.0</td>
<td>82.9±23.2</td>
<td>0.50±0.26</td>
</tr>
</tbody>
</table>

Values are mean±SD.
Angiography

All patients were sedated before imaging with 4 mg/kg nembutal PO and 3 mg/kg demerol PO. Additional sedatives, such as medazolam, were given as needed. The posteroanterior angiogram was used as a comparison for all noninvasive data (Fig 1). Injections were made either directly into the pulmonary arteries or into vessels just proximal to pulmonary artery flow (ie, ascending aorta in patients who are status post–stage I Norwood reconstruction, antecubital or subclavian vein in patients who are status post–hemi-Fontan, and so on). Cineangiograms were recorded on standard 35-mm film, and measurements were made using a Vangard XR-35 viewing system (Vangard Instrument Corp), using catheter diameter as an internal standard. Measurements of pulmonary artery size were taken along the most uniform part of the vessel just proximal to the first lobar branch. Discontinuities and stenoses were identified and recorded. The resolution is 0.7 mm (2.7 line pairs per millimeter). Because the diameter of the great vessels changes during the cardiac cycle, the dimensions at end systole and end diastole were averaged to obtain the angiographic value.

Echocardiography

Before imaging, all patients less than 3 years old were sedated with 75 to 120 mg/kg chloral hydrate PO. If more than 3 years old, depending on the ability to cooperate with the examination, the patient either was or was not sedated with chloral hydrate as described. Echocardiograms were performed within 2 weeks before cardiac catheterization. A comprehensive two-dimensional echocardiographic examination was performed on each patient, and measurements were made from leading edge to leading edge using electronic calipers from the suprasternal notch frontal position in the case of the right pulmonary artery, the suprasternal notch left anterior oblique view angling to the left in the case of the left pulmonary artery, or the high parasternal positions (Fig 2). Measurements of pulmonary artery size were taken along the most uniform part of the vessel just proximal to the first lobar branch. Hewlett Packard 77020 phased-array ultrasound systems (Sonos 500 and 1000) with 5.0-, 3.5-, or 2.5-MHz transducers were used. All studies were recorded on 1/2-in VHS format videotapes and were available for retrospective off-line analysis. Because the suprasternal notch views were used, measurement of pulmonary artery size was a function of the axial resolution of the two-dimensional image. The axial resolutions for the 5-, 3.5-, and 2.5-MHz transducers were 0.7, 0.9, and 1.1 mm, respectively. Because the diameter of the great vessels changes during the cardiac cycle, the dimensions in end systole and end diastole were averaged to obtain the echocardiographic value.

Color flow mapping was not used in the calculations of pulmonary artery size because of its poor spatial resolution (decreased line density would preclude accurate size measurements); however, it was used in the determination of branch pulmonary artery discontinuity.

Magnetic Resonance Imaging

All patients were sedated before imaging. If less than 2 years old, the patient was given either 75 to 120 mg/kg chloral hydrate PO or 2 to 6 mg/kg nembutal IV. If more than 2 years old, either 4 mg/kg nembutal PO and 3 mg/kg demerol PO or 2 to 6 mg/kg nembutal IV was administered. All patients were monitored with pulse oxymetry, nasal end-tidal CO2 monitoring, ECG, and direct visualization via closed-circuit television. All patients tolerated sedation without incident. All studies were performed on a Siemens 1.5-T Magnetom. The scanning protocol consisted of a coronal localizer (to locate the heart in the chest) followed by two sets of interleafed, T1-weighted transverse images (Fig 3) 3 to 6 mm thick, in which the distance between slices of each set of image acquisitions equaled the thickness of the slice (ie, distance
between slice centers=2×slice thickness), resulting in contiguous slices. The effective repetition time (TR) was the RR interval in milliseconds (range, 350 to 800 milliseconds), with echo time (TE) equal to 15 milliseconds, and the number of excitations equal to three (image, 128×256 pixels). This was used to clinically evaluate the entire anatomy of the cardiovascular system in the thorax.

Images containing the branch pulmonary arteries were identified, and measurements were made from leading edge to leading edge (Fig 3). Measurements of pulmonary artery size were taken along the most uniform part of the vessel just proximal to the first lobar branch. Data were then converted from pixel length to millimeters by a factor determined by the field of view and matrix size. The resolution was 1 mm.

Because magnetic resonance imaging obtains contiguous, parallel slices, the diagnosis of pulmonary artery discontinuity was aided by off-line analysis on our Sun workstation (Sun Microsystems) using an oblique sectioning program from a multidimensional, user-interactive software package called VIDAR, which was developed in our laboratory. This allowed for computer reconstruction of sagittal and coronal images from the transverse ones (Fig 4). Discontinuity was defined as nonconfluence of the pulmonary arteries in all three orthogonal planes.

Calculations

Although raw numbers were used when comparing the various imaging modalities, two indexes of pulmonary size that are in the literature were also calculated and used in the data analysis. The Nakata index uses the cross-sectional area of both branch pulmonary arteries (right [RPA] and left [LPA] pulmonary artery) indexed to the body surface area:

\[
\text{Nakata Index} = \frac{\pi \times (\text{LPA}_{\text{Diameter}}^2 + \text{RPA}_{\text{Diameter}}^2)}{4 \times \text{Body Surface Area}}
\]

and the McGoon index divides the diameter of the branch pulmonary arteries or their sum by the diameter of the descending aorta (DAo) at the level of the diaphragm:

\[
\text{RPA Index} = \frac{\text{RPA}_{\text{Diameter}}}{\text{DAo}_{\text{Diameter}}}
\]

\[
\text{LPA Index} = \frac{\text{LPA}_{\text{Diameter}}}{\text{DAo}_{\text{Diameter}}}
\]

measured coronally (across from right to left) in all imaging modalities.

Statistical Analysis

Comparison between imaging modalities was made using the method of Bland and Altman. Initially, an estimate of the bias of the imaging modality is made (ie, the difference observed in our sample population between the imaging modality and angiography [our gold standard]), and construction of a 95% confidence interval was done using the following formula using the \( t \) statistic:

\[
\text{95% Confidence Interval} = \text{Mean Difference} \pm \frac{\text{SD}}{\sqrt{n}}
\]

where mean difference is the average of the differences between the imaging modality and angiography, \( t_{0.025, n-1} \) is the
The $t$ statistic with 2.5% of the area under the $t$ distribution and $n-1$ $df$, SD is the standard deviation, and $n$ is the number of observations. If this interval contains 0, there is 95% confidence that no significant bias is present, and if it does not contain 0, then there is 95% confidence that a significant bias exists. A “prediction interval” is then constructed, which predicts where 95% of the population as a whole will fall given our data, using the following formula:

$$\text{Prediction Interval} = \text{Mean Difference} \pm 2 \times \text{SD}$$

Finally, a plot of the difference between the imaging modality and angiography versus angiography is constructed to determine how the difference varies with the size of the pulmonary artery.

Comparison of magnetic resonance imaging and echocardiographic differences with angiography was made using the paired, two-tailed Student’s $t$ test. Comparison between right and left pulmonary arteries across surgical subgroups used two-factor ANOVA with repeated measures. Pairwise comparison between different groups used two-factor ANOVA modified by the Bonferroni correction.

Both intraobserver and interobserver variability were made by the coefficient of variability, defined as

$$\text{Coefficient of Variability} = \frac{\text{Observation 1} - \text{Observation 2}}{\text{Mean of Observation 1} + \text{Observation 2}}$$

and then computing the mean±SD for each structure and imaging modality (Table 3). Interobserver variability was measured between two independent observers who were blinded to the other’s results.

**Results**

**Branch Pulmonary Artery Measurements**

Table 4 displays the raw data from angiography, the gold standard used in this study, as well as pulmonary artery size determined by magnetic resonance imaging and echocardiography. The mean values for right and left pulmonary arteries showed little difference between mean values from magnetic resonance imaging and angiography in each of the three indexes or measurements: the McGoon index (0.01 for each), Nakata index (9 mm²/m² body surface area), and absolute branch pulmonary artery size (0.21 and 0.03 mm, respectively). Echocardiographic difference between mean values was much greater than that for magnetic resonance imaging (McGoon index, by 10 times for the right pulmonary artery and 7 times for the left pulmonary artery; Nakata index, by slightly more than 3 times and, in absolute measures, by 3 times for the right pulmonary artery and 20 times for the left pulmonary artery) ($P<.05$). The mean value of the pairwise differences between magnetic resonance imaging and angiography (ie, mean difference in Table 4: mean value for all patients of the branch pulmonary artery size by magnetic resonance imaging minus the value as determined by angiography) was smaller than echocardiography for the McGoon and Nakata indexes but not for absolute size. Note, however, that the standard deviation of the pairwise differences is much greater for echocardiography than for magnetic resonance imaging ($P<.05$).

Table 5 shows the resulting 95% confidence intervals for the bias estimates and prediction intervals for magnetic resonance imaging and echocardiography. The 95% confidence interval for the bias estimate, which is a statistical measure of how each imaging modality varied from angiography in our study population, contained 0 for both magnetic resonance imaging and echocardiography, implying no statistical difference between the two compared with angiography. However, the prediction interval (a statistical measure of how
Fig 4. T1-weighted images with computer reconstructions in two orthogonal planes of a patient with hypoplastic left heart syndrome and discontinuous branch pulmonary arteries who is status post–hemi-Fontan procedure. Left, Original transverse image at the level of the pulmonary arteries, with arrow pointing to the area of discontinuity between the pulmonary arteries. The arrow labeled COR points to a coronal computer reconstruction (from transverse images) at the level of the pulmonary arteries (upper right). The arrow labeled SAG points to a sagittal computer reconstruction (from transverse images) at the level of the pulmonary arteries (lower right). Hollow arrows point to the areas of discontinuity in the computer reconstructions. AAo indicates ascending aorta; COR, coronal; LPA, left pulmonary artery; RA, right atrium; RPA, right pulmonary artery; RV, right ventricle; and SAG, sagittal.

different each imaging modality would be from angiography if we performed the imaging modality in a prospective manner to the population at large) was much different for magnetic resonance imaging and echocardiography (P<.05). Magnetic resonance imaging would be predicted to be different from angiography by approximately ±0.1 for each branch pulmonary artery in the McGoon Index, by ±160 mm²/m² body surface area in the Nakata index, and by ±3 mm for each branch pulmonary artery in absolute size. Echocardiography, on the other hand, would be different by approximately 6 times as much as magnetic resonance imaging in the McGoon index, by approximately ±100 mm²/m² body surface area more than magnetic resonance imaging in the Nakata index and in absolute size, by ±2 mm more than magnetic resonance imaging for

**TABLE 3. Interobserver and Intraobserver Variability Measured by the Coefficient of Variation for Each Structure and Imaging Modality**

<table>
<thead>
<tr>
<th>Variability</th>
<th>Structure</th>
<th>Angiography</th>
<th>Magnetic Resonance Imaging</th>
<th>Echocardiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>Interobserver</td>
<td>RPA</td>
<td>6.4±3.6</td>
<td>5.6±2.8</td>
<td>7.3±2.8</td>
</tr>
<tr>
<td></td>
<td>LPA</td>
<td>6.0±3.1</td>
<td>5.2±2.4</td>
<td>8.2±3.4</td>
</tr>
<tr>
<td></td>
<td>DAo</td>
<td>5.8±3.0</td>
<td>5.0±2.5</td>
<td>6.1±3.3</td>
</tr>
<tr>
<td>Intraobserver</td>
<td>RPA</td>
<td>4.3±2.1</td>
<td>4.0±1.5</td>
<td>5.3±2.1</td>
</tr>
<tr>
<td></td>
<td>LPA</td>
<td>4.1±2.2</td>
<td>3.1±1.4</td>
<td>5.2±2.4</td>
</tr>
<tr>
<td></td>
<td>DAo</td>
<td>4.1±2.1</td>
<td>3.3±1.2</td>
<td>4.2±2.3</td>
</tr>
</tbody>
</table>

RPA indicates diameter of right pulmonary artery; LPA, left pulmonary artery; and DAo, diameter of the descending aorta. Values are mean±SD.
Table 4. Summary of Pulmonary Artery Size Data Based on Angiography, Magnetic Resonance Imaging, and Echocardiography

<table>
<thead>
<tr>
<th>Measurement or Index</th>
<th>Angiography</th>
<th>Magnetic Resonance Imaging</th>
<th>Magnetic Resonance Imaging, Mean Difference</th>
<th>Echocardiography</th>
<th>Echocardiography, Mean Difference</th>
</tr>
</thead>
<tbody>
<tr>
<td>McGoon index, RPA</td>
<td>0.86±0.27</td>
<td>0.87±0.30</td>
<td>0.009±0.065</td>
<td>0.76±0.20</td>
<td>−0.035±0.253</td>
</tr>
<tr>
<td>McGoon index, LPA</td>
<td>0.70±0.35</td>
<td>0.71±0.34</td>
<td>0.009±0.056</td>
<td>0.63±0.19</td>
<td>−0.034±0.313</td>
</tr>
<tr>
<td>Nakata index, mm²/m² BSA</td>
<td>169.88±114.85</td>
<td>160.59±113.96</td>
<td>−9.29±81.06</td>
<td>139.84±59.96</td>
<td>−23.64±125.35</td>
</tr>
<tr>
<td>Absolute RPA size, mm</td>
<td>7.40±2.60</td>
<td>7.19±2.73</td>
<td>−0.19±1.62</td>
<td>6.70±2.63</td>
<td>−0.13±2.60</td>
</tr>
<tr>
<td>Absolute LPA size, mm</td>
<td>6.04±3.21</td>
<td>6.01±3.56</td>
<td>−0.03±1.55</td>
<td>5.45±2.21</td>
<td>−0.02±2.32</td>
</tr>
</tbody>
</table>

LPA indicates left pulmonary artery; RPA, right pulmonary artery. Values are mean±SD.

Table 5. Bias Estimates and Prediction Intervals Using 95% Confidence Limits in Determining Differences Between Angiographically Derived Pulmonary Artery Size and Magnetic Resonance Imaging or Echocardiography

<table>
<thead>
<tr>
<th>Measurement or Index</th>
<th>Bias Estimate, 95% Confidence</th>
<th>Prediction Interval, 95% Confidence</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>MRI Estimate, 95% Confidence</td>
<td>MRI Prediction Interval, 95% Confidence</td>
</tr>
<tr>
<td></td>
<td>Echocardiography</td>
<td></td>
</tr>
<tr>
<td>McGoon index, RPA</td>
<td>−0.014, 0.032</td>
<td>−0.153, 0.083</td>
</tr>
<tr>
<td>McGoon index, LPA</td>
<td>−0.011, 0.029</td>
<td>−0.180, 0.112</td>
</tr>
<tr>
<td>Nakata index, mm²/m² BSA</td>
<td>−36.53, 19.95</td>
<td>−76.58, 29.27</td>
</tr>
<tr>
<td>Absolute RPA size, mm</td>
<td>−0.252, 0.393</td>
<td>−1.21, 0.94</td>
</tr>
<tr>
<td>Absolute LPA size, mm</td>
<td>−0.584, 0.532</td>
<td>−1.00, 0.96</td>
</tr>
</tbody>
</table>

LPA indicates left pulmonary artery; RPA, right pulmonary artery.

data, and the absolute RPA size, was found to be larger than the absolute LPA size, and by ±1.5 mm more than magnetic resonance imaging for the left pulmonary artery ($P<0.05$).

Fig 5 (magnetic resonance imaging) and Fig 6 (echocardiography) display these results graphically (A and B, the McGoon index; C, Nakata index; D and E, absolute pulmonary artery size) and show how the difference between each imaging modality and angiography varies with the size of the structure being measured. Note that the y-axis scales for magnetic resonance imaging data are much smaller than those for echocardiographic data, implying less variation in magnetic resonance imaging than echocardiography across structure size. Taking that into account, note how echocardiography begins to vary to a much greater degree as the branch pulmonary artery becomes larger than magnetic resonance imaging across all indexes and measurements.

Pulmonary Artery Discontinuity and Stenoses

Magnetic resonance imaging correctly detected five of five patients with branch pulmonary artery discontinuity (Fig 4) and six of six with stenoses. Echocardiography, using two-dimensional imaging and color flow mapping, was unable to detect any nonconfluent branch pulmonary arteries with certainty and only two of six (33%) with stenoses.

Comparison Between Left and Right Branch Pulmonary Artery Size

The right pulmonary artery was found to have a larger diameter than the left pulmonary artery by absolute measures (right pulmonary artery mean diameter, 7.4 mm; left pulmonary artery mean diameter, 6.0 mm; $P=.004$, $F=9.88$, $df=29$, interaction $P=.26$) and by the McGoon index (right pulmonary artery index, 0.86; left pulmonary artery index, 0.70; $P=.002$, $F=11.97$, $df=29$, interaction $P=.24$), regardless of stage of reconstruction.

Discussion

When Choussat and Fontan published their selection criteria for the Fontan procedure in 1978, they listed in their "Ten Commandments" that the pulmonary artery-to-aortic diameter ratio be ±0.75. Since then, there has been much discussion in the literature concerning the usefulness of pulmonary artery size both as a preoperative requirement and as a predictor of outcome.

There are a number of studies that support the notion that pulmonary artery size plays an important role. Nakata et al° in 1984 initially published their index to gauge the size of the pulmonary artery and found it useful in predicting postoperative prognosis. In a review of 167 Fontan cases, Mayer et al° in 1986 found 109 (67%) exceeded one or more of the original selection criteria, and multivariate analysis showed that pulmonary artery distortion "had a significant negative impact" on survival. Norwood et al° suggested that left pulmonary artery hypoplasia may be a cause of death in some patients with hypoplastic left heart syndrome undergoing the Fontan procedure. Fontan et al° published a multicenter series of 334 patients in 1989 and concluded that "one of the most powerful risk factors
for death or takedown of the Fontan operation was the dimensions of the right and left pulmonary arteries, expressed as the McGoon ratio.”

There are a few dissenting opinions concerning the significance of pulmonary artery size.2,3 Girod et al7 used the Nakata index when measuring the pulmonary arteries during surgery and found no difference in surgical mortality between the 70 survivors and the 20 nonsurvivors. Nevertheless, they still said that “we agree with Choussat et al. and Nakata et al. that assessment of the size of the pulmonary arteries should be an integral part of the preoperative evaluation.” Bridges et al2 also used the Nakata index before surgery, measuring pulmonary arteries by echocardiography, and found no difference in “early survival” between the 23 survivors and the 6 nonsurvivors. Echocardiography, the Nakata index, or the small number of patients, which may preclude a statistical statement, all may have played a role. Bridges et al also studied a select group of patients (ie, those with hypoplastic left heart syndrome) who were different from our patients (ie, single ventricles). Bridges et al further noted that 24 of the 29 patients “underwent pulmonary artery augmentation at the time of Fontan repair,” lending credence to the notion that preoperative assessment of pulmonary artery size may affect the planning.

Fig 5. Plots of the difference between magnetic resonance imaging (MRI) and angiographic determination of pulmonary artery size (y axis) versus angiographic determination of pulmonary artery size (the gold standard, x axis) for the McGoon index (A and B), the Nakata index (C), and absolute pulmonary artery size (D and E). Note the scale of the y axis, and compare with the scale of the y axis in Fig 6. Although the data points appear to spread out as pulmonary artery size increases, it is not significant (again, note y-axis scale). RPA indicates right pulmonary artery; DAO, diameter of descending aorta; BSA, body surface area; and LPA, left pulmonary artery.

Fig 6. Plots of the difference between echocardiographic and angiographic determination of pulmonary artery size (y axis) versus angiographic determination of pulmonary artery size (the gold standard, x axis) for the McGoon index (A and B), the Nakata index (C), and absolute pulmonary artery size (D and E). Note the scale of the y axis, and compare with the scale of the y axis in Fig 5. The data points appear to spread out as pulmonary artery size increases when compared with Fig 5 (again, note y-axis scale). Abbreviations same as for Fig 5.
and ultimate conduct of the surgery. They concluded that "patients should not be excluded from consideration for Fontan’s repair solely on the basis of pulmonary artery size."

Given the usefulness of pulmonary artery size in this group of patients, noninvasive assessment of pulmonary artery size would be a useful number to factor into the Fontan patient’s equation. We therefore elected to study whether echocardiography and magnetic resonance imaging would give the same information as angiography.

Our results showing that echocardiography did not reflect the pulmonary artery size determined by angiography as well as magnetic resonance imaging may be in part due to the oblique angle with which the vessel is cut on echocardiography. A true coronal image, which is seen on the anteroposterior angiogram, is not readily obtained by echocardiography from the suprasternal notch position. Furthermore, right pulmonary artery size was better assessed by echocardiography than left pulmonary artery size, whether using the absolute measures or the McGoorn index. This may be due to the posterior course the left pulmonary artery takes and its poor visualization in that position of the thorax. It is also interesting to note that as the size of the pulmonary artery increases (eg, as patients progress through the stages of Fontan reconstruction and as they age), the agreement between angiography and echocardiography of pulmonary artery size became poorer. This may be explained by the fact that those patients with larger pulmonary arteries are generally older and bigger, which in turn increases the technical difficulty of the echocardiographic study, whereas magnetic resonance imaging does not have this handicap.

Magnetic resonance imaging was found to be superior to echocardiography in measuring pulmonary artery size for all indexes in all statistical descriptions. This was found to be the case when comparing mean values (Table 4), prediction interval (Table 5), and error across vessel size (Figs 5 and 6).

Magnetic resonance imaging agreed well with angiographic measures of pulmonary artery size, with the McGoorn index agreeing better than absolute measures (which agreed better than the Nakata index). Two explanations may be offered. The first has to do with the technique of magnetic resonance imaging, which may yield crisp images in one patient but, because of various technical factors such as the quality of ECG gating and respiratory motion, may yield less crisp images in another. This may decrease the agreement when comparing absolute values; however, when an internal standard for the imaging technique is included (eg, the diameter of the descending aorta), the agreement may be better.

Second, the pulmonary artery may not be perfectly cylindrical, and its diameter may not be identical in the anteroposterior and superoinferior planes. The anteroposterior angiogram is a coronal image, and so comparison with the transverse magnetic resonance images may not agree as well when absolute values are used. The descending aortic diameter, however, was measured coronally in both angiographic and magnetic resonance images, and indexing to this number may make the agreement better.

In the determination of pulmonary artery discontinuity and stenoses, magnetic resonance imaging outperformed echocardiography and allowed for the identification of these lesions with certainty. Although echocardiography was suggestive in one case, the diagnosis of discontinuity could not be made with certainty. In other cases, the lesion went undiagnosed until magnetic resonance imaging was used.

It is interesting to note that the absolute measurement and McGoorn index of the right pulmonary artery were statistically larger than those of the left pulmonary artery regardless of surgical stage of reconstruction. This is an important consideration to the surgeon, who would consider augmenting the branch pulmonary arteries as the patient progresses through staged Fontan reconstruction. Furthermore, because pulmonary artery size is believed to have prognostic value, we believe this finding suggests it is important to obtain adequate imaging of the left pulmonary artery.

Comparison With Previous Studies

Although multiple studies have documented the usefulness of magnetic resonance imaging in determining pulmonary artery size,19,21-29 only three reports in the pediatric literature have compared this with echocardiography.27-29 Gomes et al27 and Parsons et al28 considered a variety of congenital heart lesions, but they do not report specific measurements in their studies. Although Vick et al28 did report their measurements in 12 patients in a variety of congenital heart lesions, both they and Gomes et al27 used the Pearson correlation coefficient, not the method of Bland and Altman. All are consistent with our findings that magnetic resonance imaging is superior to echocardiography. Furthermore, our study takes a single group of patients who undergo major pulmonary artery reconstruction and shows the usefulness of magnetic resonance imaging over echocardiography in following the patient over a series of postoperative states.

Study Limitations

Only suprasternal notch imaging or high parasternal imaging was used in transthoracic echocardiography for the calculation of pulmonary artery size because of the consistently high-quality imaging of those structures in nearly all patients. It is possible that subcostal imaging or transesophageal echocardiography offers a better delineation of pulmonary artery size.

Conclusions

Magnetic resonance imaging is a useful, noninvasive tool to determine pulmonary artery size, stenosis, and branch pulmonary artery discontinuity in patients undergoing Fontan reconstruction. Although echocardiography was a fair predictor of pulmonary artery size, magnetic resonance imaging is superior to echocardiography for measurement of pulmonary artery size and definitive determination of stenosis or discontinuity. Magnetic resonance imaging may avoid unnecessary cardiac catheterization, especially in older patients (where echocardiography obtains poorer images), and may obviate the need for jugular or subclavian vein catheterization in those who have undergone bidirectional cavopulmonary anastomosis.
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References
Magnetic resonance and echocardiographic imaging of pulmonary artery size throughout stages of Fontan reconstruction.

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