Severe Congenital Pulmonic Stenosis without Marked Right Ventricular Hypertension

By JOHN M. VEROSKY, M.D., AND E. S. CROSSETT, M.D.

IN THE literature, indications given for surgical correction of congenital pulmonic stenosis have emphasized chiefly the degree of right ventricular hypertension or the gradient across the obstructed orifice.1-4 We wish to present 2 cases of severe stenosis, as judged by the size of orifice visualized at operation, in which right ventricular systolic pressure was less than 90 mm. of mercury. Both these children had decreased cardiac output and severely retarded growth. In contrast, 2 patients with equally severe stenosis are presented who had marked hypertension.

Method

All patients had investigations consisting of history, physical examination, radiographic studies of the chest, electrocardiography, and right heart catheterization. In each case the clinical findings were typical of pulmonic stenosis and will not be reiterated here. In all, pulmonic stenosis was present as the sole cardiac malformation. In one, (L.S.), a small right-to-left shunt through a patent foramen ovale was present. Catherization was performed under local anesthesia and sedation (25 mg. meperidine, 6.25 mg. chlorpromazine, and 6.25 mg. promethazine). Electrode catheters were used as an aid in locating the catheter tip. Pressures were measured by a calibrated Statham P23AA electromanometer. Oxygen content was determined with a standardized Waters cuvette. Data were recorded with an oscillographic photographic recorder (Electronics for Medicine Model PR 6).

In L.S. and K.E. cardiac output was determined by use of the Fick principle. The Gorlin formula5 was then used to calculate valve size. Systolic ejection period (S.e.p.) was timed from the pulmonary artery pressure tracing, measuring from the beginning of the systolic rise to the dicrotic notched. Right ventricular systolic mean pressure (RV$_{sm}$) was determined planigraphically from the right ventricular pressure tracing during systolic ejection.

All patients were operated upon under direct vision, by means of extracorporeal circulation; for P.M., a bubbler oxygenator was used; for the others, a multiple rotating disk oxygenator was used. Size of the pulmonary orifice was determined visually by the surgeon at the time of operation.

The data of Paul and Rudolph6 were used as a basis for normal pulmonary valve sizes. Because of the limited number of cases of any given age listed in their study, these figures can be taken as approximations only.

Weight and height were evaluated by the standards of Howard V. Meredith, Iowa Child Welfare Research Station, and the Harvard School of Public Health Studies of Child Health and Development;7 some extrapolation from these tables was necessary to evaluate L.S. Surface area was determined from the nomogram derived by R. R. Hannon from the formula of Dubois and Dubois.7

Postoperatively, the right ventricular index was calculated from catheterization data previously obtained, but the size of the pulmonary orifice observed was used in the equation,

$$\text{Right ventricular index (L/min.} / \text{M}^2) = \frac{C \sqrt{2g \pi D^4} \sqrt{RV_{sm} - PA_{sm} \sum S. e. p.}}{4} \frac{S. A. (1000)}{S. A. (1000)}$$

in which $C$ is a constant, taken as unity, $g$ is the acceleration due to gravity, $\sum S. e. p.$ is the sum of systolic ejection periods (sec./min.) and $S. A.$ is the surface area in M.$^2$

Results

Data are listed in table 1. The lack of correlation between symptoms, heart size, and severity of stenosis is well known and was not surprising. These cases, however, show a more disturbing lack of correlation between the degree of right ventricular hypertrophy electrocardiographically, right ventricular pressure, and severity of stenosis, if these factors are considered without regard to cardiac output and growth pattern.

Discussion

For an opening of given size, the Poiseuille equation (resistance equals pressure gradient divided by flow) demands that the pressure

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Table 1

<table>
<thead>
<tr>
<th>Patient</th>
<th>K. E.</th>
<th>K. C.</th>
<th>P. M.</th>
<th>L. S.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, years</td>
<td>5 5/12</td>
<td>4 9/12</td>
<td>7 5/12</td>
<td>19 3/12</td>
</tr>
<tr>
<td>Weight, pounds</td>
<td>33</td>
<td>33</td>
<td>49</td>
<td>164</td>
</tr>
<tr>
<td>-percentile</td>
<td>3rd</td>
<td>3rd</td>
<td>10th</td>
<td>75th</td>
</tr>
<tr>
<td>Height, inches</td>
<td>43</td>
<td>39</td>
<td>48</td>
<td>72</td>
</tr>
<tr>
<td>25th percentile</td>
<td>77</td>
<td>87</td>
<td>92</td>
<td>111</td>
</tr>
<tr>
<td>Surface area, M.² per cent of normal age</td>
<td>0.67</td>
<td>0.63</td>
<td>0.87</td>
<td>1.95</td>
</tr>
<tr>
<td>Type of stenosis</td>
<td>infundib.</td>
<td>valvular</td>
<td>valvular</td>
<td>valvular</td>
</tr>
<tr>
<td>Symptoms</td>
<td>mild</td>
<td>mild</td>
<td>moderate</td>
<td>severe</td>
</tr>
<tr>
<td>X-ray cardiothoracic ratio</td>
<td>0.53</td>
<td>0.53</td>
<td>0.59</td>
<td>0.58</td>
</tr>
<tr>
<td>R wave (mm.) in V₁ or V₃₅</td>
<td>16</td>
<td>25</td>
<td>25</td>
<td>22</td>
</tr>
<tr>
<td>Catheter size</td>
<td>6E</td>
<td>6E</td>
<td>7E</td>
<td>8E</td>
</tr>
<tr>
<td>Right ventricular index, L./min./M.²</td>
<td>1.55</td>
<td>—</td>
<td>—</td>
<td>0.72</td>
</tr>
<tr>
<td>Pulmonary artery pressure, mm. Hg*</td>
<td>74/10 (43)</td>
<td>84/10 (59)</td>
<td>196/4 (70)</td>
<td>145/10 (47)</td>
</tr>
<tr>
<td>RVₘₚ - PAₘₚ mm. Hg</td>
<td>12/7 (9)</td>
<td>16/11 (14)</td>
<td>6/3 (4)</td>
<td>10/3 (5)</td>
</tr>
<tr>
<td>Σ S.e.p., sec./min.</td>
<td>36.6</td>
<td>44.1</td>
<td>32.4</td>
<td>23.5</td>
</tr>
<tr>
<td>S.e.p., sec./beat</td>
<td>0.30</td>
<td>0.45</td>
<td>0.36</td>
<td>0.26</td>
</tr>
<tr>
<td>Pulmonary orifice area, cm.² calculated</td>
<td>0.12</td>
<td>—</td>
<td>—</td>
<td>0.2</td>
</tr>
<tr>
<td>Observed during surgery</td>
<td>0.16</td>
<td>0.20</td>
<td>0.13</td>
<td>0.28</td>
</tr>
<tr>
<td>Normal for age</td>
<td>1.9</td>
<td>1.8</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Pulmonary orifice diameter, mm. calculated</td>
<td>4.0</td>
<td>—</td>
<td>—</td>
<td>5.0</td>
</tr>
<tr>
<td>Observed during surgery</td>
<td>4.5</td>
<td>5</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>Normal for age</td>
<td>15.6</td>
<td>15.2</td>
<td>16</td>
<td>22.6</td>
</tr>
<tr>
<td>Right ventricular index, calculated from valve size observed during surgery, L./min./M.²</td>
<td>2.06</td>
<td>1.61</td>
<td>1.93</td>
<td>1.04</td>
</tr>
<tr>
<td>Right ventricular index, calculated from valve size observed during surgery, assuming normal weight and height, L./min./M.²</td>
<td>1.59</td>
<td>1.41</td>
<td>1.77</td>
<td>1.15</td>
</tr>
</tbody>
</table>

*Mean pressure in parentheses.

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will fall as flow decreases. In the Gorlin formula,

$$\text{Area} = \frac{\text{Flow}}{\text{constant} \times \sqrt{\text{pressure gradient}}}$$

the same relation is seen. It is thus not proper to term a stenosis as mild if the pressure is only mildly elevated because of reduction in flow. It would seem more reasonable to call a very small pulmonic opening a severe stenosis, since mathematically this will cause either severe right ventricular hypertension and strain or reduced cardiac output; in some children, the latter may prevent normal growth.

The natural history of pulmonic stenosis is still incompletely known. It has not been determined if the size of the orifice remains constant or changes with age. If it does not enlarge with age, it is clear that right ventricular pressure must increase if the cardiac output is to increase as growth requires. Until more information is available of the course of
these events, it would seem that the single most important measure of severity is the size of the opening itself.

Some cautious speculations can be made on the course of the patients presented. In P. M. the stenosis was so severe that increase in cardiac output to accompany completely normal growth was not possible. At the same time, a fair output for his moderately stunted size was maintained, at the cost of severe right ventricular hypertension, as reflected in the electrocardiogram. In L. S., the same factors were operative, except that cardiac output increased to allow good growth; then cardiac output decreased. It is very likely that progressive deterioration occurred over the recent years, since cyanosis at rest had been noted by the parents for only about 1 year. In K. E. the stenosis was so severe that neither normal growth nor normal cardiac output for her small body size could be achieved. Severe right ventricular hypertension was absent, and the R in V₁ was only moderately high. Yet, in view of her lowered cardiac output and calculated small orifice size, we rated her as severe. Apparently the R wave in V₁ can be taken as a measure of severity of the stenosis only if marked right ventricular hypertrophy had been present. In K. C. the R was very high and the stenosis severe, yet the hypertension was only moderate. It may be that once his right ventricular pressure was much higher than that observed at catheterization, and subsequently fell with reduction in cardiac output; it is probable that his stenosis has always been sufficiently severe as to prevent the increase in cardiac output necessary for normal growth and development.

Thus, various courses are possible in any one patient. We have no proof that the retarded growth observed was a result of reduced cardiac output, but this conclusion seems quite likely. All 4 patients grew in socioeconomic situations favorable to normal growth. We have no idea why some children with severe pulmonic stenosis respond mainly in the direction of retarded growth while others respond mainly with increasing right ventricular hypertension.

To emphasize the importance of evaluating the growth of the child, we have added to table 1 the results of recomputing the right ventricular index that our patients would have had if their cardiac outputs had been the same at body sizes normal for their ages. All patients had orifices approximately the same in size, and these normalized right ventricular indices are all much the same.

Before deciding to delay catheterization, one must evaluate the factors concerned: (1) symptoms, the least reliable measure, (2) heart size, similarly of limited value, since the reliability of the radiograph to determine concentric hypertrophy is low, (3) electrocardiogram, usually but not always completely reliable, since it is dependent upon right ventricular hypertrophy, and (4) growth and development. If serious abnormality is present in any of these, catheterization should be done.

Before deciding that stenosis is mild enough to delay operation, one should obtain an evaluation of the valve size, using as exact methods as possible, at the same time recognizing the influence of approximation factors and the sources of error involved in the clinical application of Gorlin’s formula. It has been widely appreciated that severe stenosis with deterioration and congestive failure, as seen in an occasional young infant, can result in only moderate right ventricular pressure readings; this situation is not likely to mislead the clinician, because the severity of the acute symptoms calls for immediate operation. Several authors¹, ³, ⁸ have noted that pressure should not be the sole criterion, but there are few explicit additional criteria. At this time we could not recommend that all cases of pulmonic stenosis be repaired, and cannot accept Zimmerman’s criterion for operation (a gradient of 20 mm. or more across the valve).³ At the same time, we believe that it is possible to delay operation unwisely in the occasional child in whom only moderate electrocardiographic and manometric abnormalities are present, if the relations outlined above are ignored. From the reports of others, it is apparent that severe physical underdevelopment is not common in congenital pulmonic steno-
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sis; Zimmerman and co-workers\(^3\) mentioned that underdevelopment may occur; Kjellberg et al.\(^9\) reported 32 cases of pulmonic stenosis, none of whom had physical dimensions below 2 standard deviations of the mean.

We are unable to correlate prognosis with definite valve sizes, but rough calculations show that it is impossible to maintain a cardiac output of over 3 liters per minute per square meter and a normal rate of growth with right ventricular hypertension below 90 mm. of mercury if the valve is less than one half the normal diameter or one fourth the normal area for age. However, our cases had valves much smaller than these conservative tolerances. It is unlikely, in our opinion, if actual calculations are carried out, that many equivocal cases will be found.

Acknowledgement

We are indebted to our anesthetist Dr. Jack Walker, and Dr. Jesse A. Hancock, Ph.D. in chemistry, for their assistance throughout various phases of this study.

Summary

Obligatory relations exist between orifice size, flow, and pressure gradient across a stenotic pulmonary valve. If the cardiac output is reduced, the patient may have severe stenosis and present only moderate right ventricular hypertension. This situation may be an acute one, which will produce progressive symptomatology, or a chronic one, which, in the growing child, will produce poor physical growth. Catheterization should be done if any of the following is present: severe symptoms, severe right ventricular hypertrophy electrocardiographically, or evidence of reduced cardiac output (such as otherwise unexplainable poor physical growth). Operation is urgently indicated if catheterization reveals either right ventricular systolic hypertension greater than 90 to 100 mm. of mercury or if there is evidence of a very small pulmonary orifice.

Summario in Interlingua

Relaciones de charater obligatori existen in casos de stenosis del valvula pulmonar inter le dimensiones del orificio, le fluxo, e le gradiente de tension ab un latere del sito stenotisate. Si le rendimento cardiac es reducet, il es possibile que le patiente ha grados sever de stenosis sed presenta solmente un moderate hypertension dextero-ventricular. Isti situation pote esser acute, produceun un symptomatologia progressive, sed ilo etiam pote esser chronic, produceun—in juveniles in crescentia—progresso crescential non satisfactori. Catheterismo es indicatur in le presentia de ulle del sequente constatationes: Symptomas sever, signos electrocardiographie de sever hypertrophie dextero-ventricular, o evidentia de un reduce rendimen cardic (como per exemplo alteremente inexplicable inadequatia del crescentia physic). Intervention chirurgic es indicate urgentemente si le cathe-
terismo revela hypertension systolie dextero-ventricular in excesso de 90 a 100 mm de Hg o si il ha provas de un micrisime orificio pulmonar.

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