Echocardiographic Determinants of Clinical Course in Infants With Critical and Severe Pulmonary Valve Stenosis

JOHN P. KOVALCHIN, MD, THOMAS J. FORBES, MD, MICHAEL R. NIHILL, MD, FACC, TAL GEVA, MD*

Houston, Texas and Boston, Massachusetts

Objectives. The purposes of this study were to determine the growth pattern of the pulmonary valve (PV) annulus and right heart structures in patients with critical and severe pulmonary stenosis (PS) after balloon dilation, and to determine any morphometric or hemodynamic differences between cyanotic infants with critical PS and asymptomatic infants with severe PS that may account for their varied clinical presentations.

Background. Growth of the PV annulus and right heart structures in patients with critical PS after balloon valvuloplasty has not clearly been defined. In addition, the anatomic and hemodynamic factors that determine whether an infant with severe PS will present with cyanosis or without symptoms are not well understood.

Methods. Measurements of the PV annulus, tricuspid valve (TV) annulus and main, right and left pulmonary arteries were obtained from initial and follow-up echocardiograms, and Z values were calculated. Hemodynamic data and balloon pulmonary valvuloplasty techniques were reviewed. Right ventricular (RV) volumes were measured from angiograms.

Results. Fourteen patients with critical PS (mean \( \pm \text{SD} \) age 0.21 \( \pm \) 0.37 months) and 20 patients with severe PS (mean age 2.6 \( \pm \) 2.9 months) were evaluated at presentation and at 32 \( \pm \) 32 and 42 \( \pm \) 32 months of follow-up, respectively. Balloon pulmonary valvuloplasty was successful in 64\% of patients with critical PS and in 90\% of patients with severe PS. The PV, TV and pulmonary arteries increased in size after balloon pulmonary valvuloplasty in both groups at a rate that paralleled or exceeded the rate of somatic growth. The initial TV diameter and RV volume were smaller in patients with critical PS than in those with severe PS (p < 0.05 and p < 0.0008, respectively).

Conclusions. After balloon pulmonary valvuloplasty in infants with critical and severe PS, right heart structures increase in size at a rate that parallels or exceeds the rate of somatic growth. The primary morphometric differences between these groups are a smaller TV diameter and RV volume in infants with critical PS. This may contribute to increased right to left atrial shunting and account for the variations in clinical presentation.

(J Am Coll Cardiol 1997;29:1095–101)
©1997 by the American College of Cardiology

Pulmonary stenosis (PS) accounts for \( \sim 8\% \) to 12\% of patients with congenital heart disease and is generally well tolerated (1–5). Severe PS with right ventricular (RV) pressure greater than or equal to systemic pressure is uncommon. Infants with severe PS can be categorized into two distinct groups based on their clinical presentation: 1) critically ill infants with cyanosis whose pulmonary blood flow may depend on ductal patency and require urgent intervention; and 2) asymptomatic infants. The anatomic and hemodynamic factors that determine the clinical presentation of infants with severe PS are not well understood.

Balloon pulmonary valvuloplasty has become the treatment of choice for patients with PS, including infants with critical and severe PS (6–8). The success rates of balloon pulmonary valvuloplasty in infants with critical PS have been reported to be 55\% to 94\%; however, restenosis has been reported in 17\% to 36\% of these patients (7–14). The pulmonary valve (PV) annulus and right heart structures have been characterized as hypoplastic in some patients with critical PS, and growth of these structures relative to somatic growth after balloon pulmonary valvuloplasty has been a subject of controversy (7,14).

The purposes of this study were: 1) to determine the growth rate of the PV and right heart structures relative to somatic growth after balloon dilation of the PV; and 2) to determine any morphometric or hemodynamic differences between infants with critical PS and those with severe PS that may account for their varied clinical presentations.

Methods

Study patients. The medical records, echocardiograms, cineangiograms and cardiac catheterization data of all patients <1 year of age with the diagnosis of PS who presented to the Texas Children’s Hospital from 1983 to 1994 were reviewed.

From the Lillie Frank Abercrombie Section of Pediatric Cardiology, Texas Children’s Hospital and Department of Pediatrics, Baylor College of Medicine, Houston, Texas; and *Department of Cardiology, Children’s Hospital and the Department of Pediatrics, Harvard Medical School, Boston, Massachusetts.

Manuscript received June 20, 1996; revised manuscript received December 20, 1996, accepted January 9, 1997.

Address for correspondence: Dr. Tal Geva, Department of Cardiology, Children’s Hospital, 300 Longwood Avenue, Boston, Massachusetts 02115. E-mail: geva_t@al.tch.harvard.edu.

©1997 by the American College of Cardiology
Published by Elsevier Science Inc.
Included in this study were all infants who had: 1) severe PS, defined as thickened, doming leaflets as seen echocardiographically or angiographically with evidence of anterograde flow across the PV and a RV to LV pressure ratio \( \geq 1 \) with an intact ventricular septum documented by cardiac catheterization; 2) a complete two-dimensional and Doppler echocardiographic examination before intervention; and 3) clinical and echocardiographic follow-up data available for review. Patients were divided into two groups according to their clinical profile at initial presentation. Group 1 included infants with critical PS, defined as severe PS in an infant with cyanosis, arterial oxygen saturation \( \leq 92\% \) or signs and symptoms of low cardiac output at initial presentation. These infants required prostaglandin \( \text{E}_1 \) infusion or other urgent intervention. Group 2 included infants with severe PS who were acyanotic and asymptomatic at initial presentation. Patients with pulmonary atresia, supravalvar PS and an Ebstein anomaly were excluded. The presence of associated infundibular stenosis due to hypertrophy was not used as an exclusion criteria. Clinical and demographic information was obtained from review of the medical records.

**Echocardiography.** Initial and follow-up echocardiograms were performed with several commercially available scanners with the transducer frequency appropriate for the patient’s size. Studies were recorded on ½-in. super-VHS videotape. All echocardiograms were reviewed, and selected still frames were identified for subsequent analysis. Measurements were performed off-line with a digitizing tablet (Summagraphic II) interfaced with a personal computer and a commercially available software package (Digisonics EchoPro System, version 3.30). Measurements were performed on the initial and follow-up echocardiograms. Each measurement was performed three times and the mean value was used for data analysis.

The PV annulus was measured in the parasternal short-axis view as the maximal distance between the hinge points of the valve leaflets in systole. The main pulmonary artery (PA) diameter was measured in the parasternal short-axis view in systole, \( \sim 1 \) cm above the valve annulus. Right and left PA diameters were measured in the high parasternal and suprasternal notch views in systole, just proximal to the first branch of each vessel. The tricuspid valve (TV) annulus was measured in the apical four-chamber view as the maximal distance between the hinge points of the valve leaflets in diastole. Linear dimensions were indexed to the square root of body surface area to allow comparison of measurements between patients of varied size (15). Corresponding \( Z \) values were calculated for the PV annulus, TV annulus and main, right and left PAs by the formula:

\[
Z = \frac{\text{Measured value} - \text{Mean value of normal control}}{\text{Standard deviation of normal control}}.
\]

The normative data are derived from measurements of 140 children without heart disease (unpublished data from Children’s Hospital, Boston, Massachusetts).

The maximal instantaneous gradient across the PV was calculated from the peak spectral Doppler velocity using the modified Bernoulli equation (16, 17). Pulmonary and tricuspid valve and PV regurgitation was graded as none, trivial, mild, moderate or severe based on the width of the regurgitant jet at its origin as seen by color Doppler flow mapping. The atrial septum was evaluated for the presence of an atrial septal defect or patent foramen ovale, and the direction of flow across the atrial communication was examined using pulsed and color Doppler flow mapping. The presence of a patent ductus arteriosus and the direction of flow were also noted.

**Catheterization.** The techniques used for balloon pulmonary valvuloplasty in our institution have been previously described in detail (18, 19). The following balloon dilation techniques were noted: single- or double-balloon dilation technique and graduated or nongraduated balloon dilation technique. A graduated technique was defined as the initial dilation being performed with a smaller diameter balloon and then progressing to additional inflations with a larger diameter balloon. The maximal balloon diameters were recorded, and in the cases in which a double-balloon technique was used, the sum of the diameters of the simultaneously used balloons was used for analysis. The maximal balloon to PV annulus ratio was calculated. The number of inflations and maximal inflation pressures were recorded. The operator and year of procedure were also noted. Right ventricular end-diastolic and right atrial pressures were recorded. Right ventricular volumes in systole and diastole were measured from the initial right ventriculograms using the biplane Simpson method and either a cardi marker or catheter size for calibration (Digisonics Inc. 1993, CathPro, Cath Lab Analysis System). Right ventricular ejection fraction was calculated as (end-diastolic volume − end-systolic volume)/end-diastolic volume. The RV to PA pressure gradient and the RV to LV pressure ratio were measured before and after balloon pulmonary valvuloplasty. In cases in which the LV was not entered, systemic pressure was used. Any complications reported in the catheterization record were noted.

**Outcome.** Balloon pulmonary valvuloplasty was attempted as the initial procedure in all patients. Success of balloon pulmonary valvuloplasty in patients with critical PS was defined by the following criteria: 1) the patient was acyanotic, allowing for the discontinuation of prostaglandin \( \text{E}_1 \) with no evidence of ductal flow; 2) the RV systolic pressure remained \( \leq \frac{2}{3} \) of systemic systolic pressure; and 3) no further intervention was required. The last two criteria defined a successful procedure in patients with severe PS.
Table 1. Demographics and Clinical Profile at Initial Presentation

<table>
<thead>
<tr>
<th></th>
<th>CPS</th>
<th>SPS</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (mo)*</td>
<td>0.21 ± 0.37</td>
<td>2.6 ± 2.9</td>
<td>&lt; 0.004</td>
</tr>
<tr>
<td>Body surface area (m²)*</td>
<td>0.23 ± 0.02</td>
<td>0.29 ± 0.09</td>
<td>&lt; 0.02</td>
</tr>
<tr>
<td>Gender ratio (M/F)</td>
<td>7:7</td>
<td>1.4:9.11</td>
<td>0.95</td>
</tr>
<tr>
<td>Systemic oxygen saturation (%)*</td>
<td>88 ± 9.3</td>
<td>92 ± 3.3</td>
<td>&lt; 0.04</td>
</tr>
<tr>
<td>PDA</td>
<td>71%</td>
<td>10%</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>PGEi</td>
<td>57%</td>
<td>0%</td>
<td>&lt; 0.0001</td>
</tr>
<tr>
<td>Intubated at catheterization</td>
<td>64%</td>
<td>25%</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Doppler gradient (mm Hg)*</td>
<td>74 ± 31.6</td>
<td>92.6 ± 26.8</td>
<td>0.09</td>
</tr>
<tr>
<td>ASD/PFO</td>
<td>100%</td>
<td>95%</td>
<td>0.59</td>
</tr>
<tr>
<td>F/U duration (mo)*</td>
<td>31.9 ± 3.29</td>
<td>42.3 ± 3.23</td>
<td>0.44</td>
</tr>
</tbody>
</table>

Data are presented as mean value ± SD or percent of patients. ASD = atrial septal defect; CPS = critical pulmonary stenosis; F/U = follow-up; PDA = patent ductus arteriosus; PFO = patent foramen ovale; PGEi = prostaglandin Ei; SPS = severe pulmonary stenosis.

Statistical analysis. Data are expressed as the mean value ± SD when appropriate. Measurements of continuous variables for each group before and after intervention were compared using a two-tailed paired Student t test. Measurements of continuous variables between groups were compared using a two-tailed unpaired Student t test. Categoric variables were compared using the chi-square analysis or Fisher exact test. The Spearman rank correlation coefficient was used to compare ordinal variables. Correlation between continuous variables was calculated using simple linear regression analysis with the least squares method. A Levenberg Marquette nonlinear curve fit was used to assess growth of right heart structures as a function of somatic growth. Interobserver variability was calculated as the difference between the two observers’ measurements divided by the mean of the two measurements. A p value < 0.05 was considered significant.

Results

Study patients. Fourteen patients with critical PS and 20 patients with severe PS were included in the study. The demographic and clinical data of the patients at initial presentation are summarized in Table 1. Patients with critical PS presented earlier, had a smaller body surface area and had a lower initial oxygen saturation, and a higher proportion of these patients required prostaglandin Ei to maintain ductal patency compared with patients with severe PS. The percentage of patients who were intubated and ventilated at the time of cardiac catheterization was higher in patients with critical PS than in those with severe PS.

Morphometric analysis. The initial and follow-up morphometric measurements for both groups are shown in Table 2. In both groups, the average initial Z values of the right heart structures were smaller than those of normal control subjects, but remained within normal limits. At follow-up of the critical PS group, the TV Z value paralleled somatic growth, the PV annulus Z value increased from −0.05 ± 0.8 to 1.0 ± 0.9 (p < 0.003), and the Z values of the main and branch PAs significantly increased to ~2 SDs above the mean of normal control subjects (Fig. 1). Similar growth patterns after balloon pulmonary valvuloplasty were found during follow-up in the severe PS group. The growth rate of the TV paralleled the rate of somatic growth, and the growth rate of the PV annulus and PAs exceeded the rate of somatic growth.

The comparison of the Z values of the initial morphometric measurements between patients with critical and severe PS showed the initial TV Z value was slightly but significantly smaller in patients with critical PS. The RV volume was also significantly smaller in patients with critical PS compared with those with severe PS (Table 2). All patients had a tripartite RV and none of the patients had a RV to coronary artery fistula. A significant linear correlation was found between the TV diameter and RV volume (r = 0.4, p < 0.04). The initial Z values for the PV annulus and main, right and left PAs were not significantly different between the two groups. At the latest follow-up, the right and left PAs were both significantly larger in the group with critical PS compared with the group with severe PS. There were no significant differences between the groups for the mean Z values of the TV, PV and main PA.

Table 2. Morphometric Variables in Infants With Critical and Severe Pulmonary Valve Stenosis

<table>
<thead>
<tr>
<th></th>
<th>Critical PS</th>
<th></th>
<th>Severe PS</th>
<th></th>
<th>Initial CPS vs. SPS (p value)</th>
<th>Follow-Up CPS vs. SPS (p value)</th>
</tr>
</thead>
<tbody>
<tr>
<td>TV diameter (cm/BSA*0.5)</td>
<td>2.4 ± 0.2</td>
<td>2.3 ± 0.2</td>
<td>2.55 ± 0.3</td>
<td>2.32 ± 0.2</td>
<td>&lt; 0.01</td>
<td>0.07</td>
</tr>
<tr>
<td>TV Z value</td>
<td>0.00 ± 0.04</td>
<td>0.2 ± 0.6</td>
<td>0.4 ± 0.6</td>
<td>0.2 ± 0.6</td>
<td>0.21</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>PV diameter (cm/BSA*0.5)</td>
<td>1.6 ± 0.2</td>
<td>1.9 ± 0.2</td>
<td>1.68 ± 0.2</td>
<td>1.81 ± 0.1</td>
<td>&lt; 0.0002</td>
<td>0.25</td>
</tr>
<tr>
<td>PV Z value</td>
<td>−0.5 ± 0.8*</td>
<td>1.0 ± 0.9</td>
<td>−0.2 ± 0.8</td>
<td>0.5 ± 0.6</td>
<td>&lt; 0.0001</td>
<td>0.23</td>
</tr>
<tr>
<td>MPA diameter (cm/BSA*0.5)</td>
<td>1.8 ± 0.3</td>
<td>2.2 ± 0.4</td>
<td>1.87 ± 0.3</td>
<td>1.95 ± 0.2</td>
<td>&lt; 0.01</td>
<td>0.79</td>
</tr>
<tr>
<td>MPA Z value</td>
<td>0.1 ± 1.1</td>
<td>2.0 ± 1.9</td>
<td>0.5 ± 0.9</td>
<td>1.1 ± 0.7</td>
<td>&lt; 0.0001</td>
<td>0.54</td>
</tr>
<tr>
<td>RPA diameter (cm/BSA*0.5)</td>
<td>1.0 ± 0.2</td>
<td>1.4 ± 0.3</td>
<td>0.99 ± 0.2</td>
<td>1.14 ± 0.2</td>
<td>&lt; 0.003</td>
<td>0.97</td>
</tr>
<tr>
<td>RPA Z value</td>
<td>−0.4 ± 0.8</td>
<td>2.1 ± 1.6</td>
<td>−0.4 ± 0.9</td>
<td>0.5 ± 1.2</td>
<td>&lt; 0.003</td>
<td>0.82</td>
</tr>
<tr>
<td>LPA diameter (cm/BSA*0.5)</td>
<td>1.1 ± 0.2</td>
<td>1.4 ± 0.2</td>
<td>1.0 ± 0.2</td>
<td>1.2 ± 0.2</td>
<td>&lt; 0.004</td>
<td>0.73</td>
</tr>
<tr>
<td>LPA Z value</td>
<td>0.1 ± 1.0</td>
<td>2.0 ± 1.6</td>
<td>0.0 ± 1.0</td>
<td>0.8 ± 1.2</td>
<td>&lt; 0.004</td>
<td>0.63</td>
</tr>
<tr>
<td>RVEDV (ml/m²)</td>
<td>29.4 ± 6.2</td>
<td>NA</td>
<td>36.5 ± 4.7</td>
<td>NA</td>
<td>&lt; 0.0008</td>
<td>NA</td>
</tr>
</tbody>
</table>

* p < 0.05 compared with normal control values. Data are presented as mean value ± SD. BSA = body surface area; CPS = critical pulmonary stenosis; LPA = left pulmonary artery; MPA = main pulmonary artery; PV = pulmonary valve annulus; RPA = right pulmonary artery; RVEDV = right ventricular end-diastolic volume; SPS = severe pulmonary stenosis; TV = tricuspid valve.
The initial RV to PA Doppler gradients are shown in Table 1. Among patients with critical PS, 13 had no pulmonary regurgitation and 1 had mild regurgitation. Among patients with severe PS, 17 had no pulmonary regurgitation, 1 had trivial regurgitation and 2 had mild regurgitation (p NS).

Mild or more tricuspid regurgitation was present in 64% of patients with critical PS compared with 25% of patients with severe PS (p 0.03). All of the patients with critical PS and 95% of the patients with severe PS had an atrial level communication (p NS) (Table 1). The direction of flow was exclusively or predominantly right to left in patients with critical PS and exclusively or predominantly left to right in patients with severe PS. Seventy-one percent of patients with critical PS and 10% of patients with severe PS had a patent ductus arteriosus (p 0.001). The direction of ductal flow was left to right in all cases.

**Interobserver variability.** The difference between two observers (J.P.K. and T.J.F.) who had no knowledge of each other’s results were examined for the echocardiographic morphometric measurements. The average difference between the observers was 5 ± 5.7%, and the correlation coefficient between the two sets of measurements was 0.94 (p < 0.0001).

**Catheterization and balloon valvuloplasty.** Balloon pulmonary valvuloplasty was performed in all patients. The catheterization data from both groups are shown in Table 3. The initial and post balloon valvuloplasty RV to PA gradient and RV to LV pressure ratio were similar in the two groups. There were no significant differences between the two groups in the hemodynamic variables examined.

**Determinants of success of balloon pulmonary valvuloplasty.** Balloon pulmonary valvuloplasty was successful in 9 (64%) of 14 patients with critical PS and in 18 (90%) of 20 patients with severe PS. Predictors of successful balloon pulmonary valvuloplasty were examined in the patients with critical PS (Table 3). The initial RV to PA pressure gradient and initial RV to LV pressure ratio were significantly lower in

![Figure 1. Scatterplot showing the Z values of the tricuspid valve (TVz) (A), pulmonary valve (PVz) (B), main pulmonary artery (MPAz) (C), right pulmonary artery (RPAz) (D) and left pulmonary artery (LPAz) (E) in patients with critical PS at presentation and during the study period. The shaded areas represent the mean value ± 2 SD of normal control values. BSA = body surface area.](image-url)
patients with critical PS who had a successful balloon pulmonary valvuloplasty. The total number of inflations and the number of inflations with the maximal diameter balloon were higher in patients who had failure of balloon pulmonary valvuloplasty compared with those who had a successful procedure, presumably reflecting a sense of inadequate relief of the obstruction in the catheterization laboratory. There were no significant differences for the remainder of the hemodynamic variables measured or for the various balloon dilation techniques. Calendar year of procedure after 1988 was predictive of success (p < 0.04) with only one failure occurring after 1988. There were no significant differences in the morphometric measurements between patients who had successful and those who had unsuccessful balloon pulmonary valvuloplasty. One patient who had a severely dysplastic PV, as defined by Ettedgui et al. (20), underwent a successful balloon valvuloplasty. In two patients who had moderately dysplastic PVs, balloon dilation was successful in one and unsuccessful in the other. Because only two failures occurred in the group with severe PS, and these were in cases in which an adequate diameter balloon could not be advanced to the heart, no predictors of successful balloon pulmonary valvuloplasty could reliably be identified in this group of patients.

Outcomes. Among the five patients with critical PS in whom balloon valvuloplasty had failed, three patients had placement of a systemic to PA shunt and two had a surgical valvotomy performed. One of the patients who underwent a shunt procedure had a small RV and eventually had a Fontan repair. Another patient who underwent a shunt procedure subsequently underwent coil occlusion of his shunt, repeat balloon pulmonary valvuloplasty and left PA stent placement, and therefore required no further surgical treatment. One of the patients had a surgical valvotomy and infundibular resection and had a complicated postoperative course with RV failure. This patient eventually died, and this was the only death in this group of patients. There were no major complications related to catheterization in this group of patients. There was no restenosis after an initially successful balloon valvuloplasty. One patient who initially had a shunt placed required repeat balloon valvuloplasty 7 years later. In patients with critical PS, the average RV to PA gradient at follow-up was 15 ± 10 mm Hg. All patients had pulmonary regurgitation at follow-up, which was mild in 11 and moderate in three patients. Of the three patients with moderate pulmonary regurgitation, there was no evidence of RV volume overload during follow-up in two patients. One patient had mild RV enlargement by echocardiography.

Among patients with severe PS, there were two failures, and both were related to the inability to advance a large enough balloon to achieve adequate dilation. One case was an extremely premature infant who had avulsion of his inferior vena cava. This patient had a Brock procedure and ligation of the inferior vena cava and eventually died. This was the only death in this group of patients. The other patient had partial relief of his obstruction and underwent successful balloon pulmonary valvuloplasty 11 months later. The only other major complication related to the catheterization in this group was in a patient who had a small cerebral infarct, which was presumed to be embolic. This patient had no long-term neurologic sequelae. Restenosis after an initially successful balloon valvuloplasty occurred in one of 18 patients. Two years after the procedure, this patient had a RV to PA pressure gradient of 75 mm Hg and underwent repeat balloon pulmonary valvuloplasty. In patients with severe PS, the RV to PA gradient at follow-up was 22 ± 15 mm Hg. All patients had trivial to mild pulmonary regurgitation at follow-up.
Discussion

This study demonstrates that the right heart structures increase in size in patients with critical and severe PS after balloon pulmonary valvuloplasty. In both groups, after ~3 to 4 years of follow-up, growth of the TV relative to somatic growth was parallel to that of the normal population. The growth of the PV annulus and PAs relative to somatic growth exceeded that of normal control subjects (Fig. 1). The observation that the growth rates of the PV annulus and PAs exceed that of the normal population may be related to post-stenotic dilation or to an increased RV stroke volume secondary to pulmonary regurgitation. In patients with critical PS, the PAs became dilated and significantly larger than the group with severe PS. This may be related to the higher degree of pulmonary regurgitation seen in these patients or to the presence of surgical systemic to PA shunts in three patients. The increased size of the PAs was not associated with any apparent clinical problem in any of the study patients, but long-term follow-up is warranted.

Previous studies. Previous studies have yielded varying reports on the growth of the PV annulus. Although Fedderly et al. (7) found that the Z values of the PV did not change significantly after balloon valvuloplasty in patients with critical PS, Tabatabaci et al. (14) found a significant increase. The Z values demonstrated in this study were slightly larger than the Z values reported in two previous studies (7,21). These small differences may be explained by the varied techniques used. The Z values of the PV annulus in these previous studies were based on normative data derived from Hanseus et al. (22), in which the measurement of the PV annulus was performed in diastole. In the present study and in the patients used for normative data, the PV annulus was measured in systole when the annulus diameter is larger.

Distinguishing factors between critical and severe PS. Patients with critical PS were distinguished from patients with severe PS by several factors. These factors included a smaller TV annulus diameter and RV volume and a higher degree of tricuspid regurgitation in patients with critical PS. As expected, all patients with critical PS demonstrated predominantly right to left atrial shunting compared with predominantly left to right shunting in patients with severe PS. Interestingly, there were no significant differences between the two groups in the hemodynamic variables measured including right atrial pressure, RV end-diastolic pressure, RV to PA pressure gradient and RV to LV pressure ratio. Unfortunately, left atrial pressure was not routinely measured at catheterization, and this would have allowed comparison of the right and left atrial pressures. Speculatively, in patients with critical PS, decreased pulmonary blood flow and subsequent decreased pulmonary venous return may affect the relative compliance of the right and left atria. Together with the higher degree of tricuspid regurgitation, smaller TV and RV, this may contribute to the differences in the degree of right to left atrial level shunting, and thus to differences in clinical presentation.

Balloon pulmonary valvuloplasty. The overall success rate of balloon pulmonary valvuloplasty for this study was 79%, which is comparable to other studies (7,9–14,21). Restenosis after successful balloon pulmonary valvuloplasty was rare and occurred in ~3% of patients, none of whom had critical PS. In patients with critical PS, a lower initial RV to PA pressure gradient and RV to LV pressure ratio were associated with successful balloon pulmonary valvuloplasty. A later year of catheterization after 1988 was also shown to be associated with success of the procedure. This may be related to operator experience or to advances in techniques or technology. These predictors of successful balloon pulmonary valvuloplasty are comparable to other studies (7,21). Previous studies have shown an association between morphometric variables and the outcome of balloon pulmonary valvuloplasty, with larger PV and TV diameters being associated with success of balloon pulmonary valvuloplasty (7,11,21). None of the morphometric variables measured in the present study were associated with the success or failure of balloon pulmonary valvuloplasty. The results of PV balloon dilation in this study, together with other reports (7,9–14), support recommending balloon dilation as the initial procedure in newborns and infants with critical and severe PS.

Conclusions. This study demonstrates that growth of right heart structures parallels or exceeds somatic growth after balloon pulmonary valvuloplasty in patients with critical and severe PS. The smaller right heart structures, higher degree of tricuspid regurgitation and subsequent larger degree of right to left atrial shunting in patients with critical PS compared with infants with severe PS may contribute to differences in clinical presentation.

References

11. Talsma M, Witsenburg M, Rohmer J, Hess J. Determinants for outcome of