Evaluation and Surgical Treatment of Pulmonary Atresia and Intact Ventricular Septum in Infancy

ALAN B. LEWIS, M.D., WINFIELD WELLS, M.D., AND GEORGE G. LINDESMITH, M.D.

SUMMARY The initial surgical approach to the infant with pulmonary atresia and intact ventricular (PA-IVS) is to establish an adequate source of pulmonary blood flow and, when possible, relieve right ventricular (RV) outflow obstruction. The selection of patients for pulmonary valvotomy, alone or in combination with a systemic–pulmonary arterial shunt, depends on the presence of an RV outflow tract and the adequacy of the RV chamber. To evaluate the size of the RV cavity in PA-IVS, an RV index (RVI) was developed using biplane angiographic measurements of the sum of the tricuspid valve annulus and the RV inflow and the RV outflow tracts. The RVI was normalized by relating it to the aortic diameter (Ao) at the diaphragm (RVI/Ao). The RVI/Ao was 13.5 ± 1.4 in 20 control subjects and only 7.3 ± 2.6 in 26 PA-IVS patients (p < 0.001), and was within the normal range in only two of the 26. Since 1976, pulmonary valvotomy plus a Blalock-Taussig shunt has been performed in 10 infants, with one death. Serial cardiac catheterizations in five of nine survivors demonstrated substantial RV growth in all, with the RVI/Ao increasing from an average of 8.0 to 12.5. In contrast, patients who underwent a shunt alone had no change in RV cavity size. We conclude that pulmonary valvotomy may be performed successfully in most PA-IVS patients, but usually must be combined with a systemic–pulmonary shunt. In a small minority of patients, a normal RV cavity, as evidenced by an RVI/Ao ≥ 11, appears to be sufficient to sustain adequate pulmonary blood flow after valvotomy alone. The RVI/Ao ratio is a simple method of quantitatively evaluating RV cavity size and is helpful in planning the initial surgical approach for these infants.

THE SURVIVAL of neonates with pulmonary atresia and intact ventricular septum (PA-IVS) is dependent upon the provision of a source of adequate pulmonary blood flow. The use of prostaglandin E1 (PGE1) is a valuable, although usually short-term, method for maintaining postnatal patency of the ductus arteriosus (PDA).1–3 Therefore, early surgical intervention is necessary to ensure a more permanent route for pulmonary flow. Most surgical approaches to the problem have been based on the pathologic classification of Green- wold et al.,4–3 which divides the right ventricle into two anatomic types: a predominant group with a small right ventricular (RV) cavity (type I) and a less common group with a normal-sized or large RV cavity (type II). Accordingly, early reports of the infants emphasized the need for a systemic arterial–pulmonary arterial shunt in the majority of patients with tiny right ventricles.6,7 Morphologic studies of PA-IVS have documented both a broad spectrum of RV cavity size8 and the ability of the right ventricle to increase after successful pulmonary valvotomy.9 Nevertheless, numerous reports have revealed high mortality rates in patients with small right ventricles in whom pulmonary valvotomy was the sole initial surgical procedure.10–13 Therefore, preoperative angiographic assessment of RV morphology12 and size9 are crucial in planning a comprehensive initial surgical approach to PA-IVS. Studies by Graham et al.9 and Patel and co-workers14 demonstrated the value of selective angiography for RV volume determinations in infants with PA-IVS. However, the application of Simpson’s rule to these often irregular and oddly shaped ventricles is of concern.12

We reviewed our experience with PA-IVS since 1970 in an effort to develop a simplified method of quantitatively evaluating RV cavity size from the angiogram. The application of this method as an adjunct to planning the initial surgical treatment of this malformation and to the postoperative evaluation of RV growth is reported.

Materials and Methods

The records of 27 infants with PA-IVS who presented to Childrens Hospital of Los Angeles between 1970 and June 1982 were reviewed. Patients with critical pulmonary stenosis without atresia, Ebstein’s malformation or other complicating cardiac lesions were excluded.

Cardiac catheterization data and RV angiograms were reviewed. Pre- and postoperative findings were compared in patients who underwent serial cardiac catheterization. The RV angiograms were recorded in the posterior-anterior and lateral projections during biplane filming. An index of RV size was developed from a modification of the morphologic measurements of Zuberbuhler and Anderson.8 The tricuspid valve annulus (TVA), RV inlet and RV outlet dimensions were measured as shown in figure 1. All measurements were made at end-diastole during sinus rhythm in both the posterior-anterior and lateral views and calibrated using a standard 1-cm grid placed at the mid-chest isocentric position. The TVA was measured at its maximum diameter from the superior to the inferior rim of the annulus. The RV inlet was measured from the midportion of the TVA line to the apex. When the apical or trabecular region was truncated, however, the RV inlet measurement extended to the inferior portion of the RV inflow tract farthest from the TVA. Care was taken to avoid inclusion of trabeculations. The RV outlet was measured from the point identified as the
RV apex to the most distal point in the RV outflow tract. The RV index (RVI) was calculated by averaging the sum of the biplane measurements using the formula:

\[
\text{RVI} = \frac{(TVA + RV\text{ inlet} + RV\text{ outlet})}{2}
\]

Current understanding of fetal flow patterns supports the proposition that the aortic diameter at the diaphragm in neonates with PA-IVS should be similar to that in normal neonates; therefore, the RVI was normalized by relating it to the diameter of the descending aorta at the diaphragm (RVI/Ao). The RVI and aortic diameter in patients with PA-IVS were matched against 20 control subjects who had undergone right- and left-heart catheterization and angiography. These patients were selected on the basis of diagnoses that should not have produced abnormalities of RV size and morphology or aortic diameter. They included seven patients with functional murmurs, one with bicuspid aortic valve with a left ventricular–aortic peak systolic pressure gradient $\leq 10$ mm Hg, nine with small ventricular septal defects or PDA with pulmonary/systemic flow ratios $\leq 1.3:1$ and normal right-sided pressures, and three with minimal pulmonary valve stenosis and a maximal RV systolic pressure $\leq 40$ mm Hg and an RV–pulmonary artery peak systolic pressure drop $\leq 10$ mm Hg. They were 3 days to 16 years old (median 1.5 years). Seven control subjects were younger than 1 year of age and had a body surface area $< 0.4$ m$^2$ at the time of catheterization; three were less than 6 months old.

The data were analyzed by linear regression analysis and $t$ test using a Hewlett-Packard desktop computer (model 9845B).

**Results**

The clinical data for the 27 infants with PA-IVS are summarized in table 1. Twenty-three infants presented with cyanosis before 1 week of age; three did not come to medical attention until they were older than 1 month.

### Table 1. Summary of Clinical Data in Infants with Pulmonary Atresia and Intact Ventricular Septum

<table>
<thead>
<tr>
<th>Category</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at initial presentation</td>
<td>n</td>
</tr>
<tr>
<td>&lt; 1 week</td>
<td>23</td>
</tr>
<tr>
<td>1–4 weeks</td>
<td>1</td>
</tr>
<tr>
<td>&gt; 1 month</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>27</td>
</tr>
<tr>
<td>Prostaglandin E$_1$ administered</td>
<td>15</td>
</tr>
<tr>
<td>Procedures</td>
<td></td>
</tr>
<tr>
<td>Balloon atrial septostomy</td>
<td>9</td>
</tr>
<tr>
<td>Systemic–pulmonary shunt</td>
<td>25</td>
</tr>
<tr>
<td>Blalock-Taussig</td>
<td>13</td>
</tr>
<tr>
<td>Waterston</td>
<td>11</td>
</tr>
<tr>
<td>Formalin infiltration PDA</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary valvotomy (± shunt)</td>
<td>11</td>
</tr>
</tbody>
</table>

**Abbreviation:** PDA = patent ductus arteriosus.

Since June 1976, all patients have been treated with PGE$_1$ to maintain postnatal patency of the ductus arteriosus. Balloon atrial septostomy had been performed in the majority of infants before 1978, but has been used in only one of 13 cases since then. A surgical systemic–pulmonary artery shunt was created in 25 of 27 patients. One child had a large PDA at 15 months of age and another child underwent a transventricular pulmonary valvotomy without an additional shunt procedure. A Waterston anastomosis was performed in 11 patients before June 1976. Since then, a right Blalock-Taussig anastomosis has been the shunt procedure of choice. Ten patients underwent a combined right Blalock-Taussig shunt and transventricular pulmonary valvotomy through a right thoracotomy. After satisfactory completion of the subclavian–pulmonary artery anastomosis, the pericardium was incised anterior to the phrenic nerve and continued well to the left to expose the RV outflow tract. A purse-string suture was placed in the surface of the right ventricle and a 15-gauge needle was inserted through the wall of the ventricle until it pierced the pulmonary valve. A small, curved hemostat was then introduced into the RV outflow tract. A purse-string suture was placed in the surface of the right ventricle and a 15-gauge needle was inserted through the wall of the ventricle until it pierced the pulmonary valve. A small, curved hemostat was then introduced into the RV outflow tract, passed through the valve and opened in the vertical and horizontal planes. The opening in the RV anterior wall was closed with the previously placed purse-string suture.

The hemodynamic and morphologic characteristics of the right ventricles, determined by cardiac catheterization and angiography, are summarized in table 2. RV peak systolic pressure was equal to or greater than systemic systolic pressure in all 21 patients in whom it

### Table 2. Hemodynamic and Angiographic Characteristics in Infants with Pulmonary Atresia and Intact Ventricular System

<table>
<thead>
<tr>
<th>Category</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>RV systolic pressure $\geq$ systemic</td>
<td>21/21</td>
</tr>
<tr>
<td>RV outflow tract present</td>
<td>21/26</td>
</tr>
<tr>
<td>RV sinusoidal-coronary communications</td>
<td>7/26</td>
</tr>
<tr>
<td>MPA present</td>
<td>25/26</td>
</tr>
</tbody>
</table>

**Abbreviations:** RV = right ventricular; MPA = main pulmonary artery.
was recorded. The RV outflow tract could be identified clearly in 21 of 26 patients. In one infant the right ventricle could not be selectively catheterized, and only a right atrial angiogram was performed. RV sinusoidal-coronary communications were present in seven infants. Contrast material usually entered both the left anterior descending and right coronary arteries. Retrograde opacification of the aortic root via the coronary arteries was also present in several instances. The main pulmonary artery was identified angiographically in 25 patients in whom the pulmonary arterial morphology could be assessed.

The early postoperative and late deaths are listed in table 3. Nine of the 27 patients (33%) have died, five during the initial hospitalization and four subsequently. Sixteen patients underwent a systemic arterial–pulmonary arterial shunt without an additional valvotomy. Of these patients, there were three early and four late deaths (44%). Ten infants underwent a shunt procedure and transventricular valvotomy. One child died early postoperatively (10%); there have been no late deaths thus far. One patient underwent pulmonary valvotomy alone. He initially did well and maintained satisfactory arterial blood gases. However, he developed progressively severe heart failure and died 2 weeks after surgery. At necropsy, a large cerebral arteriovenous malformation was found, which might have been an important factor in the development of congestive failure.

**Angiography**

A highly significant correlation was found between the aortic diameter at the diaphragm of control subjects and body surface area \((r = 0.95, p < 0.001)\). As would be predicted from the distribution of fetal blood flow, the aortic diameter in infants with PA-IVS were normal and fell at or within the 95% confidence limits of the control population in all cases (fig. 2A). The RVI in control subjects increased proportionately with the aortic diameter (fig. 2B, \(r = 0.96, p < 0.001\)). In contrast, there was no evident correlation between RVI and aortic diameter in patients with PA-IVS (\(r = 0.31, p > 0.1\)). Indeed, 22 of 26 infants in the PA-IVS group were either borderline or below the 95% confidence limits for the control population (fig. 3). The RVI/Ao for control subjects was 13.5 ± 1.4 (mean ± sd), whereas in patients with PA-IVS, the ratio was 7.3 ± 2.6 \((p < 0.001)\) and falls within the actual measured range for the control group (11.5–16.7) in only two PA-IVS patients.

Serial cardiac catheterizations were performed in eight patients, including five of the nine survivors in the pulmonary valvotomy group. The changes in RV systolic pressure are shown in figure 4A. RV pressure increased or remained essentially unchanged in the three patients who received a systemic–pulmonary arterial shunt alone. In contrast, the RV systolic pressure decreased dramatically in four of five patients who underwent pulmonary valvotomy. In the fifth patient, the RV systolic pressure increased despite pulmonary valvotomy. Nevertheless, the RVI/Ao in this child had increased substantially, from 7.9 to 15.7. A repeat pulmonary valvotomy and infundibulectomy under cardiopulmonary bypass has since been performed, and the patient is awaiting follow-up cardiac catheterization.

The RVI/Ao increased in all five patients who un-
derwent pulmonary valvotomy and serial cardiac catheterization (fig. 4B). The ratio rose from a mean of 8.0 to 12.5. While RVI/Ao was within the 95% confidence limits of the control patients in only one valvotomy patient preoperatively, it had increased to within these limits in all five valvotomy patients postoperatively. In contrast, the RVI/Ao did not change in the three patients who underwent a shunt procedure without valvotomy. Two of these three children have since undergone pulmonary valvotomy, one at 10 months and one at 7½ years of age. The older child had a cardiac catheterization at 9 years of age, which demonstrated that RV systolic pressure had fallen from 125 to 30 mm Hg (fig. 4A). Even at this relatively late age, the RV cavity appears to have retained the capacity for enlargement: the RVI/Ao increased from 6.9 to 11.5

**Discussion**

Pulmonary valvotomy has been recommended for all infants with PA-IVS in whom the RV infundibulum can be demonstrated on RV angiography to reduce RV systolic pressure and permit prograde ejection of blood.\(^{10, 12, 13}\) Our data support previous findings\(^9, 13\) that transventricular, closed pulmonary valvotomy adequately reestablishes continuity between the RV cavity and pulmonary arteries in almost all suitable patients and effectively relieves RV hypertension in the majority. However, relief of RV outflow obstruction may be inadequate in some patients, and RV hypertension may persist.\(^{13, 14}\) Massive hypertrophy of the RV wall is responsible for the encroachment and obliteration of the RV cavity.\(^{16}\) Therefore, the decrease in RV afterload is important in reducing the stimulus for RV hypertrophy or hyperplasia.\(^{15}\) Furthermore, if RV decompression by pulmonary valvotomy is not accom-
plished, RV sinusoidal–coronary shunting may continue. The portions of the right and left ventricular myocardium supplied via the right or left anterior descending coronary arteries, and occasionally the posterior descending artery, will continue to be perfused by desaturated blood during systole. This may perpetuate myocardial ischemia of both ventricles and may be a factor in the incidence of late deaths among our patients who had undergone a shunt procedure without pulmonary valvotomy.

The Right Ventricular Index

Despite a consensus in the literature about the benefits of pulmonary valvotomy in PA-IVS, controversy persists about the proper selection of patients and the need for an additional shunt. Dobell and Grignon reported survival in six of seven patients with adequate RV cavities (so-called type II) after pulmonary valvotomy alone, but other reports have stressed the need for combining a systemic–pulmonary arterial shunt with pulmonary valvotomy. Therefore, a preoperative method of assessing the size of the RV cavity and, thereby, its potential for maintaining an adequate cardiac output in the early postoperative period is essential for determining the suitability of patients for either shunt plus pulmonary valvotomy or valvotomy alone. Bull et al. recommended a morphologic classification that categorizes patients based on the presence or absence of a tricipital right ventricle: the inlet region, the trabecular (or apical) portion and the infundibular (or outlet) portion. However, a more quantitative estimate of cavity size would be helpful in assessing and comparing patients preoperatively and evaluating RV growth postoperatively. Previous studies have shown that ventricular volume measurements may be applied to infants with PA-IVS and demonstrated the variability in chamber size in these patients preoperatively and the potential for growth after valvotomy. Nevertheless, concerns persist regarding application of Simpson’s rule to these small and often distorted ventricles.

The RV length in the present report is a modification of the morphologic measurements of Zuberbuhler and Anderson. These authors related measurements of the TVA, RV inlet and RV outlet to their left ventricular counterparts in order to obtain a meaningful comparison of pathologic specimens. However, we were concerned that relating the RV measurements determined by angiography to the left ventricle would tend to underestimate RV size, since the left ventricle in PA-IVS is often larger than normal. The aortic diameter at the diaphragm appears to be a more reliable structure with which to normalize RV cavity size. The normal right ventricle appears to grow in proportion to the aortic diameter, whereas the RVI in most infants with PA-IVS is disproportionately small. Unlike volume measurements, which require outlining the entire endocardial surface of the right ventricle, the three linear measurements that constitute the RVI are less affected by the markedly irregular borders of the RV chamber. Nevertheless, the extensive trabeculations present in the apical region and, to a lesser extent, in the outflow tract often make some subjectivity unavoidable, and care must be taken to prevent overestimation.

Pre- and Postoperative Assessment of RV Size

RV angiography may be performed successfully in almost all infants with PA-IVS despite a diminutive TVA or RV cavity. The RVI could be measured in 26 of 27 infants in our series. The RVI/Ao demonstrates the broad spectrum of RV cavity size, from chambers that are less than one-third of normal to those that fall within the normal range. Although most PA-IVS patients have diminutive RV cavities, a few (two in our series) have normal-sized ventricles as assessed by the RVI/Ao ratio. This is similar to a total of 17 of 131 patients (13%) with normal RV cavity size accumulated from previously reported clinical series of PA-IVS. Therefore, despite the encouraging report of Dobell and Grignon, it appears that pulmonary valvotomy alone without an additional systemic–pulmonary arterial shunt should be reserved for a small proportion of infants with PA-IVS. Patients with an RVI/Ao > 11 appear to fall within this category. For the majority of patients with a ratio less than 1 standard deviation below the control group mean (RVI/Ao < 11), a systemic–pulmonary arterial shunt should be combined with a pulmonary valvotomy whenever a suitable RV outflow tract is identified angiographically. Since 1976, we have performed a pulmonary valvotomy in 11 of 16 patients. Review of our entire PA-IVS group indicates that a valvotomy would have been feasible in 20 of 27 patients (74%). It should be emphasized that RV size is not necessarily a predictor of the feasibility of pulmonary valvotomy; successful valvotomy was accomplished in neonates with a preoperative RVI/Ao as low as 5.2.

Follow-up cardiac catheterization in five of the eight patients who survived pulmonary valvotomy and a Blalock-Taussig shunt revealed a dramatic reduction in RV systolic pressure in four and an increase in one patient. The right ventricle, however, had increased in all five patients, from a mean RVI/Ao of 8.0 to 12.5. This improvement is even more striking when compared with the virtual absence of RV chamber development in the three patients who underwent shunting alone.

Management of PA-IVS in Neonates

We have adopted the following approach to the management of neonates with PA-IVS:

1. Intravenous infusion of PGE, is begun preoperatively and continued intraoperatively in all patients.
2. Pulmonary valvotomy is performed in patients in whom the RV outflow tract is identified angiographically. A small RV cavity alone does not preclude performing a valvotomy.
3. A Blalock-Taussig shunt is performed when the RV size appears inadequate to independently sustain sufficient pulmonary blood flow to maintain adequate systemic oxygenation in the early postoperative peri-
od. Patients with an RV/Ao less than 1 standard deviation below the mean of the control population (RV/Ao < 11) are in this category.

(4) In 8–15% of the patients, the RV cavity size is within normal limits (RV/Ao ≥ 11.0). Pulmonary valvotomy alone is advised for these infants. PGE₃ infusion may be continued for 3–5 days postoperatively to maintain adequate pulmonary blood flow via the PDA during the initial recovery period. When the child is clinically and hemodynamically stable, PGE₃ is discontinued and systemic oxygenation closely monitored. Patients who can maintain satisfactory RV output, as indicated by adequate systemic arterial oxygen tension, require no further early surgical intervention. However, if systemic arterial PO₂ falls below 30 torr, PGE₃ is restarted and a Blalock-Taussig shunt performed. Because the number of patients in this category is small, it may be several years before the validity of this approach can be determined.

(5) Postoperative cardiac catheterization is performed no later than 1 year after pulmonary valvotomy. However, in view of the inability to adequately relieve RV outflow obstruction and RV hypertension in some patients, catheterization is advisable 3–6 months postoperatively in selected cases. In evaluating the growth and development of the RV cavity, the RV/Ao ratio may be compared to both the patient’s preoperative value and the normal range.

The RV/Ao is an easy method for estimating the size of the RV cavity in patients with PA-IVS. It appears to be helpful in differentiating infants with small ventricles who require both a pulmonary valvotomy and systemic–pulmonary arterial shunt from the small group with normal-sized chambers in whom valvotomy alone may suffice.

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References
2. Lewis AB, Takahashi M, Lurie PR: Administration of prostaglan-
9. Graham TP, Bender HW, Atwood GF, Page DL, Sell CGR: Increase in right ventricular volume following valvulotomy for pulmonary atresia or stenosis with intact ventricular septum. Circulation 50 (suppl II): II-69, 1974
minations in 18 patients with pulmonary atresia and intact ventricu-
17. O'Connor WN, Cottrill CM, Johnson GL, Noonan JA, Todd EP: Pulmonary atresia with intact ventricular septum and ventriculo-
18. Gersony WM, Bernhard WF, Nadas AS, Gross RE: Diagnosis and surgical treatment of infants with critical pulmonary outflow ob-