An Evaluation of Total Correction of Tetralogy of Fallot

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Total correction of tetralogy of Fallot was first carried out by Lillehei and associates in 1954 and was presented as the treatment of choice in unselected symptomatic patients in preference to the available palliative procedures of systemic-to-pulmonary arterial anastomosis and pulmonic valvulotomy. Open-heart repair of this malformation, however, has been associated with a high mortality, from 16 to 35 per cent, as compared with the present low mortality (2 to 9 per cent) for shunting procedures.

The experience with total correction of this anomaly at the Presbyterian Hospital from 1960 to 1962 will be reported, with emphasis on the decrease in mortality rate associated with certain details in surgical management. Forty-one consecutive patients have undergone total correction without operative mortality. The postoperative results have been evaluated by clinical and x-ray studies; cardiac catheterization and selective angiography have been performed in 25 patients to obtain an objective analysis of the repair.

Material and Methods

Forty-one consecutive patients with tetralogy of Fallot underwent corrective operation from June, 1960 through July, 1962.

Prior to June, 1960, 11 patients were operated upon by means of a bubble-type pump oxygenator, as originally described by DeWall et al. There were seven operative deaths in this initial experience, resulting in a complete reappraisal of the perfusion and operative techniques.

Preoperative Assessment

The patients ranged in age from 4½ to 27 years, with an average age of 12 years at the time of operation.

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Diagnosis

The diagnosis of tetralogy of Fallot was established preoperatively in each of the 41 patients by selective biplane angiography, and was supported by right-heart catheterization in two-thirds of them. All patients had evidence of a ventricular septal defect, right ventricular outflow tract obstruction with right ventricular systolic pressure at least 75 per cent of systemic, and overriding of the aorta to a variable degree. Thirty-four patients exhibited cyanosis with arterial oxygen saturations ranging from 65 to 92 per cent at rest and hematoctrit levels from 45 to 80 per cent. One patient (E.E.) had an associated thromboctopenia with platelet levels of 10,000 to 60,000/mm³ which rose to normal following operation. Symptomatology among the cyanotic patients indicated that 19 were severely disabled with marked limitation of exercise and had a history of squatting and frequently of syncope, 11 were moderately disabled, and 4 were not restricted in ordinary activity. The seven patients who had no clinical or laboratory evidence of arterial oxygen desaturation at rest usually gave a history of cyanosis with exertion, and all had the anatomical and physiological criteria for tetralogy of Fallot described herein.

Previous Operation

Twenty-three of the cyanotic patients had undergone a total of 30 palliative operative procedures. Among the six patients who had more than one previous operation, three had bilateral Blalock anastomoses, two had one Blalock and a Broek procedure, and one had bilateral Blalock anastomoses and a Broek procedure. Of the 19 patients who were considered clinically to be severely disabled, 15 were in the group who had previous palliative procedures.

Operative Technique

Cardiopulmonary bypass has been standardized during the past two years utilizing a disk oxygenator primed with heparinized blood and one unit of low molecular weight dextran. Flow rates between 2,000 and 2,400 cc./M²/min. and hypothermia to 28 C. were employed. Perfusion time averaged 90 minutes, but occasionally extended to 150 minutes. A median sternotomy incision was used in all patients and often required considerable attention to secure adequate hemo-

*Rheomacrodex.
stasis. A vertical right ventriculotomy incision was preferred to expose the outflow tract, with a test period of occlusion of any large coronary arteries crossing this region. This incision not only gives an adequate exposure of the ventricular septal defect, but is the only incision which allows adequate resection and reconstruction of the outflow tract. The use of this incision alone has not caused serious postoperative cardiac failure and allows placement of an outflow patch when necessary, in contrast with the horizontal incision suggested by March et al.9

A Teflon felt patch secured by a series of mattress sutures about the defect has been used in all but two patients. Anoxic cardiac arrest at 28 C. has been used during placement of these sutures. Associated muscular ventricular septal defects and atrial septal defects have been repaired by direct suture. The resection of the outflow tract has been generous, including large amounts of muscle from the anterior and lateral walls (fig. 1) of the ventricle and excision of any fibrous tissue along the leading edge of the crista supraventricularis. Division of hypertrophied trabeculae carneae, as shown in figure 1, was carried out extensively and resulted in a release of the outflow tract, considerably increasing its mobility, and is essential for maximal enlargement of the right ventricular outflow tract (fig. 2).

Postoperative Care

Patients were placed in a high-humidity oxygen tent for 48 hours postoperatively. Assisted ventilation was not required except in one patient with complete heart block. Monitoring of the electrocardiogram, blood gases, pH, and venous pressure has been carried out, but the findings have not been significantly abnormal or useful in the routine management of patients without complications. Fluids were administered intravenously for 48 hours in the amount of 500 ml./M.2/24 hours with urinary volume losses added to the fluid prescription with 5 per cent dextrose in water at 12-hour intervals. Electrolytes were given in the amounts of sodium, 50 mg./M.2/24 hours, and potassium, 40 mg./M.2/24 hours, on the basis of studies previously reported.10 Blood loss was replaced volume for volume. Antibiotics consisted of penicillin and streptomycin intramuscularly for seven days. Digitalis was not used except in those patients who developed congestive cardiac failure. Ambulation was started by the sixth to seventh postoperative
Figure 2

Preoperative (A) and postoperative (B) angiocardiograms showing the enlargement of the outflow tract possible by resection alone. The absent ventricular septum noted was reconstructed with a large Teflon patch.

day, but was restricted during the first six weeks postoperatively.

Results

There has been no operative mortality in the past 41 consecutive patients. Two late deaths occurred in the group. One patient (J.C.) died of Aspergillus endocarditis, six months following operation, with vegetations on the ventricular septal patch and multiple pulmonary abscesses. The second patient (W.V.) died suddenly at play three months following reoperation for a persistent large left-to-right shunt at the site of detachment of the Teflon patch from the lower muscular septum. Autopsy revealed a complete repair, including an outflow patch, and the cause of death could not be determined.

All living patients are clinically improved and have no evidence of cyanosis. Exercise tolerance appears normal except in one (G.C.), the only patient in the group who developed a permanent atrioventricular block (complete heart block with His-bundle rhythm). Auscultation revealed residual systolic murmurs varying in intensity from grades I to III (Levine scale); short, low-pitched diastolic murmurs were heard in eight patients with pulmonary insufficiency. Roentgenograms of the chest demonstrated either no significant change or a slight increase in heart size, especially in the anteroposterior diameter, in comparison with the preoperative examination.

Twenty-five of the 34 patients in the cyanotic group have undergone postoperative catheterization and angiographic studies. Table 1 records their preoperative status and

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Table 1

Summary of Preoperative Status and Postoperative Studies of Twenty-five Patients Undergoing Total Correction of Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Patient number</th>
<th>Age</th>
<th>Clinical disability</th>
<th>Previous operations</th>
<th>Preop. arterial O₂ sat. %</th>
<th>Outflow patch</th>
<th>Postoperative status</th>
<th>Hemodynamic result</th>
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<tbody>
<tr>
<td>1-A.M.</td>
<td>10</td>
<td>Moderate</td>
<td></td>
<td>86</td>
<td>Yes</td>
<td>22/8</td>
<td>100</td>
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<tr>
<td>2-M.B.</td>
<td>16</td>
<td>Mild</td>
<td></td>
<td>85</td>
<td>Yes</td>
<td>20/7</td>
<td>98</td>
</tr>
<tr>
<td>3-R.A.</td>
<td>4½</td>
<td>Severe</td>
<td></td>
<td>0</td>
<td></td>
<td>27/8</td>
<td>96</td>
</tr>
<tr>
<td>4-M.S.</td>
<td>15</td>
<td>Severe</td>
<td>L &amp; R Blalock</td>
<td>84</td>
<td>0</td>
<td>32/10</td>
<td>96</td>
</tr>
<tr>
<td>5-G.R.</td>
<td>18</td>
<td>Severe</td>
<td>Explor. only</td>
<td>80</td>
<td>Anom. R. cor. art.</td>
<td>21/8</td>
<td>98</td>
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<tr>
<td>6-K.C.</td>
<td>6</td>
<td>Severe</td>
<td></td>
<td>80</td>
<td></td>
<td>20/8</td>
<td>96</td>
</tr>
<tr>
<td>7-G.C.</td>
<td>10</td>
<td>Severe</td>
<td>L Blalock</td>
<td>80</td>
<td>0</td>
<td>28/11</td>
<td>73/11</td>
</tr>
<tr>
<td>8-K.H.</td>
<td>9</td>
<td>Severe</td>
<td>L Blalock</td>
<td>78</td>
<td>0</td>
<td>15/3</td>
<td>22/0</td>
</tr>
<tr>
<td>9-D.P.</td>
<td>5</td>
<td>Severe</td>
<td>R Blalock</td>
<td>80</td>
<td></td>
<td>15/3</td>
<td>33/2</td>
</tr>
<tr>
<td>10-S.W.</td>
<td>15</td>
<td>Severe</td>
<td>L Blalock</td>
<td>82</td>
<td>Yes</td>
<td>24/8</td>
<td>34/8</td>
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<tr>
<td>11-J.C.</td>
<td>14</td>
<td>Severe</td>
<td>L &amp; R Blalock</td>
<td>82</td>
<td>0</td>
<td>26/7</td>
<td>32/5</td>
</tr>
<tr>
<td>12-C.H.</td>
<td>10</td>
<td>Severe</td>
<td></td>
<td>67</td>
<td>0</td>
<td>22/8</td>
<td>30/6</td>
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<tr>
<td>13-P.M.</td>
<td>11</td>
<td>Moderate</td>
<td></td>
<td>80</td>
<td></td>
<td>27/10</td>
<td>36/7</td>
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<tr>
<td>14-D.C.</td>
<td>13</td>
<td>Severe</td>
<td>L Blalock</td>
<td>75</td>
<td>0</td>
<td>27/10</td>
<td>56/0</td>
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<tr>
<td>15-A.S.</td>
<td>27</td>
<td>Moderate</td>
<td></td>
<td>92</td>
<td>0</td>
<td>13/5</td>
<td>32/2</td>
</tr>
<tr>
<td>16-M.R.</td>
<td>12</td>
<td>Moderate</td>
<td></td>
<td>85</td>
<td>0</td>
<td>25/15</td>
<td>25/0</td>
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<tr>
<td>17-N.R.</td>
<td>13</td>
<td>Severe</td>
<td>Brock; L Blalock</td>
<td>74</td>
<td></td>
<td>20/10</td>
<td>28/4</td>
</tr>
<tr>
<td>18-R.C.</td>
<td>26</td>
<td>Mild</td>
<td>Brock</td>
<td>85</td>
<td></td>
<td>26/10</td>
<td>30/9</td>
</tr>
<tr>
<td>19-E.J.</td>
<td>26</td>
<td>Severe</td>
<td>L &amp; R Blalock</td>
<td>63</td>
<td>0</td>
<td>18/10</td>
<td>20/3</td>
</tr>
<tr>
<td>20-B.M.</td>
<td>8</td>
<td>Moderate</td>
<td>L Blalock</td>
<td>85</td>
<td></td>
<td>25/10</td>
<td>71/10</td>
</tr>
<tr>
<td>21-H.W.</td>
<td>11</td>
<td>Moderate</td>
<td></td>
<td>85</td>
<td>Yes &amp; P.I.</td>
<td>25/10</td>
<td>71/10</td>
</tr>
<tr>
<td>22-D.S.</td>
<td>7</td>
<td>Severe</td>
<td></td>
<td>78</td>
<td>Yes &amp; P.I.</td>
<td>32/0</td>
<td>32/0</td>
</tr>
<tr>
<td>23-P.C.</td>
<td>8</td>
<td>Severe</td>
<td>Potts</td>
<td>81</td>
<td>Yes &amp; P.I.</td>
<td>42/10</td>
<td>42/10</td>
</tr>
<tr>
<td>24-W.V.</td>
<td>9</td>
<td>Moderate</td>
<td></td>
<td>92</td>
<td>Yes &amp; P.I.</td>
<td>64/10</td>
<td>68/7</td>
</tr>
<tr>
<td>25-S.F.</td>
<td>16</td>
<td>Mild</td>
<td>Brock</td>
<td>92</td>
<td>Yes &amp; P.I.</td>
<td>30/12</td>
<td>48/12</td>
</tr>
</tbody>
</table>

L, left; R, right; arterial O₂ sat., arterial oxygen saturation; P.A., pulmonary artery; R.V., right ventricle; QP/Qs, pulmonary/systemic flow ratio; P.I., pulmonary insufficiency; anom. R. cor. art., anomalous right coronary artery.
catheterization data. The postoperative hemodynamic status has been classified as follows: *Excellent*, if there was no residual shunt and any residual gradient less than 20 mm. Hg; *Good*, if there was a small left-to-right shunt or a residual gradient between 20 and 50 mm. Hg; and *Poor*, if a shunt and gradient were both present. It should be noted that none of the patients with a residual shunt had a pulmonary/systemic flow ratio ($Q_{Pa}/Q_{s}$) in excess of 1.5:1.

On the basis of our criteria, 16 of the 25 patients have excellent results. Five have good results. The remaining 4 patients are considered poor results, and each represents operative error because of a residual shunt. The two late deaths occurred in this group. The third patient (G.C.) had bidirectional intracardiac shunts and complete heart block. The fourth patient (A.S.), although asymptomatic, with normal heart size, is labeled a poor result hemodynamically because of a residual shunt and gradient of 30 mm. Hg.

Pulmonary valvular insufficiency was created by outflow tract reconstruction in 8 of the 41 patients, 5 of whom were restudied by catheterization (nos. 21 to 25). Seven of these patients exhibited some degree of postoperative right-heart failure requiring short-term digitalization. Cardiac size in this group has remained normal or shown only minimal enlargement. To date, there have been no complications related to the outflow patch itself, and figure 3 shows the satisfactory outflow contour at the site of replacement. The long-term effects of pulmonary valvular insufficiency are unknown, but it has been well tolerated clinically in this group up to two years postoperatively.

**Comments**

The operative risk of total correction of tetralogy of Fallot is approaching that for closure of ventricular septal defect, as the details of repair have become more clearly defined and patient selection has been adopted. Total correction has resulted in normal hemodynamics in the majority of patients, as reported here and by Kay et al. Unsatisfactory postoperative results were related directly to incomplete operative repair and hence are preventable. The problem of the small residual
shunt remains unsettled, but reoperation has not been recommended in the absence of cardiac failure and cardiomegaly.

In comparing this recent favorable experience in the surgical treatment of tetralogy of Fallot with our previous poor results, several factors besides the increased surgical experience and changes in the perfusion apparatus may have been important. Probably most essential is that the patient leave the operating table with the anomaly repaired as completely as possible, both anatomically and physiologically. Most important to this goal is accurate preoperative diagnosis, including understanding of the pathological physiology which, in this series, has been aided by selective biplane angiocardiography in all cases and, where necessary, cardiac catheterization. It should be pointed out, however, that neither the degree of dextroposition of the aorta nor the diminutive size of the main pulmonary artery or left ventricle has been regarded as an absolute contraindication to operation.

Postoperative angiocardiograms and catheterization data, as well as direct observations at operation, have shown that careful but liberal excision of muscle and fibrous tissue of the right ventricular infundibulum, together with division of hypertrophied muscular bands, can nearly always adequately widen the infundibular region. The presence of an anomalous right coronary artery crossing this area often limits the infundibular resection (fig. 4). Although the pulmonary valvular insufficiency created by outflow tract reconstruction, including the pulmonary valve and artery, has been well tolerated thus far in eight patients of this series, an outflow patch is considered undesirable unless required by the following situations: (1) extreme hypoplasia of the pulmonary annulus, usually associated with severe valvular stenosis; (2) stenosis or atresia of the main pulmonary artery beyond the level of the pulmonary veins; and (3) atrophic muscle within the infundibular chamber which would not securely hold suture

Figure 4

A. Demonstrates an anomalous right coronary artery crossing the right ventricular outflow tract. Postoperatively (B), a residual anatomical and physiological obstruction is noted in this area.
for primary closure. Its use to enlarge the infundibular outflow tract alone has not been necessary since adopting the present technique of correction.

The selection of patients with tetralogy of Fallot for open-heart surgery should be determined by age and severity of symptoms. At the present time, elective primary repair of the malformation would be recommended in cyanotic patients over the age of five years; more particularly, total correction would be deferred to the age of approximately eight to ten years, if the child were able to attend school regularly until then. A shunting procedure, preferably a subclavian-to-pulmonary arterial anastomosis, has been recommended in all severely disabled patients under five years of age. The previously constructed shunt has not increased the operative risk of total repair and may have a beneficial effect upon the pulmonary vascular bed, as suggested by the work of Ferencz.12 The use of the two-stage approach to the young, severely cyanotic patient has allowed growth and development, permitting total repair in an age period associated with a relatively low mortality.

Summary

Forty-one consecutive patients, ranging in age from 4½ to 27 years, were operated upon for complete repair of tetralogy of Fallot without an operative mortality. Postoperative catheterization of 25 patients permitted objective evaluation of the results which were, by the criteria adopted, excellent in 16 patients, good in 5, and poor in 4, including the 2 late deaths.

Surgical experience has shown the necessity for extensive resection of the right ventricular outflow tract. Prosthetic patches are considered necessary for closure of most of the ventricular septal defects encountered, but are infrequently needed for reconstruction of the right ventricular outflow tract. Its use is indicated in certain patients with hypoplasia of the pulmonary annulus or artery.

Age has been an important factor in patient selection. It is suggested that symptomatic patients under five years of age may benefit from a systemic-to-pulmonary arterial anastomosis before definitive correction is undertaken later in childhood.

References


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