Ten to Thirteen Year Follow-up on Patients after a Blalock-Taussig Operation

By Helen B. Taussig, M.D., Hilary Crawford, M.B., Salvatore Pelargonio, M.D., and Stella Zacharioudakis, M.D.

In 1956 White, McNamara, Bauersfeld, and Taussig\(^1\) reported a 5 to 8 year follow-up on 224 patients with a tetralogy of Fallot who had been improved by a Blalock-Taussig operation, and on whom we had complete information. By complete information we meant not only that we knew whether the patient was living or dead, improved or no longer improved, but also for all those who were living we knew the state of compensation, the size of the heart, and the height of the red blood cell count, of the hemoglobin, and of the hematocrit reading. In the present study we have sought the same information and have used the same criteria.

Our former study showed that 5 to 8 years after operation, 67 per cent of the entire group of patients were still doing well; 20 per cent had failed to maintain improvement, 10 per cent had had a second operation, and 10 per cent were in need of a second operation. These figures were remarkably similar to those reported by Sir Maurice Campbell\(^2\) in his follow-up study of patients over a similar period of time after operation. He had two groups of patients: those who had had a Blalock-Taussig anastomosis and those on whom Sir Russell Broek had resected the pulmonary stenosis. Both groups showed approximately the same results: in each group about 20 per cent had failed to maintain their improvement. Sir Maurice Campbell believed that in some instances the failure to maintain improvement was because the child had outgrown his anastomosis and, in other instances, because the pulmonary stenosis had increased. The latter was in all probability the principal reason for a patient failing to maintain improvement after a resection of the pulmonary

From the Department of Pediatrics, The Johns Hopkins School of Medicine, Baltimore 5, Maryland.
failure and 27 patients (12 per cent) have died during the 5 to 13 year follow-up period (table 1).

These figures clearly indicate that during the second 5-year period of observation, a considerable number of patients have failed to maintain their improvement but only a few patients have developed cardiac failure. In addition, a few patients as reported by Ross, Taussig and Evans have developed pulmonary hypertension with or without cardiac failure. In most instances, however, failure to maintain improvement has been due to a reduction in the pulmonary blood flow.

Analysis of the age of the patient in relation to the need for a second operation clearly shows that the younger the patient, the greater is the probability that he will need a second operation. Thus, of the 15 patients operated upon under 2 years of age, 11 have had a second operation. Although the group is small, it does show a very high rate of second operations. Of the 130 patients operated on between 2 to 8 years, 44 (34 per cent) have had a second operation. In contrast to these findings, of the 42 patients between 8 and 12 years, five (or only 12 per cent) had had a second operation. In our previous study, older patients had done less well. In this group too, of the 37 patients over 12 years of age, 10 (27 per cent) have had a second operation.

Second operations have been performed when failure to maintain improvement was due to decrease in the pulmonary blood flow. In most instances, this has been due to failure of the anastomosis to grow with the patient or occasionally to increase in the pulmonary stenosis. Fortunately when this occurs, the heart remains small or may even decrease in size in proportion to the growth of the chest and the lungs become clear.

As previously mentioned, in a few instances, especially when the anastomosis was too large, the patient has developed cardiac failure with or without pulmonary hypertension. In rare instances, the patient has developed pulmonary hypertension associated with great dilatation of one or both of the branches of the pulmonary artery but without cardiac enlargement. The reason for development of pulmonary hypertension in the last-mentioned group is not clear. Fortunately the group is small, approximately 1 per cent. Needless to say, patients who have developed pulmonary hypertension cannot be helped by either a second anastomosis or by correction of the malformation.

The results of the second operations (most of which were a second anastomosis) have not been as good as the results of first operations. Of the 70 patients who have had a second operation, 33 (45 per cent) obtained good results; six (9 per cent) were satisfied with the result; 13 (20 per cent) were unimproved; 14 (20 per cent) died; and four patients are too recent to permit evaluation of the result.

Correction of the malformation is clearly the ideal. Two questions arise: Is it possible? If so, at what age should total correction be performed? The answers depend not only on the skill of the surgeon to correct the malformation but also upon physiologic problems.

Lillehei, Kirklin, and others have emphasized the importance of complete relief of the pulmonary stenosis. In many instances, especially in young children and in infants, not only is the pulmonary stenosis severe but the pulmonary artery is abnormally small. Frequently it is less than one third the size of the aorta. Under such circumstances, a graft is necessary and may need to be extended into the pulmonary artery. There is serious reason to doubt that a graft will permit the right ventricle to grow and develop normally.
Table 2

Age Distribution and Time Interval between First and Second Operations

<table>
<thead>
<tr>
<th>Age at 1st operation</th>
<th>0-2</th>
<th>2-4</th>
<th>4-6</th>
<th>6-8</th>
<th>8-10</th>
<th>10-12</th>
<th>12-16</th>
<th>16+</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>15</td>
<td>45</td>
<td>48</td>
<td>37</td>
<td>28</td>
<td>14</td>
<td>18</td>
<td>19</td>
<td>224</td>
</tr>
<tr>
<td>2nd operation thru 5 yrs.</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>9</td>
</tr>
<tr>
<td>6 to 10 yrs.</td>
<td>8</td>
<td>12</td>
<td>11</td>
<td>6</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>3</td>
<td>43</td>
</tr>
<tr>
<td>11 to 13 yrs.</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>5</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td>1</td>
<td>18</td>
</tr>
<tr>
<td>Total</td>
<td>11</td>
<td>16</td>
<td>15</td>
<td>13</td>
<td>4</td>
<td>1</td>
<td>6</td>
<td>4</td>
<td>70</td>
</tr>
<tr>
<td>Died from 2nd operation</td>
<td>2</td>
<td>3</td>
<td>2</td>
<td>3</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>14</td>
</tr>
</tbody>
</table>

Broadly speaking, abnormal areas do not grow at a normal rate. This is notably true in "pure" pulmonary stenosis, both valvular and infundibular. Indeed, it is frequently because of failure of the aperture to maintain its relative size that the condition leads to progressive cardiac enlargement.

If after total correction of a tetralogy of Fallot the previous stenotic area failed to keep pace with the growth of the child, the individual would be left with a defective right ventricle and pulmonary stenosis, which may be extremely difficult to correct. For this reason, an anastomotic procedure may be the operation of choice for young patients. Therefore, let us consider the duration of the benefit derived from an anastomotic procedure.

Our first study was based on a 5 to 8 year follow-up and the present study is based on a 10 to 13 year follow-up. In both instances, we have reported the status of the patient at his most recent examination: this meant his status at the longest known time interval. Since many of the patients in the 5 to 8 year follow-up were examined 6, 7, or 8 years after operation, and in our present study a number of patients had their operation 12 and 13 years ago, we have reviewed the status of the entire group at the end of 5 years, and at the end of 10 years, and what has happened between 11 and 13 years.

Table 2 shows the number of patients in the various age groups who have required second operation within the first 5 years, and within 6 through 10 years, and those who have had their second operation 11 to 13 years after their first operation. As in all other reports, it is clear that patients who were operated on between 8 and 12 years of age have obtained the best results. In our previous study, we found that 85 per cent of children operated on between 8 and 12 years of age had maintained their improvement and none had had a second operation. In this study, none of the patients in this age group has required a second operation within 5 years and only two patients required an operation within 10 years.

The younger children have not done quite so well. Of the 85 children between 4 and 8 years of age, 28 have required a second operation; that is nearly 32 per cent. Nevertheless, only three of these children required a second operation within 5 years. These findings indicate that a Blalock-Taussig anastomosis would enable most of these children to reach 10 or 12 years of age before a second operation was necessary. Actually, the results in the 2 to 4 year age group are remarkably similar. Slightly over 33 per cent have required a second operation but only 5 per cent within the first 5 years.

The real challenge before us concerns the very young children. Young children clearly do less well than older children because of the small size of the heart and the great vessels and also because of the extreme severity of the pulmonary stenosis. Growth is a very important consideration, as regards both the results from an anastomotic procedure and correction of the malformation. In children.
under 2 years of age, our mortality rate is still high and only one third of the children who survived operation have lived to be 10 or 12 years of age.

In infancy, owing to the small size of the subclavian artery, a Potts' procedure usually gives better results than does a Blalock-Taussig anastomosis. Although in 1959 most surgeons found total correction of the malformation more difficult after a Potts' procedure than after a Blalock-Taussig anastomosis, it is reasonable to expect that this difficulty will be overcome by the time these infants require a second operation. Therefore, it seems to the authors sound to give the infant who is in difficulty the benefit of a Potts' operation and hope to permit him to reach 8 to 12 years of age before total correction need be considered.

Kirklin and Bahnsen have found that total correction is not appreciably more difficult if a child has had a Blalock-Taussig anastomosis than if no such operation has been performed. Possibly the operation may be easier because the heart is larger and the margin of safety is greater. Furthermore, the more nearly the child has attained his growth, the greater is the probability that total correction will be of permanent benefit.

In a young patient, the operation is more difficult, the mortality rate is higher, and the long-time results are less certain. In the infant with severe pulmonary stenosis or functional pulmonary atresia, it is not only necessary to insert a patch into the right ventricle but also to enlarge the pulmonary orifice, and often the patch has to be extended into the pulmonary artery. Even though connective tissue grows into the patch, it cannot be expected that the patch will grow with the individual. Consequently, there is real danger that as the child grows, the right ventricle and the pulmonary artery will not grow proportionately well and the child will suffer from the defective development of the right ventricle. For such a child, ultimate correction may well be more difficult than it would be if the child had had a previous Blalock-Taussig anastomosis.

Finally, the recent studies of Ferencz and Bahnson have shown that a Blalock-Taussig anastomosis may have a favorable effect on the pulmonary vascular bed. She has confirmed Rich's observation that almost all cyanotic children with polycythemia and reduced pulmonary blood flow show multiple thromboses and widespread evidence of recanalization in the lungs; Ferencz found, however, that in the patients who had a satisfactory anastomosis and who have died from other causes, every one of them has shown a normal pulmonary vascular bed. Total correction means the lungs suddenly receive their normal volume of blood. For this reason, the child who has had a previous operation and who has a normal pulmonary vascular bed may more readily adjust to the increased pulmonary blood flow that occurs with this operation than will the child whose lungs have not had an opportunity to recover from the injuries caused by polycythemia.

As regards other complications, as previously mentioned there were 41 deaths among the 224 patients between 5 and 10 years after operation. An analysis of the cause of death showed that the major cause of death was a second operation; of the 70 second operations, 14 died at operation; of the 24 cases of subacute bacterial endocarditis, 10 died; of the 12 cases of brain abscesses, nine have died. Thus, subacute bacterial endocarditis and brain abscess took a high toll among these patients. Four suffered from sudden unexplained death and four died of miscellaneous causes. On the cheerful side of our follow-up we know of eight marriages; three men and five women. They have had 10 children. One woman has had four children, one with a congenital malformation of the heart, but the other three are healthy children.

Finally, it is important to emphasize that our present mortality rate from a Blalock-Taussig operation on children with a tetralogy of Fallot who are between 2 and 12 years of age is 2 per cent. The present mortality rate for total correction in this age group in experienced hands is between 10 and 15 per cent. During the next 10 years the mortality rate for corrective surgery will undoubtedly
be reduced, so that the risk of the two operations will not be greater than the present risk of corrective surgery alone, and the results will be more certain. Therefore, we believe that although total correction is the ideal, an anastomosis is better for infants and small children, and total correction should be reserved for the patient who has attained or nearly attained his full growth. For older individuals, total correction restores the circulation to normal. The long-time results of excision of the stenotic area and the insertion of a patch are not known but it is to be hoped that the prognosis is excellent. It should, however, be remembered that for many the prognosis after a Blalock-Taussig operation is known to be extremely good.

Summary

Analysis of 224 patients who were living 5 years after operation showed that at the end of 10 to 13 years, 40 per cent were still doing well, 46 per cent had failed to maintain improvement, 2 per cent had developed cardiac failure, and 12 per cent had died. The majority of patients who had failed to maintain improvement suffered from increasing cyanosis and dyspnea associated with a small heart and clear lung fields; hence these patients can be helped by a second operation:—either a second anastomosis or total correction. The results of a second anastomosis have, however, not been as satisfactory as the first anastomosis. Clearly total correction is the ideal.

The age at which total correction is indicated depends not only on the skill of the surgeon but on the severity of the abnormality. The long-time results of total correction are unknown. Inasmuch as abnormal tissue does not grow as fast as normal tissue, the possibility exists that the area in which tissue has been excised and a patch inserted may fail to grow or even contract and cause the right ventricle to become abnormally small.

Analysis of the results at the end of 5, 10, and 13 years showed that only 10 patients (less than 5 per cent) had required a second operation at the end of 5 years and only 20 per cent had required a second operation in the ensuing 5 years. Older patients, i.e., those between 8 and 12 years, had done far better than younger children. Only two of a group of 38 children operated upon between 8 and 12 years of age had required a second operation within 10 years. Thus a Blalock-Taussig operation offers the child an excellent chance to attain the major part of his growth before total correction is undertaken.

A further advantage of an early anastomotic procedure is that it restores the pulmonary vascular bed to normal and thereby renders it easier for the lungs to adjust to the full circulatory load placed on them at the time of the total correction.

The conclusion is reached, therefore, that for infants and small children a Blalock-Taussig operation is preferable. For children who have attained or nearly attained full growth, total correction is the ideal.

For many patients, especially those between 8 and 12 years of age for whom total correction is not possible, a simple Blalock-Taussig operation offers an extremely good prognosis.

References

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HELEN B. TAUSSIG, HILARY CRAWFORD, SALVATORE PELARGONIO and STELLA ZACHARIAUDAKIS

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