Balloon occlusion of atrial septal defect to assess right ventricular capability in hypoplastic right heart syndrome

JOHN L. BASS, M.D., BRADLEY P. FUHRMAN, M.D., AND JAMES E. LOCK, M.D.

ABSTRACT Early surgery for forms of hypoplastic right heart syndrome may increase right ventricular size but could leave the patient with a residual right-to-left atrial shunt. Previous attempts to assess the capability of the right ventricle to accept systemic venous return have relied on angiographic estimates of tricuspid valve and right ventricular sizes. Since the minimum adequate sizes have not been established, we used a more physiologic technique of temporarily occluding the interatrial communication with a balloon-tipped catheter at cardiac catheterization in six consecutive patients. Five patients tolerated complete occlusion, although the tricuspid valve anulus diameter was less than the fifth percentile in all, and right ventricular volume was less than the fifth percentile in four. These five underwent surgical closure of an interatrial communication without evidence of postoperative systemic venous hypertension. Attempted occlusion in the sixth patient caused profound systemic venous hypoxia and surgical closure was not attempted. Temporary balloon occlusion may improve selection of patients for definitive operation.


INITIAL SURGICAL MANAGEMENT of patients with pulmonary atresia and intact ventricular septum consists of establishing adequate pulmonary blood flow and achievement of right ventricle-to-pulmonary artery continuity.1–3 This approach has been associated with an increase in right ventricular dimensions in some patients.4–5 Despite right ventricular growth there is often a residual right-to-left shunt at the atrial level as a result of “resistance” to right ventricular inflow.5–8 Factors that may be responsible for this include anatomic abnormalities such as tricuspid stenosis, tricuspid regurgitation, or a small right ventricular chamber, as well as functional abnormalities such as diminished right ventricular contractility or decreased right ventricular compliance from hypertrophy, fibrosis, or a previous ventriculotomy.5–8 Because of symptoms from hypoxia and the risk of paradoxical embolus and brain abscess, these patients must be considered for separation of systemic and pulmonary circulations. If the resistance to right ventricular inflow is too great, however, marked elevation of right atrial pressure may follow closure of the interatrial communication.

Selection of patients for separation of pulmonic and systemic circulations may be difficult. This decision generally depends on angiographic measurements of the tricuspid valve diameter,3 estimates of right ventricular volume, and the anatomy of the right ventricle. Neither the minimum tricuspid valve diameter nor the right ventricular volume that will accommodate total systemic blood flow has been established,8 and these methods do not assess the functional capacity of the right ventricle. We therefore sought a more direct and physiologic method of assessing right ventricular capability and applied the technique of temporary occlusion of the atrial septal defect with a balloon-tipped catheter. We also compared temporary atrial septal defect occlusion with conventional angiographic evaluation of the right ventricle.

Methods

Six consecutive patients, who were evaluated for elective closure of an atrial septal defect because of a residual right-to-left shunt after initial operation, formed the study group (table 1). One had tetralogy of Fallot and five had pulmonary atresia with intact ventricular septum. The patient with tetralogy of Fallot underwent surgical repair at 4 years of age, and surgery included ventricular septal defect closure and placement of a right ventricular outflow patch that did not cross the pulmonary anulus. Cyanosis persisted, and postoperative cardiac catheter-
TABLE 1
Clinical findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at study (yr)</th>
<th>Hgb (g/dl)</th>
<th>Symptoms</th>
<th>Diagnosis</th>
<th>Previous surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>9 ½</td>
<td>16.7</td>
<td>Mild exercise intolerance</td>
<td>Tetralogy of Fallot, tricuspid stenosis</td>
<td>Pulmonary valvulotomy, RV outflow patch, VSD closure</td>
</tr>
<tr>
<td>2</td>
<td>1</td>
<td>14.2</td>
<td>Asymptomatic</td>
<td>Pulmonary atresia</td>
<td>RV outflow patch</td>
</tr>
<tr>
<td>3</td>
<td>1 ½</td>
<td>15.2</td>
<td>Diaphoresis with exertion</td>
<td>Pulmonary atresia</td>
<td>Pulmonary valvulotomy</td>
</tr>
<tr>
<td>4</td>
<td>1½</td>
<td>17.9</td>
<td>Minimal symptoms</td>
<td>Pulmonary atresia</td>
<td>Pulmonary valvulotomy</td>
</tr>
<tr>
<td>5</td>
<td>2 ½</td>
<td>16.2</td>
<td>Asymptomatic</td>
<td>Pulmonary atresia</td>
<td>RV outflow patch, central shunt</td>
</tr>
<tr>
<td>6</td>
<td>10 ½</td>
<td>16.7</td>
<td>Asymptomatic</td>
<td>Pulmonary atresia</td>
<td>RV outflow patch, Waterston shunt</td>
</tr>
</tbody>
</table>

Hgb = hemoglobin; RV = right ventricular; VSD = ventricular septal defect.

ization showed low right ventricular pressures, an atrial septal defect with a right-to-left shunt, and a doming tricuspid valve. Of the five patients with pulmonary atresia and intact ventricular septum, two had a systemic-to-pulmonary artery communication placed in the first week of life, followed by establishment of right ventricle-to-pulmonary artery continuity at 3 months and 7 years of age, respectively. In the remaining three patients, a pulmonary valvulotomy was performed in two, and a transanular right ventricular outflow patch was placed in the other during the first week of life. Cardiac catheterization was performed in all six patients to determine the capability of the right ventricle to accept systemic

FIGURE 1. Roentgenogram demonstrating a modified atrial septostomy catheter, inflated with contrast medium in the left atrium and withdrawn to occlude the interatrial communication. A second catheter is positioned in the thoracic descending aorta to monitor pressure and oxygen saturation.
blood flow. Ages at cardiac catheterization ranged from 11 months to 10 years. All patients were cyanotic, with hemoglobin levels ranging from 14.2 to 17.9 g/dl. Three patients (Nos. 1, 3, and 4) had mild exercise intolerance and three were asymptomatic. All hemodynamic evaluations were performed before angiography.

**Balloon occlusion.** To determine the ability of the right ventricle to accept systemic blood flow, temporary occlusion of the interatrial communication was attempted in each patient. The occluding catheter was advanced from the systemic venous circulation into the left atrium, the balloon was inflated, and the catheter was withdrawn so as to occlude the atrial septal defect (figure 1). In two patients, the atrial septal defect was occluded with standard balloon-tipped angiocatheters (Critikon, Inc.). A large interatrial communication in the other four patients necessitated the use of specially modified atrial septostomy catheters (Edwards Laboratories). These catheters can be inflated to a larger volume (up to 4 cc with a 22 mm diameter) and have a catheter length of 100 cm so that the atrial septum can be crossed in older patients. Two patients had a systemic-to-pulmonary artery shunt that was occluded with a second balloon-tipped catheter at the time of atrial septal defect occlusion. Systemic venous pressure and saturation were measured during occlusion through a venous catheter or through a side port in the venous sheath connector. Each patient also had an arterial line placed to monitor systemic arterial pressure and saturation. Occlusion of the interatrial communication was considered complete if the right-to-left shunt was eliminated, resulting in a systemic arterial oxygen saturation of 94% or greater. We recommended surgical closure of the atrial septum in patients in whom occlusion was complete without a significant change in right atrial and aortic pressures or a fall in systemic venous oxygen saturation.

**Angiography.** Right ventricular cineangiograms were performed in each patient after hemodynamic evaluation. In patients 1 through 5, biplane angiography was performed. The angiograms were examined for tricuspid regurgitation and right ventricular anatomy. Measurements of the tricuspid valve annulus were obtained in the anteroposterior projection in diastole, with the known diameter of the catheter used to correct for magnification. These estimates were compared with the expected tricuspid valve dimension for body surface area of normal children's hearts available from the studies of Rowlatt et al., 10 corrected by a factor of 1.43 for shrinkage of preserved autopsy specimens. The deviation from the expected orifice diameter was expressed as the number of standard errors of the estimate above or below the mean (Z number). Right ventricular volumes were calculated from tracings of the right ventricle in end-diastole with a sonic pen digitizer and a Control Data Corp. computer with Graham's modification of Simpson's rule. Right ventricular volumes were expressed as a percentage of the expected volume for body surface area with equations derived by Graham et al. 11

**Results**

**Hemodynamics.** Each patient was found to have an atrial level right-to-left shunt between 24% and 45% (table 2). Right ventricular systolic pressure was normal in five patients with gradients of 1 to 6 mm Hg across the right ventricular outflow tract. One patient (No. 3) had residual suprasystemic right ventricular pressure. Right ventricular end-diastolic pressure was elevated above 10 mm Hg in two patients (Nos. 3 and 6). No significant gradient across the tricuspid valve (< 4 mm Hg) was found in four of the patients. One patient (No. 5) had a 16 mm Hg gradient across the tricuspid valve. In the remaining patient (No. 6) the right atrial a wave was consistently less than the right ventricular end-diastolic pressure.

**TABLE 2**

**Hemodynamic findings**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Time of measurement</th>
<th>Right atrial mean pressure (mm Hg)</th>
<th>Right ventricular systolic pressure (mm Hg)</th>
<th>Aortic mean pressure (mm Hg)</th>
<th>Saturation (%) Systemic venous</th>
<th>Aorta</th>
<th>Right-to-left shunt (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Baseline</td>
<td>6</td>
<td>24</td>
<td>100</td>
<td>78</td>
<td>90</td>
<td>0</td>
<td>40</td>
</tr>
<tr>
<td>2 Baseline</td>
<td>7</td>
<td>---</td>
<td>100</td>
<td>78</td>
<td>95</td>
<td>0</td>
<td>39</td>
</tr>
<tr>
<td>3 Baseline</td>
<td>5</td>
<td>20</td>
<td>95</td>
<td>63</td>
<td>83</td>
<td>0</td>
<td>35</td>
</tr>
<tr>
<td>4 Baseline</td>
<td>6</td>
<td>---</td>
<td>92</td>
<td>85</td>
<td>94</td>
<td>0</td>
<td>45</td>
</tr>
<tr>
<td>5 Baseline</td>
<td>7</td>
<td>18</td>
<td>63</td>
<td>55</td>
<td>77</td>
<td>0</td>
<td>45</td>
</tr>
<tr>
<td>6 Baseline</td>
<td>8</td>
<td>---</td>
<td>67</td>
<td>63</td>
<td>94</td>
<td>0</td>
<td>50</td>
</tr>
<tr>
<td>1 Baseline</td>
<td>12</td>
<td>24</td>
<td>75</td>
<td>58</td>
<td>87</td>
<td>0</td>
<td>24</td>
</tr>
<tr>
<td>2 Baseline</td>
<td>10</td>
<td>19</td>
<td>74</td>
<td>62</td>
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<tr>
<td>3 Baseline</td>
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<td>---</td>
<td>100</td>
<td>23</td>
<td>61</td>
<td>50</td>
<td></td>
</tr>
</tbody>
</table>
**Balloon occlusion.** Attempted balloon occlusion of the interatrial communication eliminated the right-to-left shunt in five patients, resulting in aortic saturations of 94% to 97%. Occlusion was maintained for an average of 20 min (range 5 to 40). Mean systemic venous pressure rose 1 to 2 mm Hg. Mean aortic pressure was measured during occlusion in four of the five patients and did not change significantly (average 0.0 ± 2.9 mm Hg). Systemic venous saturation rose an average of 8% in the four patients in whom it was measured. These findings were interpreted as indicating that separation of systemic and pulmonary circulations would be well tolerated.

All five of these patients have subsequently had surgical closure of an interatrial communication, and one patient underwent ligation of a systemic-to-pulmonary artery shunt. Surgery, the immediate postoperative period, and follow-up (1 to 27 months) have been uncomplicated, with relief of cyanosis and improvement of exercise tolerance in all five. No patient has had a subsequent cardiac catheterization, but mean right atrial pressures in the immediate postoperative period were low (average 9.6 mm Hg), and no clinical signs of elevated systemic venous pressure were evident.

In the remaining patient (No. 6), occlusion of the interatrial communication was not complete, although systemic venous pressure rose. After 2 min of obstruction of a shunt and the atrial septal defect, the aortic saturation fell, the systemic venous saturation plunged to 23%, and mean systemic venous pressure rose 4 mm Hg. These changes indicated that the right ventricle was incapable of accepting total systemic venous blood flow, and surgical closure of the atrial septal defect was not attempted.

**Angiography.** Angiographic estimates of the tricuspid valve anulus diameter were consistently less than expected for the patients’ body surface area. In the five patients with successful balloon occlusion, the observed tricuspid valve diameter was 2.2 to 4.1 SE below the expected mean (figure 2). The patient who failed balloon occlusion had a Z number of −11, indicating a tiny tricuspid valve. While it might be anticipated that the latter patient would be unable to accept systemic venous flow across the tricuspid valve anulus, the other five patients also had tricuspid valve diameters below the fifth percentile. In patient 5, the tricuspid valve orifice was 15 mm at surgery and the angiographic estimate of the anulus diameter was 20 mm, suggesting that the anulus diameter may not reflect the true state of the tricuspid valve.

Right ventricular volumes from right ventricular cineangiograms could be obtained in patients 1 through 5 (table 3). These volumes, expressed as a percentage of that expected for body surface area,11 ranged from 66% to 118%. With the exception of patient 2, all volumes were below the fifth percentile. In patients 2 and 5, a right ventricular outflow tract patch had been placed that was included in volume determinations (figure 3). There was no clear relationship between angiographically determined tricuspid valve diameter and right ventricular volume in this small group of patients, unlike the results of previous reports.12 In spite of the small right ventricular volumes, all five patients underwent successful surgical closure of the atrial septal defect.

Tricuspid regurgitation was minimal in all but one patient (No. 5), who had 2+ insufficiency. All three portions of the right ventricle (sinus, trabecular, and

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**FIGURE 2.** Graphic representation of the tricuspid valve annulus diameters in the six study subjects. The Z number indicates the number of standard errors of the estimate above or below the mean value expected for body surface area. The solid horizontal line represents the mean and the dotted lines represent the expected normal range.
infundibular) were present in each patient. Patient 6 had right ventricular intramyocardial sinusoids during infancy, which had resolved at postoperative cardiac catheterization.

**Discussion**

Patients with pulmonary atresia and intact ventricular septum, or other forms of hypoplastic right heart syndrome, who have undergone previous palliative surgery frequently have a residual right-to-left shunt at atrial level, resulting from resistance to right ventricular inflow. This may be a result of hypoplasia of the right ventricle with an inadequate right ventricular volume, as well as from abnormalities of the tricuspid valve. It may also be caused by diminished right ventricular compliance due to hypertrophy, fibrosis, or a previous ventriculotomy. Because of the risks of paradoxical embolus and brain abscess, as well as symptoms resulting from cyanosis and polycythemia, these patients are candidates for separation of systemic and pulmonary circulations if the tricuspid valve and right ventricle are capable of accepting systemic blood flow.

Selection of patients in whom the separation of systemic and pulmonary circuits is possible has been difficult. Angiographic estimates of tricuspid valve diameter may be misleading as the anulus, and not the true orifice, is visualized. Even when this measurement is accurate, the minimum acceptable diameter of the tricuspid valve anulus in these patients remains to be established; in the group we studied, all valve diameters were below the fifth percentile. A small or absent gradient across the tricuspid valve is also difficult to interpret when the true flow across the tricuspid valve cannot be accurately determined. Angiographic estimates of the right ventricular volume may be inaccurate in the small abnormally shaped right ventricle. Including the outermost borders of the right ventricle, as well as smoothing irregularities of the borders, may result in an overestimate of right ventricular cavity size. The volume of trabeculations included within the right ventricular cavity may vary from patient to patient. In addition, some patients have had placement of a right ventricular outflow patch to establish continuity between the right ventricle and pulmonary artery, and this nonfunctioning portion of the right ventricle must be included in volume estimates. The minimum right ventricular volume capable of accepting systemic blood flow for different body sizes re-

**FIGURE 3.** Selected diastolic frames from right ventricular cineangiograms in patient 5, both before (A) and after surgery (B). A. All three anatomic divisions of the right ventricle are present, although the trabecular portion is mostly occupied by hypertrophied muscle. B (not the same magnification as A), The right ventricular volume has significantly increased, and the trabecular hypertrophy regressed. The arrows indicate the left margin of a right ventricular outflow patch that is nonfunctioning but included in volume measurements.
mains to be established. Four of the five patients we examined who underwent successful surgical separation of systemic and pulmonary circulations had right ventricular volumes below the fifth percentile. Further, these angiographic estimates ignore right ventricular compliance that may interfere with inflow.

None of the patients we examined had significant residual tricuspid regurgitation, and no conclusions can be drawn regarding its influence on right ventricular capability. The presence of all three anatomic portions of the right ventricle does not ensure that the right ventricle will accept systemic flow, as observed in patient 6.

In contrast, balloon occlusion of the interatrial communication is a direct hemodynamic assessment of the capability of the tricuspid valve and right ventricle. The method has proved useful in evaluating several cases of isolated right ventricular hypoplasia.10-12 No assumptions are made about right ventricular geometry, and the method is not affected by the presence of a right ventricular outflow patch. There are no assumptions about the minimum acceptable size of the tricuspid valve and right ventricle. The equipment is simple and does not require sophisticated computer support.

None of the currently available methods of assessing right ventricular capability address the question of the response to an increased cardiac output during exercise. None of the patients we evaluated were exercised during occlusion. After surgical closure of their atrial septal defects, however, all five patients were asymptomatic without restrictions, including the three who had mild exercise intolerance preoperatively.

In our hands, temporary balloon occlusion predicted successful surgical closure of interatrial communications in five patients in whom conventional angiographic methods yielded tricuspid valve diameters or right ventricular volumes well below the expected normal. This method of assessing the hypoplastic right ventricle after initial palliative surgery may improve selection of candidates for definitive operation.

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Balloon occlusion of atrial septal defect to assess right ventricular capability in hypoplastic right heart syndrome.

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Circulation. 1983;68:1081-1086
doi: 10.1161/01.CIR.68.5.1081

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