Intracardiac Repair in Tetralogy of Fallot
Hemodynamic Studies Following Corrective Surgery

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SUMMARY
Experience with the first 100 cases with tetralogy of Fallot subjected to intracardiac repair are presented. In this series there was an early hospital mortality of 12 per cent and 6 late deaths occurred. Eighty out of 82 survivors were available for follow-up and 33 of these underwent postoperative catheterization studies. The functional status of the survivors revealed excellent results in 89 per cent, good in 8 per cent, fair in 1 per cent and poor in 2 per cent. This is the largest series reported from India.

Additional Indexing Words:
Hospital mortality Functional status Recatheterization data

COMPLETE REPAIR of the tetralogy of Fallot malformation remains a most challenging and difficult operation, the results of which have much improved in recent years. In 1955 intracardiac repair of this complex anomaly was first accomplished by Lillehei and associates. Surgical results and postoperative follow-up studies have been reported by several authors. Experience with the first 100 cases who underwent corrective surgery between January 1967 and December 1972 in the Department of Cardiothoracic Surgery of the Christian Medical College Hospital, Vellore, India forms the basis of this review. Experiences from this center with total correction in 45 cases from January 1967 until January 1971 have been published earlier.

Methods and Materials
Clinical Material

The ages of the patients at the time of surgery ranged between 4 and 30 years (table 1), and their weights varied from 10 to 54 kg. The primary clinical features and complications occurring prior to surgery are outlined in figure 1. Sixteen per cent of the patients were cyanotic, 6% were in congestive failure at the time of surgery, anoxic spells were present in 19% and 3% had previous brain abscesses. Previous shunt surgery had been carried out in 20%.

Preoperative cardiac catheterization and right ventricular cineangiography were carried out in every case and confirmed the clinical diagnosis. Preoperative oxygen saturation ranged between 42 per cent and 95 per cent (mean 76.7 ± 12.9%)

In recent years a left ventricular injection has also been made in order to demonstrate aortic mitral valve continuity. Cine aortography in four subjects showed evidence of mild aortic regurgitation. In two cases, selective injection of the appropriate subclavian artery was carried out to document patency of the anastomosis where serious doubts existed preoperatively.

Method of Intracardiac Repair

The technique used was uniform. Cardiopulmonary bypass was accomplished using moderate total body hypothermia with the disc oxygenator. The right ventricular chamber was exposed by a para-coronary ventriculotomy and the exposure was facilitated by intermittent cross-clamping of the aorta for 10 to 15 min. In those cases where a functioning shunt was present, it was repaired soon after institution of cardiopulmonary bypass; no difficulty was encountered during shunt closure in any case. Pulmonary valvotomy was accomplished in most instances through a pulmonary arteriotomy. An outflow gusset utilizing pericardium was necessary in 30 cases. The septal defect was closed with a woven teflon patch in each instance. In 8 cases, a patent foramen ovale was encountered and closed through a right atriotomy in 5 and through the tricuspid valve in 3. A patent ductus arteriosus present in 3 cases was closed by direct suture through the opened pulmonary artery.

Operative Findings

Of the cyanotic group of 84 patients, 20 had infundibular obstruction alone; 44 had infundibular and valvular obstruction; 18 had a fibrotic annulus in addition; and 2 showed infundibular obstruction with valvar atresia (table 2). In the cyanotic group of 16 patients, 8 had infundibular obstruction alone while the remaining subjects had both infundibular and valvar obstruction. Of the 20 patients who had undergone previous shunt surgery, the shunts were patent in 18. Four of these were Waterston shunts and 14 were Blalock-Taussig anastomoses.

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Table 1

<table>
<thead>
<tr>
<th>Age group (years)</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 5</td>
<td>7</td>
</tr>
<tr>
<td>5 - 10</td>
<td>33</td>
</tr>
<tr>
<td>11 - 20</td>
<td>47</td>
</tr>
<tr>
<td>21 - 30</td>
<td>13</td>
</tr>
</tbody>
</table>

Associated Anomalies

Associated anomalies were present in 9 per cent of cases. Four had mild aortic incompetence detected by cine aortography and confirmed during operation. Three had a patent ductus arteriosus which were closed. One subject had abnormal bronchopulmonary collaterals and one showed an absent left pulmonary artery.

Results

Hospital Mortality

The hospital mortality occurring within 30 days of corrective surgery was 12 per cent (table 3). In four cases, a low cardiac output syndrome was the causative factor and these patients succumbed in the early postoperative period. One of these was the result of inadequacies during perfusion resulting in progressive metabolic acidosis. Two of the fatal cases developed intractable pulmonary edema, and in one of these there was in addition a complete A-V dissociation. Two others died from pulmonary dysfunction. A young boy who was in refractory congestive failure following a previous Waterston shunt died 16 hours after corrective surgery from a ventricular tachyarrhythmia. Hemorrhage occurring in a friable right ventricle was the cause of death in a young boy who was in congestive failure and had aortic incompetence in addition. Disseminated intravascular coagulation ended fatally in the two remaining subjects. There was no significant correlation between hospital mortality and the degree of polycythemia (table 4).

Early Morbidity

The morbidity occurring in the early postoperative period is outlined in detail in table 5. Six subjects required reoperation for either clotted hemotorax or significant bleeding. One of these was a young woman intensely cyanosed with a preoperative platelet count of 30,000 cu mm.Transient heart block occurred in three instances.

Late Deaths

There were 6 late deaths in this series. One of them was a severely incapacitated adolescent male who developed complete heart block and died two and a half months following surgery. Another patient succumbed to a subacute bacterial endocarditis. One fatality occurred eight months after surgery from gastroenteritis. A young boy died thirty-two days following reoperation for a postoperative pseudomonas endocarditis refractory to intensive antibiotic therapy. The infected patch had been excised and another one inserted after thorough irrigation with Gentamycin solution. One young boy died suddenly seven weeks after repair. Another young boy of ten died of pneumococcal peritonitis eight months after surgery. Necropsy revealed excellent closure of the septal defect and satisfactory relief of outflow obstruction.

Follow-Up Data

Eighty of 82 survivors were available for evaluation by the cardiologists at the hospital at periodic intervals. The assessments were made according to the criteria of Baker and Hancock. The functional status

Table 2

<table>
<thead>
<tr>
<th>Group</th>
<th>No.</th>
<th>Infundibular obstruction only</th>
<th>Infundibular and valvular obstruction</th>
<th>Infundibular, valvular and fibrotic annulus</th>
<th>Infundibular obstruction and valvar atresia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cyanotic</td>
<td>84</td>
<td>20</td>
<td>44</td>
<td>18</td>
<td>2</td>
</tr>
<tr>
<td>Acyanotic</td>
<td>16</td>
<td>8</td>
<td>8</td>
<td>---</td>
<td>---</td>
</tr>
</tbody>
</table>

The cardinal clinical manifestations and complications prior to corrective surgery are illustrated.
of the survivors revealed an excellent result in 89 per cent, good in 8 per cent, fair in 1 per cent and poor in 2 per cent (fig. 2). The average period of follow-up was twelve months following corrective surgery.

Of the two in whom the results were poor, one young subject, seen 22 months following repair, was in congestive failure from a residual shunt and right ventricular outflow tract obstruction. He subsequently underwent a successful repair. Another young boy seen three months after surgery was in gross congestive failure with severe tricuspid incompetence. Recatheterization showed a significant residual shunt. At the time of initial repair, a large infracristal defect over 3 cm in diameter was closed. At the time of reoperation, exploratory cardiotomy revealed a left ventricular-right atrial shunt together with a residual ventricular septal defect and he underwent a successful repair. However, these two subjects have been followed only a short time after their second operation.

Fifteen patients showed evidence of pulmonary regurgitation. It was of a significant degree with congestive cardiac failure in five patients including two children under five years of age, all of whom had required a pericardial patch across the outflow. With carefully controlled diuretic therapy remarkable symptomatic improvement occurred in all of them.

Thirty-three of the 82 survivors underwent hemodynamic evaluation twelve to eighteen months following complete repair. In this group the incidence of residual ventricular septal defect was 6 per cent (table 6). Three patients showed evidence of a significant gradient across the outflow. Despite absence of any symptoms reoperation would be necessary.

Comments

This paper presents a review of the first one hundred cases of intracardiac repair of tetralogy of Fallot carried out in this center during a six year period between January 1967 and December 1972. The operative mortality in this series was 12 per cent. Kirklin and his associates8 quoted an early operative mortality of 15 per cent which after further experience was reduced to 7 per cent. Ikeda and Hirosawa9 reported a mortality of 19 per cent whereas Zerbini10 mentioned a figure of 13.2 per cent.

Low cardiac output syndrome continues to be the commonest cause of death following total surgical correction. Four of the 12 deaths were the result of this factor. There appeared to be no significant correlation between polycythemia and operative mortality. Table 4, which correlates polycythemia and operative mortality, reinforces the fact that out of the 100 patients, 52 patients had a hemoglobin of over 18 g % (range 18-28) with a mortality of only 13 per cent. This contrasts very strikingly with a small number of 14 patients with a hemoglobin of over 18 g % who underwent complete repair in the previous series, in which the mortality was 14 per cent. Increasing experience with regard to complete repair in the recent past has encouraged us to carry out this procedure irrespective of the degree of polycythemia.

Permanent heart block occurred in only two subjects one of whom died two and a half months after repair. In the present series where the average period of follow-up was one year, 89 per cent had excellent results as assessed by the criteria of Baker and Hancock.8 Long term functional assessment of these patients will be necessary to see if this degree of improvement will be maintained. Kirklin,1 in his much larger series, has quoted 93 per cent excellent results among the survivors during the period of follow-up.

Table 3

<table>
<thead>
<tr>
<th>Fatal Complications (Hospital Mortality)</th>
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</thead>
<tbody>
<tr>
<td>Low cardiac output</td>
</tr>
<tr>
<td>Heart block with pulmonary edema</td>
</tr>
<tr>
<td>Pulmonary edema</td>
</tr>
<tr>
<td>Disseminated intravascular coagulation</td>
</tr>
<tr>
<td>Pulmonary dysfunction</td>
</tr>
<tr>
<td>Ventricular tachyarrhythmia and cardiac arrest</td>
</tr>
<tr>
<td>Hemorrhage from friable right ventricle</td>
</tr>
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</table>

Table 4

<table>
<thead>
<tr>
<th>Correlation between Polycythemia and Operative Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>----------------------------------------------------------</td>
</tr>
<tr>
<td>Hemoglobin</td>
</tr>
<tr>
<td>&lt; 18 g %</td>
</tr>
<tr>
<td>----------------------------------------------------------</td>
</tr>
<tr>
<td>Total number of cases</td>
</tr>
<tr>
<td>Mortality</td>
</tr>
<tr>
<td>Percentage</td>
</tr>
</tbody>
</table>

Table 5

<table>
<thead>
<tr>
<th>Non-Fatal Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>----------------------------</td>
</tr>
<tr>
<td>Pulmonary infection</td>
</tr>
<tr>
<td>Congestive heart failure</td>
</tr>
<tr>
<td>Transient heart block</td>
</tr>
<tr>
<td>Permanent heart block</td>
</tr>
<tr>
<td>Reoperation for bleeding</td>
</tr>
<tr>
<td>Convulsions</td>
</tr>
<tr>
<td>Post pericardiotomy sydrome</td>
</tr>
<tr>
<td>Arrhythmias</td>
</tr>
<tr>
<td>Low cardiac output</td>
</tr>
<tr>
<td>Pulmonary edema</td>
</tr>
</tbody>
</table>
RESULTS AT FOLLOW-UP
OF 80 OUT OF 82 PATIENTS
DURATION - 6 MONTHS TO 5½ YEARS

<table>
<thead>
<tr>
<th>POST OPERATIVE GRADE</th>
<th>NO. OF PATIENTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>EXCELLENT</td>
<td>32</td>
</tr>
<tr>
<td>GOOD</td>
<td>11</td>
</tr>
<tr>
<td>FAIR</td>
<td>7</td>
</tr>
<tr>
<td>POOR</td>
<td>1</td>
</tr>
</tbody>
</table>

Figure 2
Photograph illustrating follow-up data in 82 survivors, 80 of whom were assessed in the institution while the remaining two were followed by the referring physician.

In our study, 33 patients underwent catheterization twelve to eighteen months following corrective surgery. It was gratifying to note that results could be graded as excellent or good in 25 of the 28 severely cyanotic patients who underwent total surgical correction in this series.

The incidence of residual ventricular septal defect was only 6 per cent. Meticulous technique with interrupted sutures is of vital importance in reducing the incidence of residual ventricular septal defects. Gotsman et al. reported a residual shunt in 6 per cent of their series. Lillehei et al. mentioned an incidence of 21 per cent in 69 patients catheterized one to seven years following corrective operation. Three of our 33 cases showed a significant gradient across the outflow tract. Long term hemodynamic follow-up is being carried out. Exercise testing has not been done in this group of patients under review.

There were 15 patients who showed evidence of pulmonary regurgitation, in 12 of whom it was considered mild. The recent reports of Bristow et al. that pulmonary regurgitation has been tolerated well up to ten years postoperatively is very reassuring to the surgeon, that whenever necessary an outflow gusset should be utilized.

Seven per cent of our patients were under 5 years of age. All of them have done well. Burnell and co-workers and Dobell and his co-workers have had excellent results in children in this age group.

References

Table 6
Recatheterization Data in 33 Patients: Hemodynamic Status

<table>
<thead>
<tr>
<th>Grades</th>
<th>Ventricular septum</th>
<th>RV outflow</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excellent*</td>
<td>no shunt</td>
<td>&lt; 20 mm. Gdt.</td>
<td>19</td>
</tr>
<tr>
<td>Good</td>
<td>no shunt</td>
<td>20 - 50 mm. Gdt.</td>
<td>9</td>
</tr>
<tr>
<td>Fair</td>
<td>no shunt</td>
<td>&gt; 50 mm. Gdt.</td>
<td>3</td>
</tr>
<tr>
<td>Poor</td>
<td>significant shunt</td>
<td>Qp/Qs 2:1 and/or 50 mm. Gdt.</td>
<td>2</td>
</tr>
</tbody>
</table>

*Excellent or good result: 25/28 cyanotic group. Excellent or good result: 3/5 acyanotic group.

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