Critical Pulmonary Stenosis with a Diminutive Right Ventricle in Neonates

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SUMMARY

Thirteen of 19 neonates seen in the first months of life with critical pulmonary stenosis (PS) and an intact ventricular septum had a diminutive right ventricle (RV) evidenced by electrocardiographic and angiocardiographic criteria. All were critically ill and at catheterization had hypoxemia secondary to right-to-left shunting at the atrial level, and RV hypertension. Right ventricular angiogram was useful to evaluate RV size and differentiate the children with PS from those with pulmonary atresia.

Pulmonary valvulotomy was the procedure of choice and in ten children the operation resulted in marked improvement with disappearance of cyanosis and cardiomegaly within one year. All are well six months to eight and one half years later. Two children had an additional unrecognized infundibular stenosis and required a shunt after valvulotomy because of persistent cyanosis. One child had a shunt without valvulotomy and died three years later in congestive failure.

The three children who have been recatheterized at two to three years postoperatively have RV pressures less than 60 mm Hg. In two the right ventricles more than doubled in size but remain small.

These children are critically ill and a good salvage rate depends on rapid evaluation and valvulotomy.

Additional Indexing Words: Right ventricular volumes Pulmonary valvulotomy

Severe right ventricular (RV) outflow tract obstruction with an intact ventricular septum is a common cause of cyanosis in newborns with congenital heart disease (CHD). The entity is represented by a spectrum ranging from pulmonary atresia (PA) and extreme RV hypoplasia, to severe pulmonary stenosis (PS) with a normal size or dilated RV. Several recent publications attest to the high medical and surgical mortality of infants with pulmonary atresia and an intact ventricular septum with a small or normal size RV.

While results of valvulotomy in infants with PS and a normal-sized right ventricle have been good, some authors have commented on the poor prognosis in infants with PS and hypoplasia of the RV.

In a previous report from this institution, only one of 19 infants with severe pulmonary stenosis had hypoplasia of the RV. Since then we have seen 12 additional neonates with this problem. This report describes the clinical profile, hemodynamic studies, surgical results and follow-up care on these 13 neonates with PS and a diminutive RV.

Material and Methods

Infants under one month of age were considered to have critical pulmonary stenosis if cardiac catheterization disclosed RV systolic pressure equal to, or greater than systemic arterial pressure, and right-to-left shunting at the atrial level; and angiography demonstrated a patent though restrictive pulmonary valve and an intact ventricular septum. The RV was considered to be
diminutive if there was absence of the usual RV predominance on electrocardiogram (R/S ratio \(\leq 1\) in \(V_1\) and \(\geq 1\) in \(V_6\)), and hypoplasia of the RV cavity could be demonstrated on angiogram.

Thirteen of 19 neonates (11 males and two females) with critical PS seen at The Children's Hospital Medical Center between January, 1964, and December, 1972, had a diminutive RV by these criteria, and these patients form the basis of this report.

Histories, a physical examination, a 12 lead electrocardiogram and a chest X-ray were obtained on all infants. Cardiac catheterization with selective RV angiography was performed within a few days of admission in all, and RV volume determination calculated by the area-length method\(^15\) could be obtained in ten.

All patients underwent surgery soon after catheterization. Postoperative physical examination, electrocardiograms, and chest X-rays were available for all patients, and serial arterial oxygen saturation were determined by Waters Ear Oximeter (model XP 65B) in 11 of 13. The duration of follow-up was six months to eight and one-half years (median of 24 months). Postoperative catheterizations were available in five–two in the immediate postoperative period, two at 24 and 36 months of age, and one immediately postoperatively and again at 26 months of age.

Results

All 13 neonates were initially hospitalized in the first week of life. The prenatal histories were unremarkable, and all were full-term infants of normal birth weight. Apgar scores were normal, but cyanosis was noted within the first two days of life. A heart murmur was noted in the delivery room in three, and by two days of age in the rest. On initial examination by a cardiologist all 13 were cyanotic and had a harsh systolic murmur of moderate intensity (Grade 2–3/6) along the left sternal border. In ten the murmur was thought to be ejection in quality while in three others it was thought to be more regurgitant in nature and suggested tricuspid incompetence. None of the infants had a thrill and in only two was an ejection click detected. The second heart sound was single in eight and split with a decreased pulmonary component in five.

Electrocardiograms on admission disclosed a QRS axis of +30° to +110° (fig. 1) and in 11/13, right atrial hypertrophy (P wave taller than 2.5mm and

Figure 1

Left: Distribution of the mean QRS axis in the frontal plane in 13 infants. Right: Representative electrocardiogram from one of the infants (no 4) at three days of age. Note the QRS axis of +75°, right atrial hypertrophy, and left ventricular predominance for age.
peaked in II, III, V_{4R} or V_{1}). In ten the QRS axis was less than +90°. The R/S ratio in V_{1} varied from 0.3 to 1.0 and in V_{6} from 1.0 to 10.0. Chest X-rays disclosed cardiomegaly in 12, but in only two was the heart markedly enlarged.

Preoperative cardiac catheterization (table 1) revealed hypoxemia with arterial saturations ranging from 20–86% (median 64%) due to right-to-left shunting at the atrial level. RV peak systolic pressure ranged from 70–186 mm Hg. The RV end-diastolic pressure was less than 10 mm Hg in six infants, and over 15 mm Hg in the two (6, 7). In these two infants the lowest peak systolic pressure suggested that the ventricle was incapable of generating a higher pressure despite maximal valve stenosis.

No attempt was made to enter the pulmonary artery since it has been previously observed that even a small catheter can almost totally obstruct the pinpoint opening of a severely stenotic valve.\textsuperscript{14} Right ventricular angiograms (fig. 2) demonstrated a small RV cavity in all babies and this was confirmed by calculation of RV volume in ten. RV end-diastolic volume varied between 16 to 30 ml/m^2 with a mean of 22 ml/m^2 (normal = 39 ± 6\textsuperscript{15}; \textit{P} ≤ 0.01 paired \textit{t} test). RV ejection fraction ranged from 50 to 88% and was within two \textit{SD} of normal in seven of the ten patients in whom it was measured (\textit{N} = 66 ± 7%).\textsuperscript{15}

All patients underwent surgery soon after catheterization (table 2). Ten children had a pulmonary valvulotomy, seven by transventricular and three by transpulmonary approach. All operations except one were under inflow occlusion and hyperbaric oxygenation of 30 lbs/square in. No major postoperative complications occurred, and there has been no early or late mortality.

Early in the series (1965), one child (9), underwent ascending aorta to right pulmonary artery (RPA) anastomosis and atrial septectomy. It was felt at that time that the RV was not large enough to sustain a normal pulmonary flow. Experience with subsequent infants suggests that this was probably not the case. This child died (our only fatality in this series) three years postoperatively in severe congestive failure with aspiration pneumonia.

Two children (10, 13) underwent valvulotomy (one transventricular and one transpulmonary arterial) and remained very cyanotic in the immediate postoperative period. Repeat catheterization revealed an open pulmonary valve but residual obstruction angiographically located at the infundibular level (fig. 3). Since at the time we were not doing intracardiac operations on small infants, it was elected to do an ascending aorta to RPA anastomosis. Both of these children are doing well several months after surgery.

The ten children who had valvulotomy alone have been followed from 5 to 104 months (median 30). All are now active, asymptomatic, and acyanotic (fig. 4). Systolic murmurs are present in all and four have developed a diastolic murmur suggestive of pulmonary regurgitation. Two infants

### Table 1

<table>
<thead>
<tr>
<th>Patient #</th>
<th>Age (Days)</th>
<th>Inhaled \textsubscript{O}_2 %</th>
<th>\text{O}_2 Saturation %</th>
<th>RV, RA, PV, LA</th>
<th>\text{FA or UA} \text{RV}</th>
<th>\text{FA or UA} \text{RV}</th>
<th>RVEDV \text{ml/m}^2</th>
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<td>125/10</td>
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Abbreviations: SVC = superior veno cava; RA = right atrium; PV = pulmonary vein; LA = left atrium; FA = femoral artery; UA = umbilical artery; RV = right ventricle; RVEDV = right ventricular end-diastolic volume; RVESV = right ventricular end systolic volume; RVEF = right ventricular ejection fraction; normal values\textsuperscript{2}; RVEDV = 30 ± 8; RVEF = 0.66 ± 0.07.
Figure 2

Right ventricular (RV) angiogram in the anteroposterior and lateral projections on patient 3. Above: At two weeks of age, 11 days following pulmonary valvulotomy. Note the diminutive size of the RV. Below: At 26 months of age. The RV cavity has increased in size.

Five children have been recatheterized—two in the immediate postoperative period, two at 24 and 36 months of age, and one in the immediate postoperative period and again at 26 months of age. All have RV peak systolic pressure less than 60 mm Hg.

have systolic thrills at the left upper sternal border and two others have developed electrocardiographic evidence of RV hypertrophy. Heart size is normal in all but the youngest patient, now six months of age, who continues to have cardiomegaly on X-ray.
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Table 2
Operative Procedures Performed for Critical Pulmonary Stenosis

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<tr>
<th>Surgical Procedure</th>
<th>Number</th>
<th>Deaths</th>
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<td>10</td>
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<tr>
<td>Transventricular</td>
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<td>0</td>
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<tr>
<td>Transpulmonary arterial 3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Shunt and atrial septal defect creation</td>
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<td>1 (late)</td>
</tr>
<tr>
<td>Valvulotomy followed by shunt</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>13</td>
<td>1</td>
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</tbody>
</table>

Hg. In two of the children with late catheterizations (3, 5) the RV end-diastolic volume has more than doubled to 49 and 47 ml/m² (fig. 2).

Discussion

Although it is usually possible to differentiate critical RV outflow obstruction with an intact ventricular septum from other cyanotic heart disease in the newborn infant before cardiac catheterization, it is often impossible to differentiate PS with a diminutive RV from pulmonary atresia. Both usually present with cyanosis and a systolic murmur. The electrocardiogram in both groups shows a QRS axis of +30° to +90°, RAH, and diminished RV forces; and on chest X-ray there is cardiomegaly with diminished pulmonary flow. The presence of a split second sound or a variable ejection click in PS may be helpful, but in many instances only the angiographic demonstration with contrast material passing across the pulmonary valve, can differentiate between the two conditions. It is not altogether surprising, therefore, that in only two of our cases of pulmonary stenosis was the correct diagnosis made prior to catheterization.

The differentiation between neonates with PS and a diminutive RV from those with PA is quite important since their prognoses are so different. The long-term survival of children with PA is probably less than 20%. While some have reported similar discouraging results for PS with hypoplasia of the RV, and others have suggested a two-stage procedure for repair, our results, and those of Mustard, Jain, and Trusler and Miller et al., suggest a more favorable prognosis. In our group, relief of the valvar obstruction was followed by a dramatic clinical improvement. In the infants with

Figure 3
Right ventricular angiogram in the lateral projection on patient 10 one day after pulmonary valvulotomy. Note the severe infundibular obstruction during systole. The pulmonary valvulotomy appears adequate. There is a chest tube draining the right pleura. (RV = right ventricle; PA = main pulmonary artery; INF = infundibular obstruction).
smaller initial right-to-left shunts, the cyanosis disappeared prior to discharge from the hospital. In infants with more severe hypoplasia and a large right-to-left shunt, the cyanosis disappeared within a year of surgery (fig. 4). Persistent right-to-left shunting at the atrial level following relief of PS has been described in older children and adults, but in none of our cases was the RV cavity so small or noncompliant that it could not accommodate a relatively normal RV stroke volume within a few months following valvulotomy.

The operative procedure of choice in these infants is pulmonary valvulotomy with inflow occlusion. We prefer, as does Mustard, an approach through the main pulmonary artery (Swan) exposing the valve from above when the main pulmonary artery is of adequate size. We have also had good results, as have others, using a transventricular approach. All but one of our operations were done under hyperbaric oxygenation (30 lbs/square in). Its beneficial effect on improving myocardial oxygenation has been reported, but whether this alone accounts for our good results remains speculative.

While up to 40% of newborns with pulmonary atresia with intact ventricular septum are thought to have significant infundibular obstruction, (unpublished data: Ando, M. and Van Praagh, S.) this has, to the best of our knowledge, not been previously reported to be a significant problem in neonates with PS. Although several authors have noted that there may be some infundibular narrowing, they all followed the advice of Engle et al. in abstaining from infundibular resection on the premise that the hypertrophy will regress with time. In two of our cases, valvulotomy alone did not suffice, and the infants remained severely cyanotic and acidotic. They improved dramatically after a systemic to pulmonary arterial shunt and are now alive and well at six and seven months of age. Closure of the shunt and resection of the infundibulum with cardiopulmonary bypass is planned for the near future.

Whether the right ventricle will increase in size after relief of the obstruction remains an important and unanswered question. In two of our infants (3 and 5) the RV volume data calculated from the postoperative angiogram showed an increase in right ventricular end-diastolic volume from 16 and 19 ml/m² to 49 and 47 ml/m², respectively. While this is at first glance very encouraging, Graham et al. has shown that normally the RV end-diastolic volume (EDV) increases from 39 ml/m² in infancy to 70 ml/m² in older children. This normal increase in EDV has previously been reported for left heart volumes and has been associated with the increased stroke volume that accompanies the normal slowing of resting heart rate in the first few years of life. Thus, while in one of our patients RV end-diastolic volume has more than doubled and in one it tripled, it remains abnormally low in both.

Although it has been assumed that the diminutive RV is hypoplastic, the small RV cavity may be due to marked hypertrophy of the moderator band and the trabeculae impinging on a relatively normal size RV. The angiograms show small RV cavities during systole. In diastole, however, some contrast material can often be seen between the hypertrophied trabeculae. The estimate of RV volume either grossly or by measurement probably underestimates the actual or potential size of the RV. After adequate surgical relief of the outflow obstruction, the hypertrophied muscle may regress and the observed RV cavity size increase (fig. 2).

The diminished RV potentials recorded at the surface are probably not due to diminished muscle mass but rather to a small RV cavity. An increase in intracavitary volume tends to augment the radially directed dipoles and thus increases the recorded precordial voltages (Brody effect). Conversely, diminished cavity may be expected to decrease radial forces and increase tangential forces which are cancelled out. In spite of increased RV wall thickness the net effect is a decrease in precordial voltages. This may explain the LV predominance on the initial ECG in these infants. Following surgical relief of the obstruction the ECG may not be a
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reliable tool for assessing the RV pressure. If the intracavity volume remains small, even when significant residual gradient exists, right ventricular hypertrophy may not be apparent. Evaluation of the surgical results must be based on other clinical criteria or at cardiac catheterization.

In conclusion we would like to emphasize as others have before, that these neonates are critically ill and that survival depends on rapid clinical evaluation, catheterization with RV angiography, and prompt valvulotomy.

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