Right Ventricular Volume Determinations in 18 Patients with Pulmonary Atresia and Intact Ventricular Septum

Analysis of Factors Influencing Right Ventricular Growth

RAMAN G. PATEL, M.B., ROBERT M. FREEDOM, M.D., C.A.F. MOES, M.D., KENNETH R. BLOOM, M.D., PETER M. OLLEY, M.D., W.G. WILLIAMS, M.D., GEORGE A. TRUSLER, M.D., AND RICHARD D. ROWE, M.D.

SUMMARY Right ventricular growth was assessed angiographically in 18 patients with pulmonary atresia, intact ventricular septum, and hypoplastic and hypertensive right ventricle. A variety of surgical procedures were performed. In only 12 patients (66.7%) was right ventricular-pulmonary artery continuity achieved (group 1). Nine of these 12 patients persisted with systemic or suprasystemic right ventricular pressures. Among the six patients in whom right ventricular-pulmonary artery continuity was not achieved (group 2), all maintained suprasystemic right ventricular pressures.

Right ventricular growth was assessed in groups 1 and 2. The patients were also subdivided according to the qualitative degree of tricuspid regurgitation as determined angiographically on right ventricular cineangiograms at the preoperative catheter study. Right ventricular growth to normal levels as evidenced by change in right ventricular end-diastolic volume was rarely observed in group 2 patients. Among the four patients with severe tricuspid regurgitation and a large tricuspid valve, right ventricular growth to normal levels was achieved whether they were in group 1 or group 2. Right ventricular growth is thus predicated on numerous morphologic factors in these patients. However, reconstitution of right ventricular-pulmonary artery continuity and a nonobstructive tricuspid valve are probably two of the more important factors.

ALTHOUGH the morphologic, clinical, angiographic and surgical features of the patient with pulmonary atresia, intact ventricular septum and small or diminutive right ventricle are extensively recorded, surgical results in this group of patients are, for the most part, disappointing. Survival for even a short time depends on augmentation or maintenance of an adequate pulmonary blood flow, and several institutions have summarized their experience in this regard. Yet long-term functional and hemodynamic improvement, predicated in part on right ventricular growth, is seldom achieved.

To extend these observations, we analyzed the hemodynamics, angiograms, angiographically derived ventricular volumes, and types of surgical intervention in 18 patients with pulmonary atresia, intact ventricular septum and small right ventricle who survived initial operation and have had at least one postoperative complete hemodynamic and angiographic investigation.

Materials and Methods

Eighteen patients with pulmonary atresia, intact ventricular septum, and hypoplastic and hypertensive right ventricle form the basis of this study. All had an initial hemodynamic and angiographic investigation, followed by surgical intervention, and finally a postoperative complete catheter hemodynamic and angiographic investigation. Some of these patients have been included in previous reports from this institution. Six of the patients were boys and 12 were girls. The age at the initial study ranged from 1 day to 7 months (median 1 day; mean 20 days), and only two were older than 1 month. At the second cardiac catheter investigation, their ages ranged from 7 months to 9 years (median 2½ years; mean 3 years).

All 18 patients underwent balloon septostomy and five were treated by prostaglandin E₁ and E₂ infusion before surgery. The number and types of surgical procedures are presented in table 1. At the initial operation, a transarterial pulmonary valvotomy was performed in 15 patients, and the transventricular approach was used in one. Twelve of the 18 patients (61%) had a pulmonary valvotomy combined with a systemic-pulmonary artery shunt as the initial surgical therapy. One patient initially had only a pulmonary valvotomy but subsequently underwent a cavopulmonary shunt. Five patients initially underwent a systemic-to-pulmonary artery anastomosis without pulmonary valvotomy. Percardial reconstruction of the right ventricular outflow tract was performed as a subsequent operation in six patients (ranging in age...
from 4 months to 10 years; median 3 years), with two survivors (currently ages 6 and 7 years).

Right and left ventricular or aortic pressures were measured at both cardiac catheterizations using standard fluid-filled catheters. Complete oximetric data were obtained using Water's Oximeter (The Waters Co.) or co-oximeter 1L 82 (Instrumentation Lab), and arterial PO₂ (mm Hg) was recorded in all patients.

Biplane right and left ventricular cineangiograms were recorded at 60 frames/sec in both anteroposterior and lateral projections and were used for volume determinations. Right and left ventricular ejection fractions were derived from these data. When magnification could not be corrected for by the usual 1.0-cm grid, we used the vertebral grid system for correction of magnification as described by Wagner and colleagues. Right ventricular volumes were calculated from tracings of cineangiograms of the right ventricle in end-diastole and end-systole by tracing with a sonic pen interfaced with Nova Computer, using Graham's modification of Simpson's rule. Similar to the technique advocated by Graham and co-workers, the outermost borders of the right ventricle were traced for volume determinations, even though this might result in overestimation of right ventricular volumes. Right ventricular volumes were corrected for surface area and were also expressed as a percentage of the expected normal using the regression equations derived by Graham et al.

Measurements of right ventricular inflow were obtained from biplane cineangiograms in the anteroposterior projection in diastole (fig. 1). The amplitude of the tricuspid valve, derived from standard echoangiographic techniques, was obtained in 11 of the 18 patients. Necropsy examination was performed in six of the eight patients who died after the postoperative hemodynamic and angiocardiographic investigation. The diameter of the tricuspid valve was recorded, and the postmortem dimensions were compared with the normal values of Rowlett et al.

Right ventricular growth was then evaluated in two major groups of patients. Group 1 includes patients in whom right ventricular-pulmonary artery continuity was surgically achieved, as demonstrated unequivocally by the postoperative hemodynamic and angiocardiographic study. Patients in whom right ventricular-pulmonary continuity was not achieved comprise group 2 (tables 1–3).

We also assessed the role of tricuspid regurgitation qualitatively at the initial catheter study in promoting right ventricular growth. Since all 18 patients had pulmonary atresia, we hypothesized that there might be a relationship between the degree of tricuspid
regurgitation, tricuspid valve or annulus size and ultimately, right ventricular size and growth. Tricuspid regurgitation was assessed qualitatively by selective right ventricular cineangiocardiograms filmed at 60 frames/sec. Only ectopic-free frames were evaluated for regurgitation. Tricuspid regurgitation was judged independently by three of the authors as mild, indicating faint opacification of the right atrium; moderate if there was denser opacification of a somewhat larger right atrium; and severe if there was very dense opacification of a very enlarged right atrium.

In the tables, mild tricuspid regurgitation is indicated by the designation A, a moderate degree of tricuspid regurgitation by B, and severe regurgitation by C. Thus group 1A, B, or C indicates patients in whom right ventricular–pulmonary artery continuity was surgically achieved, and in whom the initial catheter study indicated mild, moderate, or severe tricuspid regurgitation. Group 2A or C identifies patients in whom continuity was not achieved, and in whom tricuspid regurgitation was similarly judged as mild or severe. No patient had right ventricular–pulmonary artery discontinuity and moderate tricuspid regurgitation. (This notation is used throughout the text and in tables 1–3.)

Results

Identifying data, separating of patients in two groups 1 and 2 (presence or absence of right ventricular–pulmonary artery continuity), and their subgroups based on the initial degree of tricuspid regurgitation, types of surgery and pre- and postoperative right ventricular pressures are shown in table 1. Right ventricular–pulmonary artery continuity was established in 12 of the 16 patients who had initial pulmonary valvotomy (group 1). Preoperatively, 11 of these 12 had suprasystolic right ventricular pressures (ranging from 115–208% of peak systemic systolic pressure [mean 143%]). Postoperatively, right ventricular pressures were still either systemic or suprasystolic in nine patients (ranging from 115–186% of peak systemic systolic pressure). In two patients, the right ventricular peak systolic pressures was 54 mm Hg. The pulmonary artery was not probed during the initial study. Peak systolic pulmonary artery pressures ranged from 14–75 mm Hg (mean 31.8 mm Hg) at the postoperative investigation.

Six patients (33%) had right ventricular–pulmonary artery discontinuity (group 2) at the postoperative cardiac catheterization. Severe infundibular pulmonary artery discontinuity or atresia had precluded attempted pulmonary valvotomy in five of these six. Right ventricular pressure remained suprasystolic in all six group 2 patients (table 1).

Systemic arterial PO2 ranged from 17–31 torr (mean 24 torr) at the initial study in group 1 (before prostaglandin administration), and at the subsequent study ranged from 32–74 torr (mean 58 torr). Group 2 had a similar initial range (24–32 torr, mean 28 torr); at the subsequent study, systemic PO2 was lower, ranging from 25–48 torr (mean 40 torr).

Right Ventricular Volume Determinations

Preoperative right ventricular volumes calculated by Simpson’s rule and modified by Graham et al51, 60 reflect the severity of right ventricular hypoplasia (tables 2 and 3). Right ventricular end-diastolic volumes (RVEDV) were less than 2.0 ml in four patients (tables 2 and 3). Since RVEDV increases with age and body surface area, the right ventricular volumes are expressed as ml/m2 as well as percentage of expected normal, using the regression equations of Graham and colleagues51, 60 (tables 2 and 3).

The smallest RVEDVs were observed in the patients with mild and moderate tricuspid regurgitation (and presumably more obstructive tricuspid valves). Thus, patients in groups 1A, 1B and 2A, as shown in table 2 (individually) and summarized by groups in table 3, had the smallest RVEDV per square meter at the initial study, ranging from 11–22 ml/m2, or 19–45% of normal (tables 2 and 3). The changes in RVEDV vs body surface area are shown in figures 2A and B. In figure 2, the patients are divided into groups 1 and 2 and subgroups A–E. Right ventricular growth was maximal in patients with the most severe tricuspid regurgitation, whether or not right ventricular–pulmonary artery continuity was achieved. Right ventricular growth was least in group 2 patients who had mild tricuspid regurgitation (fig. 2B; table 3). Similarly, right ventricular growth was unsatisfactory in patients with a perforate but severely obstructive
right ventricular outflow tract and mild or moderate tricuspid regurgitation. Although an increment in right ventricular dimensions was observed in patients with initially moderate degrees of tricuspid regurgitation and continuity between the right ventricle and the pulmonary artery, right ventricular dimensions assumed nearly normal values at the postoperative study in only five patients (figs. 2A and B; tables 2 and 3).

Right ventricular ejection fraction was subnormal at the initial study and improved in most groups (table 3). The ejection fraction in the presence of tricuspid regurgitation may be spuriously high, but the relative improvement is encouraging. A summary of pre- and

<table>
<thead>
<tr>
<th>Group</th>
<th>Case</th>
<th>Age</th>
<th>SA (m²)</th>
<th>TVA (mm)</th>
<th>RV volume (ml/m²) ED</th>
<th>RVED volume (% of normal)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1A</td>
<td>1</td>
<td>1 day</td>
<td>0.19</td>
<td>8</td>
<td>14.2</td>
<td>33</td>
</tr>
<tr>
<td></td>
<td>8 mos</td>
<td>0.35</td>
<td>12</td>
<td>14</td>
<td>31</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>4 mos</td>
<td>0.24</td>
<td>8</td>
<td>13.8</td>
<td>33</td>
</tr>
<tr>
<td></td>
<td>5 yrs</td>
<td>0.59</td>
<td>12</td>
<td>17.8</td>
<td>32</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>1 day</td>
<td>0.19</td>
<td>9</td>
<td>17.8</td>
<td>45</td>
</tr>
<tr>
<td></td>
<td>5 yrs</td>
<td>0.64</td>
<td>11</td>
<td>19.6</td>
<td>35</td>
<td></td>
</tr>
<tr>
<td></td>
<td>4</td>
<td>1 day</td>
<td>0.21</td>
<td>—</td>
<td>16.6</td>
<td>41</td>
</tr>
<tr>
<td></td>
<td>1 yr, 10 mos</td>
<td>0.45</td>
<td>11</td>
<td>25.5</td>
<td>54</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2 yrs, 10 mos</td>
<td>0.49</td>
<td>12</td>
<td>27.5</td>
<td>52</td>
<td></td>
</tr>
<tr>
<td>1B</td>
<td>5</td>
<td>3 wks</td>
<td>0.2</td>
<td>12</td>
<td>7.6</td>
<td>19</td>
</tr>
<tr>
<td></td>
<td>6 mos</td>
<td>0.32</td>
<td>15</td>
<td>27.5</td>
<td>53</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1 day</td>
<td>0.22</td>
<td>15</td>
<td>15</td>
<td>36</td>
<td></td>
</tr>
<tr>
<td></td>
<td>7 mos</td>
<td>0.31</td>
<td>17</td>
<td>41.3</td>
<td>98</td>
<td></td>
</tr>
<tr>
<td></td>
<td>6 mos</td>
<td>0.28</td>
<td>11</td>
<td>20.7</td>
<td>44</td>
<td></td>
</tr>
<tr>
<td></td>
<td>4 yrs, 8 mos</td>
<td>0.61</td>
<td>17</td>
<td>38.5</td>
<td>74</td>
<td></td>
</tr>
<tr>
<td></td>
<td>8</td>
<td>1 day</td>
<td>0.19</td>
<td>15</td>
<td>21.7</td>
<td>33</td>
</tr>
<tr>
<td></td>
<td>7 yrs</td>
<td>0.83</td>
<td>22</td>
<td>36.8</td>
<td>63</td>
<td></td>
</tr>
<tr>
<td></td>
<td>9</td>
<td>1 day</td>
<td>0.17</td>
<td>14</td>
<td>7.1</td>
<td>26</td>
</tr>
<tr>
<td></td>
<td>4 yrs, 3 mos</td>
<td>0.61</td>
<td>17</td>
<td>23.7</td>
<td>47</td>
<td></td>
</tr>
<tr>
<td>1C</td>
<td>10</td>
<td>1 day</td>
<td>0.22</td>
<td>16</td>
<td>23.3</td>
<td>49</td>
</tr>
<tr>
<td></td>
<td>1 yr, 1 mo</td>
<td>0.31</td>
<td>20</td>
<td>63.8</td>
<td>142</td>
<td></td>
</tr>
<tr>
<td></td>
<td>11</td>
<td>1 wk</td>
<td>0.22</td>
<td>17</td>
<td>23.2</td>
<td>57</td>
</tr>
<tr>
<td></td>
<td>6 mos</td>
<td>0.31</td>
<td>20</td>
<td>85.1</td>
<td>192</td>
<td></td>
</tr>
<tr>
<td></td>
<td>12</td>
<td>1 day</td>
<td>0.21</td>
<td>16</td>
<td>16.2</td>
<td>40</td>
</tr>
<tr>
<td></td>
<td>3 mos</td>
<td>0.23</td>
<td>19</td>
<td>43.4</td>
<td>111</td>
<td></td>
</tr>
<tr>
<td>2B</td>
<td>13</td>
<td>1 day</td>
<td>0.18</td>
<td>7</td>
<td>8.9</td>
<td>17</td>
</tr>
<tr>
<td></td>
<td>7 mos</td>
<td>0.23</td>
<td>8</td>
<td>9.5</td>
<td>22</td>
<td></td>
</tr>
<tr>
<td></td>
<td>14</td>
<td>1 mo</td>
<td>0.23</td>
<td>10</td>
<td>17.3</td>
<td>46</td>
</tr>
<tr>
<td></td>
<td>4 yrs, 3 mos</td>
<td>0.64</td>
<td>14</td>
<td>14.8</td>
<td>27</td>
<td></td>
</tr>
<tr>
<td></td>
<td>15</td>
<td>4 days</td>
<td>0.22</td>
<td>12</td>
<td>8.7</td>
<td>21</td>
</tr>
<tr>
<td></td>
<td>3 yrs, 5 mos</td>
<td>0.58</td>
<td>15</td>
<td>23.6</td>
<td>45</td>
<td></td>
</tr>
<tr>
<td></td>
<td>16</td>
<td>1 day</td>
<td>0.17</td>
<td>8</td>
<td>20.5</td>
<td>53</td>
</tr>
<tr>
<td></td>
<td>2 yrs</td>
<td>0.41</td>
<td>12</td>
<td>20.7</td>
<td>46</td>
<td></td>
</tr>
<tr>
<td></td>
<td>17</td>
<td>1 day</td>
<td>0.21</td>
<td>8</td>
<td>15.4</td>
<td>41</td>
</tr>
<tr>
<td></td>
<td>4 yrs, 3 mos</td>
<td>0.72</td>
<td>11</td>
<td>23.8</td>
<td>42</td>
<td></td>
</tr>
<tr>
<td></td>
<td>9 yrs</td>
<td>0.89</td>
<td>12</td>
<td>23</td>
<td>38</td>
<td></td>
</tr>
<tr>
<td>2C</td>
<td>18</td>
<td>1 day</td>
<td>0.21</td>
<td>15</td>
<td>20.5</td>
<td>51</td>
</tr>
<tr>
<td></td>
<td>9 wks</td>
<td>0.36</td>
<td>18</td>
<td>50.5</td>
<td>138</td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: SA = surface area; TVA = tricuspid valve annulus; RV = right ventricular; ED = end-diastole; RVED = right ventricular end-diastolic.
Table 3. Summary of Right Ventricular End-systolic and End-diastolic Volume Measurements and Right Ventricular Ejection Fraction Based on Right Ventricular-to-Pulmonary Artery Patency (Group 1) and Right Ventricular-to-Pulmonary Artery Nonpatency (Group 2) at the Second Study

<table>
<thead>
<tr>
<th>Group</th>
<th>Preoperative values</th>
<th>Postoperative values</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>RVESV (ml/m²)</td>
<td>RVEDV (% normal)</td>
</tr>
<tr>
<td>1A</td>
<td>n = 4</td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>(15.6)</td>
<td>(38)</td>
</tr>
<tr>
<td>1B</td>
<td>n = 5</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(14.4)</td>
<td>(31.6)</td>
</tr>
<tr>
<td>1C</td>
<td>n = 3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(20.9)</td>
<td>(48.6)</td>
</tr>
<tr>
<td>2A</td>
<td>n = 5</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(14.2)</td>
<td>(35.6)</td>
</tr>
<tr>
<td>2C</td>
<td>n = 1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(20.6)</td>
<td>(51)</td>
</tr>
</tbody>
</table>

Subgroups A, B, and C are based on the tricuspid valve measurements and the degree of tricuspid valve incompetence as indicated.

Abbreviations: RVESV = right ventricular end-systolic volume; RVEDV = right ventricular end-diastolic volume; EF = ejection fraction.

Figure 2. A) Right ventricular (RV) end-diastolic volumes in 12 patients with pulmonary valve patency at postoperative catheter study (group 1) vs surface area. Open squares represent group 1A, closed squares group 1B, and open triangles group 1C. TVA = measured tricuspid valve annulus; TI = tricuspid regurgitation; RV/PA = right ventricular-pulmonary artery. B) RV end-diastolic volumes in six patients without pulmonary valve patency at postoperative catheter study (group 2). Closed triangles represent group 2A, and the open circles group 2C.
postoperative RVEDVs in each subgroup is shown in figure 3. The ejection fraction showed little change in group 2A.

The Tricuspid Valve: Correlation between Angiographically and Echocardiographically Derived Dimensions and Correlation with Right Ventricular Dimensions and Growth

The tricuspid annulus was measured in each patient in an anteroposterior right ventricular cineangiocardiogram corrected for magnification, and these values were compared with the echo measurement of the tricuspid valve amplitude (fig. 4). Good correlation \( r = 0.79 \) was achieved. Although the number of patients in each subgroup was small, the patient assessed qualitatively as having only mild tricuspid regurgitation had the smallest tricuspid valve dimension as measured from magnification corrected right ventriculogram (i.e., groups 1A and 2A; tables 2 and 3). Slight overlap in angiographically derived tricuspid valve dimension was found between groups 1A and 1B. Similarily, in the patients with qualitatively the most severe degrees of tricuspid regurgitation, the angiographically derived tricuspid valve measurement had the largest dimension. Thus, group 1A had the smallest tricuspid valve measurements; group 1B had moderate tricuspid valve dimensions, and Group 1C, the largest tricuspid valve dimensions. Therefore, these groups also represent relative tricuspid valve dimension and can be evaluated in terms of change in right ventricular dimension (tables 2 and 3; figs. 2A and B, 3 and 4).

When the anteroposterior measurement of the tricuspid valve ring was less than 12 mm, the right ventricular volume was the smallest (group 1A, table 2), and the degree of tricuspid regurgitation was assessed as mild (implying a very obstructive tricuspid valve). This dimension was consistently 9–11 mm initially, and 11–12 mm at the subsequent study (table 2). Figures 2, 3 and 5 show that among patients with mild tricuspid regurgitation and a small tricuspid annulus, right ventricular growth was not achieved. In contrast, the largest tricuspid annulus size, ranging from 16–18 mm at the initial study and 19–20 mm at the postoperative study, were observed in the three group 1 patients with qualitatively severe tricuspid regurgitation. These three patients did achieve a normal right ventricular dimension (figs. 2, 3 and 6). Among five other group 1 patients, the tricuspid valve annulus size ranged from 11–15 mm at the initial study and 15–22 mm at the postoperative study. Among five of six group 2 patients in whom the initial degree of tricuspid regurgitation was assessed as mild, the tricuspid annulus size was initially 7–12 mm and 8–15 mm subsequently. In the only group 2 patient in whom the initial degree of tricuspid regurgitation was assessed as severe (patient 18, tables 1 and 2), the tricuspid annulus initially measured 15 mm and postoperatively was 18.0 mm.

The tricuspid valve orifice was measured at postmortem examination in six patients (table 2). While the actual dimensions obtained by the three methods (angiography, echocardiography and postmortem) are not the same, a distinction can be made between those with a small tricuspid valve annulus and those with a large tricuspid valve annulus. Angiocardiography measures the annulus size; at postmortem, the tricuspid orifice is measured. The echocardiogram only reflects the amplitude of the tricuspid valve motion, and we believe that the amplitude is reduced in patients with tricuspid stenosis. The amplitude is probably nearer normal in those with severe tricuspid incompetence. A comparison of tricuspid valve an-

![Figure 3](http://circ.ahajournals.org/)

**Figure 3.** Mean right ventricular (RV) end-diastolic volumes expressed as percentage of expected volumes in the various groups. Open squares represent group 1A; closed squares, group 1B; open triangles, group 1C; closed triangles represent group 2A; and open circles, group 2C. Abbreviations: see figure 2.
Discussion

Graham and colleagues have emphasized the inability to characterize right ventricular size correctly without volume measurements, and have called attention to errors incurred by using either routine chest

Myocardial Sinusoidal–Coronary Artery Communications

Ten of the 18 patients had extensive myocardial sinusoidal–coronary artery communications. These seemed to be the most extensive in those patients with the most diminutive and hypertensive right ventricles (groups 1A and 2A). In nine patients at the initial study, dense opacification of the left anterior descending coronary artery with retrograde opacification of the aortic sinus of Valsalva was recorded. In only one patient was dense retrograde visualization of the right coronary artery achieved by right ventricular angiocardiography. Of the 12 group 1 patients, these sinusoids persisted in only two. Among the six group 2 patients, four had these abnormal communications initially; at the second study, little change in the angiocardiographic appearance had occurred (fig. 5). In four patients, right ventricular growth had occurred, and it appeared that the area of growth incorporated the region of extensive myocardial sinusoidal–trabecular spaces (fig. 6). At the initial study, there was no difference in peak systolic pressure among patients with and those without sinusoids.

Figure 4. A comparison of tricuspid valve annulus dimension measured from anteroposterior right ventriculogram vs tricuspid valve amplitude obtained from M-mode echocardiogram. X represents group 1A; open circles, group 1B; closed circles, group 1B; and closed diamonds, group 2A.

Figure 5. Case 17. Anteroposterior right ventriculograms in a patient with pulmonary atresia, intact ventricular septum and diminutive right ventricle (RV). The tricuspid valve dimension is small (black arrows) and the initial degree of tricuspid regurgitation was assessed as mild. Pulmonary artery patency was not achieved as determined at the postoperative catheter study. There was minimal change in right ventricular dimension, and myocardial sinusoids persisted. A) Neonatal study. B) Anteroposterior right ventriculogram at 9 years, with persistent severe ventricular underdevelopment.
radiography or qualitative assessment of right ventricular angiograms as indicators of right ventricular dimensions. We are also aware of the real and potential errors of applying currently used right ventricular volume quantitative methods to the diminutive, bizarrely shaped right ventricle, with its "astral" projection of sinusoids, fistulous coronary artery communications and trabecular spaces. In Graham's study, right ventricular shape was characterized as not grossly abnormal in five of seven patients. This experience is somewhat unusual and reflects the relatively small number of patients studied. We agree with Graham that the Simpson's rule method is theoretically more applicable to the abnormally shaped ventricle. Furthermore, we have included the outermost borders of the right ventricle in our images for calculation and have attempted to smooth out any irregularities in these borders, partially obviating objections to performing these measurements on an irregularly shaped ventricle. Such a method, however, probably does overestimate right ventricular cavity size. Thus, this quantitative approach must be interpreted within the limitations of the method. Previous studies have suggested that right ventricular dimensions tend to increase after achievement of right ventricular-pulmonary artery continuity. Several mechanisms may cause enhanced right ventricular filling and, presumably, growth, including: 1) improved right ventricular emptying; 2) ameliorating functional and organic obstruction at tricuspid valve/annulus level, which augments right ventricular preload; 3) reducing right-to-left shunting at the atrial level; 4) improving right ventricular compliance; and 5) augmenting right ventricular volumes by varying degrees of pulmonary and tricuspid valve regurgitation. All of these factors (and possibly more) are important to augmenting right ventricular filling; but if right ventricular emptying is not significantly improved, these other factors may prove trivial.

Right ventricular-pulmonary artery continuity was surgically achieved in 12 patients (group 1), but in only two were the postoperative right ventricular pressures significantly systemic. Coincident with the relief of severe right ventricular hypertension in these two patients was an increase in right ventricular dimensions. Our own disappointing results made it difficult to assess the role that the establishment of such continuity and relief of severe right ventricular hypertension have on right ventricular growth. However, one can provide data on the morphologic basis for failure to achieve right ventricular-pulmonary artery continuity. Among patients with pulmonary atresia and intact ventricular septum, it is the exception rather than the rule to have isolated pulmonary valve fusion and atresia with a widely patent and nonobstructive pulmonary infundibulum. Both morphologic observations and the double-catheter angiographic techniques advocated by Freedom et al. have demonstrated to advantage the severe infundibular hypoplasia and underdevelopment that often accompany pulmonary valve atresia. Among our 18 patients, severe infundibular obstruction or atresia accompanied pulmonary valve atresia in 14 patients (78%). A transarterial pulmonary valvotomy was performed in 12 patients, and in two, right ventricular hypertension was significantly relieved. The morphologic observations of Arom and Edwards help to clarify the reasons for these surgical results. Their study of the relationship between right ventricular muscle bundles and pulmonary valve among 18 patients with pulmonary atresia and intact ventricular septum suggested that there is only a small segment of the atretic valve that is in direct contact with the right ventricular cavity. They observed that the least prominent subpulmonary muscle bundle is related to the anterior pulmonary cusp and sinus, and that a valvotomy through the region of the anterior cusp and sinus is more apt to provide direct continuity with the right ventricle. If the double-catheter technique is performed at the initial catheter study, this should demonstrate the morphologic bases for the pulmonary atresia. Certainly, the longer the segment of infundibular atresia, the less likely that satisfactory right ventricular-pulmonary artery continuity will be established by valvotomy.

Although we can hope to achieve right ventricular pulmonary continuity with decompression of the hypertensive right ventricle with surgical intervention, only in the past few years have we begun to appreciate fully the role of the tricuspid valve in this disorder. The tricuspid valve among patients with pulmonary atresia and intact ventricular septum can have a wide range of morphologic features. The valve can be grossly insufficient, with features of Ebstein's anomaly or profound dysplasia. In some patients, despite pulmonary atresia, right ventricular pressures will be systemic or even "low" in a neonate, with a normal or enlarged right ventricular cavity and extreme tricuspid valve regurgitation. In these patients, the right ventricular myocardium may be extremely thinned or deficient, as in Uhl's anomaly. The therapeutic problems posed by these patients are considerably different from those in the patient with a grossly small and hypertensive right ventricle, and will not be considered further.

However, among patients with pulmonary atresia, intact ventricular septum and small or diminutive right ventricle, the tricuspid valve usually assumes a dominant obstructive role. Although some observers have suggested that the tricuspid valve in this situation is hypoplastic, though normally formed, our experience and an extensive literature strongly dispute this. The tricuspid valve ring is usually grossly underdeveloped, assuming a circumference proportionate to the right ventricular cavity size. The free valve margin is often thickened; the chordal attachments are poorly excavated, and the papillary muscles are often hypoplastic. Our observations, based on a necropsy study of 60 patients, suggest that inflow obstruction at the tricuspid annulus and valve level pose a serious, if not irreparable disturbance, es-
FIGURE 6. Case 8. Pre- and postoperative anteroposterior and lateral right ventricular cineangiograms showing that excellent right ventricular growth was achieved after neonatal pulmonary valvotomy and Pott's shunt. A and B) Anteroposterior and lateral right ventricular (RV) cineangiogram in a neonate. In B, the tricuspid valve is well seen (black arrows) and the distal infundibulum is considerably narrowed. C and D) Anteroposterior and lateral angiograms at 7 months. Excellent growth was achieved, and the pulmonary artery (PA) opacifies from the right ventriculogram.

especially in the patient with a diminutive right ventricular cavity. Even with relief of severe right ventricular hypertension in some of these patients, profound obstruction at right ventricular inflow will preclude restoration of a hemodynamically normal circulation. Although our data are still preliminary, it is important to try to quantitate tricuspid annulus (and valve) size, as it may have relevance to predicting right ventricular growth.

Inadequate right ventricular distensibility or compliance may also prevent right ventricular filling even when the right ventricle is satisfactorily decompressed. This lack of compliance may result from a combination of severe myocardial hypertrophy, fibrosis, and endocardial fibroelastosis and sclerosis. Although inadequate right ventricular compliance may be reversible, long-standing right ventricular hypertension and myocardial free-wall and septal hypertrophy may certainly alter right (and left) ventricular myocardial oxygen supply-demand ratio, culminating in ischemia, cell death and fibrosis. Such changes are probably not fully reversible, and underscore the need for early right ventricular decompression.

Anomalous myocardial sinusoidal–coronary artery fistulous communications complicate pulmonary atresia, intact ventricular septum and hypoplastic, hypertensive right ventricle in 8–30% of patients. Initially regarded as a pathologic curiosity, they may have a more ominous role. We suggest that extensive fistulous communications with the coronary arteries, particularly the left, may predispose the left ventricular subendocardial myocardium to ischemia and
fibrosis. Such fistulous communications do serve as one site for egress of right ventricular blood, and either the left, or less often the right, and infrequently both coronary arteries in a patient may participate in these primitive communications (although some sinusoids appear to terminate blindly into the myocardium). Rarely, as in the case reported by Lenox and Briner, the proximal connections between aorta and coronary arteries will be absent, with right ventricular sinusoidal communications providing continuity with the coronary arteries. In some patients with these fistulous communications, the involved coronary arteries may have a variety of histopathologic alterations, including nodularity, beading, and obliterator endarteritis, all of which serve mechanically to compromise oxygen transport.

With pulmonary atresia, a nearly competent, obstructive tricuspid valve, and a grossly underdeveloped right ventricle, right ventricular systolic ejection time is markedly prolonged, and retrograde filling of the coronary arteries, and their aortic origin and sinus of Valsalva can be demonstrated by selective right ventricular angiograms. This well-documented phenomenon can and does occur during aortic diastole and thus during maximal coronary artery perfusion. One would anticipate that oxygen transport to that portion of the left ventricle supplied by the involved segment of the left anterior coronary artery would be either marginal or inadequate, resulting in a discrepancy between myocardial oxygen demand and delivery. The histopathologic correlates to this imbalance, i.e., left ventricular myocardial ischemia, necrosis and fibrosis, have been amply documented in this disorder.

The morphogenesis of these communications rests with the abnormal persistence of the embryonic pattern of myoarchitecture with its lacunary blood supply. Such postnatal persistence of spongy myocardium with its embryonic blood supply is most frequently recorded in patients with pulmonary atresia, intact ventricular septum and small and hypertensive right ventricle; but these findings have also been observed in patients with severe aortic stenosis, endocardial fibroelastosis, anomalous left coronary artery from the pulmonary artery, obliterator fibroelastosis of the coronary arteries, complex congenital heart disease associated with dextrocardia and situs inversus, and in three patients with an imperforate tricuspid valve, right ventricular tensor apparatus, and congenital absence of the pulmonary valve. One assumes that with effective right ventricular decompression, obliterator of the fistulous communication will result, and that right ventricular growth is accompanied by incorporation of the embryonal pattern of myoarchitecture into a more normal right ventricle. Our unpublished observations, and those of others, suggest that the spongy myocardial and sinusoidal–coronary artery communications will be absorbed into the ventricular mass, with relief of right ventricular hypertension. The natural history of this primitive myocardial architecture thus needs further definition.

In conclusion, it is probable that long-term functional and hemodynamic improvement in these patients is predicated on 1) establishing right ven-
tricular–pulmonary artery continuity, with significant reduction in right ventricular hypertension; the direct corollary to this is promoting both right ventricular emptying and filling and, therefore, growth; 2) ameliorating functional and organic disturbances of right ventricular inflow; 3) preventing right ventricular endomyocardial sclerosis and fibrosis and thus minimizing alterations in right ventricular compliance; and 4) protecting the integrity of left ventricular subendocardial myocardium, potentially jeopardized by the cumulative effects of chronic hypoxemia, volume overload and by alterations of myocardial perfusion resulting from the presence of right ventricular myocardial sinusoid–coronary artery communications, and coronary arterial distortions intrinsic to and secondary to these anomalous communications. Obviously, adequate “early” decompression of the diminutive or small and hypertensive right ventricle is mandatory if most of the above objectives are to be achieved, but the appropriate timing for this is uncertain. Our recent experience has suggested that the fragile neonate does not tolerate pericardial right ventricular outflow tract reconstruction. Thus, our medical and surgical approach to these infants includes: 1) balloon septostomy at the initial diagnostic cardiac catheterization; 2) prostaglandin infusion before and during cardiac surgery,23 3) transpulmonary valvotomy and construction of a systemic–pulmonary artery anastomosis.27 Then, at or before 6 months of age, we advocate a second complete hemodynamic and angiocardiographic investigation. If right ventricular–pulmonary artery continuity has not been achieved, or if severe right ventricular hypertension persists (right ventricular pressure greater than 70% systemic), right ventricular outflow tract reconstruction with autologous pericardium is performed with resection of infundibular muscle. During the same procedures, the tricuspid valve ring is measured, and if the tricuspid valve approximates 60% or more of the normal, the patent foramen ovale is carefully narrowed, but not closed. The latter maneuver is obviously designed to augment right ventricular filling, although in some patients with a grossly obstructive tricuspid valve ring and apparatus, narrowing of the foramen ovale is not indicated. Finally, we would suggest a third full cardiac catheter assessment 1 year to 18 months after right ventricular outflow tract reconstruction. We recognize that significant obstruction at the tricuspid annulus or valve level may preclude restoration of a hemodynamically normal circulation despite normal right ventricular pressures. The tricuspid valve could be oversewn, with continuity between right atrium and either right ventricle or pulmonary artery provided by a Fontan or Kreutzer type operation.88, 89

References
25. Miller GAH, Restifo M, Shinebourne EA, Paneth M, Joseph MC, Lenox SC, Kerr IH: Pulmonary atresia with intact ven-
tricular septum and critical pulmonary stenosis presenting in first month of life. Investigation and surgical results. Br Heart J 35; 9, 1973


50. Aziz KU, Olley PM, Rowe RD, Trusler GA, Mustard WT: Survival after systemic to pulmonary arterial shunt in infants less than 30 days old with obstructive lesions of the right heart chambers. Am J Cardiol 36: 479, 1975

51. Graham TP Jr, Bender HW, Atwood GF, Page DL, Sell CGR: Increase in right ventricular volume following valvulotomy for pulmonary atresia or stenosis with intact ventricular septum. Circulation 50 (suppl II): II-69, 1974

52. Luckstead EF, Mattioli L, Crosby IK, Reed W, Diehl A: Two stage palliative surgical approach for pulmonary atresia with intact ventricular septum (type I). Am J Cardiol 29: 490, 1972


55. Sauder U, Mocellin V, 508 720, 1971


69. Goeckel RJ, Carlsson E: Calculation of right and left cardiac volumes. Invest Radiol 2: 360, 1967


73. Arcilla RA, Gasul BM: Congenital asplasia or marked hypoplasia of the myocardium of the right ventricle (Uhl's anomaly). J Pediatr 58: 381, 1961
74. Oakley CM, Brainbridge MV, Bentall HH: Reversed interatrial shunt following complete relief of pulmonary valve stenosis. Br Heart J 26: 662, 1964
76. Haworth SG, Shinebourne EA, Miller GAH: Right to left interatrial shunting with normal right ventricular pressure. Br Heart J 37: 386, 1975
Right ventricular volume determinations in 18 patients with pulmonary atresia and intact ventricular septum. Analysis of factors influencing right ventricular growth.
R G Patel, R M Freedom, C A Moes, K R Bloom, P M Olley, W G Williams, G A Trusler and R D Rowe

Circulation. 1980;61:428-440
doi: 10.1161/01.CIR.61.2.428

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/61/2/428