Long-Term Results of Aortic-Pulmonary Anastomosis for Tetralogy of Fallot
An Analysis of the First 100 Cases Eleven to Thirteen Years after Operation

By Milton H. Paul, M.D., Robert A. Miller, M.D., and Willis J. Potts, M.D.

The classical subclavian-pulmonary and aortic-pulmonary anastomoses were devised as palliative surgical maneuvers for the tetralogy of Fallot complex to augment pulmonary blood flow and to increase systemic arterial saturation. These shunt procedures, performed during the past 14 years, have provided reasonably good health to thousands of patients with severe cyanosis and physical limitation.

More recently, intracardiac surgical techniques supported by an extracorporeal pump-oxygenator circulation have provided a "corrective" repair for this lesion. Although a moderately high initial surgical mortality has persisted in many clinics, it seems apparent that this form of open intracardiac repair will become the treatment of choice in suitably selected patients.

The present report is concerned with the long-term results of one form of shunt procedure, the aortic-pulmonary anastomosis, as a surgical therapy for tetralogy of Fallot.

Clinical Material
Our study is concerned with the first 100 consecutive patients with a diagnosis of tetralogy of Fallot upon whom an aortic-pulmonary anastomosis could be performed. The diagnostic term, tetralogy of Fallot, cannot be too rigidly interpreted in respect to this group of patients, since the diagnosis at this early period (1946-1948) of our congenital heart surgery program was completely without assistance from angiocardiography or catheterization.

This initial group of patients, operated upon at The Children's Memorial Hospital in 1946-1948, was comprised in general of (a) older children in rather desperate condition who had somehow remained alive, and (b) a considerable number of infants and smaller children for whom aortic-pulmonary anastomosis was especially devised. Eleven patients were below 12 months of age, 20 patients between 12 and 36 months, 32 patients between 3 and 6 years of age, and 28 over 6 years of age. Severe cyanosis, extreme limitation in locomotion, and frequent squatting were the rule rather than the exception.

Persistence and good fortune have enabled us to obtain follow-up information in 1959 on 92 of the first 100 consecutive patients operated on in 1946-1947 (table 1). Three fourths of the surviving patients have had clinical, electrocardiographic, and x-ray evaluations in our own clinic. The others were evaluated on the basis of telephone interviews as well as written questionnaires and, whenever possible, recent chest x-rays, and reports of physical examination were provided by the family physician.

Operation
An aortic-pulmonary anastomosis was performed in each of these 100 patients as the initial surgical therapy.

The size of the anastomotic channel was carefully planned by the use of calipers to measure the length of the incisions in the aorta and pulmonary artery. At first, these incisions were made 8 mm. long and this provided an anastomotic channel 5 mm. in diameter. Marked postoperative enlargement in the heart of the fourth patient in this series (J. P. fig. 1) prompted shortening these incisions to 6.3 mm., which produced a channel diameter of 4 mm. We have continued to make the incision 6.3 mm. long in all children except infants, in whom they are made 5 mm. long, since in some instances considerable growth of the anastomotic channel can be anticipated.

To our knowledge, only three patients in this group have required a second operation to relieve recurrent cyanosis. The clinical

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Marked cardiac enlargement and congestive heart failure following surgical anastomosis with long-term poor (group III) clinical result and severe pulmonary artery hypertension. Preoperative (1-22-47), early postoperative (6-25-47), and long-term follow-up (8-14-58) x-rays.

Table 1

Analysis of the First 100 Consecutive Patients with a Diagnosis of Tetralogy of Fallot upon Whom an Aortic-Pulmonary Anastomosis Could Be Performed

<table>
<thead>
<tr>
<th>100 Consecutive aortic-pulmonary anastomoses for tetralogy of Fallot (1946-1949)</th>
<th>92 Cases followed (1946-1959)</th>
<th>8 Cases lost</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>73 Living contacts</td>
<td>19 Dead</td>
</tr>
<tr>
<td></td>
<td>34 Physiological studies</td>
<td>39 Clinical evaluation</td>
</tr>
</tbody>
</table>

details are presented in an earlier follow-up report. It should be noted here, however, that in two of these three cases the aortic-pulmonary anastomosis was performed on the right side in the presence of a right aortic arch, a procedure long abandoned because of technical difficulties.

Mortality
Ninety-two of the first 100 patients are accounted for in Table 1 and of this number.
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19 are known to be dead. Nine of the 100 patients subjected to an aortic-pulmonary anastomosis in this early period (1946-1948) died at operation or in the immediate postoperative period.

The over-all operative mortality through the years for this surgical procedure in approximately 700 patients has been 9 per cent. In patients under 3 years of age the operative mortality has persistently averaged 12 to 15 per cent, and in patients over 3 years of age it has been 3.8 to 4.0 per cent.

After leaving the hospital improved by surgery an additional 10 patients are known to have died in the period from 1946 to 1959. Table 2 briefly indicates the known causes of death in this group and the interval between surgery and death.

Clinical Evaluation

Our major interest in this study was to determine the clinical results in those 73 patients known to be alive 11 to 13 years after an aortic-pulmonary anastomosis was performed for tetralogy of Fallot. The evaluation of the postoperative status presented in table 3 is primarily based upon criteria suggested by Potts et al. and Taussig and Bauersfeld. These criteria are obviously not an accurate measure of the cardiovascular status of a given individual, since they are based solely upon the degree of relief of cyanosis and a subjective evaluation of general clinical improvement and increased exercise tolerance. Such important factors as the presence of marked cardiac enlargement, severe pulmonary hypertension, and aneurysmal dilatation of the pulmonary arteries are not taken into consideration. This evaluation, however, does represent the past and present status of the patient with regard to his ability to carry out day-to-day normal activities.

Group I patients are those who show little or no cyanosis at rest and slight cyanosis during exercise. They are able to attend public school, college, or work, and in general do what other children or young adults do with the exception of the more violent forms of activity. Many, however, will admit under careful questioning that they tire more easily than their schoolmates or friends. In this present report we have separated from group I, a IA group, which represents those individuals who can carry out rather extreme activity with what appears to be a normal degree of fatigue. These activities include manual farming, ballet dancing, skiing, and long-distance bicycling.

Figure 2

Normal heart size and excellent (group IA) clinical result 13 years after aortic-pulmonary anastomosis surgery. Preoperative (9-10-46), and long-term follow-up (9-14-59) x-rays.
It seems appropriate, although somewhat repetitious, to review briefly the clinical course of the first patient upon whom an aortic-pulmonary anastomosis was performed. 

Diane S., age 21 months, weighing 18 lbs., was admitted to The Children's Memorial Hospital September 9, 1946. She was deeply cyanotic, unable to stand, and, following the slightest exertion, even too rapid eating, became unconscious. Her red blood cell count was 10,300,000 per mm. \(^3\) Clinical and laboratory data confirmed the diagnosis of tetralogy of Fallot.

On Friday September 13, 1946, an aortic-pulmonary anastomosis was performed. The anastomotic channel theoretically was made 5 mm. in diameter, but actually must have been considerably smaller. During the first operation of this kind anxiety about possible hemorrhage or later leakage undoubtedly led to too snug sutures and to irregular placement of sutures.

She had a surprisingly uneventful postoperative recovery. Within a few months she gained considerable weight and learned to walk. Needless to say, this patient has been followed most intently during the past 13 years.

In figure 2 are shown roentgenograms of her heart immediately before operation, two weeks later, and 13 years after operation. A rather high pitched, continuous murmur has persisted unchanged throughout these years. This child, now a bright-eyed, attractive, 15-year-old girl, engages in all the regular and irregular activities common to this age group. During the summer of 1959, with one of her unhandicapped friends, she bicycled 30 miles in 1 day. Parental displeasure with this unauthorized escapade did not seem to lessen her pleasure in being able to demonstrate to herself and to her parents that she was capable of such strenuous activity. It is unfortunate that all patients subjected to shunt procedures cannot have equally good results.

Group-II patients may tire easily on moderate exercise. All show moderate to marked improvement over their condition before surgery; however, some have persistent, mild cyanosis at rest, and all have definite decreased exercise tolerance. In group III are classed those patients who are unable to perform normal daily activities without considerable fatigue.

Eleven to 13 years following the aortic-pulmonary anastomosis surgery two thirds of the patients can be classified (table 3) as having good or moderately good results (group IA, I). The earlier clinical evaluation (1954) gave a somewhat higher percentage of good results. Approximately one third of the patients are classified now as group II, fair results. In the poor-result group III are two patients: one who has severe pulmonary hypertension and pulmonary vascular obstruction and one who still has an inadequate pulmonary blood flow despite a second shunt procedure.

**Roentgenographic Findings**

Recent roentgenograms of the chest were available in 58 of the 73 contacted, living patients and cardiac enlargement was evaluated both by cardiothoracic ratio measure-
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Figure 4

Electrocardiographic evaluation in 13 patients with tetralogy of Fallot 11 to 13 years after aortic-pulmonary anastomosis.

Table 3
Clinical Evaluation in "Tetralogy of Fallot" Eleven to Thirteen Years after Aortic-Pulmonary Anastomosis

<table>
<thead>
<tr>
<th>Group</th>
<th>1959 (73 Patients)</th>
<th>1959 (73 Patients)</th>
<th>1954 (86 Patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group</td>
<td>Excellent</td>
<td>7</td>
<td>10%</td>
</tr>
<tr>
<td>Group</td>
<td>1 Good</td>
<td>42</td>
<td>58%</td>
</tr>
<tr>
<td>Group</td>
<td>II Fair</td>
<td>22</td>
<td>30%</td>
</tr>
<tr>
<td>Group</td>
<td>III Poor</td>
<td>2</td>
<td>2%</td>
</tr>
</tbody>
</table>

Table 4
X-ray Evaluation in Tetralogy of Fallot Eleven to Thirteen Years after Aortic-Pulmonary Anastomosis (Fifty-Eight Patients)

<table>
<thead>
<tr>
<th>Cardiac size</th>
<th>Patients</th>
<th>Pulmonary artery size</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>5</td>
<td>Normal or slight enlargement</td>
<td>14</td>
</tr>
<tr>
<td>Slight enlargement</td>
<td>17</td>
<td>1+</td>
<td>22</td>
</tr>
<tr>
<td>1+</td>
<td>28</td>
<td>2+</td>
<td>17</td>
</tr>
<tr>
<td>2+</td>
<td>8</td>
<td>3+</td>
<td>5</td>
</tr>
<tr>
<td>3+</td>
<td>0</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 5
Cardiac Catheterization Findings in Tetralogy of Fallot Eleven to Thirteen Years after Aortic-Pulmonary Anastomosis (Eighteen Patients)

<table>
<thead>
<tr>
<th>Pulmonary pressure Mn. Hg (mean)</th>
<th>Patients</th>
<th>Pulmonary flow L./min./M.²</th>
<th>Patients</th>
<th>Pulmonary resistance Units</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;20</td>
<td>10</td>
<td>4—6</td>
<td>8</td>
<td>&lt;3</td>
<td>15</td>
</tr>
<tr>
<td>20—30</td>
<td>5</td>
<td>6—8</td>
<td>3</td>
<td>3—5</td>
<td>1</td>
</tr>
<tr>
<td>30—50</td>
<td>3</td>
<td>8—14</td>
<td>7</td>
<td>&gt;5</td>
<td>2</td>
</tr>
</tbody>
</table>

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ments and clinical estimate (table 4). The majority of the patients had a slight to moderate cardiac enlargement after more than 10 years of a functioning aortic-pulmonary anastomosis. There were five patients with normal heart size (fig. 2), and none was found living at present with extreme cardiac enlargement.

Slight cardiac enlargement is anticipated in the early postoperative period if satisfactory clinical improvement is to occur. Although the heart size following any cardiac operation is undoubtedly an important factor in prognosis, there was not always a good correlation in this group with the clinical, symptomatic evaluation. One half of the patients with moderate (2+) cardiac enlargement had a good (group I, IA), long-term clinical result.

Dilatation and progressive enlargement of the main pulmonary artery branches were not uncommon and, if extreme, (fig. 3) represented a problem at the time of corrective surgery. Twenty-two patients demonstrated a considerable degree of pulmonary artery dilatation (2+, 3+).

**Electrocardiographic Findings**

In figure 4 the present electrocardiographic status in 45 patients is summarized. Although preoperative electrocardiograms with complete precordial chest leads are not available from the 1946-1948 period, it is reasonable to assume that almost all of these patients had the typical right ventricular hypertrophy pattern of tetralogy of Fallot.

One third of the patients studied have maintained this typical right ventricular hypertrophy pattern with tall R waves in the right precordial leads and deep reciprocal S waves over the left precordium. Combined ventricular hypertrophy is now present in two thirds of the patients, indicating that there has been a considerable increase in left ventricular work following the surgical anastomosis with its attendant increase in pulmonary blood flow. This is well illustrated in the lower electrocardiogram, which has tall R waves in the right precordial leads, V₄R and V₁, and also extremely tall, R waves in the left precordial leads, V₅ and V₆.

**Physiologic Studies**

A small, unselected group of 27 patients were studied by right heart catheterization. The pulmonary artery was entered in 18 patients, and the aorta in 21 patients. Since we were particularly interested in an assessment of the pulmonary pressure, blood flow, and vascular resistance, the data obtained in these 18 patients are summarized in table 5. The difficulty of obtaining adequate, representative blood sampling from the pulmonary artery in the presence of a large aortic-pulmonary anastomosis must be considered in evaluating some of the measurements of pulmonary blood flow and pulmonary vascular resistance. The majority of the pulmonary artery blood samples were obtained, however, from the nonanastomosed pulmonary artery branch.

After 11 to 13 years of a functioning aortic pulmonary anastomosis 10 patients had pulmonary artery mean pressures of less than

**Table 6**

<table>
<thead>
<tr>
<th>% Saturation</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>91-94</td>
<td>12</td>
</tr>
<tr>
<td>86-90</td>
<td>8</td>
</tr>
<tr>
<td>81-85</td>
<td>4</td>
</tr>
<tr>
<td>&lt;80</td>
<td>3</td>
</tr>
</tbody>
</table>
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20 mm. Hg and only three patients had grossly elevated mean pressures (45, 75, 85 mm. Hg). The pulmonary blood flow estimates indicate the wide range (4 to 14 L./min./M.²) present in these 18 patients. Finally there were 15 patients with normal pulmonary vascular resistance and three patients with estimated resistances (5.0, 15.8, and 18.0 units) greater than the normal level of approximately 3 units.

Table 6 illustrates that almost one half of this group had a resting arterial oxygen saturation at or above 90 per cent. The effect of mild exercise (3 minutes of continuous bicycling movements of the legs) in nine patients with resting arterial oxygen saturations above 90 per cent is illustrated in figure 5. The first seven patients were all classified in clinical group I (good) and all demonstrated a fall in oxygen saturation into the 80 to 90 per cent saturation range. The last two patients were in clinical group II (fair) and both demonstrated a more pronounced fall.

Discussion

The present long-term (11-13 year) follow-up study reinforces the findings presented in an earlier (6-8 year) report on the first 100 consecutive patients operated upon at The Children's Memorial Hospital for tetralogy of Fallot during 1946-1948. The data concerning operative mortality, over-all mortality, and incidence of good results from several other large, but not homogeneous series⁸,⁷ are in approximate agreement with our figures of 9, 19, and 70 per cent for these three categories.

Although it has become quite clear that open-heart resection of the infundibular obstruction and closure of the interventricular septal defect is the ideal procedure for tetralogy of Fallot, it seems equally clear, at this time, that an anastomatic operation remains the procedure of choice for the severely cyanotic infant, perhaps under the age of 2 or 3 years, who is subject to repeated syncopal attacks and needs immediate surgical relief. In this severely cyanotic young group, are the patients who present an unusually high surgical risk from open-heart corrective surgery. As yet undefined factors, such as marked aortic overriding, a small left ventricular chamber, and certainly pulmonary hypoplasia bordering on atresia, contribute to reported mortalities of about 50 per cent in this age group. Age-dependent difficulties are also reflected in the mortality statistics on shunt procedures, with the mortality ranging from 9 to 17 per cent below 3 years of age as opposed to 2.5 to 4.5 per cent in older patients.⁷ The long-term clinical results appear to be less age-dependent than the mortality rate; nevertheless, a higher percentage of good results was obtained in patients over 6 years of age (table 7).

A review of the long-term clinical course of these patients indicates that a considerable degree of stabilization resulted 1 to 2 years after the aortic-pulmonary anastomosis had been performed. It further appears that the long-term poor results were most likely related to the presence of too large an anastomosis, leading either to left ventricular over-
work and congestive heart failure or to severe pulmonary hypertension and probably progressive pulmonary vascular obstruction and a decreasing aortic-pulmonary shunt. A counterpart to these clinical implications of too large an anastomosis may be found in the experimental results of Heath et al., who found severe pulmonary vascular pathologic changes analogous to hypertensive pulmonary vascular disease in a dog who had an aortic-pulmonary anastomosis for nearly 4 years before necropsy.

Since it seems likely that the aortic-pulmonary anastomosis will, at least for the present, continue to serve as a valuable salvage operation for a small segment of patients with tetralogy of Fallot and for certain other complex cyanotic lesions (pulmonary atresia, tricuspid atresia, transposition of the great vessels with severe pulmonary stenosis), some further discussion is directed toward the factors leading to difficulties.

The critical size of an aortic-pulmonary anastomosis for optimum relief of cyanosis requires meticulous surgical technic. Even if the initial anastomotic channel is of ideal size, there remains the problem of growth of the anastomosis with the passage of time. Some preliminary experimental studies performed on rapidly growing pigs indicate that progressive enlargement of the anastomosis does occur; however, about half of the animals showed no growth in the orifice size.

It must further be observed that improvement in the arterial oxygen saturation above a conservative level is purchased only at the cost of a markedly increased pulmonary blood flow, an obviously large aortic-pulmonary anastomosis, and a markedly increased left ventricular work load. Born et al. have shown that, assuming a relatively constant oxygen consumption before and after operation, the resultant arterial oxygen saturation ($C_1$) is dependent upon the preoperative arterial oxygen saturation ($C_0$) and the increase in pulmonary blood flow $\frac{\alpha}{\beta}$ that surgery has provided. In figure 6 the curved line indicates that a preoperative arterial saturation of 70 per cent would be increased to approximately 87 to 90 per cent if the pulmonary blood flow were increased approximately threefold. The steeply ascending slope of the pulmonary blood flow curve in the region of 90 per cent arterial oxygen saturation indicates that any further significant increase in arterial saturation to the point of abolishing clinical cyanosis is derived from a markedly increased pulmonary blood flow and left ventricular work load.

In reviewing the long-term clinical results of the aortic-pulmonary anastomosis for tetralogy of Fallot it is important to consider the problems to be anticipated when corrective open-heart surgical repair is undertaken in such patients. At the present time the technical problems of isolating and "taking down" a large, functioning aortic-pulmonary anastomosis are considerably greater than those of a subclavian-pulmonary anastomosis. In two patients from our 1946-1948 group open-heart "corrective" surgery has been performed with good clinical results. Continuing experience with this reoperation problem, however, has indicated a high operative morbidity and mortality. Thus, in circumstances in which early shunt surgery is indicated and future open-heart surgery is anticipated, the subclavian-pulmonary anastomosis, if surgically possible, may be the preferable procedure.

Summary

The first 100 consecutive patients with a diagnosis of tetralogy of Fallot upon whom an aortic-pulmonary anastomosis could be performed were subjected to a clinical and physiologic evaluation 11 to 13 years after operation.
At this long-term follow-up 92 patients were traced. Nine patients died at the time of surgery and, in the period from 1946 to 1959, 10 patients died, most often from congestive heart failure. To our knowledge only three patients have required a second shunt operation to relieve recurrent cyanosis. The clinical results were considered good or excellent in 68 per cent of the survivors, fair in 30 per cent, and poor in 2 per cent.

The long-term poor results and mortality were usually associated with too large an initial anastomosis leading to either left ventricular overwork and congestive heart failure or severe pulmonary hypertension and progressive pulmonary vascular obstruction.

Right heart catheterization studies indicated that in a group of 18 patients in whom the pulmonary vascular bed could be evaluated, 15 patients had a normal pulmonary vascular resistance after 11 to 13 years of a clinically adequate aortic-pulmonary shunt.

The role of the shunt procedures as a valuable salvage operation at the present time for a select segment of patients with tetralogy of Fallot and certain other complex cyanotic lesions is reemphasized.

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