Diagnosis and Surgical Treatment of Infants with Critical Pulmonary Outflow Obstruction

Study of Thirty-four Infants with Pulmonary Stenosis or Atresia, and Intact Ventricular Septum

By Welton M. Gersony, M.D., William F. Bernhard, M.D., Alexander S. Nadas, M.D., and Robert E. Gross, M.D.

SUMMARY
The records of 15 infants with pulmonary atresia and 19 infants with critical pulmonic stenosis and intact ventricular septum, all under 1 year of age, have been reviewed. Symptoms started earlier and were more severe in the group with pulmonary atresia. All patients showed arterial unsaturation, and many had congestive heart failure. The electrocardiogram was the most helpful tool in differentiating pulmonary atresia from pulmonic stenosis with intact ventricular septum. The presence of P pulmonale, significant right axis deviation, and right ventricular hypertrophy favors pulmonic stenosis. At cardiac catheterization, patients belonging to both groups showed right ventricular hypertension of significant degree, but this was more marked in infants with pulmonic stenosis. Results of surgery for pulmonary atresia with intact ventricular septum, by a variety of methods, have been uniformly poor up to the present. Direct attack on the pulmonary outflow tract, when possible, may offer more promise for the future. Pulmonary valvotomy is the operation of choice for pulmonic stenosis and the results are good.

Additional Indexing Words:
Systemic-pulmonary artery shunt
Transventricular valvotomy
Hyperbaric surgery
Hypoxic syncope

Congenital heart disease
Cyanosis

RECENTLY, the results of open-heart surgery at this institution in children over 1 year of age with pulmonic stenosis and an intact ventricular septum have been reviewed. Of interest now is a survey of our experience with infants under 1 year of age who had severe pulmonic stenosis or atresia, without a ventricular septal defect. This group of infants is considered separately from older patients with similar problems because of the more severe, often emergency, nature of the disease in the first year of life.

The present study will discuss the clinical profile and differential diagnosis of pulmonic stenosis and pulmonary atresia with intact ventricular septum and then the surgical techniques and results.

Pulmonary atresia with intact ventricular septum has been defined by Edwards and associates, as a condition which consists of complete obstruction of the pulmonary valve, two distinct ventricles, and a patent tricuspid...
orifice guarded by a valve. This entity has been classified by Greenwold\textsuperscript{3} according to the size of the right ventricle. When a diminutive right ventricular chamber and thick ventricular wall are present, the condition is referred to as type I, whereas when the ventricular cavity is normal or large the condition is designated as type II. Since, without operative intervention, the anomaly leads inevitably to death at an early age,\textsuperscript{4} attempts at surgery have been undertaken in several centers but with universally poor results.\textsuperscript{5-8}

In pulmonary stenosis with intact ventricular septum, although the pulmonary orifice is severely obstructed, there is always a distinct opening in the valve. Two ventricles are present, and there is a patent tricuspid valve. It should be emphasized that whereas older children and adults who undergo pulmonary valvotomy may have moderate, severe, or critical obstruction, no infant in our institution is subjected to surgery unless the stenosis is considered to be critical. Although these infants without surgery are likely to succumb early, survival time is considerably longer than that of infants with pulmonary atresia.\textsuperscript{9} Surgical intervention for pulmonic stenosis, although hazardous, has met with considerably greater success than that for pulmonary atresia.\textsuperscript{9-11}

**Methods**

Thirty-four patients (16 males and 18 females) representing all of the infants who underwent cardiac surgery at this institution for pulmonary atresia (15) or stenosis (19) with intact ventricular septum from January 1953 to July 1, 1965, were included in the study. In 13 of the 15 infants with pulmonary atresia it was classified as type I and in two as type II. All but one patient with pulmonic stenosis had a normal or large right ventricular cavity diagnosed by angiography, surgery, or autopsy. The age distribution at the time of operation is presented in figure 1. Histories, physical examinations, chest x-rays, and 12-lead electrocardiograms were available for all patients. The criteria utilized for the electrocardiographic diagnosis of ventricular and atrial hypertrophy were those defined by Nadas.\textsuperscript{12} Preoperative cardiac catheterization was carried out on 18 of 19 patients with pulmonic stenosis and 11 of 15 with pulmonary atresia. The five other infants were judged to be too ill to undergo catheterization, and surgery was performed on the basis of clinical evaluation. The diagnosis was confirmed in all instances either by selective right ventricular angiography or at postmortem examination. Autopsy was available on 17 of the 18 patients who succumbed.

The patients with pulmonic stenosis and pulmonary atresia operated on in the hyperbaric chamber were subjected to oxygen ventilation at an environmental pressure of 30 to 40 pounds per square inch. Following stabilization at the working pressure, aortic blood samples were analyzed, and 22 to 60 mm Hg increments in oxygen tension were noted\textsuperscript{13} before the operative procedures were undertaken.

Postoperative physical examinations, electrocardiograms, and x-rays were available on 12 of 16 surviving patients (11 with pulmonic stenosis and one with pulmonary atresia). One patient was lost to follow-up and for three patients (two with pulmonic stenosis and one with pulmonary atresia), insufficient time has elapsed since surgery to allow definitive evaluation. The duration of follow-up ranged from 2 months to 8 years. Cardiac catheterization was performed postoperatively in three of the 12 survivors for whom results can be determined. In addition, two of the infants with pulmonic stenosis who later expired were catheterized after the first of two surgical procedures.

**Results**

**Preoperative and Postoperative Evaluation**

**Signs and Symptoms**

Pulmonary Atresia: The presence of anoxic spells was a common clinical feature in the
Initial signs and symptoms in 34 patients at the time of surgery. Spells = anoxic episodes; CHF = congestive heart failure; FTT = failure-to-thrive syndrome; PA = pulmonary atresia; and PS = pulmonic stenosis.

Figure 2

Intensity of the systolic murmur in 34 patients with pulmonary atresia and pulmonic stenosis before surgery and in 12 patients with pulmonic stenosis after surgery. PA = pulmonary atresia; PS = pulmonic stenosis; and PS (P.O.) = pulmonic stenosis, postoperative patients.

Figure 3

The patients with pulmonic stenosis had more intense systolic murmurs at the left upper sternal border (fig. 3) than those with atresia; no continuous murmurs were audible. Pulmonic closure was usually not audible although, rarely, a faint delayed P2 was heard. Contrary to reported results in older individuals,1 the murmur of pulmonic stenosis did not decrease in intensity postoperatively in these infants, a finding also reported by Mustard and associates.8 The postoperative

Pulmonary atresia group (fig. 2). The apparent absence of the failure-to-thrive syndrome, noted in these patients, may be due to the fact that 11 of the babies were evaluated when less than 2 weeks of age. Two of the four older infants, however, did display a poor growth pattern.

Cardiac examination revealed no murmur, or only an insignificant one, in 40% of the patients with pulmonary atresia (fig. 3). The remaining infants had grade 2 to 3 murmurs, presumably due to the presence of tricuspid regurgitation.12 In three instances, the murmur was continuous, and this was thought to indicate a patent ductus arteriosus. Pulmonic closure sound was absent in all of the patients.

After surgery, the one infant with an adequate follow-up had complete regression of all cardiac signs and symptoms and has had a normal growth and development.

Pulmonic Stenosis. The patients with pulmonic stenosis tended to develop their symptoms at a later age (fig. 1). The only difference in symptomatology in these infants from those in the pulmonary atresia group was the rarity of anoxic spells (fig. 2).

The patients with pulmonic stenosis had more intense systolic murmurs at the left upper sternal border (fig. 3) than those with atresia; no continuous murmurs were audible. Pulmonic closure was usually not audible although, rarely, a faint delayed P2 was heard. Contrary to reported results in older individuals,1 the murmur of pulmonic stenosis did not decrease in intensity postoperatively in these infants, a finding also reported by Mustard and associates.8 The postoperative
had evidence of the failure to thrive syndrome.

**Electrocardiographic Features**

**Pulmonary Atresia.** The electrocardiogram was the most important differential diagnostic tool among these patients (figs. 4 and 5). In the entire group of 15 infants with pulmonary atresia, right atrial hypertrophy ("P pulmonale") was present in only five. In type I pulmonary atresia, left ventricular hypertrophy was present in 10 of 13 instances and the QRS axis tended to be normal or deviated only slightly to the right. By contrast, in both cases of type II pulmonary atresia, right ventricular hypertrophy was present, with increased right precordial voltage and more prominent right axis deviation. The paradox, in type I pulmonary atresia, of a markedly thickened right ventricular wall with complete lack of right ventricular potentials may be explained by the absence of significant right ventricular end-diastolic volume vital for the development of surface potentials. In the one infant with type I pulmonary atresia who survived a Brock procedure, right precordial potentials developed, and the axis in
the frontal plane shifted rightward postoperatively.

**Pulmonic Stenosis.** Seventeen of 19 patients with pulmonic stenosis had right atrial hypertrophy on the electrocardiogram at initial evaluation. One of the other two had a nodal rhythm with an absent P wave. Eighteen patients had right ventricular hypertrophy with definite right axis deviation. This then would indicate the presence of a thick right ventricular wall as well as a normal or large-sized cavity. The infant without electrocardiographic evidence of right ventricular hypertrophy presumably had a diminutive right ventricular cavity as has been reported occasionally in pulmonic stenosis.4,16

"P pulmonale" disappeared in seven of the 11 surviving patients with pulmonic stenosis with adequate follow-up; right ventricular hypertrophy completely regressed in six. In two cases, mild right ventricular hypertrophy persisted and in three instances the residual right ventricular hypertrophy was severe. One of the latter group (the single case with greater than 30 mm of voltage in lead V1 postoperatively) underwent elective open-heart surgery at the age of 4½ years, followed by complete regression of electrocardiographic evidence of right ventricular hypertrophy.

**Radiological Findings**

**Pulmonary Atresia.** Cardiomegaly was present in x-rays of all of the patients with pulmonary atresia, and in nine of the 15 infants, the enlargement was marked. The pulmonary artery segment was invariably concave, and the pulmonary vasculature was interpreted as decreased in each instance.

The single long-term surgical survivor in the pulmonary atresia group had complete regression of his severe cardiomegaly and decreased pulmonary vasculature.

**Pulmonic Stenosis.** Ten of 19 patients with pulmonic stenosis had massive cardiac enlargement and the remainder had less striking cardiomegaly. Eleven patients had decreased pulmonary vasculature by x-ray, and in seven instances the markings were described as normal. In contrast to the situation in older children, the main pulmonary artery segment was only rarely prominent in these infants with severe pulmonic stenosis.

In the 11 surviving patients with pulmonic stenosis who have had adequate follow-up, the heart size has regressed appreciably. The cardiac silhouette has returned completely to normal in six patients, and in five, mild or borderline cardiomegaly has remained. Pulmonary vasculature was interpreted as normal in all of these infants on follow-up, and in only two was the main pulmonary artery segment slightly prominent.

**Cardiac Catheterization Data**

**Pulmonary Atresia.** The infants with pulmonary atresia were almost always severely unsaturated at the time of catheterization (table 1) and right-to-left shunts were found to occur through the atrial septum. Right ventricular pressure was increased in all but one patient (fig. 6). In most instances, it was impossible to judge from angiography how much distance was present from the blind right ventricular outflow tract to the lumen of the main pulmonary artery. This was primarily due to the difficulty in delivering enough contrast material into the pulmonary tree by means of an inadequate patent ductus arteriosus.

The two surviving postoperative patients in the pulmonary atresia group have not as yet been restudied.

**Pulmonic Stenosis.** All of the infants with critical pulmonic stenosis undergoing preoperative cardiac catheterization also had arterial oxygen unsaturation, although this was

<table>
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<th>Table 1</th>
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<tr>
<td><strong>Arterial Oxygen Saturation Data at Preoperative Cardiac Catheterization</strong></td>
</tr>
<tr>
<td>---</td>
</tr>
<tr>
<td>&gt; 95%</td>
</tr>
<tr>
<td>Pulmonic stenosis</td>
</tr>
<tr>
<td>Pulmonary atresia</td>
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</tbody>
</table>

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not as severe as in the patients with pulmonary atresia (table 1). The right-to-left shunts occurred across a patent foramen ovale or atrial septal defect in this group as well. Right ventricular hypertension was in general more marked than in the pulmonary atresia group (fig. 6). In contrast to older children with pulmonic stenosis, there was no correlation between the pressure in the right ventricle and the height of the R wave in lead V1 on the electrocardiogram in the present series of infants.

Immediate postoperative cardiac catheterizations were obtained in two patients with pulmonic stenosis in whom clinical improvement was not evident. One of these was the patient in whom preoperative catheterization had not been done. Both displayed severe residual right ventricular pressure elevation (170 and 155 mm Hg) and both eventually died, one while waiting for reoperation and the other after reoperation.

Late postoperative catheterizations were carried out in three patients with pulmonic stenosis, in two because of suspected significant residual right ventricular hypertension. The right ventricular systolic pressure in these cases was 98 and 100 mm Hg, respectively, compared to 140 and 170 mm Hg preoperatively. In the third patient who was restudied electively, the hemodynamics and arterial O2 saturations were normal.

Operative Indications for Infants with Pulmonary Atresia and Pulmonic Stenosis with Intact Ventricular Septum

Pulmonary Atresia

All infants with a clinical diagnosis of pulmonary atresia, preferably confirmed at cardiac catheterization and angiography, should be treated surgically. As indicated in the introduction, early death in these babies is a virtual certainty without operation.

Pulmonic Stenosis

Clearly, not all babies with a clinical diagnosis of pulmonic stenosis will be investigated by cardiac catheterization during infancy. Many of these patients have only mild or moderate obstruction; physiological studies as well as surgery may safely be postponed, with benefit, until they are older. Indications for cardiac catheterization in babies with the clinical diagnosis of pure pulmonic stenosis, according to the material presented, will include: symptoms (congestive heart failure, anoxic spells, and cyanosis); electrocardiographic findings (severe right ventricular hypertrophy); and roentgenograms (at least moderate cardiac enlargement). If the patient is critically ill, the experienced pediatric cardiologist may suggest operative intervention without physiological studies. In situations in which the diagnosis or the severity of the condition is in doubt, cardiac catheterization is mandatory. Right ventricular pressures above systemic level, the presence of

Figure 6

Right ventricular peak systolic pressure in mm Hg recorded at cardiac catheterization prior to surgery in 28 patients.

Figure 7

Operative procedures and results of surgery in 34 patients.
PULMONARY OUTFLOW OBSTRUCTION

Table 2

<table>
<thead>
<tr>
<th>Postoperative Results</th>
<th>Pulmonary atresia</th>
<th>Pulmonic stenosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total operated on</td>
<td>15</td>
<td>19</td>
</tr>
<tr>
<td>Outcome</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Excellent*</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>Good†</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Fair‡</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Improved, but</td>
<td></td>
<td></td>
</tr>
<tr>
<td>inadequate length</td>
<td></td>
<td></td>
</tr>
<tr>
<td>of follow-up§</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Died</td>
<td>13</td>
<td>5</td>
</tr>
</tbody>
</table>

*Seven patients with normal ECG and x-rays (one with normal findings on postoperative cardiac catheterization).
†Two patients with mild right ventricular hypertrophy on ECG.
‡Two patients with right ventricular pressure between 90 and 100 mm Hg at postoperative catheterization and one patient who required reoperation.
§Four patients who improved clinically postoperatively, but in whom adequate long-term follow-up data are not available.

Table 3

<table>
<thead>
<tr>
<th>Causes of Postoperative Death</th>
<th>Pulmonary atresia</th>
<th>Pulmonic stenosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arrest during operation</td>
<td>5 (all N)</td>
<td>3</td>
</tr>
<tr>
<td>Within 24 hrs P.O.</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>CHF</td>
<td>2 (N)</td>
<td></td>
</tr>
<tr>
<td>Arrest</td>
<td>2 (1N-1H)</td>
<td></td>
</tr>
<tr>
<td>Hypotension</td>
<td>1 (H)</td>
<td></td>
</tr>
<tr>
<td>Within 1 mo P.O.</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Chronic infection &amp; anoxia</td>
<td></td>
<td>Persistent</td>
</tr>
<tr>
<td>Anoxia</td>
<td></td>
<td>obstruction</td>
</tr>
<tr>
<td>Within 2 mo P.O.</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Anoxia</td>
<td></td>
<td>Persistent</td>
</tr>
<tr>
<td>Obstruction</td>
<td></td>
<td>obstruction</td>
</tr>
<tr>
<td>Total</td>
<td>13</td>
<td>5</td>
</tr>
</tbody>
</table>

Abbreviations: N = normobaric; H = hyperbaric; CHF = congestive heart failure; P.O. = postoperative.

a sizable atrial right-to-left shunt, markedly diminished pulmonary flow, and increased right ventricular end-diastolic pressure, all serve as indications for early surgery.

Surgery

Pulmonary Atresia

The surgical procedures undertaken and their results are presented in figure 7 and tables 2 and 3.

A variety of approaches was attempted in infants with pulmonary atresia because of the lack of satisfactory results with any one method (fig. 7). The mortality was extremely high; only two of 15 patients may be considered as surgical survivors. One infant (operated on under normobaric conditions) is well 3 years after transventricular valvotomy. The second survivor, operated upon under hyperbaric conditions, is doing well but is still cyanotic 6 months after a Blalock shunt procedure. Both living patients have type I pulmonary atresia.

Of the 13 infants who died, eight were operated on without hyperbaric support and all died on the operating table or within 24 hours after operation; one of these deaths occurred during a second shunt procedure performed 1 month after the initial shunt.
In the group of five patients who died following surgery under hyperbaric conditions, none died on the operating table and only one died within 24 hours after surgery. Two survived their initial procedure, but succumbed following a second operation after failure to maintain clinical improvement. The two remaining patients are classified as "late" deaths, having died 4 and 7 weeks postoperatively. One of these failures was due, in part, to a wound infection.

**Pulmonic Stenosis**

Among the group of 19 infants with pulmonic stenosis, five succumbed. Mortality was not related to age or weight at surgery. All infants underwent transventricular valvotomies (Brock procedures), and in all but one instance the stenosis was felt to be valvar. Thirteen patients were operated on under normobaric conditions, and in one hypothermia and inflow occlusion were employed. Six other patients underwent inflow occlusion (at normal temperature) in a hyperbaric chamber at pressure of 4.0 atmospheres absolute. In two of these, additional pulmonary valvotomies were performed through pulmonary arteriotomy incisions.

The five fatalities were all in the normobaric group. Three infants expired during the operative procedure (hypoxic cardiac arrest), and a fourth baby died during repeat surgery undertaken 2 weeks after the first operation because of a worsening of the clinical condition and increased right ventricular pressure established at cardiac catheterization. The fifth infant succumbed, unexpectedly, 2 months after what must be considered an inadequate pulmonary valvotomy.

Of the eight living patients operated on under normobaric conditions, three are considered to have had an excellent result and three are classified as having a fair result (table 2). One of the latter infants, at age 4½ years, required open-heart valvotomy, which was successful. The remaining two infants were clinically much improved in the immediate postoperative stage, although not enough data are available to evaluate their long-term status.

The six patients who were operated upon under hyperbaric conditions all survived and three are classified as having excellent results. Two are considered to have good results, and the remaining infant is improved clinically 3 months postoperatively. These cases represent the last six patients, under 1 year of age, who have undergone surgery for pulmonic stenosis at this institution.

**Pathology**

Postmortem examination was carried out in 12 of 13 fatal cases of pulmonary atresia and in all five cases of valvar pulmonic stenosis. Autopsy protocols were analyzed in each instance and in 10 of the cases of pulmonary atresia the heart specimens were reviewed by one of the authors (W. M. G.).

**Pulmonary Atresia**

The cardiac findings in the infants with pulmonary atresia are presented in table 4.

<table>
<thead>
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<th>Table 4</th>
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<tr>
<td><strong>Cardiac Anatomy in Twelve Patients with Pulmonary Atresia and Intact Ventricular Septum</strong></td>
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</table>

<table>
<thead>
<tr>
<th>Description</th>
<th>Main pulmonary artery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Small</td>
<td>Adequate</td>
</tr>
<tr>
<td>Right ventricular cavity, small; tricuspid orifice, small</td>
<td>2*</td>
</tr>
<tr>
<td>Right ventricular cavity, adequate; tricuspid orifice, small</td>
<td>0</td>
</tr>
<tr>
<td>Right ventricular cavity, adequate; tricuspid orifice, adequate</td>
<td>2</td>
</tr>
</tbody>
</table>

*Main pulmonary artery not patent in one case.
†Two of these may be classified as type II.
The size of the tricuspid orifice, right ventricular cavity, and main pulmonary artery was variable, and the right heart did not necessarily fall into either the "hypoplastic" or the "dilated" category. Instead there was a spectrum of right ventricular cavity size as well as variation in the degree of tricuspid stenosis. In two instances, however, the right ventricular chamber was sufficiently large to be considered characteristic of type II pulmonary atresia. The usual marked hypertrophy of the right ventricular wall was present in every case, and the opening in the atrial septum, usually a patent foramen ovale, was wide in all but one instance.

**Pulmonic Stenosis**

In each of the five patients with pulmonic stenosis who came to postmortem examination, the pulmonary orifice was critically obstructed. In two cases, significant right ventricular endocardial fibroelastosis was present and in three the right ventricular cavity was considered to be decreased in size. The tricuspid valves were normal in structure, but thickened in two instances. Examination of the atrial septum revealed a dilated patent foramen ovale in every specimen.

**Discussion**

A review of the literature⁴, ⁶, ¹⁰, ¹⁵, ¹⁸, ¹⁹ and the present series, revealed a total of 123 reported cases of pulmonary atresia with intact ventricular septum. Among these, 96 (80%) were type I and 27 (20%) were type II. Outside of the two survivors in the present series only one successful surgical result has been well documented—a successful transventricular valvotomy in an infant with type II pulmonary atresia.⁶ Keith and associates⁸ mentioned three of eight survivors of surgery but did not discuss the details of the surgical procedure, nor whether the malformations were considered to be type I or II pulmonary atresia. Despite these poor results, the 100% mortality rate occurring in those infants who are not operated on⁴ makes it imperative that an attempt be made to intervene surgically.

The primary cause of death in patients with pulmonary atresia and intact ventricular septum is hypoxia which leads to spells and congestive heart failure. Right ventricular pressure is almost invariably elevated in these infants. Surgery must be aimed not only at increasing arterial oxygen saturation by increasing pulmonary blood flow, but also, if possible, at decompressing the right ventricle. In theory, the most physiological surgical approach to gain these two objectives would be a direct attack on the right ventricular outflow tract. Including our one case of successful transventricular valvotomy in an infant with type I pulmonary atresia, there are now two of seven survivals reported using this approach.⁶, ⁹, ¹⁰ Unfortunately, it is often not feasible to carry out this operation and it is difficult to distinguish those infants in whom a Brock procedure would be technically possible.

One must have detailed knowledge about the size of the right ventricular cavity and the tricuspid orifice. Although pulmonary atresia with intact ventricular septum is rather arbitrarily divided into two types according to the size of the right ventricular cavity, the distinction between the two is not always anatomically clear cut. After a review of our autopsy material, it appears rather that the size of the right ventricular chamber represents more of a continuum ranging from a diminutive, teardrop-sized cavity to the more dilated right ventricle typical of type II pulmonary atresia. These latter patients are the best candidates for a transventricular valvotomy. However, the infants labeled as having type I pulmonary atresia still may have a reasonable-sized right ventricular chamber and this clinical diagnosis based on the electrocardiogram does not in itself rule out the possibility of a Brock procedure being carried out. This is especially true in the first week of life.⁶, ¹⁵ The status of the tricuspid valve also varies greatly. The orifice may be severely stenotic, but on occasion may be almost completely normal. The main pulmonary artery should be evaluated, if possible. This vessel is almost always patent but again variability exists, the lumen being
either reasonably wide or, for practical purposes, so narrow as to rule out the possibility of enlargement. The pulmonary artery should, if possible, be visualized by retrograde filling with dye from a patent ductus, which is invariably present in these infants. The distance between the main pulmonary artery and the right ventricle may then be estimated.

Although the pulmonary valve is represented by a thin fibrous membrane, the channel from the right ventricle may be long and extremely narrow. From the review of our autopsy material and angiographic data, it appeared that a Brock procedure could have been attempted on at least six of the 15 patients (including the successful case).

Theoretically the Glenn procedure would be the second choice for these infants, because it accomplishes simultaneously both an increase in pulmonary flow and right ventricular bypass. However, the poor results reported with this operation in the first 6 months of life, due to technical problems, and the actual reported failure in 17 of 18 infants with pulmonary atresia under 2 years of age in a collected series, contraindicate its use at the present time.

A systemic-pulmonary artery shunt is the remaining operative possibility. The problem of the failure to decompress the right ventricle and the technical problems in anastomoses preclude a great deal of enthusiasm for this procedure. It is of interest that the single patient in this series with a normal right ventricular pressure is also the only long-term (3-year) surgical survivor in a total of 14 reported cases. This indirectly illustrates the importance of right ventricular decompression in those infants with right ventricular hypertension.

We feel that if angiography reveals an adequately large ventricular cavity, with a short (1 to 2 mm) segment of atresia, a Brock procedure should be attempted. With the introduction of hyperbaric surgery in our institution, we hope that results with this direct approach to the right ventricular outflow tract may prove to be more fruitful.*

If a Brock procedure is not feasible, a systemic-pulmonary arterial shunt seems to be the only second choice at the present time. A combination of a shunt between the aorta and the right pulmonary artery, with creation of an atrial defect, seems a promising approach.

Critical pulmonic stenosis in infants leads to death at a young age. Mortality is related to anoxia and right ventricular failure, and as with pulmonary atresia, but with more hope of success, the aim of surgery must be to increase pulmonary blood flow as well as to decompress the right ventricle. There is general agreement, therefore, that pulmonary valvotomy, either transventricular or transarterial, is the best surgical approach in infants with severe pulmonic stenosis. Results, however, have been variable and published 8, 22 In the present series, Brock (transventricular) procedures were done in all instances, with open transpulmonary valvotomy carried out (in addition) in two of the six hyperbaric cases. Direct valvotomy was delayed until the pulmonary orifice had been enlarged sufficiently (by a Brock procedure) to insure good heart action and prevent severe arterial desaturation and subsequent cardiac arrest. In many instances, under normobaric conditions, hypoxic arrest occurred as the pericardium was opened. Successful resuscitation was accomplished after valvotomy in most cases, but this type of exitus still accounted for the majority of deaths in patients operated upon under normal atmospheric pressures in this series (four of five). Since no infant who underwent hyperbaric surgery died, the improvement in myocardial oxygenation obtained seems to be a definite advantage of this technique. Other factors which may account for success in both survival and long-term results in the hyperbaric groups include: (1) a prolongation of inflow occlusion time at body temperature of 35

*Since completion of this paper, this approach has been attempted in three infants, with success in two.
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to 37 C; and (2) strict attention to the acid-base status of the patient, both during operation and in the critical early postoperative period.

If possible, surgery should be carried out in infants with pulmonic stenosis before the onset of congestive heart failure. In this series, four of five deaths occurred among the nine patients who were in cardiac failure at the time of operation. However, if congestive heart failure is present and if clinical judgment indicates that there is time for medical maneuvers, decongestive measures must be applied vigorously. An occasional infant may have to be treated as a surgical emergency, the diagnosis being based on clinical evaluation only.

Surgical results with this lesion are encouraging, even in the presence of severe obstruction. Mirowski and associates22 found that incomplete relief of pulmonic stenosis is more likely in the more severe cases, but in the present study, this was not true. Engle and associates23 also did not relate poor long-term results to severity of stenosis in a large group of infants and children.

A crucial question in any infant with pulmonic stenosis as well as pulmonary atresia is whether there is an associated ventricular septal defect. Cardiac catheterization must be particularly oriented toward ruling out this possibility, because if a ventricular septal defect is present, we would much prefer to perform a systemic-pulmonary artery (Blalock) shunt and plan complete repair at a later date.

References


Rewards of Practice

One does not escape from being a patient because of being also a physician, and for my part I am glad to confess my sense of enjoyment in such visits, and how I have longed to keep my doctor at my side and to decoy him into a protracted stay. The convalescence he observes is for him, too, a pleasant thing. He has and should have pride in some distinct rescue, or in the fact that he has been able to stand by, with little interference, and see the disease run its normal course. I once watched a famous surgeon just after he had done a life-saving operation by dim candle-light. He stood smiling as the child’s breath came back, and kept nodding his head with pleasant sense of his own competence. He was most like a Newfoundland dog I once had the luck to see pull out a small child from the water and on to a raft. When we came up, the dog was wagging his tail and standing beside the child with sense of self-approval in every hair. The man wagged his head; the dog wagged his tail. Each liked well what he had done.—S. WEIR MITCHELL: Doctor and Patient, ed. 4. Philadelphia and London, J. B. Lippincott Company, 1904, p. 58.
Diagnosis and Surgical Treatment of Infants with Critical Pulmonary Outflow Obstruction: Study of Thirty-four Infants with Pulmonary Stenosis or Atresia, and Intact Ventricular Septum
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