Influence of Right Heart Size on Outcome in Pulmonary Atresia With Intact Ventricular Septum

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Background. Neonates with pulmonary atresia and intact ventricular septum (PA-IVS) are frequently born with hypoplastic right heart structures that must grow after right ventricular decompression (RVD) procedures for a complete two-ventricle physiology to be achieved. Previous authors have asserted that neonatal right heart size or morphology will predict right heart growth potential. Since 1983, our bias has favored early RVD regardless of initial right heart size. In 1986, we recognized a subset of patients with coronary artery abnormalities associated with poor outcome after RVD and have defined these patients as having a right ventricular-dependent coronary circulation (RVDCC).

Methods and Results. To assess the influence of right heart size on outcome independent of the presence of RVDCC, we measured echocardiographic right ventricular (RV) dimensions in 37 neonates with adequate studies presenting between 1983 and 1990. Coronary artery anatomy was adequately assessed by angiography in 36. RV volume and tricuspid valve (TV) diameter were significantly smaller in patients with RVDCC than in those without. However, there was no statistically significant association between RV volume or TV diameter and survival among patients with or without RVDCC. Among 29 patients without RVDCC, 23 of 24 (95.8%) who achieved RVD are alive compared with 1 of 5 (20%) who did not achieve RVD (P=.001). Twenty-one of the 23 survivors have a complete two-ventricle physiology with low right atrial pressure. Among 7 patients with RVDCC, 2 patients who underwent RVD died early of left ventricular failure, whereas 4 of 5 who did not undergo RVD have survived single ventricle palliation.

Conclusions. Small right heart size is associated with RVDCC but is not associated with survival in PA-IVS. Patients without RVDCC have improved survival after RVD regardless of neonatal right heart size. (Circulation. 1993;88[part 1]:2248-2256.)

Key Words • pulmonary atresia • right ventricle • congenital heart surgery

In the ideal surgical outcome for patients with pulmonary atresia with intact ventricular septum (PA-IVS), the right ventricle provides the total pulmonary blood flow at a low filling pressure with no residual right-to-left shunt. At birth, however, patients with PA-IVS have varying degrees of right ventricular (RV) and tricuspid valve (TV) hypoplasia as well as systolic and/or diastolic dysfunction. Whether the right ventricular outflow tract (RVOT) should be opened in the neonatal period depends to a significant extent on the potential of the right heart structures to grow and develop. Some groups have asserted that initial right heart size and/or morphology will predict whether the ventricle will grow. These groups have suggested that neonatal RV anatomy can predict subsequent right heart function.

Since 1983, our institutional bias has been to attempt RV decompression in all neonates with PA-IVS regardless of RV size, TV size, RV anatomy, coronary fistulae, or coronary stenoses. In 1986, this strategy was amended based on coronary artery anatomy. We recently published our results defining patients with RV-dependent coronary circulation (RVDCC) as those with right ventricle to coronary artery fistulae plus stenoses of both the right and left coronary systems. It is our current policy to attempt RV decompression in all patients with PA-IVS who do not have a RVDCC irrespective of right heart size. The purpose of this investigation is to (1) use echocardiographic data to evaluate the relation between neonatal RV or TV size and outcome in patients with PA-IVS and (2) assess the success of our current surgical strategy and the resources used in following this strategy.

Methods

Patient Selection

All patients with the diagnosis of PA-IVS who presented for initial management to the Children's Hospital, Boston, between January 1983 and January 1990 were identified by a search of the computerized cardiology data base.

Echocardiographic Measurement of Neonatal RV Volume and TV Diameter

Two-dimensional echocardiograms were performed using either a Hewlett-Packard 77020A ultrasound sys-
system or an Acuson cardiac imager with a 5-MHz transducer. Using a Dextra D-200 cardiac analysis system (Dextra Medical Inc, Long Beach, Calif), the endocardial border of each right ventricle was traced at end diastole in the subxiphoid long- and short-axis planes. The RV diastolic volume was estimated using a Simpson’s rule algorithm and was indexed for body surface area. The maximum TV annulus was measured in the apical four-chamber view.

Patients in whom there was inadequate visualization of the endocardial border of the right ventricle at end diastole and/or inadequate visualization of the TV on neonatal echocardiogram were subsequently excluded. Measurements of RV volume and TV annulus diameter were obtained solely for the purpose of this study by two reviewers (T.M.G., A.M.).

Assessment of RV Anatomy

We assessed RV anatomy using preoperative neonatal right ventricular angiograms. Patients were classified as having a unipartite, bipartite, or tripartite right ventricle based on the presence or absence of RV inflow, apex and outflow portion, as previously described by Bull et al.14

Assessment of Coronary Anatomy

Preoperative coronary angiograms were reviewed solely for the purpose of this study by two investigators (T.M.G., V.S.M.). Coronary artery anatomy was classified according to a predetermined protocol as (1) fistulae alone, (2) fistulae plus stenoses to a single coronary artery, and (3) fistulae plus stenoses to multiple coronary arteries. In one patient (patient 5) without neonatal angiograms, coronary anatomy was assessed by postmortem exam. Patients were defined as having RV-dependent coronary circulation (RVDCC) when coronary artery stenoses of both the right and left coronary systems were present, as described previously.42

Definition of Successful RV Decompression

Successful RVOT reconstruction, or RV decompression, was defined as achievement of RV pressure that was less than one half of systemic pressure at or before 2 years of age.

Determination of Outcome

The hospital medical records for all patients were reviewed for surgical procedures, cardiac catheterizations, and clinical course, including death. Follow-up times are defined as age on January 1, 1992, the date of initial medical record review.

Resource Implications

To estimate the health care resources used by our current surgical strategy, we computed the total number of surgical procedures, cardiac catheterizations, echocardiograms, and in-hospital weeks used per survivor for (1) all patients without RVDCC in whom RV decompression was attempted and (2) all patients with RVDCC who were managed with a single ventricle palliation without RV decompression.

Statistical Analysis

ANOVA was used to compare continuous variables between survivors and nonsurvivors in the group as a whole and in those patients without RVDCC. The nonparametric rank-sum test was used to compare continuous variables between survivors and nonsurvivors in patients with RVDCC. Comparisons of dichotomous variables were made using a Fisher’s exact test. The relation between indexed RV volume and TV diameter was assessed using a Pearson linear correlation coefficient. Ordinal logistic regression was used to adjust for the influence of RV volume on the relation between RV decompression and survival in patients without RVDCC. A P value of .05 was considered sufficient evidence to reject the null hypothesis of no association between variables.

Results

Patient Characteristics

Of 46 patients with PA-IVS identified by computer search, 9 (19.6%) were subsequently excluded from analysis because of inadequate echocardiographic studies for RV volume and TV diameter measurement. The characteristics of the remaining 37 patients are summarized in Tables 1 and 2. The characteristics of the 9 patients excluded from the data analysis are summarized in Table 3.

Lack of Relation Between RV Volume or TV Diameter and Survival

There were 9 deaths in the total group of 37, for an overall mortality of 24.3%. For the group as a whole, the differences in indexed RV volume and TV annulus diameter between the survivors and nonsurvivors were not statistically significant (10.9±5.7 mL/m², mean±SD, vs 7.7±5.8 mL/m² and 11.0±3.8 mm vs 8.5±2.0 mm; P=NS).

Indexed RV volumes for the 37 patients ranged from 1.0 to 22.1 mL/m², and TV diameters ranged from 4.3 to 20.0 mm. In our laboratory, estimated normal values for RV volumes range from 30 to 50 mL/m²; similarly, normal newborn tricuspid valve dimensions range from 9 to 17 mm. Linear regression analysis revealed a

<table>
<thead>
<tr>
<th>Table 1. Patient Characteristics (n=37)</th>
</tr>
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<tbody>
<tr>
<td>Indexed RV volume, mL/m² (mean±SD)</td>
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<tr>
<td>TV annulus diameter, mm (mean±SD)</td>
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<tr>
<td>RV anatomy</td>
</tr>
<tr>
<td>Tripartite</td>
</tr>
<tr>
<td>Bipartite</td>
</tr>
<tr>
<td>Coronary artery anatomy</td>
</tr>
<tr>
<td>Normal</td>
</tr>
<tr>
<td>Abnormal</td>
</tr>
<tr>
<td>Fistulae only</td>
</tr>
<tr>
<td>Fistulae + stenosis of 1 coronary artery</td>
</tr>
<tr>
<td>Fistulae + stenosis of right and left coronaries</td>
</tr>
<tr>
<td>Follow-up time, y (mean±SD, range)</td>
</tr>
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</table>

RV indicates right ventricle and TV, tricuspid valve.
**TABLE 2. Neonates With Echocardiograms Adequate for Analysis**

<table>
<thead>
<tr>
<th>Patient</th>
<th>RVVI, mL/m²</th>
<th>TVD, mm</th>
<th>RVDCC</th>
<th>Age at Successful RVD</th>
<th>Initial Surgical Procedure</th>
<th>Status</th>
<th>Age at Follow-up (or Death), y</th>
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<tr>
<td>1</td>
<td>4.3</td>
<td>10.0</td>
<td>No</td>
<td>12 mo</td>
<td>PV*/LMBTs</td>
<td>A1</td>
<td>7.8</td>
</tr>
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<td>2</td>
<td>5.5</td>
<td>7.0</td>
<td>No</td>
<td>4 d</td>
<td>RVOT/PMBT</td>
<td>A1</td>
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</tr>
<tr>
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<td>7.6</td>
<td>7.0</td>
<td>No</td>
<td>6 d</td>
<td>PV</td>
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<td>8.3</td>
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<td>4</td>
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<td>PV*/LMBTs</td>
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<td>(0.3)</td>
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</tr>
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<td>PV*</td>
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<td>6.7</td>
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</tr>
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<td>PV*</td>
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<td>5.6</td>
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<td>2 d</td>
<td>PV</td>
<td>A1</td>
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</tr>
<tr>
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<td>9.0</td>
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<td>12.0</td>
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<td>RVOT/PMBT</td>
<td>A1</td>
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</tr>
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<td>(1.3)</td>
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<td>31</td>
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<td>No</td>
<td>3 d</td>
<td>RVOT/PMBT</td>
<td>DD</td>
<td>(6 d)</td>
</tr>
<tr>
<td>32</td>
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<td></td>
<td>RMBT/PA</td>
<td>DS</td>
<td>(12 d)</td>
</tr>
</tbody>
</table>

A1 indicates alive with atrial septal defect and shunt closed; A2, alive with atrial septal defect; A3, alive with Glenn; AF, alive with modified Fontan; AS, alive with shunt; DD, dead after right ventricular decompression; DF, dead with modified Fontan; DS, dead with shunt; PA, pulmonary artery; RMBT, right (modified) Blalock-Taussig shunt; RVD, right ventricular decompression; RVDCC, right ventricular-dependent coronary circulation; RVOT, transannular right ventricular outflow tract patch; RVV, right ventricular volume; RVVI, right ventricular volume indexed; PV, pulmonary valvotomy; and TVD, TV annulus diameter.

*Incomplete RVD.
†Inadequate angiogram.
TABLE 3. Neonates With Echocardiograms Inadequate for Analysis

<table>
<thead>
<tr>
<th>Patient</th>
<th>RVVI, mL/m²</th>
<th>TVD, mm</th>
<th>RVDCC</th>
<th>Age at Successful RVD</th>
<th>Initial Surgical Procedure</th>
<th>Status</th>
<th>Age at Follow-up (or Death), y</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>...</td>
<td>...</td>
<td>...</td>
<td>2 d</td>
<td>RVOTP/F-PDA</td>
<td>A1</td>
<td>9.0</td>
</tr>
<tr>
<td>2</td>
<td>...</td>
<td>...</td>
<td>...</td>
<td>...</td>
<td>PV*/LMBTSS</td>
<td>DS</td>
<td>(2.3)</td>
</tr>
<tr>
<td>3</td>
<td>...</td>
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<td>2 d</td>
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<td>A1</td>
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</tr>
<tr>
<td>4</td>
<td>...</td>
<td>...</td>
<td>...</td>
<td>12 mo</td>
<td>PV*</td>
<td>A1</td>
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</tr>
<tr>
<td>5</td>
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<td>DC</td>
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</tr>
<tr>
<td>7</td>
<td>...</td>
<td>...</td>
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<td>(14 d)</td>
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<td>9</td>
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<td>...</td>
<td>4 d</td>
<td>PV/RBTS</td>
<td>A1</td>
<td>9.9</td>
</tr>
</tbody>
</table>

A1 indicates alive with atrial septal defect and shunt closed; AF, alive with modified Fontan; AS, alive with shunt; d, dead; DD, dead after right ventricular decompression; DC, dead with collateral; DF, dead with modified Fontan; DS, dead with shunt; F-PDA, formal infiltration of patent ductus arteriosus; PA, pulmonary artery; RMBTS, right (modified) Blalock-Taussig shunt; RVD, RV decompression; RVDCC, right ventricular-dependent coronary circulation; RVOTP, transannular RV outflow tract patch; RVV, right ventricular volume; RVVI, right ventricular volume indexed; PV, pulmonary valvotomy; and TVD, TV annulus diameter.

*R* indicates alive.

†Pulmonary blood supply via aortopulmonary collaterals (no surgery).

A significant correlation between indexed RV volume and TV annulus diameter (*r*=.73).

**RVDCC and RV Size**

Thirty-six of the 37 patients (97.3%) had RV angiograms of sufficient quality to determine coronary artery anatomy. Seven of these 36 patients (19.4%) were classified as having RVDCC, whereas 29 patients (80.6%) did not have RVDCC. Both the indexed RV volume (3.8±1.7 mL/m² vs 11.9±5.0 mL/m², *P*=.001) and the TV annulus diameter (6.9±1.6 mm vs 11.5±3.2 mm, *P*=.013) were significantly smaller in the patients with RVDCC in comparison to those without RVDCC. These data are shown in Fig 1.

The single patient (patient 13) whose coronary artery anatomy could not be determined had an indexed RV volume of 1.5 mL/m² and a TV diameter of 7.0 mm. This patient underwent initial placement of a right modified Blalock-Taussig shunt (BTS) without RV decompression and died at 25 days of life.

**Outcome of Patients With RVDCC**

Of the 7 patients with RVDCC, 2 underwent neonatal RV decompression before 1986 (see Fig 2). Both of these patients died within 18 hours of surgery with clinical and echocardiographic evidence for left ventricular dysfunction (patients 5 and 6). Of the 5 remaining patients, 4 (80%) are alive with a single ventricle palliation (1 Fontan, 3 BTS) at a follow-up of 5.5±3.1 years (patients 8, 9, 10, 11). One patient (20%) died late, 1.3 years after placement of a right BTS (patient 14). There was no statistically significant difference in indexed RV volume (3.1±1.0 mL/m² vs 4.8±2.0 mL/m²) or TV annulus diameter (5.5±0.5 mm vs 7.1±0.7 mm) between survivors and nonsurvivors among patients with RVDCC (*P*=NS).

**Outcome of Patients Without RVDCC**

Of the 29 patients without RVDCC, RV decompression was attempted in 26 (89.7%) and was successful in 24 (82.8%). Twenty-three of the 26 (89%) who had attempted RV decompression are alive in comparison to 1 of 3 (33.3%) who did not undergo decompression (*P*=.07). Twenty-three of 24 (95.8%) who achieved successful RV decompression are alive in comparison to 1 of 5 (20%) who were not successfully decompressed (*P*=.001). Thus, achievement of a successful RV decompression was highly related to survival among patients without RVDCC. See Fig 2.

Of the 5 patients who did not achieve a successful RV decompression, 3 did not undergo RV decompression because of the choice of their individual physician, and 2 had an unsuccessful operative decompression. One infant in the "physician’s choice" group had severe tricuspid incompetence producing RV pressures less than 50% of systemic pressures despite pulmonary atresia. This patient died early from right heart failure (patient 33). A second patient who did not undergo RV decompression based on physician's choice subse-
quently died after a modified Fontan operation (patient 12). Of the 2 patients with unsuccessful operative decompression, both had a single surgical procedure with no further attempts at RV decompression. One child was found to have intracavity RV muscle bundles causing persistent severe RV hypertension 1 year after placement of an RV outflow tract patch; the patient died suddenly awaiting surgical relief (patient 30). A second patient underwent an initial unsuccessful decompression procedure and died of fungal sepsis 12 days after surgery (patient 32). Of this group who never achieved successful RV decompression, one patient is alive at a follow-up of 7.0 years. This patient did not undergo an attempt at RV decompression based on physician's choice and has subsequently undergone a modified Fontan procedure (patient 7).

Of the 24 patients who achieved a successful RV decompression, 17 (70.8%) achieved decompression after a single surgical procedure, 3 with RV transannular pericardial outflow tract patch reconstruction alone, 11 with RV outflow tract patch with BTS, and 3 with pulmonary valvotomy with or without BTS. Seven of these 24 (29.2%) required two operations to decompress their right ventricles. In 6 of the 7 requiring two operations to decompress the right ventricle, the initial attempt was a pulmonary valvotomy. Subsequent successful RV decompression was achieved via RV transannular pericardial outflow tract patch in 6 and transcatheter dilation of the RVOT in 1. Pulmonary valvotomy was successful as an initial surgical procedure in only 3 of 9 patients (33%) compared with RV transannular pericardial outflow tract patch as an initial successful surgical procedure in 14 of 15 (93.3%, \(P<.004\)). Overall, successful RV decompression in these 24 patients was achieved at a median age of 4 days (range, 2 days to 23 months).

Of the 24 patients who achieved a successful RV decompression, 23 (95.8%) are alive at a mean follow-up of 5.0 years (range, 2.2 to 8.6). Twenty-one of these 23 (91.3%) have achieved a complete two-ventricle repair with closure of both residual atrial septal defects and systemic to pulmonary artery shunts. Angiograms showing RV growth for one patient in this group are shown in Fig 3. One patient has a residual small right-to-left atrial shunt (patient 23), and one has a right Glenn anastomosis (patient 4). Eventual com-
Lack of Relation of RV Volume or TV Diameter to Outcome Among Patients Without RVDCC

Patients without RVDCC who did not undergo RV decompression tended to have smaller indexed RV volumes than patients who underwent RV decompression (9.4±6.4 mL/m² vs 12.4±4.8 mL/m², \( P = .23 \)). However, there was no statistically significant difference between indexed RV volume (12.2±5.0 mL/m² vs 10.7±6.0 mL/m²) or TV annulus diameter (11.7±3.5 mm vs 10.0±1.8 mm) among survivors and nonsurvivors by univariate analysis. In a regression model adjusting for indexed RV volume, achievement of successful RV decompression remained highly associated with survival (\( P = .006 \)). Kaplan-Meier survivor curves for patients without RVDCC who did and did not achieve successful RV decompression are shown in Fig 4.

Resource Implications

These data indicate that optimum strategy for patients without RVDCC is decompression, whereas optimum strategy for patients with RVDCC is probably single ventricle palliation. Thus, 31 of 36 patients (86.1%) were treated according to an optimum surgical strategy. Twenty-six patients without RVDCC underwent RV decompression procedures with 23 survivors. These 26 patients used a total of 45 surgical procedures, 67 catheterizations, 151 echocardiograms, and 111 in-hospital weeks. Therefore, the strategy of RV decompression for patients without RVDCC consumed, on average, 2.0 operations, 2.9 catheterizations, 6.6 echocardiograms, and 4.8 in-hospital weeks per survivor. Similarly, 5 patients with RVDCC did...
Discussion

Previous authors have suggested that various preoperative measures of RV or TV hypoplasia might predict which patients with PA-IVS had right ventricles that were capable of growth to normal size and thus which patients would benefit from early RV decompression. Most studies have determined that patients with significant RV and TV hypoplasia did poorly and have concluded that tiny right heart structures were incapable of normal growth with preserved long-term RV function. Although a few studies have documented normal growth of markedly hypoplastic right ventricles after decompression in some patients, it is still debatable whether RV decompression and survival among patients with PA-IVS. This study does, however, show a significant association between both indexed RV volume and TV diameter and the presence of coronary artery fistulae with stenoses to multiple coronary arteries (RVDCC), which we have previously shown contributes to mortality in these patients.

In contrast, this study shows no statistically significant association between indexed RV volume or TV diameter and survival among patients with PA-IVS. This study does, however, show a significant association between both indexed RV volume and TV diameter and the presence of coronary artery fistulae with stenoses to multiple coronary arteries (RVDCC), which we have previously shown contributes to mortality in these patients.

In addition, patients who did not have RV-dependent coronary circulation but achieved successful RV decompression had a markedly improved survival (95.8%) compared with similar patients who did not achieve successful decompression (20%). Moreover, despite the presence of patients with very hypoplastic right heart structures (minimum indexed RV volume, 4.3 mL/m²; minimum TV diameter, 4.3 mm) in this group, most patients (21 of 23, 91.3%) currently have evidence for excellent RV function at a mean follow-up of 5.0 years, as evidenced by their having achieved a complete two-ventricle repair, at low right atrial pressure. Although 2 patients had evidence of TV stenosis after RVD, this did not preclude eventual attainment of a complete two-ventricle repair.

Also, among the small group of patients with RV-dependent coronary circulation in this series, patients who underwent RV decompression died soon after surgery with evidence for severe left ventricular dysfunction, whereas 4 of 5 patients who did not undergo RV decompression are alive with a single ventricle palliation.

Study Limitations

Certain limitations must be recognized in this study. First, it is a retrospective design. Second, pulmonary atresia with intact ventricular septum is a rare disease, and the number of patients with markedly hypoplastic right ventricles is limited. In addition, since the strategy of our institution is to decompress all right ventricles in this disease except in the presence of a RVDCC, the number of patients in the nondecompressed group is obviously small, thereby limiting the statistical power of our analyses. Finally, 9 patients of our total patient population were excluded from analysis secondary to inadequate neonatal echocardiograms for assessment of right ventricle size. Since the clinical course of these 9 patients was similar to the clinical course in the 37 patients analyzed, the exclusion of these patients from the analysis does not seem to alter the inferences.
Despite these limitations in sample size and selection, several conclusions appear warranted.

Conclusions

These results support our current hypothesis that coronary artery anatomy and not RV or TV hypoplasia predicts which patients with PA-IVS will do well after early RV decompression. Earlier authors may have concluded that outcome after RV decompression in patients with PA-IVS could be predicted by RV and TV size because of the frequent presence of coronary artery fistulae with stenoses of multiple coronary arteries in patients with markedly hypoplastic right hearts.

These results also support our current strategy for aggressive angiographic determination of coronary artery anatomy in neonates with PA-IVS and early RV decompression using RV outflow tract patch for all patients without RV-dependent coronary circulation, independent of RV or TV size. Assessment of management strategy for patients with RV-dependent coronary circulation is limited by the small number of patients with RVDCC in this series. However, the available data suggest that single ventricle palliation without RV decomposition may be an appropriate management strategy for these patients.

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