Percutaneous Balloon Valvotomy in Pulmonary Atresia With Intact Ventricular Septum
Impact on Patient Care

Tilman Humpl, MD; Björn Söderberg, MD; Brian W. McCrindle, MD, FRCPC; David G. Nykanen, MD, FRCPC; Robert M. Freedom, MD, FRCPC; William G. Williams, MD, FRCSC; Lee N. Benson, MD, FRCPC

Background—Pulmonary atresia with intact ventricular septum (PA-IVS) is a rare congenital lesion with high mortality. Therapy was exclusively surgical until recently, when the use of radiofrequency-assisted perforation of the atretic valve was introduced as a treatment option. This study analyzes the outcomes and morphological changes to right heart structures after percutaneous perforation and balloon dilation of the atretic valve.

Methods and Results—Between April 1992 and August 2000, 30 patients with PA-IVS underwent attempted percutaneous valve perforation and balloon dilation of the pulmonary valve. Longitudinal echocardiographic measurements of the tricuspid valve diameter, right ventricular length and area were recorded. Z scores were calculated according to published formulas. Perforation was achieved in 27 patients. In 14 patients a modified Blalock-Taussig shunt was performed between 2 and 24 days after valve dilation. There were 3 early and 2 late deaths. Among the survivors (follow-up time of 1 to 87 months), 16 patients had a biventricular circulation, 3 a 1½-ventricle circulation, and 1 a Fontan operation. Four patients are awaiting further palliation. There was no significant change of the tricuspid valve Z score or right ventricular length Z score with time.

Conclusions—Percutaneous balloon valvotomy is an effective treatment strategy for patients with PA-IVS provided that there is a patent infundibulum and a lack of a right ventricle–dependent coronary circulation. Despite the observation that right heart growth does not increase with body growth in early follow-up, it appears adequate to maintain a biventricular circulation in many patients. (Circulation. 2003;108:826-832.)

Key Words: pediatrics • catheterization • valvuloplasty

Pulmonary atresia with intact ventricular septum (PA-IVS) is a rare and enigmatic disorder with significant morphological heterogeneity.1 In the absence of a right ventricle (RV)–dependent coronary circulation, decompression of the RV is a component of a treatment algorithm that attempts to salvage the right heart as a component of a biventricular or so-called 1½ ventricular repair.2 An alternative to primary surgical decompression strategies is the use of percutaneous laser or radiofrequency (RF)–assisted perforation of the atretic valve and subsequent balloon dilation.3,4 This study reviews the clinical outcomes and morphological changes to the tricuspid valve and RV after a percutaneous perforation and balloon dilation strategy.

Methods

Patient Population
From April 1992 through August 2000, 50 neonates were diagnosed with membranous atresia of the pulmonary valve with an intact ventricular septum, as defined by 2D transthoracic echocardiography. Of this group, 30 patients underwent attempted percutaneous RF-assisted valvotomy and balloon dilation of the pulmonary valve. The remaining 20 patients were treated following a surgical strategy (univentricular palliation).5 The criterion for not proceeding with a catheter-based approach was the presence of an RV-dependent coronary circulation (n=13), severely attenuated RV cavity, and/or a severely stenotic/hypoplastic infundibulum (n=7).

Diagnostic angiography was performed in all cases (biplane right ventriculogram and careful delineation of the course and distribution of any coronary vessels supplied from the RV cavity; frequently, a left ventriculogram was also performed defining the course and distribution of the coronary arteries as they arose from the aorta). An RV-dependent coronary circulation was defined as the presence of proximal coronary artery atresia, stenoses, or interruptions, and distal supply originating from the RV was not considered RV dependent.5,6 Provocative maneuvers to induce potential ischemia if the right ventricular cavity was temporarily decompressed were not undertaken. Rarely, selective coronary angiography or antegrade balloon occlusion angiography was required. All patients had situs solitus, levocardia, concordant atrioventricular and ventriculoarterial connections, a left aortic arch, and confluent pulmonary arteries.

Received January 7, 2002; de novo received March 11, 2003; revision received May 16, 2003; accepted May 29, 2003.
From the Departments of Pediatrics and Surgery, Divisions of Cardiology (T.H., B.S., B.W.M., D.G.N., R.M.F., L.N.B.) and Cardiovascular Surgery (W.G.W.), The Hospital for Sick Children, The University of Toronto School of Medicine, Toronto, Ontario, Canada.
Correspondence to Lee N. Benson, MD, FRCPC, The Hospital for Sick Children, Division of Cardiology, 555 University Ave, Toronto, ON M5G 1X8, Canada. E-mail benson@sickkids.ca

© 2003 American Heart Association, Inc.
Circulation is available at http://www.circulationaha.org

DOI: 10.1161/01.CIR.0000084548.44131.D1
Patient Characteristics
There were 16 (53%) males and 14 (47%) females. The median age at presentation was 1 day (range 0 to 4 days), and the median age at intervention 2 days of age (range 0 days to 22 months). Median weight was 3.28 kg (range 2.2 to 7.6 kg), median length 50 cm (range 46 to 76 cm), and median body surface area 0.22 m² (range 0.17 to 0.4 m²). In 11 (37%) patients, right ventriculocoronary connections were detected angiographically.

Procedural Characteristics
All patients underwent cardiac catheterization (see above) under general anesthesia with ductal patency maintained with a prostaglandin E₁ infusion. There were minor modifications of the management over the time period, and the earlier technique is described elsewhere. A 4F or 5F right coronary catheter (2.5 curve, Cordis, Inc) was maneuvered into the infundibular area preloaded with a side arm back bleed tap (Cook, Inc), allowing intermittent contrast injections. The stiff end of a 0.014-inch coronary wire (USCI Angiographics, Inc) was used in the first 2 patients to perforate the atretic valve, an 0.018-inch RF guide wire (PA 120, Opsyka, Inc) in the subsequent 6 patients, and a 0.024-inch RF perforation wire (Baylis Medical) in the remaining patients, connected to an RF generator (Radionics, Inc [n = 6] or Baylis Medical [n = 22]). RF energy (3 to 10 W/s) was delivered for 2 to 5 seconds, and the wire was advanced toward the valve membrane, terminating the energy after it appeared that the wire had entered the main pulmonary artery. A coaxial catheter (Baylis Medical) was advanced over the perforation catheter into the main pulmonary artery, allowing passage of a 0.014-inch coronary wire (Wisdom, Cordis) through the arterial duct into the descending aorta. To improve trackability, the wire was maneuvered through the descending aorta into one of the femoral arteries. Externally, the femoral artery was digitally compressed, fixing the wire in place. If this was not possible, and the initial low-profile balloon could not cross the valve plate, the femoral artery (n = 8), or umbilical artery (n = 4) entered with a 4F sheath, and the guide wire snared (10 mm diameter, Microvena Corp). Thereafter, over this wire, serial balloon dilation catheters (Cordis Corp) were introduced across the pulmonary valve plate, and dilations were sequentially performed with increasing balloon diameters to ~20% larger than the estimated valve diameter obtained from the right ventriculogram (first dilation, 2.5 to 7.0 mm; final dilation, 6.0 to 10.0 mm). Pressures in the descending aorta and RV were measured before and after the final balloon dilation. An RV angiogram was performed to assess the adequacy of the outflow reconstruction, status of coronary sinusoids (if patent), degree of tricuspid valve regurgitation, and RV size and function before catheters and sheaths were removed. All patients received antibiotic prophylaxis; heparin sulfate was only given when needed.

Anatomic Measurements
For assessment of the growth of the RV, all echocardiographic studies were reviewed. Two-dimensional, spectral, and color Doppler studies were performed using either a Hewlett-Packard (models 1000, 2500, or 5500) or Advanced Technology Laboratories (Philips, Best, models ATL HDI Ultramark 9 or ATL HDI 5000) ultrasound system with transducer frequencies appropriate for patient size. Measurements were performed offline using electronic calipers (Hewlett-Packard Sonos 5500) and included the following: (1) the diameter of the tricuspid valve in early diastole (from inner edge to inner edge), (2) the RV length (tricuspid valve annulus to apex at ventricular end diastole), and (3) the RV area at end diastole with the maximal area bordered by RV endocardium (all measurements in the apical 4-chamber view). The tricuspid valve Z score, RV length Z score, and RV area Z score were calculated according to published formulas. The presence and degree of pulmonary insufficiency were determined from the initial echocardiographic study after dilation.

Data Analysis
Data are described as frequencies; means with SDs and medians with ranges are given as appropriate. Changes with dilation were sought with paired t tests. Trends and factors related to serial echocardiographic measurements were sought in repeated-measures mixed linear regression analysis, using a compound symmetry covariance matrix, and multiple logistic analysis was performed between groups where indicated. All analyses were performed using SAS statistical software version 8 (SAS Institute, Inc) using default settings.

Results

Acute Procedural Results
Antegrade perforation of the atretic pulmonary valve was attempted with the stiff end of a coronary guide wire in 2 and with an RF wire in 28; these attempts were unsuccessful in 3 of these latter patients (see below).

Case Descriptions of Failed Procedures
The first patient (age 13 days) had a prior diagnostic catheterization and a balloon atrial septostomy performed. The neonate had a small but developed pulmonary infundibular chamber leading to an atretic pulmonary valve, and RV coronary artery sinusoids were detected. A modified right Blalock-Taussig shunt was performed, but a decision was subsequently made to electively attempt to perforate the pulmonary valve before discharge (tricuspid valve Z score, −3.8). It was not possible to obtain a stable position of the right coronary catheter within the pulmonary infundibulum before attempted perforation. As the catheter was being removed from the RV, the child developed 2:1 heart block with hypotension, which rapidly deteriorated to asystole. The patient died in the catheterization laboratory despite attempted resuscitation.

The second patient was a former preterm child with cerebral palsy. The parents originally declined any cardiac intervention but reconsidered the option of a transcatheter procedure at 22 months of age. Transcutaneous systemic arterial saturation ranged between 78% and 80%, and the arterial duct was patent. During the procedure, the RF wire perforated the inferior lateral margin of the main pulmonary artery resulting in a dissection toward the pulmonary artery confluence. While the child was stable, retrograde cannulation of the 2-mm arterial duct allowed balloon angioplasty, improving resting saturations from 59% to 94%. Extension of the dissection obstructed the main and left pulmonary artery, and the child was emergently transferred to the operating room for an uneventful surgical outflow tract patch and bidirectional cavopulmonary anastomosis.

The final patient (age 3 days) was referred to surgery after perforation of the main pulmonary artery by the RF wire with tamponade and was stabilized with a percutaneous pericardiocentesis. A surgical pulmonary valvotomy, repair of the perforation, and arterial duct ligation were performed the following day without complication.

Hemodynamics
Before dilation, the mixed venous and systemic saturations were 59±9 and 93±11%, respectively, mean right atrial pressure 7±3 mm Hg and RV and aortic systolic pressures 104±30 and 64±11 mm Hg respectively. After the dilation, the RV and aortic pressures were 48±20 and 54±11 mm Hg, respectively. The median RV/systemic pressure ratio before the procedure was 1.65 (range 0.8 to 2.3, mean 1.61±0.36)
and 0.79 (range 0.4 to1.6, mean 0.73±0.30) after the procedure. The arterial duct was patent in all patients.

Complications
Loss of the arterial pulse occurred in 4 neonates (all having 4F sheaths placed), and returned with heparin therapy over the following 24 hours. Seven patients had rhythm disturbances (transient atrial flutter or heart block), cardiac perforation occurred in 6 patients (pneumomediastinum in 1, pericardial effusion in 3, and main pulmonary artery perforation in 2), 1 neonate developed a main pulmonary artery aneurysm, and another developed neonate severe tricuspid regurgitation.

There was 1 death in the catheterization laboratory, described above. Another patient died 7 days after the procedure of presumed sepsis, and 1 patient died at the age of 7 weeks, after surgical RV outflow tract repair. This patient underwent RF perforation on day 3 of life, but because of persistently elevated RV pressures, repeat balloon dilation was unsuccessful as a result of the dynamic nature of subpulmonary obstruction. The child developed a myotic aneurysm of the main pulmonary artery and bilateral pulmonary artery thromboses. Surgery was performed with an RV outflow tract reconstruction, resection of the main pulmonary artery aneurysm, left pulmonary artery pericardial patch angioplasty, tricuspid valve annuloplasty, and a fenestrated atrial septal patch. The postoperative course was complicated by low cardiac output, and the child died on the first postoperative day.

Late deaths occurred in an additional 2 patients; 1 patient died 18 months after a modified Blalock-Taussig shunt with presumed shunt occlusion, and the patient with cerebral palsy (noted above) 2 years after surgery (right ventricular outflow tract repair and bidirectional cavopulmonary anastomosis), of noncardiac causes.

Clinical Outcomes
In 14 patients, a modified Blalock-Taussig shunt was performed between 2 and 24 days (mean 9.1±6.8) after valve dilation. After a complicated interventional procedure, the RV outflow tract was reconstructed within 48 hours in 2 patients. In 2 additional patients, an RV outflow tract reconstruction was necessary at 7 weeks and 6 months respectively. In 21 (69%) patients there was a communication at atrial level. In 14 patients, a modified Blalock-Taussig shunt was performed between 2 and 24 days (mean 9.1±6.8) after valve dilation. After a complicated interventional procedure, the RV outflow tract was reconstructed within 48 hours in 2 patients. In 2 additional patients, an RV outflow tract reconstruction was necessary at 7 weeks and 6 months respectively. In 21 (69%) patients there was a communication at atrial level.

There was 1 death in the catheterization laboratory, described above. Another patient died 7 days after the procedure of presumed sepsis, and 1 patient died at the age of 7 weeks, after surgical RV outflow tract repair. This patient underwent RF perforation on day 3 of life, but because of persistently elevated RV pressures, repeat balloon dilation was unsuccessful as a result of the dynamic nature of subpulmonary obstruction. The child developed a myotic aneurysm of the main pulmonary artery and bilateral pulmonary artery thromboses. Surgery was performed with an RV outflow tract reconstruction, resection of the main pulmonary artery aneurysm, left pulmonary artery pericardial patch angioplasty, tricuspid valve annuloplasty, and a fenestrated atrial septal patch. The postoperative course was complicated by low cardiac output, and the child died on the first postoperative day.

Late deaths occurred in an additional 2 patients; 1 patient died 18 months after a modified Blalock-Taussig shunt with presumed shunt occlusion, and the patient with cerebral palsy (noted above) 2 years after surgery (right ventricular outflow tract repair and bidirectional cavopulmonary anastomosis), of noncardiac causes.

Clinical Outcomes
In 14 patients, a modified Blalock-Taussig shunt was performed between 2 and 24 days (mean 9.1±6.8) after valve dilation. After a complicated interventional procedure, the RV outflow tract was reconstructed within 48 hours in 2 patients. In 2 additional patients, an RV outflow tract reconstruction was necessary at 7 weeks and 6 months respectively. In 21 (69%) patients there was a communication at atrial level.

Anatomic Data
Tricuspid valve diameter, RV length, and RV area were smaller than in the normal newborn, and as a result, the respective Z scores were more negative (Table 2). The initial Z scores of the tricuspid valve, RV length, and RV area were significantly correlated with one another (P<0.001).

As shown in Table 3, the initial Z scores for tricuspid valve, RV length, and RV area were less negative for patients pursuing on a biventricular circulation, whereas there was a trend in patients with other types of circulation to have more negative Z scores.

Independent Predictors of a Biventricular Repair or Need for Further Intervention
The presence of RV sinusoids (OR=8.54 [95% CI=1.05–69.8]) and an elevated RV systolic pressure (OR=1.095 [95% CI=1.011–1.187]) were predictive of failure to achieve a biventricular repair. Only the preintervention RV systolic pressure (OR=1.055 [95% CI=1.008–1.094]) was weakly predictive of the need for further intervention after the initial procedure (Tables 4 and 5).

### Table 1. Measurements and Anatomical Z Scores of Right Ventricular Structures at Presentation (n=30)

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tricuspid valve</td>
<td></td>
</tr>
<tr>
<td>Diameter, cm</td>
<td>0.97±0.27</td>
</tr>
<tr>
<td>Z score</td>
<td>−1.33±1.58</td>
</tr>
<tr>
<td>Right ventricle</td>
<td></td>
</tr>
<tr>
<td>Length, cm</td>
<td>1.57±0.44</td>
</tr>
<tr>
<td>Length Z score</td>
<td>−5.93±2.47</td>
</tr>
<tr>
<td>Area, cm²</td>
<td>1.82±0.70</td>
</tr>
<tr>
<td>Area Z score</td>
<td>−4.50±2.18</td>
</tr>
</tbody>
</table>
Independent Predictors of Longitudinal Changes in Right Heart Dimensions

**Z Score of the Tricuspid Valve Diameter**

There was no relationship with age at echocardiographic study, indicating no significant change in the Z score diameter with time. There was also no effect of the RV pressure or RV/systemic pressure ratio after the valve dilation and no significant difference in longitudinal measurements between patients who did and those who did not achieve a final biventricular repair. The only significant predictor of subsequent measurements was the initial Z score, with mixed linear regression estimates as follows: intercept, −1.3612±0.3329 (SE), and initial Z score, 0.7171±0.3329 (P<0.0001).

**Z Score of the RV Length**

There was no relationship with age at echocardiographic study, indicating no significant change in Z score of length with time. There was also no effect of RV pressure or RV/systemic pressure ratio after dilation and no significant difference in longitudinal measurements between patients who did and those who did not achieve a final biventricular repair. The only significant predictor of subsequent measurements was the initial Z score, with mixed linear regression estimates as follows: intercept, −2.6818±0.5549, and initial Z score, 0.5938±0.08484 (P<0.0001).

**Z Score of the RV Area**

There was also no effect of RV pressure or RV/systemic pressure ratio after dilation, and no significant difference in longitudinal measurements between patients who did and those who did not achieve a final biventricular repair. Both initial Z score and age were significant independent predictors of the subsequent measurement, although there was significant interaction as illustrated in Figure 1, with mixed linear regression estimates as follows: intercept, −1.8802±0.4662; initial Z score, 0.6410±0.0916 (P<0.0001); age at echo, per 1-year increment, −0.3030±0.0794 (P=0.0003); and interaction term, −0.0568±0.0160 (P=0.0007).

Figure 1 shows that early after RF perforation there is a convergence of Z scores of RV length initially, but then no

### Table 3. Comparison of Initial Z Scores With the Most Recent Clinical Status

<table>
<thead>
<tr>
<th>Type of circulation</th>
<th>Tricuspid Valve Z Score</th>
<th>Right Ventricular Length Z Score</th>
<th>Right Ventricular Area Z Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biventricular (n=16)</td>
<td>−3.03±2.51</td>
<td>−5.30±2.78</td>
<td>−3.96±2.46</td>
</tr>
<tr>
<td>1½ ventricle (n=3)</td>
<td>−9.28±2.04</td>
<td>−7.84±1.46</td>
<td>−4.08±1.46</td>
</tr>
<tr>
<td>Bidirectional cavopulmonary anastomosis (n=3)</td>
<td>−8.39±6.74</td>
<td>−8.78±1.56</td>
<td>−6.53±1.96</td>
</tr>
<tr>
<td>Fontan (n=1)</td>
<td>−5.9</td>
<td>−7.43</td>
<td>−5.82</td>
</tr>
<tr>
<td>Modified Blalock-Taussig shunt (n=1)</td>
<td>−7.29</td>
<td>−6.09</td>
<td>−4.28</td>
</tr>
<tr>
<td>Lost to follow-up (n=1)</td>
<td>−4.29</td>
<td>−5.60</td>
<td>−3.90</td>
</tr>
<tr>
<td>Death (n=5)</td>
<td>−6.01±3.90</td>
<td>−5.92±2.33</td>
<td>−5.71±1.52</td>
</tr>
</tbody>
</table>

### Table 4. Comparison Between Patients With and Without Surgery*

<table>
<thead>
<tr>
<th>Gender</th>
<th>With Surgery n=16</th>
<th>Without Surgery n=14</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>6</td>
<td>10</td>
<td>0.09</td>
</tr>
<tr>
<td>Female</td>
<td>10 (63%)</td>
<td>4 (29%)</td>
<td></td>
</tr>
<tr>
<td>Mean weight (kg)</td>
<td>3.56±1.19</td>
<td>3.13±0.48</td>
<td>0.14</td>
</tr>
<tr>
<td>Mean BSA (m²)</td>
<td>0.22±0.03</td>
<td>0.18±0.08</td>
<td>0.06</td>
</tr>
<tr>
<td>Mean age (days)</td>
<td>2 (1 to 13)†</td>
<td>1 (&lt;1 to 4)</td>
<td>0.006</td>
</tr>
<tr>
<td>Mean pre RVSP (mm Hg)</td>
<td>116±28</td>
<td>92±27</td>
<td>0.03</td>
</tr>
<tr>
<td>Mean pre RVSP/AoP</td>
<td>1.71±0.35</td>
<td>1.50±0.35</td>
<td>0.11</td>
</tr>
<tr>
<td>RV sinusoids</td>
<td>9/16 (56%)</td>
<td>2/14 (14%)</td>
<td>0.03</td>
</tr>
<tr>
<td>Median balloon diameter (mm)</td>
<td>4 (3 to 6) (n=12)</td>
<td>3.5 (2 to 8)</td>
<td>0.72</td>
</tr>
<tr>
<td>Mean post RVSP (mm Hg)</td>
<td>65±20 (n=13)</td>
<td>44±14 (n=13)</td>
<td>0.005</td>
</tr>
<tr>
<td>Mean post RVSP/AoP (mm Hg)</td>
<td>1.00±0.34 (n=13)</td>
<td>0.73±0.17 (n=12)</td>
<td>0.03</td>
</tr>
<tr>
<td>Mean initial Z TV (mm)</td>
<td>−6.52±4.12</td>
<td>−3.20±2.49</td>
<td>0.02</td>
</tr>
<tr>
<td>Mean initial Z RV length (mm)</td>
<td>−6.49±2.15</td>
<td>−5.61±2.84</td>
<td>0.35</td>
</tr>
<tr>
<td>Mean initial Z RV area (mm²)</td>
<td>−5.16±1.69</td>
<td>−4.12±2.58</td>
<td>0.20</td>
</tr>
</tbody>
</table>

BSA indicates body surface area; RVSP, right ventricular systolic pressure; AoP, systemic systolic pressure; RV, right ventricle; and TV, tricuspid valve.

*Includes the 5 patients who died and the 1 patient lost to follow-up at latest known status.
†Excluded 1 patient, age 680 days.
change in Z score over time, remaining markedly below normal. The flaring at the beginning of the curves could possibly represent some inaccuracy in the early measurements when the structure would be smallest, and a small amount of error may have a larger effect on the Z score. Growth of right heart structures is proportionate to the initial values and remains below normal. This study shows that initial Z scores are good predictors of subsequent Z scores and that “catch-up” growth toward a more normal Z score appears not to occur.

Discussion

Transcatheter approaches to opening the atretic pulmonary valve in PA-IVS constitute a relatively recent technical advance, applied to a relatively rare lesion, and thus lack reasonable patient numbers and follow-up. Data addressing the growth of RV structures (tricuspid valve, chamber dimensions) in patients achieving a biventricular circulation using this strategy have not been analyzed previously. Surgical repair for the neonate with PA-IVS is associated with a high morbidity, a 1-year mortality of 52%, and a 4-year survival of 64% from a recent surgical multicenter study. In a highly selective patient population, 98% survival can be achieved. Final-stage surgery for these patients incorporates all potential circulations, including a biventricular or 1½-ventricle repair or univentricular palliation.

The choice of treatment algorithm depends on the estimated adequacy of the RV to cope with systemic venous return, the size of tricuspid valve, and the status of the coronary arteries. In the presence of adequate right heart structures, surgical strategies have generally required the creation of a modified Blalock-Taussig shunt in the newborn period to maintain pulmonary artery blood flow in addition to an RV outflow tract reconstruction. In the presence of inadequate right heart anatomy or coronary artery anomalies, patients are managed with staged procedures leading to a univentricular circulation.

Clearly there are benefits and disadvantages to a catheter-based approach, compared with a surgical strategy, in patients who will eventually require a univentricular repair. In a recent editorial, Cheatham suggested that, as long as there was a tripartite RV and a well-formed infundibulum, a tricuspid valve annulus ≥11 mm, and membranous atretic pulmonary valve annulus ≥7 mm, transcatheter therapy should be performed. However, no supportive data for these recommendations were given. Our intervention was not predicated on predefined anatomic measurements, only on the presence of a patent tract to the valve plate and the absence of RV coronary blood flow dependence.

| TABLE 5. Comparison Between Patients With and Without a Further Intervention |
|-----------------------------|-----------------------------|-----------------------------|
| Intervention               | No Intervention             | P                           |
| n=21                       | n=9                         |                             |
| Gender                      |                             |                             |
| Male                        | 10                          | 6                           | 0.44 |
| Female                      | 11 (52%)                    | 3 (33%)                     |     |
| Mean weight (kg)            | 3.57±1.05                   | 3.02±0.53                   | 0.15 |
| Mean BSA (m²)               | 0.22±0.03                   | 0.16±0.09                   | 0.007|
| Mean age (days)             | 2 (1 to 13)                 | 1 (<1 to 4)                 | 0.13 |
| Mean pre RVSP (mm Hg)       | 114±26                      | 82±27                       | 0.005|
| Mean pre RVSP/AoP           | 1.68±0.34                   | 1.46±0.39                   | 0.14 |
| RV sinusoids                | 11/21 (52%)                 | 0/9 (0%)                    | 0.02 |
| Mean post RVSP (mm Hg)      | 61±19 (n=18)                | 42±14 (n=8)                 | 0.03 |
| Median balloon size (mm)    | 4 (3 to 6) (n=17)           | 3.5 (2 to 8)                | 0.75 |
| Mean post RVSP/AoP (mm Hg)  | 0.92±0.31 (n=18)            | 0.75±0.21 (n=21)            | 0.21 |
| Mean initial Z RV length (mm)| -6.3±2.3                   | -5.6±3.00                   | 0.55 |
| Mean initial Z RV area (mm²)| -5.0±1.8                    | -3.9±2.9                    | 0.23 |

Abbreviations as in Table 4.

Longitudinal measurements of Z score of the right ventricular area vs patient age. Early after RF perforation, there is a convergence of Z scores of right ventricular length initially, but then no change in Z score over time, with scores remaining markedly below normal. Flaring at the beginning of curves could represent some inaccuracy in early measurements when the structure would be smallest, where a small error may have a larger effect on Z score. Curved solid line indicates predicted model, with curved dashed lines enclosing a 95% confidence interval.
Humpl et al Balloon Valvotomy in Pulmonary Atresia 831

Although the intervention can be achieved without thoracotomy or cardiopulmonary bypass, complications include potential femoral arterial occlusion, cardiac perforation with possible tamponade, and rhythm disturbances, all of which are in general self-limiting, but occurred in 25% of our cohort. Primary transcatheter valvotomy strategy permits (as does surgery) forward blood flow early in life through the RV outflow tract, which may encourage pulmonary artery growth and, with acquired pulmonary insufficiency, potential RV growth. Ovaert et al noted, from their review after valve surgery and, with acquired pulmonary insufficiency, potential RV outflow tract, which may encourage pulmonary artery growth does surgery) forward blood flow early in life through the RV outflow tract, which may encourage pulmonary artery growth and, with acquired pulmonary insufficiency, potential RV growth. Ovaert et al noted, from their review after valve surgery (Table 2) and indeed were well below normal. In this regard, Hanséus et al showed, for patients managed with a surgical algorithm, that newborns with a very hypoplastic RV almost always have normalized values for RV size after 52 months (range 18 to 87 months). The best chamber growth was achieved in patients who underwent RV outflow reconstruction in the neonatal period.

There is debate as to the best method to assess RV size and its adequacy for a biventricular repair as well as how to monitor growth of the chamber and valve. As noted by Ovaert et al, normal tricuspid valve growth might not be necessary for an RV-competent circulation, to maintain the pulmonary circulation, and the initial size of the tricuspid valve might be only a weak indicator of outcome. An indirect assessment was proposed by Minich et al, using a tricuspid/mitral valve ratio of >0.5 as a predictor for a successful biventricular repair. The question was raised whether the initial size of the RV was significant in defining the treatment algorithm, particularly as it was difficult to estimate its size or volume. In patients achieving a biventricular circulation from our data series, the tricuspid valve Z score was lower than in patients with other management formats, whereas RV length and Z scores were not that different between the groups (Table 2) and indeed were well below normal.

The central question therefore is whether there are appropriate anatomic criteria to patient selection, for a particular treatment algorithm. Comparing the mean initial Z scores of the tricuspid valve (Table 3), between patients who did not have a surgical procedure (−3.20±2.49) and those who did (−6.52±4.12, P=0.02), there were no differences noted. On the other hand, the Z value for the tricuspid valve in patients who underwent a further intervention versus the Z value for those with no further intervention was significantly different (Table 5, P=0.04). This suggests that a subgroup of patients can be defined who would be expected to avoid any further intervention beyond the initial procedure. Nevertheless, with the diverse spectrum of morphologies seen in this disorder, it is difficult to predict, at the time of presentation, whether the RV will have the potential for accommodating systemic blood return.

As underscored by our data, management of newborns with PA-IVS has changed. Only those patients with very diminutive right-sided structures, a severely attenuated infundibulum, or coronary artery anomalies were not considered as candidates for a catheter-based valvotomy. Also shown in this data set, there was little growth of RV structures over time, apart from the RV area, which showed an initial decrease followed by more stable development. There was, however, enlargement of the tricuspid valve and RV length, and the dimensions paralleled normal ones. Despite apparently inadequate right heart size, a biventricular repair was achievable in 53% of patients. This suggests that a “normal”-sized right heart is not required to maintain at rest a normal pulmonary blood flow with an acceptable right atrial pressure.

The requirement of a shunt following successful RV decompression varies among clinical units. In children born in Sweden with PA-IVS between 1980 and 1999, when transcatheter management was not available, a systemic to pulmonary shunt was created in 93% of patients, whereas the number of shunts constructed in our institution in this population was 46%. Whereas 44% of the patients undergoing a successful interventional valvotomy did not require a surgical shunt procedure in 1 series, 84% of the patients reported by Alwi et al achieved a biventricular circulation without a shunt. Initially, in our unit, a shunt was offered with failure of the first attempt at weaning prostaglandins, but more recently a policy has evolved allowing the patients a longer time period to accommodate and improve right ventricular compliance. Persistent forward and regurgitant flow through the opened pulmonary valve may allow the RV to adapt to the circulation and potentially grow, although a “normal” size may not be reached.

Finally, in those patients who, after RV decompression, persist with an inadequate RV inflow or chamber to achieve a biventricular or 1½-ventricle circulation, the reduction in RV pressure remains advantageous in the univentricular circulation. A low RV pressure allows for a non-isometrically contracting chamber and eliminates the development of subaortic stenosis.

In conclusion, technical success was achieved with valve perforation and balloon dilation in 27 of 30 patients. If the goal was to achieve a biventricular repair, then 14 patients were an unequivocal success, 9 patients not requiring any further intervention and 5 only a Blalock-Taussig shunt, which was later taken down. Patients with a less negative tricuspid valve Z score tended to achieve a biventricular repair (Tables 3 and 5). Percutaneous balloon valvotomy appears effective in palliation, achieving an RV circulation in a selected population with PA-IVS. The technique can be considered as an alternative to surgical intervention in patients with a well-formed tricuspid valve with a patent infundibulum, and without an RV-dependent coronary circulation.

References


17. Cheatham JP. To perforate or not to perforate—that’s the question…. or is it? Just ask Richard! Cathet Cardiovasc Diag. 1997;42:403–404.


Percutaneous Balloon Valvotomy in Pulmonary Atresia With Intact Ventricular Septum:
Impact on Patient Care
Tilman Humpl, Björn Söderberg, Brian W. McCrindle, David G. Nykanen, Robert M. Freedom,
William G. Williams and Lee N. Benson

_Circulation_. 2003;108:826-832; originally published online July 28, 2003;
doi: 10.1161/01.CIR.0000084548.44131.D1
_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2003 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

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/108/7/826

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published
in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial
Office. Once the online version of the published article for which permission is being requested is located,
click Request Permissions in the middle column of the Web page under Services. Further information about
this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org//subscriptions/