Corrective Surgery for Tetralogy of Fallot

Evaluation of Results

By Earle B. Kay, M.D., Cid Nogueira, M.D., David Mendelsohn, Jr., M.D., and Henry A. Zimmerman, M.D.

A SUFFICIENT NUMBER of patients with tetralogy of Fallot have now had definitive correction by open-heart surgery, made possible by the use of the pump-oxygenator and extracorporeal circulation, to make a comparative analysis of the results attained between open definitive correction of the defects and the results of the earlier bypass shunt procedures of the Blalock-Taussig 1 or Potts 2, and the pulmonary valvulotomy or infundibulectomy technic initiated by Brock 3, and perhaps to delineate the current role of these available procedures to the present care of patients with this condition.

The increased pulmonary blood flow afforded by systemic-pulmonary artery anastomosis and infundibulectomy was a dramatic development in the palliation of these otherwise hopeless children. Follow-up studies 4-6 of the patients so treated have demonstrated in ensuing years an increasing diminution of what appeared to be good palliation during the first several years following surgery from late complications such as bacterial endocarditis, cerebral abscesses, heart failure, and thrombosis of the anastomosis. Later morbidity or death has been due to the patient outgrowing the anastomosis with recurrence of the original disability; or in some instances due to the development of pulmonary hypertension caused by further increase in the size of the anastomosis with growth.

Pulmonary valvulotomy or infundibular resection represented a further step forward in a more objective treatment of tetralogy of Fallot. According to Campbell and Deucher 7 however, the late results of the direct and indirect methods were very similar. Loss of improvement following valvulotomy or infundibular resection was due either to recurrence of the obstruction in the right ventricular outflow tract or to the development of a large left-to-right shunt created by the operation because of the inability with this technic to correct associated ventricular septal defect.

The development of safe extracorporeal technics, the advances in intracardiac corrective surgery, and the use of cardioplegia have made possible the complete repair of tetralogy of Fallot 8, 9 In view of the conflicting results reported with the palliative procedures, and the scarcity of reports in the literature of postoperative hemodynamic evaluation in patients submitted to a corrective operation by the open technic, this report appears to be pertinent.

The criteria for postoperative evaluation of benefit gained in the past have for the most part been the clinical condition of the patient, the status of cyanosis, the degree of polycythemia, and the hemoglobin level. The evaluation of the late results attained by the open corrective method for tetralogy of Fallot should also include hemodynamic improvement as demonstrated by cardiac catheterization studies in addition to the clinical evaluation.

Series

This report concerns the repair of tetralogy of Fallot in a series of 50 consecutive patients. The method of correction is described and the results are analyzed. Complete evaluation including cardiac catheterization studies of a group of patients operated upon 1 to 5 years previously is presented.
TETRALOGY OF FALLOT

In all patients the diagnosis of tetralogy of Fallot was clinically established and complemented by laboratory studies. All patients were cyanotic. Polycythemia and high hemoglobin levels were always present. Clubbing was present in all patients but the two very young ones. The age varied from 12 months to 42 years, and the majority of the patients were in the younger age group. Four patients had had previous Blalock operations. In three of them the anastomosis was patent. One patient had had a Brock procedure with a good initial result but with gradual recurrence of symptoms 3 years after surgery.

Preoperative Evaluation

Although the diagnosis of tetralogy of Fallot can be established clinically in the majority of instances, a more objective preoperative evaluation is indispensable in order to gain more information about the hemodynamic alterations and to provide details about the anatomic variations of the defects. Electrocardiographic and roentgenologic examinations, as well as catheterization studies were performed in each patient in this series. For the past several years cineangiographic studies have been done routinely. This is the most objective method of obtaining reliable information preoperatively concerning the anatomic condition of the pulmonary outflow tract. The visualization of the ventricular septum and the ventricular septal defect aided in establishing the differential diagnosis between tetralogy of Fallot and patients with a single ventricle and pulmonary stenosis. It further provided information relative to atresia of the pulmonary outflow tract and pulmonary artery. It aided in evaluating the efficacy of corrective surgery when associated with marked pulmonary atresia in instances of pseudotruncus arteriosus.

Contrary to the policy of others, small infants (less than 20 lb. of body weight) have been followed by a careful medical supportive management. The risk of any kind of surgery in these patients has been great and does not, in our minds, justify surgical intervention before these children grow larger. These cyanotic infants in the past have been frequently hospitalized for acute supportive therapy during the first year of life. During the past 5 years only one infant died during this first year of life under this policy. All others survived until the time they became better risks for surgical treatment. The performance of shunt operations in this group of small infants has not been satisfactory due to the high operative mortality and incidence of thrombosis of the anastomosis.

Operative Approach

A bilateral thoracotomy was used initially. During the last 2 years a longitudinal sternotomy has been employed. Postoperatively, pain and pulmonary complications were less with the sternal-splitting incision. The pleural spaces were kept closed until the end of surgery, avoiding manipulation and trauma of the lungs. At the end of surgery the right pleural space was opened to provide thoracotomy drainage of the mediastinal structures.

Extracorporeal circulation for all patients was provided with a Kay-Cross pump oxygenator. One of the superficial femoral arteries was cannulated for perfusion. The management of these patients and the physiologic controls of the perfusion have been previously reported. Heparin (1 mg./lb.) was given prior to cannulation. With the patient on total heart-lung bypass, elective cardiac arrest was induced, the cardiotomy was performed, and the defects were repaired under direct vision. In the three patients in whom a previous Blalock anastomosis was still patent, the shunt was interrupted at the moment the bypass was started.

Elective cardiac arrest was employed to obtain a bloodless, motionless field, which afforded perfect identification and repair of the ventricular septal defect. In 42 patients potassium chloride was the cardioplegic drug. In all instances cardiac rhythmicity was restored without difficulty. In the past year, however, hypothermic cardiac arrest has been used because it afforded greater myocardial protection without the supposed deleterious effects of potassium arrest. Frequently, pericardial irrigation with the same solution was used to supplement this method for continued
hypothermia. Intracardiac repair of tetralogy of Fallot is a time-consuming procedure (45 to 60 minutes) and extra protection to the myocardium was gained by this means of applying hypothermia.

The cardiomyotomy was made in the outflow tract of the right ventricle and carefully extended to permit good exposure of the intracardiac structures. Anomalous branches of the coronary arteries may cross this area that should not be sacrificed without proper evaluation. The infundibulum and excess muscle mass obstructing the ventricular outflow tract was resected under direct vision and a pulmonary valvulotomy was performed when necessary.

The ventricular septal defect was accurately identified and a small sump was introduced into the left ventricle, via the septal defect, in order to maintain a bloodless field. This same sump was used for decompression of the left ventricle during the recovery phase of the elective cardiac arrest.

Individual 4-0 silk sutures were placed at the edges of the septal defect and used for suturing an Ivalon® patch. Special care was exercised in order to avoid inclusion of elements of the conductive system. The careful placement of interrupted sutures and the use of a patch in the repair of the defect were essential factors in the prevention of operative heart block. Atrioventricular block associated with closure of the septal defect has never occurred. Disturbances in the right ventricular conduction system have been frequent after repair of tetralogy of Fallot, and have been attributed to extensive resection of the infundibulum. In tetralogy of Fallot, the ventricular septal defect was generally due to a complete absence of the membranous septum. In our experience, simple suture closure used in a few instances has not been as satisfactory a method of closure. The use of an Ivalon patch insured a permanent closure of the septal defect without tension, and the interrupted sutures used for its fixation avoided damage to the conduction system. Before the ventricular septal defect was completely closed, the aortic clamp was released and the coronary flow was re-established. If ventricular fibrillation occurred and did not revert spontaneously, electric defibrillation (60 to 100 volts at 0.1 second) was used.

During elective hypothermic cardiac arrest, the temperature of the myocardium was monitored by a Yellow Springs needle thermistor. Electric defibrillation was attempted only after the myocardial temperature reached 32 C. When a good heart beat was re-established, the sump was removed from the left ventricle and the remaining sutures were tied.

In spite of extensive infundibular resection, the majority of the patients (70 per cent) had an elliptical patch of heavily compressed Ivalon (10 to 1 mm.) sutured to the edges of the cardiomyotomy in the ventricular outflow tract to increase further the size of this chamber and to avoid residual obstruction (fig. 1). Upon occasions it has been necessary to extend this plastic graft for varying distances across the pulmonary valve and into the pulmonary conus in order to obtain a satisfactory right ventricular pulmonary artery pathway. The correction of the pulmonary stenosis was a vital phase of the repair of tetralogy of Fallot. Complete closure of the septal defect, without assuring an adequate ventricular pulmonary artery outflow tract, precipitates right ventricular failure in the immediate

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*Commercial brand of polyvinol sponge.


Table 1

Hemodynamic Studies before and after Surgery Demonstrating the Benefit Obtained by Open Correction of Tetralogy of Fallot

<table>
<thead>
<tr>
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</tr>
</thead>
<tbody>
<tr>
<td>R.F.</td>
<td>20</td>
<td>128/16</td>
<td>75/-3</td>
<td>—</td>
<td>40/12</td>
<td>69.4</td>
<td>96.5</td>
<td>Small L-to-R shunt</td>
<td>Previous Blalock (thrombosed)</td>
</tr>
<tr>
<td>G.S.</td>
<td>6</td>
<td>110/-10</td>
<td>27/1</td>
<td>—</td>
<td>26/8</td>
<td>48.5</td>
<td>96.7</td>
<td>No shunt</td>
<td>Previous Brock</td>
</tr>
<tr>
<td>K.H.</td>
<td>6</td>
<td>77/8</td>
<td>45/0</td>
<td>23/10</td>
<td>36/9</td>
<td>75.6</td>
<td>94.9</td>
<td>No shunt</td>
<td></td>
</tr>
<tr>
<td>J.E.</td>
<td>6</td>
<td>135/-10</td>
<td>38/0</td>
<td>15/3</td>
<td>32/10</td>
<td>32.4</td>
<td>93.4</td>
<td>No shunt</td>
<td></td>
</tr>
<tr>
<td>L.McK.</td>
<td>3</td>
<td>102/-7</td>
<td>35/-2</td>
<td>—</td>
<td>17/7</td>
<td>79.2</td>
<td>93.3</td>
<td>No shunt</td>
<td></td>
</tr>
<tr>
<td>R.R.</td>
<td>19</td>
<td>129/-2</td>
<td>65/0</td>
<td>—</td>
<td>50/12</td>
<td>72.3</td>
<td>92.4</td>
<td>L-to-R shunt</td>
<td></td>
</tr>
<tr>
<td>D.L.</td>
<td>6</td>
<td>126/-3</td>
<td>28/0</td>
<td>8/3</td>
<td>26/9</td>
<td>76.5</td>
<td>94</td>
<td>No shunt</td>
<td></td>
</tr>
<tr>
<td>K.H.</td>
<td>12</td>
<td>85/11</td>
<td>40/-2</td>
<td>15/4</td>
<td>20/6</td>
<td>81.6</td>
<td>96.7</td>
<td>No shunt</td>
<td>Previous Blalock (open)</td>
</tr>
<tr>
<td>J.M.</td>
<td>14</td>
<td>112/-5</td>
<td>35/0</td>
<td>—</td>
<td>25/9</td>
<td>77</td>
<td>96</td>
<td>No shunt</td>
<td>Previous Blalock (open)</td>
</tr>
<tr>
<td>B.R.</td>
<td>7</td>
<td>110/-1</td>
<td>30/0</td>
<td>—</td>
<td>27/8</td>
<td>81</td>
<td>95</td>
<td>No shunt</td>
<td>Previous Blalock (open)</td>
</tr>
<tr>
<td>H.C.</td>
<td>6</td>
<td>110/-5</td>
<td>40/0</td>
<td>—</td>
<td>26/9</td>
<td>52.3</td>
<td>93.4</td>
<td>No shunt</td>
<td></td>
</tr>
<tr>
<td>N.E.</td>
<td>12</td>
<td>105/0</td>
<td>31/0</td>
<td>15/6</td>
<td>24/9</td>
<td>84</td>
<td>97.4</td>
<td>No shunt</td>
<td></td>
</tr>
<tr>
<td>M.F.</td>
<td>4</td>
<td>125/0</td>
<td>22/0</td>
<td>—</td>
<td>21/8</td>
<td>57.3</td>
<td>99</td>
<td>No shunt</td>
<td></td>
</tr>
<tr>
<td>K.T.</td>
<td>6</td>
<td>112/0</td>
<td>28/0</td>
<td>32/10</td>
<td>26/7</td>
<td>81</td>
<td>96</td>
<td>No shunt</td>
<td></td>
</tr>
<tr>
<td>R.P.</td>
<td>8</td>
<td>110/-2</td>
<td>32/0</td>
<td>—</td>
<td>30/10</td>
<td>80.7</td>
<td>95</td>
<td>No shunt</td>
<td></td>
</tr>
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</table>

Postoperative period. This was the main cause of failure initially when grafting of the outflow tract was not used.

After the definitive surgery was accomplished and the heart was able to take over the circulation, protamine was given (1 mg./lb.) and the cannulae were removed. Careful hemostasis was necessary. It is our impression that cyanotic patients have a higher bleeding tendency than acyanotic ones. Often it was necessary to give these patients larger doses of protamine and vitamin K1 to re-establish the clotting mechanism.

Pressures were routinely taken in the right ventricle and pulmonary artery before and after the bypass in order to evaluate the repair of the pulmonary stenosis. Postoperatively special care was taken to maintain a normal blood volume by replacing thoracotomy drainage with fresh whole blood. In addition, five per cent glucose was given during the first 48 hours (500 ml./M.2 of body surface). The patients were placed in an Eliot hood, where a high concentration of oxygen (75 to 85 per cent) was maintained during the first 24 hours. The airway was kept clean, and tracheal aspiration was performed with a laryngoscope as indicated. Tracheotomy was employed only once early in this series.

Results of Corrective Surgery

The over-all 5-year mortality was 18 per cent. During the last 2 years, 20 patients have had complete correction of tetralogy of Fallot with an operative mortality of 15 per cent. The higher mortality rate observed during the first 21/2 years was inherent in the developmental phases of extracorporeal circulation and surgical techniques for repair of this complex malformation. Forty-one of the fifty patients are alive and well. Of the nine operative deaths, those occurring early in the series were for the most part due to poor correction of the infundibular stenosis in the presence of a complete closure of the septal defect. Attempts to correct hearts with too atretic pul-
monary arteries were the next frequent cause of failure. Three patients, also among the first 25 cases operated upon, died in consequence of anuria secondary to high hemolysis during perfusion. With added experience and improvement in perfusion technics anuria is no longer a complication. A 12-month-old baby succumbed on the third postoperative day as a result of a subdural hematoma. Only one patient died in the late postoperative period. This was a 20-year-old patient who died 2½ years postoperatively after a short hospitalization for bacterial endocarditis. During the previous 2½ years he was able to attend school and carry on normal physical activities. Postmortem examination revealed that the ventricular septal defect was not completely closed and there was evidence of subacute bacterial endocarditis involving the tricuspid valve. The Ivalon grafts in the right ventricular outflow tract and ventricular septum were well incorporated, pliable, and covered by endothelium. There was no evidence of calcification or aneurysmal dilatation of the grafts.

The remaining 41 patients with tetralogy of Fallot operated upon during this 5-year period are alive, asymptomatic, and have normal physical activity. The cyanosis and polycythemia have disappeared and the hemoglobin levels are normal. Postoperative roentgenologic studies revealed normal-sized hearts and normal pulmonary vasculature. Complete postoperative evaluation including hemodynamic studies have now been performed in 15 of these patients from 1½ to 3 years postoperatively. The pertinent data are reported in table 1.

An analysis of these 15 patients demonstrates that in 13 of them a complete hemodynamic correction was accomplished. In two patients, a small left-to-right shunt still existed. One of these patients (R. R.) although clinically well for 2½ years, developed subacute bacterial endocarditis and died as noted above. This was the only late death in the series. The other patient (R.F.) with a residual left-to-right shunt also has a gradient between right ventricle and pulmonary artery (33 mm. Hg). He has had excellent clinical improvement, is asymptomatic, and pursues normal activity, over 3 years following surgery. The arterial saturation has increased from 69.4 to 96.5 per cent.

Conclusion

Definitive surgical correction of tetralogy of Fallot by the open technic has not been accompanied by a higher operative mortality than that associated with the shunt operations. As a result of the surgical technics being directed at the correction of the defects, greater improvement approximating normal is attained.

Summary

Fifty patients with tetralogy of Fallot have had surgical correction of their complex defects made possible by the open technic during the past 5 years. The overall 5-year operative mortality was 18 per cent. This was reduced to 15 per cent during the past 2 years in 20 patients. Forty-one of the patients are alive, asymptomatic, and have normal physical activity. Fifteen patients have had cardiac evaluation studies including cardiac catheterization from 1½ to 3 years postoperatively. Thirteen patients had normal cardiac hemodynamics. In only two patients was there evidence of incomplete hemodynamic improvement even though marked clinical improvement was gained.

References

4. Lillehei, C. W.: In discussion of Kirklin et al. 5.


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**On Permanent Patency of the Mouth of the Aorta, or Inadequacy of the Aortic Valves**

*By Dominic John Corrigan, M.D.*

One of the Physicians to the Charitable Infirmary, Jervis Street, Dublin; Lecturer on the Theory and Practice of Medicine; Consulting Physician to St. Patrick's College, Maynooth

The morbid affections of the valves and aorta permitting this regurgitation are the following.

1st, The valves may be absorbed in patches, and thus become reticulated and present holes, through which the blood flows back into the ventricle...

2nd, One or more of the valves may be ruptured; the ruptured valves, when pressed, flapping back into the ventricle instead of catching and supporting the column of blood in the aorta, the blood then regurgitating through the space left by the broken valves...

3rd, The valves may be tightened or curled in against the sides of the aorta, so that they cannot spread across its mouth; and an opening is then left between the valves, in the centre of the vessel, through which the blood flows freely back into the ventricle...

4th, The valves without any proper organic lesion may be rendered inadequate to their function by dilatation of the mouth of the aorta. The aorta, affected by aneurism, or dilated, as it frequently is in elderly persons, about its arch, will sometimes have the dilatation extending to the mouth of the vessel, and in such a case, the valves become inadequate to their function, not from any disease in themselves, but from the mouth of the aorta dilating to such a diameter, as to render the valves unable to meet in its centre; the blood then, as in the other instances, regurgitates freely into the ventricle.
Corrective Surgery for Tetralogy of Fallot: Evaluation of Results
EARLE B. KAY, CID Nogueira, DAVID MENDELSOHN, JR. and HENRY A. ZIMMERMAN

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