Pulmonary Atresia and Intact Ventricular Septum: Surgical Management Based on a Revised Classification


SUMMARY Sixty patients with pulmonary atresia and intact ventricular septum (PA:IVS) presenting from 1970 to 1980 are reviewed. Three groups of patients are discussed: those with tripartite right ventricles, those with no trabecular portion to the cavity, and those with either trabecular or infundibular portions. The decrease in early mortality for neonates with PA:IVS since 1977 (one death in 15 patients) supports our current management policy of preoperative prostaglandin E1 infusion with transpulmonary valvotomy (for patients with an infundibular cavity) combined with a left modified Blalock-Taussig shunt using a Gore-Tex prosthesis. Tricuspid valve growth, estimated by serial angiograms in 12 patients, was greater if right ventricle-to-pulmonary artery continuity was established. Later definitive repair was attempted in nine patients, with two early deaths; five underwent right ventricular outflow tract reconstruction and four had modified Fontan procedures. Neonates with critical pulmonary stenosis are also discussed. Their neonatal mortality (nine deaths in 20 patients) was similar to that of comparable patients with PA:IVS, but their actuarial survival at 5 years (55%) was superior (36% at 5 years).

ACCOUNTS of the management of pulmonary atresia with intact ventricular septum (PA:IVS) often constitute a catalog of early and late failures of surgical treatment.

1-3 The dismal outlook in this condition has been attributed to the associated right ventricular hypoplasia. In a recent anatomic and angiographic study, we reported that cavitary hypoplasia in PA:IVS is related to massive hypertrophy of the right ventricular wall and proposed a classification of the condition based on the tripartite approach to right ventricular morphology. We distinguished three types of ventricles: those with generalized cavitary hypoplasia, those without a trabecular portion to the cavity and those in which both trabecular and infundibular portions are entirely overgrown by hypertrophied myocardium.

With this appreciation of the variety of ventricular morphology encountered, we have reviewed the results of our surgical management of PA:IVS over the past 10 years. This retrospective study forms the basis of a prospective management protocol both for the neonatal period, when mortality is high, and for definitive repair later. The question of right ventricular growth and the comparability of critical pulmonary stenosis to PA:IVS are also discussed.

Methods
The records of 81 patients admitted to the Hospital for Sick Children from 1970 to 1980 inclusive were examined. Sixty patients had PA:IVS and 21 had critical pulmonary stenosis. Patients with critical pulmonary stenosis were included only if they required operation in the first month of life, with suprasystemic pressures in the right ventricle, right-to-left shunting at atrial level and a pinhole patency of the pulmonary valve at angiography. Patients with PA:IVS were included regardless of age at initial operation. Even in the presence of extreme right ventricular hypoplasia, both lesions were considered amenable to surgical treatment throughout the period reviewed.

Details of the use of prostaglandin infusion, the na-
ture of the initial operation, the age at which it was performed, and survival were noted. Early mortality included all deaths during the first hospital admission. Details of subsequent operations and the duration of follow-up of survivors were also recorded.

Catheterization data and right ventricular angiocardioograms were reviewed. The right ventricular pressures of patients who had undergone serial studies were noted. On right ventricular angiocardiograms, the diameter of the tricuspid annulus in diastolic frames was estimated on both anteroposterior and lateral views, using the known diameter of the catheter as magnification factor. Serial angiocardiograms gave serial estimates of tricuspid annulus diameter in 12 patients. These estimates were related to surface area and to the data on the tricuspid valve dimensions of the normal child’s heart available from the studies of Rowlatt et al. Their study presents measurements of the tricuspid valve circumference taken at autopsy of 83 children of various ages so that the normal relationship of tricuspid valve diameter to body surface area can easily be derived from the data. To render the angiographic estimates of tricuspid valve diameter in PA:IVS comparable to the autopsy data of Rowlatt et al., we established that a correction factor of 1.43 must be introduced, as autopsy specimens shrink on preservation.

Contingency tables were analyzed using the chi-square test and actuarial survival curves were constructed according to the method described by Grunke and Starr.

To improve our understanding of the relative contributions of atrial and ventricular systolic work to pulmonary blood flow, pulmonary artery pressure tracings from patients after various types of surgical repair were studied.

In formulating guidelines for the definitive surgical treatment of PA:IVS with right ventricular and tricuspid valve hypoplasia, one must determine when a repair requiring the whole cardiac output to cross the tricuspid valve during ventricular diastole is likely to be successful. Under these circumstances, the tricuspid valve diameter and gradient will be important determinants of whether the ultimate right atrial pressure will be tolerable. Accepting the limitations of the formulæ of Gorlin and Gorlin, and assuming a heart rate of 100 beats/min, we have derived the theoretical relationship between tricuspid valve diameter, tricuspid valve gradient and cardiac output. Thus, preoperative (angiographic) or intraoperative assessment of the tricuspid valve dimensions allows prediction of the diastolic gradient across the valve over a physiologic range of cardiac output.

**Results**

Among the 60 patients with PA:IVS, six died before surgery could be arranged; of the remaining 54, 46 underwent urgent surgery in the first month of life. Seven patients had their first palliative operation after the neonatal period and one patient had a Fontan procedure at age 8 years without previous surgery.

**The Neonatal Period**

**PA:IVS**

Pulmonary blood flow was increased by one of three methods, alone or in combination:

1. Pulmonary valvotomy and/or infundibulotomy. This was carried out on cardiopulmonary bypass in four patients. Closed pulmonary valvotomy was performed using the transventricular approach in 12. (2) Systemic-to-pulmonary shunts. These were aortopulmonary in 16 patients and between the subclavian and pulmonary arteries in 15. (3) Manipulation of the ductus arteriosus. Temporary ductal patency was maintained by prostaglandin infusion in 15 patients and by formalin infiltration in four.

Seven neonates required a second operation within the first month of life because the first operation did not provide an adequate pulmonary blood flow (e.g., a shunt procedure prompted by clinical deterioration after formalin infiltration of the ductus arteriosus).

In summarizing the early mortality figures, we divided the patients into two groups: those operated before and those after July 1977 (table 1). This date corresponds to the introduction of changes in our management policy: prostaglandin infusion during the perioperative period, a transpulmonary valvotomy through a left thoracotomy incision for patients with an infundibular portion to the right ventricle, and the use of the modified Blalock-Taussig shunt as the shunt of choice. Early surgical mortality fell from 55% to 7% with these changes ($\chi^2 = 8.82, p < 0.01$).

**Table 1. Early Mortality in Neonates with Pulmonary Atresia with Intact Ventricular Septum**

<table>
<thead>
<tr>
<th>Operation</th>
<th>Before June 1977</th>
<th>Total</th>
<th>Died</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transventricular valvotomy alone</td>
<td>9</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>Right shunt alone*</td>
<td>11</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Transventricular valvotomy +</td>
<td>7</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>right shunt*</td>
<td>4</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Open heart repair</td>
<td>31</td>
<td>17</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Operation</th>
<th>After June 1977</th>
<th>Total</th>
<th>Died</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transpulmonary valvotomy ±</td>
<td>5</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>formalin infiltration of ductus</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Blalock or modified</td>
<td>3</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Blalock alone</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulm. valvotomy, left modified</td>
<td>7</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Blalock</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Mainly Waterston but includes five Blalock shunts.
Prostaglandin E₁ was administered by infusion at a dose of up to 0.1 μg/kg/min before and during operation in only one patient (3%) before July 1977 and in 14 patients (87%) since then. Only one patient has died preoperatively since July 1977.

Transpulmonary valvotomy was performed under direct vision with the main pulmonary artery clamped proximal to its bifurcation so as to maintain perfusion of both right and left pulmonary arteries by the ductus arteriosus. Though transpulmonary valvotomy was less commonly performed without an additional shunt in neonates with PA:IVS, only one of 12 patients died, compared with 11 of 16 patients who underwent transventricular valvotomy with or without a shunt procedure.

The left modified Blalock-Taussig shunt, interposing a Gore-Tex prosthesis between the subclavian and pulmonary arteries, has been our shunt of choice when combined with a transpulmonary valvotomy. Seven such combined procedures have been performed since July 1977, with no deaths. In contrast, four of seven patients who underwent simultaneous transventricular valvotomy and right-sided shunt procedures (Waterston or Blalock shunt) died. For patients who received only a shunt, the early mortality for classic or modified Blalock-Taussig shunts (five cases without death during the 10-year period) has been superior to that for the direct aortopulmonary anastomosis (nine patients with three early deaths) and for formalin infiltration of the ductus, which has provided very poor short- and medium-term duct patency rates in our hands.

Four neonates with PA:IVS were operated on cardiopulmonary bypass, all before July 1977. In all, the patent foramen ovale and patent ductus arteriosus were closed and an infundibular patch was used to relieve the right ventricular outflow tract obstruction. Only one patient survived long-term.

The early mortality for the different types of PA:IVS is shown in table 2. The later mortality is shown in table 3.

The highest early mortality (50%) was among the neonates with tripartite ventricular cavities. Almost half of this group had surgery to relieve the right ventricular outflow obstruction without a systemic-to-pulmonary shunt at initial operation. Only two patients thus treated survived. The proportion of patients in the other two groups who did not receive a primary shunt was much smaller (two of 22 patients). The disappointing results in the patients with tripartite ventricles suggests that reliable augmentation of pulmonary blood flow requires a systemic-to-pulmonary connection, however favorable the right ventricular anatomy appears.

Four patients who survived the neonatal period with no infundibular portion to the right ventricular cavity and no right ventricle to pulmonary artery continuity have required second systemic-to-pulmonary shunts. Three patients with patent right ventricular outflow tracts have undergone a second closed valvotomy. These second palliative operations have carried no hospital mortality.

**Critical Pulmonary Stenosis**

Twenty-one neonates with critical pulmonary stenosis have been discharged from the Hospital for Sick Children from 1970 to 1980. One patient died preoperatively. The early surgical mortality for the remainder is summarized in table 4.

Mortality has thus been comparable to the corresponding group with PA:IVS (χ² = 2.49, p > 0.2) in the first month of life. However, there have been no late deaths among the patients who had critical pulmonary stenosis during the neonatal period. The two survivors of closed valvotomy alone in the first month of life have both required relief of residual right ventricular outflow obstruction on cardiopulmonary bypass 7 months and 2 years after the initial surgery. The interatrial communications were closed in both cases and both patients have done well.

**Definitive Repair**

Definitive repair outside the neonatal period was attempted in nine patients with PA:IVS between 1970 and 1980 (table 5).

Two patients in whom inlet, trabecular and infundibular portions of the ventricular cavity were present and right ventricle-to-pulmonary artery continuity was established in the neonatal period had repair in the second year of life. In both patients, previous systemic-to-pulmonary shunts and the patent foramen ovale were closed. The residual pulmonary stenosis was treated by transpulmonary excision of the valve in one patient and by commissurotomy in the other. Neither patient had a ventricular incision. Both patients were discharged from hospital, but one died later in right-sided congestive heart failure.

Three patients in whom an inlet and infundibular portion to the cavity were distinguishable on the right ventricular angiogram but in whom no trabecular cavity was present were repaired after right ventricle-to-pulmonary artery continuity had been established in

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**Table 2. Early Mortality in Neonates with Pulmonary Atresia with Intact Ventricular Septum According to Ventricular Morphology**

<table>
<thead>
<tr>
<th>Description</th>
<th>Total</th>
<th>Died</th>
</tr>
</thead>
<tbody>
<tr>
<td>Three portions</td>
<td>24</td>
<td>12</td>
</tr>
<tr>
<td>No trabecular cavity</td>
<td>11</td>
<td>3</td>
</tr>
<tr>
<td>No infundibular cavity</td>
<td>11</td>
<td>3</td>
</tr>
<tr>
<td>No trabecular cavity</td>
<td>46</td>
<td>18</td>
</tr>
</tbody>
</table>

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**Table 3. Later Outcome of Survivors of Neonatal Surgery for Pulmonary Atresia with Intact Ventricular Septum**

<table>
<thead>
<tr>
<th>Description</th>
<th>Alive, no further operation</th>
<th>Alive, after further operation</th>
<th>Dead, no further operation</th>
<th>Dead, after further operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alive, no further operation</td>
<td>10</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alive, after further operation</td>
<td>9</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dead, no further operation</td>
<td>4</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dead, after further operation</td>
<td>5</td>
<td></td>
<td></td>
<td>28</td>
</tr>
</tbody>
</table>
the neonatal period. One patient had a valvotomy on cardiopulmonary bypass, but the patent foramen ovale was not closed. She remains asymptomatic 7 years later. In two others, the residual right ventricular outflow obstruction was relieved using a patch and the foramen ovale closed. Both patients suffered severe low cardiac output postoperatively and one died. Reconstruction of the right ventricular outflow using an infundibular patch demands closure of the interatrial communication to prevent massive right-to-left shunting at the atrial level in the early postoperative period.

Four patients in whom no trabecular portions to the right ventricular cavity had been established by the ages of 4–12 years underwent Fontan procedures. Valved atriopulmonary conduits with tricuspid valve closure were used in two patients. Conduits from the right atrium to the right ventricle-to-pulmonary artery junction using the conduit itself to patch the right outflow tract with the tricuspid valve left patent were used in the others. One of the latter patients died after surgery. Extensive sinusoids had been seen filling the aorta in the preoperative right ventricular angiogram, and at autopsy, severe myocardial fibrosis and arterial wall thickening was seen in the left ventricle. The other three patients made good postoperative recoveries, although one died 7 months later of miliary tuberculosis possibly related to the homograft conduit. The remaining two patients are active.

**Overall Survival**

Actuarial survival data for patients who presented with PA:IVS, including those who died preoperatively and those who did not require surgery until after the first month of life, are presented in figure 1. Actuarial survival for the patients with PA:IVS and critical pulmonary stenosis who required surgery in the first month of life shown in figure 2.

**Tricuspid Valve Growth**

Serial angiographic estimates of tricuspid valve diameter were available in 12 patients. In six, right ventricle-to-pulmonary artery continuity had been achieved by the time of recatheterization, although only two had right ventricular pressures below systemic levels. Their tricuspid valve growth with increasing body surface area is illustrated in figure 3, and compared to the 95% confidence limits around the normal tricuspid valve diameters derived from the data of Rowlatt et al.3 Three demonstrated tricuspid valve growth at or above the normal rate. In six patients, including two in whom pulmonary valvotomy was attempted, no right ventricle-to-pulmonary artery continuity was demonstrated on restudy. Their tricuspid valve growth is illustrated for comparison. None showed even a normal rate of growth.

**Pulmonary Artery Pressures**

Four postoperative pulmonary artery pressure tracings of patients with PA:IVS, along with the corresponding right-sided angiocardiograms, are illustrated in figure 4. The first patient had a tripartite ventricle with a tricuspid valve within the 95% confidence limits for normal as a neonate. Transventricular valvotomy without shunt was performed, and the right ventricular

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**TABLE 4. Early Mortality in Neonates with Critical Pulmonary Stenosis**

<table>
<thead>
<tr>
<th>Operation</th>
<th>Before June 1977</th>
<th>Total</th>
<th>Died</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transventricular valvotomy alone</td>
<td>6</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Transventricular valvotomy + right shunt*</td>
<td>5</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>11</strong></td>
<td><strong>7</strong></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Operation</th>
<th>After June 1977</th>
<th>Total</th>
<th>Died</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transpulmonary valvotomy alone</td>
<td>2</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Transpulmonary valvotomy + modified Blalock shunt</td>
<td>2</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Open heart repair†</td>
<td>5</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>9</strong></td>
<td><strong>2</strong></td>
<td></td>
</tr>
</tbody>
</table>

*Three Waterston, two Blalock. †All had an outflow patch.

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**TABLE 5. Definitive Repair of Pulmonary Atresia with Intact Ventricular Septum**

<table>
<thead>
<tr>
<th>Operation</th>
<th>Total no. of patients</th>
<th>Deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tripartite ventricle</td>
<td>Valvotomy on bypass; PFO closed</td>
<td>2</td>
</tr>
<tr>
<td>No trabecular cavity adequate tricuspid valve</td>
<td>Valvotomy ± patch ± PFO closed</td>
<td>3</td>
</tr>
<tr>
<td>No trabecular cavity small tricuspid valve</td>
<td>Modified Fontan procedure</td>
<td>2</td>
</tr>
<tr>
<td>No trabecular or infundibular portion</td>
<td>Fontan procedure</td>
<td>2</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>9</strong></td>
<td><strong>2</strong></td>
</tr>
</tbody>
</table>

Abbreviation: PFO = patent foramen ovale.
angiogram at the age of 8 years (fig. 4A) suggests that ventricle and tricuspid valve have grown. The configuration of the pulmonary artery pressure tracing is normal. The second patient had no trabecular portion to the ventricular cavity, but a patent outflow tract by age 2 years. Repair consisted in patch relief of the residual obstruction and closure of the foramen ovale. Besides the ventricular wave, a peak corresponding to atrial systole appears in the pulmonary artery pressure tracing (fig. 4B). The third patient had no trabecular portion to the right ventricular cavity, and the tricuspid valve was considered too small to carry the whole cardiac output in diastole. A modified Fontan procedure, anastomosing the distal end of the conduit to the right ventricle–pulmonary artery junction was performed. Atrial and ventricular systole both appear to contribute to pulmonary artery flow (fig. 4C). The fourth patient had only an inlet portion to the right ventricular cavity by age 5 years. An atriopulmonary conduit operation with tricuspid valve closure was performed. Atrial systole produces some pulsatility in the pulmonary artery pressure (fig. 4D).

Tricuspid Valve Gradient

The constraints on the circulation produced by tricuspid valve hypoplasia are illustrated in figure 5. According to Rowlett et al.,9 the mean tricuspid valve diameter for a patient of surface area 0.5 m² is about 20 mm, so a valve 14 mm in diameter would be hypoplastic. Although a diastolic gradient of less than 1 mm Hg would be present across the hypoplastic valve at rest (e.g., cardiac output 1.5 l/min and cardiac index 3 l/min/m²), the abnormality would become much more significant with increasing cardiac output. Moreover if atrial systole must contribute to pulmonary artery flow (fig. 4B), the right atrial pressure must be high, exceeding the pulmonary artery pressure during ventricular diastole. A restrictive tricuspid valve would then constitute an additional burden for right atrial pressure and function.

**Discussion**

Surgical management of PA-IIVS in the neonate and the definitive repair in childhood are closely related to right ventricular morphology and size.

**Management in the Neonate**

In neonates, the treatment must increase the pulmonary blood flow and enable the right ventricle to serve the pulmonary circulation whenever possible.

The dramatic fall in early mortality in our series since 1977 is attributable to a more reliable and durable augmentation of the pulmonary blood flow before, during and after operation. This has been achieved by the pre- and perioperative use of prostaglandin, by use of the modified Blalock-Taussig shunt as the shunt of choice and by the recognition that pulmonary valvotomy is rarely sufficient as an isolated procedure. Prostaglandin infusion allows the patients to come to surgery less acidotic and better oxygenated.11 The modified Blalock-Taussig is superior to the Blalock shunt. In neonates, it creates less distortion of the pulmonary artery, it has a high patency rate9 and it is practical on either side of the aortic arch and can thus be readily combined with a transpulmonary valvotomy performed through a left thoracotomy. Theoretically, its hemodynamic characteristics are such that the flow is regulated by the diameter of the subclavian artery.

Early pulmonary valvotomy for patients who have a right ventricular infundibulum appears essential if the right ventricle is to have the opportunity to grow (fig. 3). However, in contrast to the experience of Dobell and Grignon,12 closed valvotomy has proved disastrous as an isolated procedure in our hands. Only eight of 20 neonates with pulmonary atresia or stenosis with intact ventricular septum survived closed valvotomy without shunt. Ironically, patients managed in this way all had favorable anatomy (tripartite ventricles and adequate tricuspid valves). We now recognize that closed valvotomy does have limitations: Relief of the right ventricular outflow obstruction is rarely complete and there is a high incidence of late restenosis and even occlusion, and patients often remain severely cyanotic for several days after valvotomy. Because we do not perform isolated transpulmonary valvotomy, we cannot prove that the transpulmonary approach is safer than the transventricular approach, but believe that trauma and edema around the infundibulum produced by the transventricular approach may compromise

![Figure 1](http://circ.ahajournals.org/)

**Figure 1.** Actuarial survival for patients with pulmonary atresia and intact ventricular septum, 1970–1980.

![Figure 2](http://circ.ahajournals.org/)

**Figure 2.** Actuarial survival for patients with pulmonary atresia (pa) and intact ventricular septum and critical pulmonary stenosis (ps) who required surgery as neonates.
right ventricular ejection in the short term. While temporary prostaglandin infusion may protect against postoperative hypoxia, the incompleteness of the relief of the obstruction and uncertainty of the long-term results of valvotomy demand a more permanent increase in the pulmonary blood flow beyond the neonatal period. We believe that all patients with PA:IVS and hypoplastic tricuspid valve who have an anatomy suitable for a transpulmonary valvotomy should also have a systemic-to-pulmonary artery shunt. This philosophy has been advocated by Malm et al. for some years, but they suggest a right anterior thoracotomy to perform a Waterston shunt, with extension of the incision across the sternum to perform a transventricular valvotomy.

A complete repair consisting of relief of the right ventricular outflow obstruction and closure of the patent foramen ovale and the patent ductus arteriosus could be considered in neonates with a tripartite ventricle and adequate tricuspid valve size. Some patients with critical pulmonary stenosis and intact ventricular septum qualify for such treatment, and mortality has been 20% in our hands. Some groups recommend a complete excision of the pulmonary valve on cardiopulmonary bypass or inflow occlusion, leaving the patent foramen ovale open as an option for patients with PA:IVS and a hypoplastic right ventricle. However, to achieve a saturation of 80% with the blood ejected by the right ventricle as the only blood flow to the lungs, the right ventricular stroke volume must be at least 56% of the left ventricular stroke volume. This theoretical calculation assumes a cardiac index of 3 l/

min/m² and a systemic arteriovenous oxygen difference of 5 ml of oxygen per liter. In practice, only hearts with three portions to the right ventricular cavity would be large enough to contemplate pulmonary valve excision alone. Furthermore, an incision in the infundibulum could jeopardize both ventricular function and compliance. We believe this accounts for the picture of severe or fatal right ventricular failure (if the foramen ovale has been closed) or postoperative hypoxemia (if the foramen ovale has been left open) that we have observed after transannular patch enlargement performed in the neonatal period. Thus, we recommend pulmonary valve excision only when the right ventricular outflow tract obstruction is confined to the valve.

For neonates without an infundibular portion to the cavity, the entire systemic venous return, with the exception of blood reaching the aorta via sinusoids, reaches the left atrium through the patent foramen ovale, and pulmonary blood supply depends entirely on systemic-to-pulmonary connections. Since pulmonary valvotomy is impractical in these patients, we recommend a right-sided systemic-to-pulmonary artery shunt to avoid intraoperative distortion of the patent ductus arteriosus. Since the pulmonary atresia is not relieved and the patient’s life still depends on an adequate interatrial communication, some have recommended balloon atrial septostomy at initial cardiac catheterization. We have not routinely performed septostomy, although interatrial pressure gradients during atrial systole are often recorded. We consider balloon septostomy for patients in whom valvotomy is not fea-

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**Figure 3.** Tricuspid valve growth (angiographic estimates) with increasing body surface area (BSA). The stippled panel encloses the upper and lower 95% confidence limits for tricuspid valve diameter (tv dia) of normal patients derived from the autopsy data of Rowlatt et al. RV = right ventricle; PA = pulmonary artery.
possible when there is a significant pressure gradient between the two atria. This would be unlikely to add to the operative risk, although creation of a large atrial defect might impair the development of right atrial hypertrophy, which may be advantageous to a later atrio-pulmonary connection.

**Definitive Repair**

Although right ventricular growth has been documented in PA:IVS and related to early valvotomy or important tricuspid incompetence, there is little indication as to the proportion of ventricles that can be expected to attain near-normal dimensions. Similarly, while there has been some discussion of the complete repair of PA:IVS along the same lines as pulmonary stenosis in patients in whom ventricular and tricuspid valve growth has been achieved, in our experience most of the survivors of early operation reach childhood with severe hemodynamic and anatomic abnormalities. Residual right ventricular outflow tract obstruction with some degree of hypoplasia of the cavity and fibrosis of the ventricular wall are the most common features of these hearts. Right-to-left shunts at atrial level with or without left-to-right shunts outside the heart (ductus or surgically constructed shunts) are also often present. It is in the rational management of patients whose early palliative surgery, while ensuring survival into childhood, has provided less than optimal results in terms of right ventricular growth, that the tripartite approach to ventricular classification has been most helpful.

In a minority of survivors, a second operation may not be indicated because the repair achieved in the neonatal period has been as complete as it can be. This is the case for the patient in whom the right ventricular cavity and tricuspid valve have grown sufficiently after valvotomy and in whom there is no significant remaining obstruction to right ventricular outflow (fig. 4A).

For other patients, the aim of the definitive repair is a complete separation of the pulmonary and systemic circulations. Depending on the right ventricular size and morphology, four options are available:

1. When attempts to induce tricuspid valve growth have succeeded and the ventricle is tripartite, any residual right ventricular outflow tract obstruction can be relieved by valvotomy with or without infundibular resection and patch enlargement. The patent foramen ovale is closed. After such a repair, there should be a normal ventricular systolic pulse in the pulmonary artery.

2. The tricuspid valve has grown after valvotomy, but
there is no trabecular portion of the right ventricular cavity. Such ventricles are all effectively hypoplastic, their stroke volume being less than that of the left ventricular stroke volume. If the tricuspid valve is not too restrictive (fig. 4B), these patients are treated by relief of the right ventricular outflow obstruction and closure of the patent foramen ovale. After complete relief of the right ventricular outflow tract obstruction, these hearts are hemodynamically similar to those described by Van der Hauwaert and Michaelsson. Using the term isolated right ventricular hypoplasia, this group described a patient with a right ventricle without trabecular portion but with no outflow tract obstruction. The child was cyanotic and had a right-to-left shunt at atrial level but normal right ventricular pressures. After closure of the interatrial defect, a striking feature of the pressure trace in the pulmonary artery was the appearance of “a” waves corresponding to the diastolic opening of the pulmonary valve during atrial contraction. To reproduce these hemodynamics in the repair of PA:IVS, atrial systole must efficiently contribute to pulmonary blood flow. To achieve this, there must be a low pulmonary artery pressure and resistance, a complete relief of the right ventricular outflow tract obstruction and no gradient of any significance across the tricuspid valve.

If the tricuspid valve is so small that it would be too restrictive to allow the atrial “a” wave to be transmitted to the pulmonary artery, a valved conduit can be anastomosed from the right atrium to the right ventricle–pulmonary artery junction. Thus, the conduit relieves the right ventricular outflow tract gradient, provides an unobstructive atrio pulmonary connection and still allows the circulation to benefit from the right ventricular systolic contribution (fig. 4C).

Since clinical experience with small tricuspid valves is limited, we suggest using the information shown in figure 5 to decide which tricuspid valves are “too restrictive.” A tricuspid valve annulus across which there would be more than a 3-mm Hg diastolic gradient on exercise might be considered inadequate for the types of repair suggested in (1) or (2). Such rigorous criteria are necessary because the right atrial pressure obligatorily exceeds the pulmonary artery pressure in the presence of right ventricular hypoplasia.

For patients without an infundibular cavity or those in whom attempts to induce tricuspid valve growth have failed, a separation of the pulmonary and systemic circulations can be achieved using an atro pulmonary conduit as suggested by Fontan for tricuspid atresia. As for the Fontan operation, such atro pulmonary conduits should be considered only in the presence of adequate pulmonary arteries, a low pulmonary vascular resistance and normal left-sided hemodynamics.

Critical Pulmonary Stenosis

Critical pulmonary stenosis, which has hemodynamic findings identical to those of PA:IVS, has been considered by some to be identical in practice and by others to be importantly different. Our morphologic and angiographic studies suggest that deficiency of the trabecular portion of the right ventricle is much less common in critical pulmonary stenosis, these ventricles being usually tripartite. In our hands, early mortality for surgery in the neonatal period has been similar in critical pulmonary stenosis and PA:IVS, though this may be attributable to misplaced confidence in pulmonary valvotomy performed as an isolated procedure for many patients with tripartite ventricles.

The main difference between the two conditions in our series lies in the late mortality figures. There may have been no late deaths in patients presenting with pulmonary stenosis as neonates. Late attrition of patients with PA:IVS accounts for progressive divergence of the actuarial curves. While the key to survival for the neonate resides in the prompt augmentation of the pulmonary blood flow, the long-term results may be more related to the morphology and the size of the ventricle.

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