Long-Term (10 to 13 Years) Follow-Up Study after Transventricular Pulmonary Valvulotomy for Pulmonary Stenosis with Intact Ventricular Septum

By M. Mirowski, M.D., Kishor D. Shah, M.D., Catherine A. Neill, M.D., and Helen B. Taussig, M.D.

In the last 15 years surgical methods for the relief of the pulmonic obstruction in patients with pulmonary stenosis and intact ventricular septum have been successfully developed. Although the open-heart technic with the use of cardiopulmonary bypass is today the procedure of choice, the classic Brock's transventricular valvulotomy is still preferred for small infants. The final conclusion as to the respective advantages and disadvantages of these procedures cannot be reached without long-term follow-up of the operated patients. The purpose of this study is to present an evaluation of 46 patients, 10 to 13 years, following transventricular pulmonary valvulotomy for pulmonary valvar stenosis with intact ventricular septum.

Material

Fifty-three consecutive patients with valvular pulmonary stenosis and intact ventricular septum were submitted to transventricular valvulotomy at the Johns Hopkins Hospital between November 1949 and January 1952. Forty-six patients survived the operation and are the subject of the present report. There were 18 males and 28 females in this group. At the time of surgery their ages ranged between 6 months and 33 years, the mean age being 11 years and the median 8.8 years. The age distribution is shown in table 1. Valvular pulmonary stenosis was the only known cardiac malformation present in these patients with the exception of one with an associated atrial septal defect and of three patients who subsequently were found to have an associated infundibular stenosis; one of these three had previously undergone elsewhere ligation of what was thought to be a patent ductus arteriosus; actually the left pulmonary artery had been ligated in error. None of these patients had infundibular resection. Two patients in this series had had previous anastomotic procedures: one a Blalock-Taussig anastomosis and the other a Pott's procedure.

All the 46 patients who survived surgery were alive as of October 1961. Virtually complete information was obtained concerning all but one patient; this patient, however, is known to have had a poor result from surgery and is awaiting a second operation elsewhere. Twenty-six patients have been examined personally by the authors; 20 were examined by the patients' cardiologist, and their evaluations together with the x-rays and electrocardiograms, were forwarded to the authors. The follow-up period in this series ranges between 10 years and 12½ years, the mean figure for the whole group being 11 years and 2 months. Seven patients needed further cardiac surgery. In this report the status of these patients is based on their condition prior to any further surgery.

Results

The relevant preoperative and postoperative data as well as the long-term results after pulmonary valvulotomy are presented in figures 1, 2, and 3, respectively.

Symptomatology

The group as a whole was preoperatively severely incapacitated. According to the classification of the American Heart Association, ten patients were in class IV, 17 in class III, and 24 in class II; in three infants the functional capacity could not be determined. All patients showed a striking functional improvement after surgery. At the end of the follow-up period 37 patients (80 per cent) lead virtually normal lives and were in class I. Eight patients were in class II and only one was severely incapacitated (class III).
Table 1

Long-Term Results after Transventricular Pulmonary Valvulotomy for Pulmonary Stenosis According to the Age at the Time of Surgery

<table>
<thead>
<tr>
<th>Age at surgery</th>
<th>No. of patients</th>
<th>Excellent results</th>
<th>Good results</th>
<th>Poor results</th>
<th>PSD*</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
<td>%</td>
<td>No.</td>
<td>%</td>
<td>No.</td>
</tr>
<tr>
<td>6-14</td>
<td>22</td>
<td>10</td>
<td>45</td>
<td>7</td>
<td>32</td>
</tr>
<tr>
<td>15-33</td>
<td>12</td>
<td>4</td>
<td>33</td>
<td>3</td>
<td>25</td>
</tr>
<tr>
<td>Total group</td>
<td>46</td>
<td>20</td>
<td>44</td>
<td>12</td>
<td>26</td>
</tr>
</tbody>
</table>

*PSD, poststenotic dilatation of pulmonary artery.

Physical Findings

Cyanosis, which prior to surgery was present in 35 patients (76 per cent), was still present in eight patients at the time of this evaluation.

Congestive failure had occurred in 24 patients (52 per cent) prior to the operation. In every instance the patient regained compensation following valvulotomy. One patient, in whom the stenosis was not adequately relieved, suffered a recurrence of cardiac failure 9 1/2 years after surgery.

A systolic thrill over the pulmonary area was felt preoperatively in all but five patients. At the present follow-up a systolic thrill was still palpable in 25 of the 45 patients in whom these data were available. An ejection-type systolic murmur, best heard over the pulmonary area, was present preoperatively in all patients. The intensity of this murmur, according to Levine's criteria, was grade V in three patients, grade IV in 38, grade III in four, and grade II in one patient. This murmur persisted after surgery in all patients but its intensity decreased: it was grade V in two patients, grade IV in 11, grade III in 14, grade II in 15, and grade I in three patients. No definite correlation could be found between the postoperative changes in the intensity of the systolic murmur and the clinical improvement.

The murmur of pulmonary insufficiency was audible in one patient prior to surgery and one other had a questionable early diastolic murmur. Postoperatively, an early diastolic murmur suggestive of pulmonary insufficiency appeared in 13 patients (29 per cent).

Radiologic Data

The size of the heart was evaluated in terms of being normal, moderately enlarged, and markedly enlarged. A cardiothoracic ratio of 58 per cent in infants, 52 per cent in children, and 48 per cent in adults was considered to be the upper limit of normal. A markedly enlarged heart was present when the cardiothoracic ratio exceeded 65 per cent in infants and 57 per cent in older children and adults. Prior to surgery, only nine patients had a heart within normal limits in size; 24 patients showed moderate cardiac enlargement, and 13 suffered from marked cardiac enlargement. After surgery, 29 patients had a heart of normal size and 16 still showed moderate cardiac enlargement.

Poststenotic dilatation of the pulmonary artery was the rule; it occurred preopera-
respectively in all but two instances. The pulmonary artery was moderately prominent in 31 patients, markedly so in 11, and extremely prominent in one patient. At the time of the present evaluation the poststenotic dilatation was found to be moderately prominent in 23 patients, markedly prominent in 18, and extremely prominent in two patients; in two patients no poststenotic dilatation was apparent.

**Electrocardiographic Data**

The electrocardiographic diagnosis of right atrial enlargement, right ventricular hypertrophy, and intraventricular conduction disturbances was based upon the criteria of Sodi-Pallares. Prior to surgery electrocardiographic evidence of right atrial enlargement was present in all but five patients. At the time of present evaluation 17 patients still showed signs of moderate right atrial enlargement. There was no significant difference in the values of the average mean frontal P axis before and after surgery (+47° and +45°, respectively). The preoperative distribution and the postoperative changes of the mean frontal P axes are shown in figure 4.

The degree of right ventricular hypertrophy was graded as mild, moderate, and severe; this classification was based mainly on the importance of the repolarization changes. The right ventricular hypertrophy was considered as severe when the ST segment showed marked depression with negative secondary T-wave configuration over the right precordium. Right ventricular hypertrophy was moderate when such repolarization changes were slight; it was considered as mild when the secondary repolarization changes were absent. On the basis of this classification, prior to surgery, right ventricular hypertrophy was severe in 33 patients, moderate in four, and mild in nine. At the time of this study the signs of right ventricular hypertrophy had completely disappeared in 17 patients. Twelve patients still showed mild right ventricular hypertrophy, eight showed moderate hypertrophy, and eight others showed severe right ventricular hypertrophy. The average mean frontal QRS axis, which preoperatively was +120°, had shifted to +75° at the time of the present evaluation, and the average mean frontal T axis shifted from −21° to +32°. The preoperative distribution and the postoperative changes of the mean frontal QRS and T axes are shown in figure 4.

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Before the operation only one patient had a complete right bundle-branch block and four patients had a pattern of incomplete right bundle-branch block. These intraventricular conduction disturbances persisted unchanged by the operation in all but one patient; in this instance the incomplete right bundle-branch block disappeared. In addition, 23 patients developed incomplete right bundle-branch block after surgery.

**Hemodynamic Data**

A comparative study of the preoperative and postoperative hemodynamic data was not always possible, since cardiac catheterization was not a routine procedure, either before or after operation. Thirty-four patients had right heart catheterization before the operation; in 28 (82 per cent) the right ventricular pressure was above 100 mm Hg and in half of them this pressure ranged between 160 and 223 mm Hg. Postoperatively, right heart catheterization was performed in 25 patients, only 19 of whom had catheterization prior to surgery. In these 25 patients the postoperative right ventricular pressure ranged from 32 to 170 mm Hg; in six instances it was below 50, in 11 between 50 and 100, and in eight the right ventricular pressure was above 100 mm Hg. It should be noted that the postoperative hemodynamic studies were performed only in patients without significant subjective and objective improvement or in those who showed some persistent cardiac abnormalities; only occasionally was right heart catheterization undertaken primarily to prove that the stenosis had been relieved.

**Long-Term Results**

The long-term results of the pulmonary valvulotomy were classified as excellent, good, or poor. The results were considered excellent when the analysis of the available clinical and laboratory data indicated that the pulmonary obstruction was virtually relieved. In the absence of hemodynamic evidence, the complete regression of right ventricular hypertrophy on the electrocardiogram was considered to be the most reliable criterion for a successful operation. The results were classified as good when there was a remarkable functional and objective improvement though the electrocardiographic evidence of residual right ventricular hypertrophy persisted. The results were poor when it was obvious that the relief of stenosis was unsatisfactory or when the available data suggested that the patient needed further surgery.

According to the above criteria, 20 patients (44 per cent) had excellent postoperative re-
sults (fig. 3). Three of these patients showed electrocardiographic evidence of mild to moderate right ventricular hypertrophy; their hemodynamic studies, however, demonstrated that the right ventricular pressure was virtually normal but only slightly elevated (32, 43, and 55 mm. Hg, respectively). Twelve patients (26 per cent) were considered to have good results.

Twelve patients (26 per cent) had poor results. These patients preoperatively were extremely incapacitated. Immediately after surgery they all had shown a significant subjective improvement; in some of them this improvement had persisted for a number of years. The cyanosis disappeared completely in two patients and cardiac failure was relieved in four. Four patients showed electrocardiographic evidence of regression of right ventricular hypertrophy. In the majority of cases the surgical notes gave no indication that the stenosis was not satisfactorily corrected. At the time of this evaluation, however, decreased physical tolerance was a common complaint. Eight patients still presented a variable degree of cyanosis, six had cardiomegaly, and eight showed persistent electrocardiographic abnormalities. Seven patients in this group have already been reoperated upon, six with the use of cardiopulmonary bypass and one under hypothermia; there were two postoperative deaths in this group. One of the other five is scheduled for reoperation elsewhere and the remaining four are not interested in further surgery at the present time.

One patient in this group had an unusual type of pulmonary stenosis in that calcification of the pulmonary valve was found at the initial operation. Shortly after surgery this patient developed an aneurysm at the site of the ventriculotomy. An attempt 11 years after the Brock procedure to excise the aneurysm and to relieve the residual pulmonary stenosis resulted in death. This patient and another patient who developed an aneurysm at the site of ventriculotomy after transventricular valvulotomy done elsewhere will be reported separately.13

Two patients postoperatively developed a conspicuous progressive dilatation of the pulmonary artery as illustrated in figure 5; these patients are not included in any of the above-described groups because, aside from the dilatation of the pulmonary artery, they have had excellent results. One of these patients has a right ventricular pressure of 44/6/4 mm. Hg, with a gradient of 30 mm. Hg across the pulmonary valve; he works as a truck driver. The second patient has a normal electrocardiogram; she has recently had two uneventful pregnancies.

Discussion

In the evaluation of the long-term results of transventricular pulmonary valvulotomy a few considerations concerning the composi-

Figure 5

Progressive conspicuous dilatation of the pulmonary artery in a patient who otherwise had an excellent result from transventricular pulmonary valvulotomy. A, prior to surgery; B, 1 year after valvulotomy; C, 12 years after valvulotomy.
tion of our group must be kept in mind. First, the patients in this study were the earliest to undergo pulmonary valvulotomy at this hospital. Since then not only has the surgical skill been greatly perfected but also the indications for surgery have been markedly extended; at the present time operation is frequently recommended for asymptomatic patients with only moderate stenosis. In contrast to this, our series was composed of patients with very severe pulmonary stenosis and consequently represented an unusually high risk group. Indeed, congestive cardiac failure, cyanosis, and severe functional incapacity were repeatedly observed in these patients, whereas today such symptomatology is unusual.

It is noteworthy that all the patients who survived the operation were alive 10 or more years after the procedure. The incidence of the excellent results is between 44 to 48 per cent. The latter figure includes the two patients with conspicuous dilatation of the pulmonary artery in whom the pulmonary obstruction was otherwise satisfactorily relieved. Only 12 patients had a poor result, and even these patients had derived functional and objective improvement from the operation. Indeed, all of the patients who had poor results from valvulotomy were sufficiently improved to be able to wait for many years for surgery to be further perfected.

As previously stated, the 12 patients classified as having good results had remarkable functional improvement but still showed evidence of right ventricular hypertrophy. It is quite possible that a few of these patients may still have some residual pulmonary obstruction and may require further surgical intervention. It is probable, however, that the majority of them will never have a sufficiently important gradient across the pulmonary valve to justify a second operation.

The absence of routine hemodynamic studies before and after pulmonary valvulotomy is a limitation of this report. Inasmuch as reaetheterization was performed only when specifically indicated, the hemodynamic data on those patients who were reaetheterized do not reflect the postoperative improvement of the entire group. Furthermore, the majority of patients who were reaetheterized had this procedure performed shortly after surgery. Even though valvular pulmonary stenosis has been successfully relieved, it is now well known14, 15 that for months and sometimes for years after closed valvulotomy the right ventricular pressure may remain elevated. With the passage of time, however, the muscular obstruction responsible for this residual high ventricular pressure may gradually regress. Such was the course of events in one of our patients who was followed closely because of the postoperative development of increasing poststenotic dilatation of the pulmonary artery (fig. 5). One month after valvulotomy his right ventricular pressure had dropped from the preoperative level of 143/0 to 80/8 mm. Hg. Five years later this pressure was found to be 86/0; finally, 11 years after the valvulotomy his right ventricular pressure was only 44/3/6 mm. Hg.

It should be emphasized, however, that the hemodynamic proof of the relief of the pulmonary obstruction is scarcely necessary in patients in whom the electrocardiogram has shown a complete regression of right ventricular hypertrophy. Complete regression of the electrocardiographic evidence of right ventricular hypertrophy virtually excludes the existence of a significant gradient across the pulmonary valve. On the other hand, evidence of residual right ventricular hypertrophy on the electrocardiogram can definitely persist in the presence of a normal right ventricular pressure. This was seen in two patients in our series.

Analysis of the factors that may influence the long-term results of transventricular valvulotomy showed that the incidence of excellent results was higher in the younger age group than in older patients (table 1). A similar observation has been noted by Campbell.15 There was no significant difference in the long-term results according to the sex of the patient. The three major factors that ap-
pear to affect the success of the operation are the functional capacity of the patient, the presence or absence of cyanosis, and the state of compensation.

The preoperative functional capacity has a direct relation to the long-term benefits (table 2): only 12 per cent of the patients who had functional capacity class II had poor results, whereas this incidence increased to 35 per cent in patients whose functional capacity was class III; both the patients who were in class IV had poor results from surgery.

The influence of cyanosis is shown in table 3. The incidence of poor results is considerably higher in cyanotic patients than in those without cyanosis. Congestive cardiac failure also reduces the chance of excellent results (table 4).

It is important to note that none of the three patients who had an associated infundibular stenosis obtained excellent or even good results from surgery: two had poor results and the third developed progressive dilatation of the pulmonary artery.

The development of pulmonary insufficiency has been reported as relatively frequent after both closed and open valvulotomy,\textsuperscript{15-17} its clinical significance in relation to the long-term results of surgery is still debated. Campbell\textsuperscript{13} believes that postoperative pulmonary

<table>
<thead>
<tr>
<th>Preoperative functional capacity</th>
<th>No. of patients</th>
<th>Excellent results No. %</th>
<th>Good results No. %</th>
<th>Poor results No. %</th>
<th>PSD* No. %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class II</td>
<td>24</td>
<td>10 42</td>
<td>9 38</td>
<td>3 12</td>
<td>2 8</td>
</tr>
<tr>
<td>Class III</td>
<td>17</td>
<td>8 47</td>
<td>3 18</td>
<td>6 35</td>
<td></td>
</tr>
<tr>
<td>Class IV</td>
<td>2</td>
<td></td>
<td></td>
<td>2 100</td>
<td></td>
</tr>
<tr>
<td>Undetermined</td>
<td>3</td>
<td>2 67</td>
<td></td>
<td>1 33</td>
<td></td>
</tr>
<tr>
<td>Total group</td>
<td>46</td>
<td>20 44</td>
<td>12 26</td>
<td>12 26</td>
<td>2 4</td>
</tr>
</tbody>
</table>

*PSD, poststenotic dilatation of pulmonary artery.

<table>
<thead>
<tr>
<th>Group</th>
<th>No. of patients</th>
<th>Excellent results No. %</th>
<th>Good results No. %</th>
<th>Poor results No. %</th>
<th>PSD* No. %</th>
</tr>
</thead>
<tbody>
<tr>
<td>With cyanosis</td>
<td>35</td>
<td>16 46</td>
<td>9 26</td>
<td>10 28</td>
<td></td>
</tr>
<tr>
<td>Without cyanosis</td>
<td>11</td>
<td>4 37</td>
<td>3 27</td>
<td>2 18</td>
<td>2 18</td>
</tr>
<tr>
<td>Total group</td>
<td>46</td>
<td>20 44</td>
<td>12 26</td>
<td>12 26</td>
<td>2 4</td>
</tr>
</tbody>
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<table>
<thead>
<tr>
<th>Group</th>
<th>No. of patients</th>
<th>Excellent results No. %</th>
<th>Good results No. %</th>
<th>Poor results No. %</th>
<th>PSD* No. %</th>
</tr>
</thead>
<tbody>
<tr>
<td>With CCF\textsuperscript{1}</td>
<td>24</td>
<td>8 33</td>
<td>9 38</td>
<td>6 25</td>
<td>1 4</td>
</tr>
<tr>
<td>Without CCF</td>
<td>22</td>
<td>12 55</td>
<td>3 14</td>
<td>6 27</td>
<td>1 4</td>
</tr>
<tr>
<td>Total group</td>
<td>46</td>
<td>20 44</td>
<td>12 26</td>
<td>12 26</td>
<td>2 4</td>
</tr>
</tbody>
</table>

\textsuperscript{1}CCF, congestive cardiac failure.
*PSD, poststenotic dilatation of pulmonary artery.

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insufficiency is harmful and should be avoided to obtain a good lasting result. On the other hand, Blount et al.\textsuperscript{17} considered that in most instances pulmonary insufficiency is without hemodynamic significance. The analysis of the long-term results in our 13 patients who developed pulmonary insufficiency shows that nine of them (69 per cent) belong to the "excellent result" group and only two to the "poor result" group (15 per cent). Since the incidence of the "excellent result" and the "poor result" groups in the total series is 44 per cent and 26 per cent, respectively, it appears that pulmonary insufficiency has apparently not altered the postoperative course of these patients. Nevertheless, in the absence of routine hemodynamic studies it may be premature to infer that pulmonary insufficiency has no hemodynamic significance. In this respect it may be of interest to observe that 10 of these 13 patients with pulmonary insufficiency also postoperatively developed an incomplete right bundle-branch block. Indeed, in this series the incidence of incomplete right bundle-branch block in patients with postoperative pulmonary insufficiency was found to be almost twice as high as in patients without this complication. The difference between these two figures is statistically significant ($p < 0.05$). Since incomplete right bundle-branch block is frequently observed in patients with diastolic overloading of the right ventricle,\textsuperscript{18, 19} its high incidence in the first group of patients may be related to the diastolic overloading secondary to pulmonary insufficiency.

**Summary**

A long-term follow-up study on 46 patients with pulmonary stenosis and intact ventricular septum who survived transventricular pulmonary valvulotomy performed at the Johns Hopkins Hospital between 1949 and 1951 was carried out; the mean follow-up period was 11.2 years. Preoperatively these patients were severely incapacitated, 76 per cent were cyanotic, 80 per cent had cardiomegaly, and 52 per cent were in congestive cardiac failure.

All 46 survivors of the pulmonary valvulotomy were alive at the time of the present evaluation. In almost half of these patients the long-term results were excellent and the pulmonary stenosis appeared to be virtually relieved. The results were good in 26 per cent of the patients; these patients also had remarkable subjective and objective improvement but still presented signs of residual right ventricular hypertrophy on the electrocardiogram. In 26 per cent of patients the long-term results were poor; seven of these patients have been reoperated upon. Twenty-nine per cent of the patients developed pulmonary insufficiency after surgery; there was no evidence, however, that this complication affected the long-term results.

The long-term studies indicate that for patients with mild to moderate pulmonary valvular stenosis a transventricular pulmonary valvulotomy gives extremely satisfactory results. For patients with congestive cardiac failure, cyanosis, or severe incapacity, the long-term results frequently are less satisfactory.

**References**

9. BLALOCK, A.: Personal communication.

Effect of Electricity on Muscular Motion

What no one had attempted before Galvani, was now accomplished by his sagacity and ingenuity, that we may have muscles disposed to contraction by the mere passage of a spark, although they may be either situated far remote from the conductor, or surrounded on all sides with sheets of glass. And those contractions in enclosed frogs remote from the conductor are excited by the action of either artificial or atmospheric electricity.

Muscles immersed, for example, in an atmosphere of extrinsic positive electricity, cannot themselves escape being electrified unless they find the opposite electricity, or exude electricity introduced into them; while this is happening, muscular motions are excited. Therefore communicated electricity does not itself produce the contraction, but, disturbing from its equilibrium the electricity in the muscles, conduces to the production of contraction.—LUIGI GALVANI. Commentary on the Effect of Electricity on Muscular Motion. Translated by ROBERT MONTRAVILLE GREEN, M.D. Cambridge, Massachusetts, Elizabeth Licht, Publisher, 1953, p. 6.
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