Imaging of Pulmonary Venous Pathway Obstruction in Patients After the Modified Fontan Procedure

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To evaluate the efficacy of chest roentgenograms and echocardiograms in identifying pulmonary venous pathway obstruction in patients after the modified Fontan procedure, the records of 297 patients who underwent 307 Fontan procedures between 1984 and 1990 were reviewed. Twelve cases of pulmonary venous pathway obstruction documented by autopsy (3 cases) or cardiac catheterization (9 cases with an A wave gradient ≥4 mm Hg) were found in 10 patients (mean age 43 ± 28 months). The mechanisms of obstruction included narrow pulmonary vein ostia in six cases, narrow left atrial outlet in four and atrial baffle obstruction in two. Three causes of obstruction were present in one case. No patient had pulmonary venous congestion on chest roentgenograms.

Pathway diameters indexed to $\sqrt[3]{\text{body surface area}}$ were found to be $6.8 \pm 1.2 \text{mm} / \sqrt[3]{\text{m}^2}$ in the subcostal frontal view, $7.9 \pm 0.6 \text{mm} / \sqrt[3]{\text{m}^2}$ in the subcostal sagittal view and $6.5 \pm 1.7 \text{mm} / \sqrt[3]{\text{m}^2}$ in the apical "four-chamber" view. These values differed significantly from those in 11 age-matched patients undergoing the Fontan procedure without pulmonary venous pathway obstruction documented by catheterization ($p = 0.001$).

With pulsed Doppler ultrasound, there was a relatively narrow range of velocities distal to the obstruction (1.3 to 2.5 m/s). In five of the seven cases with pulsed Doppler measurements, flow was continuous and the Doppler spectral recordings were not phasic.

Thus, in patients who have undergone the Fontan procedure and have pulmonary venous pathway obstruction 1) chest roentgenography cannot be used as a screening tool; 2) distal velocities as low as 1.3 m/s occur, usually with midsystolic and diastolic forward flows; and 3) pathway diameters indexed to $\sqrt[3]{\text{body surface area}}$ may be used as an output-independent parameter to cross-check Doppler data.

Methods

Study patients. We reviewed the records of all 297 patients who underwent 307 Fontan procedures between January 1984 and December 1990. All patients underwent cardiac catheterization, chest X-ray study and echocardiographic examination before both the original or repeat Fontan operation. Cardiac malformations included tricuspid atresia, pulmonary atresia, hypoplastic left heart syndrome, other "single ventricle" lesions and complex anomalies such as double-outlet right ventricle with complete common AV canal.

Types of Fontan procedures. Four types of Fontan procedures were performed during the study period (Fig. 1). In
one (Fig. 1A), a systemic venous "tube" is created along the lateral atrial wall (by insertion of a hemicylindrical piece of polytetrafluoroethylene), baffling the inferior vena cava and hepatic venous blood to an anastomosis between the superior vena cava-right atrial junction and the right pulmonary artery. In another type (Fig. 1B), a right atrium to right pulmonary artery anastomosis is created and a large patch baffles pulmonary venous blood through the original atrial septal defect to the atroventricular valve associated with the systemic pumping chamber. C, Used in some cases of tricuspid or pulmonary atresia, a right atrium to pulmonary artery anastomosis is created and the atrial septal defect is closed. D, Used in tricuspid atresia with mild or moderate right ventricular hypoplasia, a right atrial to right ventricular anastomosis is created and the ventricular septal defect is closed.

Entrance criteria into study. The reference standards for the diagnosis of pulmonary venous pathway obstruction were 1) cardiac catheterization with intraoperative confirmation, or 2) autopsy (Table 1). An A wave gradient across the pulmonary venous pathway ≥4 mm Hg was considered evidence of pulmonary venous pathway obstruction. If no A wave measurement was obtained (as in some cases of pulmonary vein ostial stenosis), a mean gradient anywhere across the pulmonary venous pathway ≥4 mm Hg was used as an entrance criterion (Table 2). Gradients were measured from either the pulmonary capillary wedge position to the left atrium or ventricle (using the end-diasolic pressure) or from the left atrium to the ventricle (Fig. 2). The range of A wave gradients was 4 to 17 mm Hg and that of the mean gradient was 10 to 15 mm Hg. Mean (±SD) cardiac index was 2.6 ± 0.8 liters/min per m² (range 1.5 to 3.9) (Table 2).

Nine of 12 cases of pulmonary venous pathway obstruction were diagnosed before death. Although all children underwent cardiac catheterization, two of the three autopsy cases had not shown an A wave gradient ≥4 mm Hg or a mean gradient ≥4 mm Hg (Table 2, Patients 5 and 6) at catheterization and the diagnosis was made only at autopsy. One patient (Patient 9) did in retrospect meet cardiac catheterization criteria, but this fact was recognized only at postmortem examination. Postmortem pulmonary venous pathway diameter (relative to the body surface area) smaller than the standards for the normal aortic valve diameter used in our Cardiac Registry was considered evidence of pulmonary venous pathway obstruction. Table 3 summarizes the autopsy findings. There are no standard criteria for assessing individual pulmonary vein ostial stenosis. Finally, in one postmortem case with total anomalous pulmonary venous connection, there was a pulmonary venous confluence to left atrial anastomosis that was less than the normal aortic root diameter for body surface area.

Imaging. Chest X-ray films taken within 2 days before the diagnosis of pulmonary venous pathway obstruction were examined for evidence of pulmonary venous congestion. Echocardiograms performed in the 2 weeks before the diagnosis of pulmonary venous pathway obstruction or cardiac catheterization were reviewed and analyzed. All echocardiographic studies were performed on phased-array Hewlett-Packard ultrasound machines (Sonos 1000 and 500) using 5-, 3.5- or 2.5-MHz transducers. We measured the pulmonary venous pathway at its narrowest point in three
different views: subcostal frontal, subcostal sagittal and apical “four-chamber” (Fig. 3). Dimension measurements were then normalized by indexing to $\sqrt{V}$ body surface area (14).

**Pulsed Doppler interrogation** was used in studies after January 1985. We measured the velocity at multiple sites along the pathway and evaluated the temporal pattern of Doppler spectral recordings to determine whether the velocity was phasic or flow was continuously anterograde (Fig. 4). Doppler color ultrasound imaging using 2.5- or 3.5-MHz transducers was routinely performed in studies performed after January 1987 and guided the pulsed Doppler interrogation (Fig. 5).

**Control group.** Eleven consecutive patients who had undergone the modified Fontan procedure (mean age 46 ± 38 months) and were documented not to have pulmonary venous pathway obstruction by catheterization between January 1989 and December 1990 were chosen from the computerized data base for study as control subjects (Table 4). All follow-up studies—chest X-ray films, echocardiograms, and cardiac catheterization at The Children’s Hospital of Philadelphia—were performed within 2 weeks of each other in all patients, and no patient had anatomic abnormalities of the reparative operation. We compared pathway diameters measured by echocardiography in control subjects (indexing to $\sqrt{V}$ body surface area) with those in our patients documented to have pulmonary venous pathway obstruction by catheterization. In the control group, mean cardiac index was 2.5 ± 0.9 liters/min per m$^2$ (range 1.1 to 4.7). Neither age nor cardiac index in the control subjects differed significantly from those in patients who had undergone the Fontan procedure and had pulmonary venous pathway obstruction.

**Statistics.** To compare mean values between patients who had undergone the Fontan procedure with pulmonary venous pathway obstruction and the control subjects, the unpaired two-tailed Student $t$ test was used. To compare the incidence of pleural effusion between the two groups, the Fisher exact test was used. Unless otherwise indicated, data are expressed as mean value ± SD.

**Results**

**Prevalence.** Twelve cases of pulmonary venous pathway obstruction were found in 10 (3.4%) of the 297 children who underwent the Fontan procedure in our institution (mean age 43 ± 28 months). The time interval between the Fontan procedure and the diagnosis of obstruction was 246 ± 90

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**Table 1.** Initial Patient Malformation, Study Entry Criteria and Type of Fontan Procedure and Pulmonary Venous Pathway Obstruction

<table>
<thead>
<tr>
<th>PVPO No.</th>
<th>Pt No.</th>
<th>Diagnosis</th>
<th>Fontan Procedure</th>
<th>Catheterization</th>
<th>Autopsy</th>
<th>PVPO Type</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>1</td>
<td>HLHS</td>
<td>A</td>
<td>Y</td>
<td>—</td>
<td>LA outlet</td>
</tr>
<tr>
<td>2</td>
<td>2</td>
<td>HLHS</td>
<td>B</td>
<td>Y</td>
<td>—</td>
<td>LA outlet</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>HLHS</td>
<td>A</td>
<td>Y</td>
<td>—</td>
<td>LA outlet and atrial patch</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>HLHS</td>
<td>B</td>
<td>Y</td>
<td>N</td>
<td>Atrial patch</td>
</tr>
<tr>
<td>5†</td>
<td>5†</td>
<td>DORV [S.D.A], single LV, subpulmonary stenosis, right AVV stenosis, single RCA, L SVC → CS, LIAA</td>
<td>C</td>
<td>N</td>
<td>Y</td>
<td>LA outlet</td>
</tr>
<tr>
<td>6</td>
<td>6</td>
<td>DORV [S.D.D], left AVV stenosis and subpulmonary stenosis</td>
<td>B</td>
<td>N</td>
<td>Y</td>
<td>Atrial patch</td>
</tr>
<tr>
<td>7</td>
<td>7</td>
<td>TOA [S.D.D], atrial and pulmonary stenosis</td>
<td>B</td>
<td>Y</td>
<td>—</td>
<td>Pulmonary vein</td>
</tr>
<tr>
<td>8</td>
<td>8</td>
<td>DILV [S.D.S], aortic atresia, right AVV hypoplasia</td>
<td>A</td>
<td>Y</td>
<td>—</td>
<td>Pulmonary vein</td>
</tr>
<tr>
<td>9</td>
<td>9</td>
<td>Heterotaxy, DORV [A.D.D], CCACV, aortic atresia, TAPVC to mesenteric vein, coarctation</td>
<td>A</td>
<td>N</td>
<td>Y</td>
<td>Pulmonary vein</td>
</tr>
<tr>
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<td>10</td>
<td>Heterotaxy, DORV [A.D.D], CCACV</td>
<td>A</td>
<td>Y</td>
<td>—</td>
<td>Pulmonary vein</td>
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<tr>
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<td>Y</td>
<td>—</td>
<td>Pulmonary vein</td>
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<tr>
<td>12</td>
<td>10</td>
<td>Heterotaxy, DORV [A.D.D], CCACV</td>
<td>A</td>
<td>Y</td>
<td>—</td>
<td>Pulmonary vein</td>
</tr>
</tbody>
</table>

*For a description of the types of Fontan procedures, see text and Figure 1. Patient 5 underwent patch closure of the right atrioventricular valve (AVV). Left atrial (LA) outlet obstruction was caused by the coronary sinus baffle into the left atrium, obstructing flow to the left atrioventricular valve. A, B and C correspond to the Fontan procedures shown in Figure 1. A, B and C, respectively: (A.D.D) = situs ambiguous of the viscera and atria, ventricular D loop, D transposed or malposed great arteries (meaning aortic valve to the right of the pulmonary valve); CCACV = complete common atrioventricular canal; DILV = double-inlet left ventricle; DORV = double-outlet right ventricle; HLHS = hypoplastic left heart syndrome; LIAA = left-sided juxtaposition of the atrial appendages; L SVC = left superior vena cava to coronary sinus; LV = left ventricle; N = no; Pt = patient; PVPO = pulmonary venous pathway obstruction; RCA = right coronary artery; (S.D.A) = situs solitus of the viscera and atria, ventricular D loop, ambiguous malposed great arteries (meaning aortic and pulmonary valves in an anteroposterior relation to each other); (S.D.D) = situs solitus of the viscera and atria, ventricular D loop, D transposed or malposed great arteries (meaning aortic valve to the right of the pulmonary valve); (S.D.S) = situs solitus of the viscera and atria, ventricular D loop, situs solitus normally aligned great arteries; TAPVC = total anomalous pulmonary venous connection; TOA = transposition of the great arteries; Y = yes.*
days (range 13 to 1.084). The patients' native lesion, type of Fontan procedure, pulmonary venous pathway obstruction mechanism and entrance criteria are summarized in Table 1.

**Study group characteristics.** Five of these 10 children had undergone the Fontan repair shown in Figure 1A. four had that shown in Figure 1B, one had the reconstruction depicted in Figure 1C (Patient 5, who also underwent right AV valve patch closure) and none had had a procedure of the type shown in Figure 1D. Patient 5 had undergone the reconstruction shown in Figure 1A and had a total of three instances of pulmonary venous pathway obstruction (each occurring approximately 1 year apart), accounting for the additional two cases of obstruction (Patient 10, Table 1). The initial malformations included four cases of hypoplastic left heart syndrome, two of double-outlet right ventricle and pulmonary stenosis, two of double-outlet right ventricle and complete common AV canal, one of double-inlet left ventricle and aortic atresia and one of transposition of the great arteries accompanied by pulmonary and mitral stenosis.

The etiology of pulmonary venous pathway obstruction included narrowed pulmonary vein ostium (n = 6), narrowed left atrial outlet (area of original atrial septal defect or left AV valve) Fig. 6 (n = 4) and obstruction caused by the atrial baffle (n = 3); in one case, obstruction was due to both left atrial outlet and atrial baffle obstruction (Table 1, Fig. 1A). Patient 5 had left atrial outlet obstruction on the basis of the coronary sinus (which accepts left superior vena cava blood flow) bulging anteriorly and superiorly, obstructing the free flow of blood from pulmonary veins to the left AV valve.

**Imaging**

Chest roentgenograms. No chest X-ray film revealed a unilateral or bilateral increase in pulmonary venous markings in the lung fields over those observed in previous films. None displayed a fine, diffuse, linear reticular pattern, Kerley B lines or perihilar congestion obscuring heart borders, all of which would be common findings in patients with two ventricles and pulmonary venous obstruction. In 9 (90%) of the 10 patients with pulmonary venous pathway obstruction, the chest X-ray film showed pleural effusion; in contrast, only 2 of the 11 control patients had pleural effusion (p = 0.002). A typical chest X-ray film from a patient with pulmonary venous pathway obstruction (Patient 10) is shown in Figure 7.

Two-dimensional echocardiographic measurements (Table 2). In the patients with pulmonary venous pathway obstruction, pathway diameter measured 6.8 ± 1.2 mm/Vm³ in the subcostal frontal view, 7.9 ± 0.6 mm/Vm³ in the subcostal sagittal view (Fig. 3) and 6.5 ± 1.7 mm/Vm³ in the apical “four-chamber” view. In the 11 control subjects pathway diameter measured 15 ± 2.7 mm/Vm³ in the subcostal frontal view, 15 ± 2.9 mm/Vm³ in the subcostal sagittal view and 14 ± 3 mm/Vm³ in the apical “four-chamber”
Figure 2. Patient 1. Cardiac catheterization tracings from a patient with pulmonary venous pathway obstruction after the Fontan procedure: measurements are made at 50 scale. Pullback from the native left atrium (A) to the native right atrium (B) across the atrial septal defect shows a pressure decrease from an A wave of 27 mm Hg (mean 14) to an A wave of 14 mm Hg (mean 10).

Table 3. Autopsy Results From Three Patients With Pulmonary Venous Pathway Obstruction

<table>
<thead>
<tr>
<th>Autopsy No.</th>
<th>Pt No.</th>
<th>Body Surface Area</th>
<th>Smallest Pathway Diameter</th>
<th>Normal Aortic Root Diameter</th>
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<tbody>
<tr>
<td>1</td>
<td>5</td>
<td>0.87</td>
<td>5</td>
<td>13 ± 1</td>
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<tr>
<td>2</td>
<td>6</td>
<td>0.88</td>
<td>6</td>
<td>12 ± 1</td>
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<tr>
<td>3</td>
<td>9</td>
<td>0.36</td>
<td>6</td>
<td>9 ± 1</td>
</tr>
</tbody>
</table>

Smallest pathway diameter is that found at autopsy and measured in mm. Normal aortic valve diameter is also measured in mm (mean ± SD) and is from the standards used in the Cardiac Registry. Note that the study group values are far below 2 SD. Pt = patient.

obstruction. Distal velocities ranged from 1.3 to 1.6 m/s, whereas in patients with left atrial outlet or atrial baffle obstruction, distal velocities ranged from 1.7 to 2.5 m/s.

Of the seven patients with pulsed Doppler measurements, five had continuous flow and the Doppler spectral recordings were not pheastic. The remaining two had no hemodynamic data that distinguished them from the others. The 11 control patients did not have complete Doppler interrogation of the pulmonary venous pathway.

Symptoms. Table 5 lists the symptoms that led to medical evaluation of the patient. The most common symptom was persistent effusion, either pleural, pericardial or peritoneal.

Discussion

Chest roentgenograms. In patients with pulmonary venous pathway obstruction and a pulmonary pumping chamber, the lung fields on the chest X-ray film (7,8) reflect prominent venous engorgement of the pulmonary veins and show a fine, diffuse, linear reticular pattern that fans out from the pulmonary hilus. Kerley B lines may also be present with perihilar congestion, which results in obscured cardiac borders. Hyperinflation of the lungs may also be seen (15).

The manifestations of pulmonary venous pathway obstruction may not be similar in the patient who has undergone a Fontan procedure in which flow through the lungs is achieved by “passive” means (that is, low pulmonary vascular resistance, negative intrathoracic pressure during inspiration and a compliant systemic ventricle). In such a situation, the hydrostatic pressure for forward flow may not induce changes on the chest X-ray film seen in pulmonary venous obstruction in patients with two ventricles. Decreased oncotic pressure resulting from altered permeability of the capillary wall, as evidenced by the tendency of patients who have undergone the Fontan procedure to develop protein-losing enteropathy, may also play a role (16,17).

None of the patients we reviewed who had undergone the Fontan procedure had any evidence of pulmonary venous congestion by plain X-ray film, although 99% had pleural
isolated, transitory pleural effusion was not in itself considered a sign of pulmonary venous obstructive physiology. The symptom that consistently led to medical evaluation was persistent (>3 weeks) serous cavity effusion. Roentgenography is therefore a poor screening test for pulmonary venous pathway obstruction.

**Two-dimensional echocardiographic measurements.** Pathway diameters indexed to $/sqrt{S}$ body surface area (14) significantly differed between patients who had undergone the Fontan procedure with and without pulmonary venous pathway obstruction. In patients who have undergone the Senning procedure for transposition of the great arteries (two-ventricle physiology), pulmonary venous pathway measurements (22) in patients with obstruction are different from those without obstruction. Our control patients who had undergone the Fontan procedure had a slightly larger normalized pathway diameter than did patients who were found to have an unobstructed pulmonary venous pathway after the Senning operation ($15.7 \pm 2.7$ mm vs. $10.8 \pm 1.7$ mm in the subcostal frontal view and $15 \pm 2.9$ mm vs. $11.2 \pm 2.3$ mm in the subcostal sagittal view).
Figure 5. Patient I. Doppler color flow map from a patient who underwent the Fontan procedure shown in Figure 1A and left atrial outlet obstruction. The view is subcostal frontal. Note the aliasing in the area of the atrial septal defect. Small white arrows point to the atrial baffle. aLA = anatomic left atrium; aRA = anatomic right atrium.

the subcostal sagittal view, respectively, indexed to body surface area. Our patients with pulmonary venous pathway obstruction had perhaps a slightly smaller normalized pathway diameter than that of patients who had undergone the Senning operation and had been documented to have pulmonary venous pathway obstruction (6.8 ± 1.2 vs. 8 to 12 mm in the subcostal frontal view and 7.9 ± 0.6 vs. 10 to 12 mm in the subcostal sagittal view, respectively, all measurements being indexed to body surface area), although the number of patients is small.

Doppler echocardiography. On Doppler echocardiographic study in patients with pulmonary venous pathway obstruction and a pulmonary pumping chamber, estimated right ventricular systolic pressure is elevated. Vick et al. (10) reported that in 12 patients with a two-ventricle circulation and severe pulmonary venous obstruction (defined as a mean catheterization pressure gradient >16 mm Hg), all had a Doppler-determined maximal velocity jet ≥ 2 m/s. They state that “the maximal Doppler velocities of patients with a mean catheterization pressure gradient of <16 mm Hg overlapped with maximal Doppler velocities of patients who had no mean pressure gradient... It was not possible to distinguish patients with milder degrees of obstruction from patients with no obstruction on the basis of peak Doppler flow velocity alone.” In our study of patients undergoing the Fontan procedure with pulmonary venous pathway obstruction, only one had a catheterization gradient >16 mm Hg, and therefore the study of Vick et al. (10) is not comparable to ours. The study of Vick et al. (10) also makes no statement about a step-up in velocity across sites of obstruction or about velocity proximal to sites of narrowing. However, in our patients with a pulmonary venous pathway gradient >4 mm Hg distal velocities as low as 1.3 m/s were observed. In evaluating a patient for pulmonary venous pathway obstruction, the use of distal velocity alone would therefore lead to a false negative diagnosis. In view of this observation, we recommend not neglecting the proximal velocity if the Bernoulli theorem is utilized.

In five of seven patients, the Doppler spectral recording

Table 4. Echocardiographic and Cardiac Catheterization Data From 11 Control Patients (without pulmonary venous pathway obstruction after the Fontan procedure)

<table>
<thead>
<tr>
<th>Pt No.</th>
<th>Native Lesion</th>
<th>Echocardiography</th>
<th>Catheterization</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>DORV (S.D.D), PS, LIAA</td>
<td>12/12/13, A = 10, m = 7</td>
<td>10, CI = 3.1</td>
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<tr>
<td>2</td>
<td>DORV (S.D.D), MA</td>
<td>11/12/13, m = 11</td>
<td>10, CI = 2.2</td>
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<tr>
<td>3</td>
<td>HLHS</td>
<td>14/12/18, m = 6</td>
<td>6, CI = 4.7</td>
</tr>
<tr>
<td>4</td>
<td>HLHS, TAPVC</td>
<td>14/12/18, m = 9</td>
<td>8, CI = 1.8</td>
</tr>
<tr>
<td>5</td>
<td>TGA (S.D.D), SI Vent, hypo RV, multiple VSDs</td>
<td>15/12/11, A = 6, m = 4</td>
<td>8, CI = 2.5</td>
</tr>
<tr>
<td>6</td>
<td>PA, hypo RV</td>
<td>16/10/17, m = 7</td>
<td>3, CI = 3.3</td>
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<tr>
<td>7</td>
<td>TGA (S.L.L), single LV</td>
<td>17/12/17, m = 16</td>
<td>16, CI = 1.1</td>
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<tr>
<td>8</td>
<td>HLHS</td>
<td>18/12/17, A = 5, m = 3</td>
<td>8, CI = 2.4</td>
</tr>
<tr>
<td>9</td>
<td>TGA (S.L.L), single LV</td>
<td>19/12/13, m = 8</td>
<td>12, CI = 2.4</td>
</tr>
<tr>
<td>10</td>
<td>HLHS</td>
<td>15/12/14, m = 10</td>
<td>15, CI = 2.3</td>
</tr>
<tr>
<td>11</td>
<td>HLHS</td>
<td>12/1/13, m = 1</td>
<td>2, CI = 2</td>
</tr>
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</table>

Pathway diameters are in the following order: subcostal frontal; subcostal sagittal; and are measured in mm. PCWP = end-diastolic pressure; PS = pulmonary stenosis; TLPA = transposed left pulmonary artery; PA = pulmonary atresia; RA = right atrium; RA = right atrial dilatation; LA = left atrial dilatation; MP = malposed great arteries; VSD = ventricular septal defect; LV = left ventricle; IV = inferior vena cava; RV = right ventricle; TM = tricuspid stenosis; TA = tricuspid atresia; TV = truncus arteriosus; PA = pulmonary artery; SI Vent = superior vena cava. All catheterization data are measured in mm Hg. EDP = end-diastolic pressure; hypo RV = hypoplastic right ventricle; MA = mitral atresia; PA = pulmonary atresia; PS = pulmonary stenosis; S.D.L = situs solitus of the viscera and atria; VSD = ventricular septal defect.
showed anterograde nonphasic flow continuously during the cardiac cycle distal to the obstruction. This differs
from the normal biphasic pulmonary venous flow pattern in patients who have undergone the Fontan procedure
of right atrium to pulmonary artery anastomosis without pulmonary venous pathway obstruction (11) and
the normal pulmonary venous flow pattern in a two-ventricle circulation (23,24). We cannot compare our
maximal velocity data because Smallhorn et al. (11) did not describe the maximal velocities observed; how-

ever, our waveform shapes were similar to the high velocity turbulent flow described with pulmonary vein ostial or
pulmonary venous pathway stenosis in patients with a pulmonary pumping chamber (10-13).

Figure 6. Patient I. Slightly right anterior oblique (A) and lateral (B) left atrial angiograms from a patient who underwent the Fontan
procedure shown in Figure 1A and had left atrial outlet obstruction. White arrows point to the area of obstruction. LA = anatomic left
atrium; RA = anatomic right atrium; RV = right ventricle.

Figure 7. Patient III. Typical chest X-ray film from a patient with
pulmonary venous pathway obstruction after the Fontan procedure.
This patient underwent the procedure shown in Figure 1A and had
pulmonary vein stenosis. As was usual in our series, there is no
evidence of any pulmonary venous congestion on this film (that is,
prominent venous engorgement of the pulmonary veins, a fine,
diffuse, linear reticular pattern that fans out from the pulmonary
hilum, Kerley B lines or peripheral congestion that results in obscured
cardiac borders or hypoinflation of the lungs).

Figure 8. Two-dimensional echocardiographic data comparing path-
way diameters in patients with pulmonary venous pathway obstruc-
tion after the Fontan procedure (circles) with those of the control
patients without obstruction (triangles). Pulmonary venous pathway
measurements are noted on the ordinate in mm/m². The three
views analyzed are noted on the abscissa. Note that the patients with
pulmonary venous pathway obstruction have measurements well
below those of the normal group.

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Table 5. Reason for Cardiac Catheterization or Symptoms Before Death

<table>
<thead>
<tr>
<th>PVPO No</th>
<th>Pt No</th>
<th>Reason for Cardiac Catheterization Symptoms Before Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1</td>
<td>Inability to be weaned from ventilator, \url{pleural effusion}</td>
</tr>
<tr>
<td>2</td>
<td>2</td>
<td>Inability to be weaned from ventilator, \url{pleural effusion, low output}</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>\url{Persistent effusion}</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>\url{Respiratory distress, pleural effusion, hepatomegaly, ascites}</td>
</tr>
<tr>
<td>5</td>
<td>5</td>
<td>\url{Persistent chylothorax}</td>
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<td>6</td>
<td>6</td>
<td>\url{Persistent pleural and pericardial effusion}</td>
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<td>7</td>
<td>Abdominal pain and distention, ascites</td>
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<tr>
<td>8</td>
<td>8</td>
<td>\url{Persistent pleural effusion}</td>
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<td>9</td>
<td>\url{Cardiopulmonary arrest, pleural effusion}</td>
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</tr>
<tr>
<td>12</td>
<td>10</td>
<td>\url{Persistent pleural effusion, lethargy, poor feeding}</td>
</tr>
</tbody>
</table>

Persistent effusion is defined as effusion present for >3 weeks. Abbreviations as in Table 1.

Postmortem findings. Our autopsy patients showed that a smaller-than-normal pathway diameter may be present even when the cardiac catheterization data show a mean gradient <4 mm Hg.

Onset of pulmonary venous pathway obstruction. The interval from the time of the Fontan operation to diagnosis of pulmonary venous pathway obstruction ranged from 13 to 1,084 days. Pulmonary venous pathway obstruction can develop over the course of years and does not necessarily manifest within the first few months after operation.

Limitations of the study. In a retrospective analysis, there is potential bias in analysis of echocardiographic and radiographic data. We did not control the ventilatory patterns during Doppler echocardiography. Because positive pressure is a major effect on pulmonary artery flow, it is possible that it might affect pulmonary venous pathway flow patterns.

No data are available on patients with pulmonary venous pathway obstruction who may be asymptomatic or who may have died without medical evaluation or autopsy. There are also no quantitative criteria to use for pulmonary vein ostial stenosis at autopsy. For all these reasons, our estimate of the prevalence of pulmonary venous pathway obstruction may be artificially low.

Proximal velocities were not obtainable in the case of pulmonary vein ostial stenosis. Recognition of this variety of pulmonary venous pathway obstruction might be better approached by transesophageal echocardiography.

Problems in comparing absolute Doppler measurements include the fact that the incident Doppler beam may be imperfectly aligned with the direction of blood flow (>20°) and that patients with a low output state may have a much lower flow velocity in the pulmonary venous pathway. A two-dimensional echocardiographic pathway dimension measurement may therefore serve as a relatively output-independent parameter to cross-check Doppler data.

Conclusions. Pulmonary venous pathway obstruction was present in ≥3% of our patients who had undergone the Fontan procedure during the years from 1984 to 1990. Chest roentgenography was a poor screening test for such obstruction in these patients. Two-dimensional echocardiographic measurements of pathway dimension (indexed to the body surface area) appear to differentiate patients with those without pulmonary venous pathway obstruction after the Fontan procedure. Pulsed Doppler echocardiography reveals that the maximal velocity at the narrowing sites can be as low as 1.3 m/s and the spectral tracing may be nonphasic. Both the area proximal to and that distal to the narrowing should be scrutinized if the Bernoulli theorem is utilized. Prospective application of these ultrasound techniques should definitively settle whether the pulmonary venous pathway in the patient who had undergone the Fontan procedure can be reliably identified noninvasively. Finally, autopsy can show a small pulmonary venous pathway orifice, even when the mean gradient at cardiac catheterization is <4 mm Hg.

References

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