# Surgical Management of the Tetralogy of Fallot:

## Influence of a Previous Systemic-Pulmonary Anastomosis on the Results of Open Correction

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TWENTY years have passed since Blalock and Taussig described an operation for relief of cyanosis in patients with tetralogy of Fallot.[4] Thousands have benefited from this procedure which constitutes one of the most significant modern concepts introduced into surgery. The first correction of the tetralogy was performed by Scott in 1954 employing hypothermia,[9] and the following year Lillehei reported correction employing extracorporeal circulation.[6] Although the early mortality with open correction was appreciable, in the succeeding decade the risk associated with this procedure has decreased sharply. Experience has shown that total correction of the tetralogy performed in infants and small children is possible but is accompanied by greater technical problems, increased morbidity, and higher mortality. These are factors which have limited the use of open correction in this age group. For these reasons most observers recommend that a pulmonary-systemic anastomosis be created as the first procedure in infants or young children who require operation prior to the age of five years. It is for these patients, as well as the large number who already have such shunts, that emphasis is placed upon the importance of an assessment of the effects of a prior Blalock-Taussig anastomosis on subsequent surgical correction.

The present series of patients having open correction of tetralogy of Fallot is of interest because 71% had a previous shunt operation. The study is directed primarily to an evaluation of the effects of a previous systemic-pulmonary anastomosis on the results of ultimate total correction. As the shunt procedure was originally performed to relieve serious symptoms produced by the tetralogy of Fallot, usually in infancy or early childhood, this group is characterized by the most serious *anatomical* malformations.

### Materials and Methods

The patients in this report represent a consecutive series of all patients with total correction operated upon with a diagnosis of tetralogy of Fallot at the Johns Hopkins Hospital from January 1, 1962 to July 1, 1964. Ages ranged between 5 and 54 with the majority in the 12- to 20-year group. This series represents a continuation of the original study reported by Bahnson and associates.[3] Cardiac catheterization and cineangiography or selective angiocardiography was used to confirm the clinical diagnosis and to demonstrate anatomical defects. The majority of patients were followed from early childhood by the Harriet Lane Cardiac Clinic and 71% had previous shunt procedures. The shunt procedure had proven its effectiveness in maintaining

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these children for many years, and the majority entered the hospital for *elective* correction during vacation periods or following completion of high school.

A vertical screen modification of the original Gibbon-Mayo pump-oxygenator was employed. The pump was primed with 1,500 cc. of fresh heparinized blood and 2,500 cc. of dextrose 5% in 0.2% NaCl. The body temperature during perfusion was lowered to 32–33° with intermittent aortic occlusion during repair of the ventricular septal defect. The subclavian-pulmonary artery shunts were ligated after initiation of bypass, and this portion of the procedure rarely provided difficulty. On one occasion the subclavian artery was inadvertently entered on its posterior surface. The bleeding was controlled by the finger without interrupting the flow through the shunt to the lungs. Cardiopulmonary bypass was begun immediately and with temporary aortic occlusion, the perforation in the subclavian artery was easily controlled.

The ventriculotomy was made either transversely or longitudinally, depending upon anatomic factors, in the outflow tract of the right ventricle. If valvular pulmonic stenosis was present, it was relieved by commissural incisions. Every attempt was made to preserve a functioning pulmonic valve. A Teflon felt prosthesis was used for closure of the ventricular septal defect. If the pulmonic stenosis could not be relieved adequately by valvotomy and infundibulectomy, a pericardial patch was used to enlarge the outflow tract. If the pulmonary artery or valve annulus were small, the patch was extended to the bifurcation of the pulmonary artery. Relief of the pulmonic stenosis and reduction of right ventricular pressure were considered mandatory, and right ventricular-pulmonary artery gradient should be less than

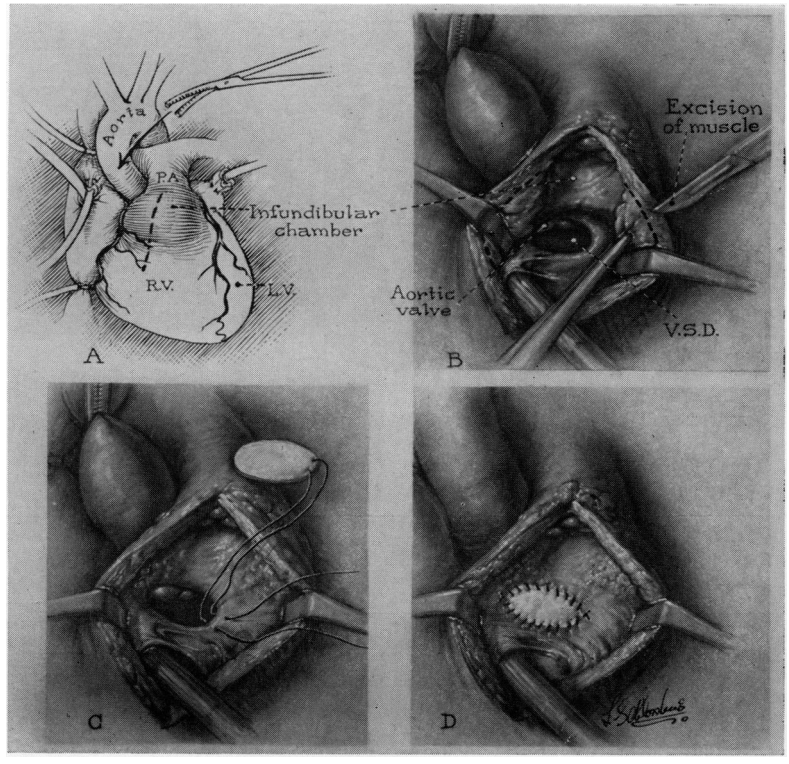


FIG. 1. Method for complete correction of tetralogy of Fallot. A) Cannulation of the superior and inferior venae cavae for extracorporeal circulation. The infundibular chamber of the right ventricle is shown. B) The outflow tract of the right ventricle is opened. The hypertrophied muscle characteristic of the infundibular stenosis is excised. The ventricular septal defect is shown and the proximity of the aortic valve may be seen. C) The plastic patch is being placed to close the ventricular septal defect. The aorta is temporarily occluded to provide a bloodless operative field. D) Completion of the placement of the prosthesis.

TABLE 1. *Open Operation for Correction of Tetralogy of Fallot*

| | Results Related to Age | | | | | |
|---|---|---|---|---|---|---|
| | 5–10 | 11–15 | 16–21 | 22–32 | Over 32 yrs. | Entire Group |
| Total | 15 | 28 | 37 | 12 | 6 | 98 |
| Survived | 13 | 24 | 34 | 11 | 4 | 86 |
| Mortality % | 13 | 14 | 8 | 8 | 33 | 12 |

50 mm. Hg. The steps in the operative technic are illustrated in Figure 1.

## Results

The statistics indicate a definite relationship of age to survival as shown in Table 1. Twenty of 98 patients were clinically acyanotic although all had right-to-left shunts as shown by decreased systemic arterial oxygen saturation with insignificant left-to-right shunts. Those classified as *severely* cyanotic had hematocrits of 70–90 and had *considerable* exercise intolerance. Survival as related to degree of cyanosis is shown in Table 2. Seventy-one per cent of the 98 patients had previous shunt operations (Table 3). The majority of the shunts were performed at least 10 years prior to total correction and had provided the patients an active childhood. Clinical histories following shunt procedures were similar in the majority. The patients did *well* for 3 to 5 years following the systemic-pulmonary anastomosis and were considered *satisfactory* results for an additional several years. After that time exercise intolerance, dyspnea, and cyanosis returned with greater prominence and was progressive. While the shunt was open as indicated by a continuous murmur and by patency on the angiocardiogram, the patient appeared to outgrow the anastomosis and symptoms of cyanotic heart disease returned.

TABLE 2. *Correction of Tetralogy of Fallot. Relationship of Severity of Cyanosis to Survival*

| | Survival | | Deaths | | |
|---|---|---|---|---|---|
| Cyanosis | Number | % | Number | % | Total |
| None | 20 | 100 | 0 | 0 | 20 |
| Moderate | 37 | 88 | 5 | 12 | 42 |
| Severe | 29 | 81 | 7 | 19 | 36 |
| Totals | 86 | 88 | 12 | 12 | 98 |

TABLE 3. *Correction of Tetralogy of Fallot. Patients with Previous Systemic-Pulmonary Anastomoses*

| | No Shunt | | Shunt | | |
|---|---|---|---|---|---|
| Cyanosis | Number | % | Number | % | Total Patients |
| None | 15 | 75 | 5 | 25 | 20 |
| Moderate | 10 | 24 | 32 | 76 | 42 |
| Severe | 3 | 8 | 33 | 92 | 36 |
| Totals | 28 | 29 | 70 | 71 | 98 |

Of interest is the observation that in most patients the subclavian artery was found to be smallest at the *site* of the anastomosis, a factor of undoubted significance in the return of symptoms (Fig. 2). The mortality was least in the group of patients having *one* previous shunt procedure, but was highest in the small number with *two* shunts (Table 4). Three of the patients with two previous shunts had an *end-to-end* anastomosis on one side. In these patients no attempt was made to reanastomose the "systemic lung" to the pulmonary artery. Thus, after total correction all of the venous return went through the lung with the previous *end-to-side* subclavian to pulmonary artery anastomosis.

In the 78 patients with clinical *cyanosis* a prosthesis was used to enlarge the right ventricular or pulmonary outflow tract in 51 (Table 5). The mortality in this group was less when a prosthesis was used, especially in those with *severe* cyanosis. The pulmonic valve was preserved if possible as a partially functioning valve has been

TABLE 4. *Correction of Tetralogy of Fallot. Mortality*

| | No Shunt | One Shunt | Two Shunts | Total |
|---|---|---|---|---|
| Patients | 28 | 65 | 5 | 98 |
| Deaths | 4 | 7 | 1 | 12 |
| Percentage | 14 | 11 | 20 | 12 |

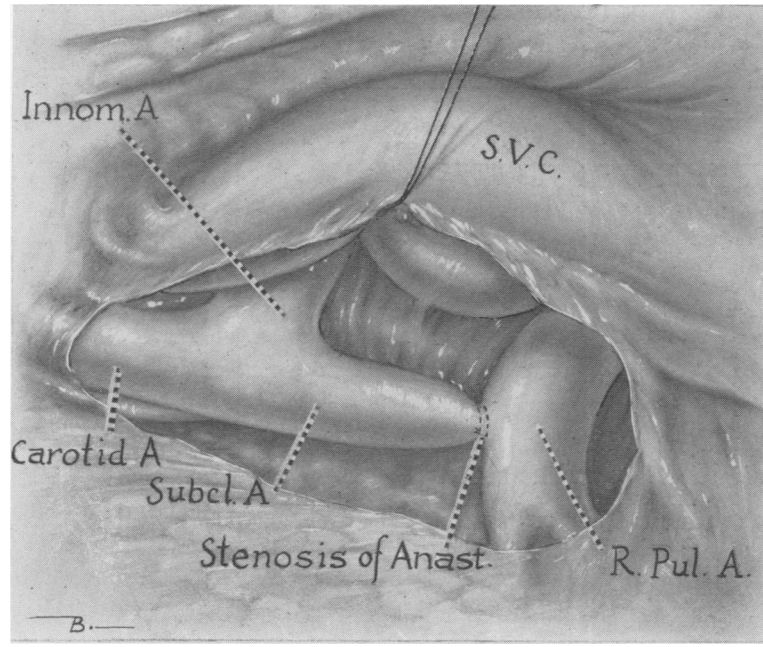


FIG. 2. Subclavian-pulmonary artery anastomosis illustrating stenosis at anastomotic site. The diameter of the anastomosis has failed to enlarge in proportion to the growth of the vessels.

shown experimentally to reduce the amount of regurgitation and in turn to decrease right ventricular work.[1] It is apparent that patients with a very small annulus or total pulmonary valve atresia are left with gross pulmonary insufficiency after a prosthetic patch is extended across the valve ring to the bifurcation of the pulmonary artery. The clinical courses of these patients with outflow patches extending out to the bifurcation of the pulmonary artery have shown minimal disability thus far. However, in a few patients right-sided failure has been observed.

The total hospital mortality was 12% and the causes of death are listed in Table 6. Hemorrhage after operation occurred in two patients, and one death was due to severe pulmonary hypertension. The most frequent cause of death was low cardiac output failure. Some of these patients had residual systolic gradients of 45–70 mm. Hg across the pulmonary valve and inadequate relief of pulmonary stenosis was probably responsible at least in part for the myocardial failure. Heart block occurred in three patients. One of these reverted spontaneously three days after operation; one expired with low output and myocardial failure; and the third subsequently required insertion of a permanent pacemaker. Two patients were thought to

TABLE 5. *Correction of Tetralogy of Fallot. Relation of Outflow Tract Prosthesis to Survival*

| | Moderate Cyanosis | | | Severe Cyanosis | | |
|---|---|---|---|---|---|---|
| | Number | Deaths | Per cent Survival | Number | Deaths | Per cent Survival |
| Excision & valvotomy | 15 | 2 | 87 | 12 | 4 | 67 |
| Outflow tract patch | 17 | 1 | 94 | 14 | 1 | 93 |
| Outflow patch across Pulmonary valve | 10 | 2 | 80 | 10 | 2 | 80 |
| Totals | 42 | 5 | 88 | 36 | 7 | 81 |

TABLE 6. *Total Correction of Tetralogy of Fallot Jan. 1962–June 1964 inclusive: Hospital Deaths*

| Cause of Death | Number of Cases |
|---|---|
| Low output cardiac failure | 5 |
| Post-operative bleeding | 2 |
| Pulmonary hypertension | 1 |
| Glomerulonephritis (one month after operation) | 1 |
| Myocardial failure (adequate anatomic correction) | 2 |
| Massive hemoptysis (etiology unknown) | 1 |
| Total | 12 |

have an open ventricular septal defect and one required reoperation. Assisted respiration through the endotracheal tube was utilized until the patient was fully awake and able to breath adequately with the usual time of ventilatory support being 3 to 4 hours. If the patient did not breath adequately after this period, the endotracheal tube was left in place and assisted ventilation continued for periods up to 36 hours. If mechanical ventilation was clearly required after this time tracheostomy was performed.

## Discussion

Total correction of tetralogy of Fallot is now performed frequently and with an acceptable mortality. The present problem concerns the age of the patient at the time of open correction in relation to the ultimate result. It has been shown in several series that a higher mortality can be expected with total correction in the small cyanotic child whereas a shunt procedure can be performed in the same group with a low operative mortality. The question is whether the shunt produces changes in the heart and lungs which are beneficial and which reduce mortality at the time of subsequent correction.

The decrease in mortality during the past several years associated with open correction of the tetralogy of Fallot has been impressive. In recent series, Kirklin [5] and Malm [7] each report a mortality of 7%, and Bahnson [2] has described a consecutive group of 60 patients with 2 deaths (3%). The reason for the decreased mortality appears to be a combination of factors. Prevention of postoperative bleeding, decreased use of outflow tract prostheses (especially across the pulmonary valve ring), improved postoperative care, and prevention of air emboli have been emphasized. Severe anatomic malformations, including aberrant coronary vessels and complete obstruction of the outflow tract of the right ventricle continue to represent difficult problems.[8]

The degree to which a shunt aids in preparation of the pulmonary vasculature for subsequent total correction is difficult to evaluate. Fortunately, in patients with subclavian to pulmonary artery shunts and total correction, there has been no significant problem with elevation of pressure in the pulmonary artery. This would indicate that the lungs can accept the total cardiac output without difficulty and that the systemic shunt does not produce permanent changes in the pulmonary arterioles. In fact, the development of pulmonary hypertension following a subclavian to pulmonary artery anastomosis has rarely been observed. This probably is related to the failure of the shunt to enlarge proportionately as the patient grows (Fig. 1). Pulmonary hypertension has developed following an aortic-pulmonary artery anastomosis (Potts) as the size of this opening may enlarge significantly with growth of the patient.

The absence of left-sided failure after total correction in patients with previous shunt operations substantiates the theory that the shunt exerts a favorable influence on the left ventricle. The angiocardiograms in the patients having had previous shunt procedures have shown a *normal* or *large* left ventricular cavity. Some consider the finding of a small and underdeveloped left ventricle as demonstrated by angiocardi-

ography to be an indication for a shunt procedure rather than total correction. Under such circumstances it is hoped that increased blood flow returning to the left heart will produce enlargement. Experience has shown that an underdeveloped left ventricle is often present in tetralogy of Fallot and that total correction in such patients may be associated with low cardiac output and pulmonary edema. There is now clear evidence that relief of pulmonic stenosis is important in reducing the incidence of *right-sided* failure and the subsequent low cardiac-output syndrome. Reconstruction of the outflow tract of the right ventricle alone with a prosthesis has rarely resulted in serious complications. Extending the prosthesis across the annulus results in pulmonary insufficiency which is more likely to produce complications and every attempt should be made to avoid this or to reduce the insufficiency to a minimum. However, if the patch is not of adequate size and a significant residual gradient results, right-sided failure with subsequent low cardiac output is associated with a poor prognosis and appreciable increase in operating mortality.

The majority of the patients in this series having a previous shunt operation had total correction between the ages of 12 and 20. Postoperative hemorrhage is uncommon at this age and technical aspects of performing the repair seemed most favorable in this group. Closure of the previous subclavian-to-pulmonary artery anastomosis is not difficult and has not increased the operative mortality of cases. The patients with previous shunts were more cyanotic and had more complicated anatomical abnormalities than those without earlier operation. Yet, the hospital mortality was less in the patients having previous shunt procedures. It would appear that a previous shunt does not adversely affect the results of open correction and may prepare the left ventricle and the pulmonary circulation for a better response to the subsequent total correction operation. Therefore, these results suggest that subclavian-pulmonary shunts represent the procedure of choice for those children requiring repair prior to the age of five years.

## Summary

The results of total correction of tetralogy of Fallot in a consecutive group of patients are presented. A reduction in mortality and improved technical results have characterized the recent surgical experience. The major cause of death was low cardiac output and could be related in some instances to inadequate relief of pulmonary stenosis. Use of a prosthesis for repair of the outflow tract did not increase operative mortality. The over-all survival rate was 88%. A majority of the patients had previous shunt operations for relief of cyanosis, and there was a lower mortality in this group. These data suggest that a previous shunt procedure may exert a favorable influence on subsequent correction, and such a shunt appears indicated in infants and small children who require operation for severe symptoms under the age of five. The mortality in this series was 12% and was affected by the extremes of age, the severity of cyanosis, and the more extreme malformations such as complete outflow obstruction of the right ventricle. The age group 15 to 32 years, comprising half the group, had the most favorable results with a mortality of 8%.

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## Discussion

Dr. Philip R. Allison (Oxford, England): Dr. Mahorner, gentlemen. I think that Dr. Sabiston has been very helpful in dividing the patients with Fallot's tetralogy into groups of deeply cyanosed, moderately cyanosed and not cyanosed. Those of us who have seen a fair number of these patients are really almost beginning to think that Fallot's tetralogy shouldn't be grouped under this one heading. Almost every patient we see differs from the other. This is reflected in different mortality rates.

I was interested that Dr. Sabiston found the stenosis in the subclavian artery at the point at which it joins the pulmonary artery after a Blalock operation. We found the same thing. And in one or two we were able to examine more closely, we found marked intimal thickening at this point. It may be that this is not a matter of an anastomosis that fails to grow with the child, but an anastomosis which does in fact close off by thrombus formation and endothelialization.

The other thing that came out from reading this manuscript was that in the patients in whom an extensive outflow patch had to be inserted from the right ventricle through the pulmonary artery there was an appreciable morbidity and mortality over and above the others. I think this does lend some weight to the fact that complete pulmonary incompetence is being recognized more and more as a rather serious situation. In the early days we tended to rationalize and say that pulmonary incompetence was of no great significance.

The third thing I wonder if Dr. Sabiston would consider worth deducing from his results is that insofar as the operative mortality is increased in the deeply cyanosed patients (either those primarily deeply cyanosed or those becoming so through closing off of the Blalock shunt) would this not be an indication for total correction at the earliest possible stage of deterioration in those patients who have had a shunt. Thank you.

Dr. Grady Hallman (Houston): I would like to compliment Dr. Sabiston and his colleagues on their excellent presentation and to show you some of the means we have utilized at the Texas Children's Hospital for classifying patients with tetralogy of Fallot and how this has influenced our plan of management.

(Slide.) The first slide summarizes the classification in the series over a 9-year period ending last year and shows that of a total of 319, the vast majority were so-called cyanotic or typical tetralogy of Fallot patients with peripheral arterial desaturation and all the hallmarks of the disease.

We divided these further into those who had had a previous shunt and those who underwent total correction as a primary procedure. Note that in line with Dr. Sabiston's findings, there is not much difference between these two groups and that the average mortality is about 17%. Only 54 of the 319 patients had the so-called acyanotic type of tetralogy. These patients had all the anatomic features of tetralogy but with less severe pulmonary stenosis, so that there was either a left to right shunt or a balanced shunt and not much desaturation of the peripheral arterial blood. The mortality in this group was only 3.7%, modifying the total overall mortality so that it is 15%.

(Slide.) One can utilize various means for further subdividing those patients with cyanotic tetralogy. One of these is to classify them according to the degree of aortic override. A moderate degree is shown here. There is a progressive increase in the degree to which the aorta is shifted toward the right ventricle in the other patients illustrated. This modifies the repair in that the placement of the patch must be in an oblique fashion. In a patient with severe aortic override, when the patch