Primary Total Correction of Tetralogy of Fallot in Children Less Than Four Years of Age

By Albert D. Paciento, M.D., L. M. Bargerone, Jr., M.D.,
and John W. Kirklin, M.D.

SUMMARY

Twenty-two consecutive severely symptomatic infants and small children less than 4 years of age (mean age 25.5 months, range 3 to 48 months) have presented for initial surgical treatment of the tetralogy of Fallot at the University of Alabama Medical Center between September 1, 1971 and January 31, 1973. One was considered unsuitable for complete repair because of massive thrombosis of the inferior vena cava, and was successfully treated by a right Blalock-Taussig operation. Twenty-one were treated by primary intracardiac repair. One patient (age 24 months) died, a hospital mortality rate of 4.8%. Mean value for the RV/LV peak pressure ratio immediately after repair was 0.55. An outflow patch was placed across the valve ring in one patient, and should have been placed in one other. Heart block has not occurred. Profound hypothermia and circulatory arrest (Kyoto-Barratt-Boyes technique) was used in 9 patients, and cardiopulmonary bypass with profound hypothermia in 12. We discuss the controversy concerning initial management of infants and small children severely symptomatic from the tetralogy of Fallot, and conclude that primary intracardiac repair is advisable in centers prepared for infant intracardiac surgery.

Additional Indexing Words:
Cardiopulmonary bypass  Hypothermia

Since September 1, 1971 we have applied a policy of primary intracardiac repair for all infants and small children severely symptomatic from the tetralogy of Fallot. Patients with absence of anatomic continuity between right ventricle and pulmonary artery (pseudotruncus arteriosus) are not included in this report. This change in patient management program from that which we had previously used was made because of the disadvantages inherent in the two-stage approach, increased experience with intracardiac surgery in infants, and the excellent results being obtained with this policy by Barratt-Boyes. This paper presents our experiences during the first 17 months of this program.

Material and Methods

A total of 30 severely symptomatic infants and children less than four years of age with tetralogy of Fallot were seen consecutively at the University of Alabama Hospital from September 1, 1971 to January 31, 1973. Surgery was advised in all cases. Nine patients had previously-constructed, poorly or non-functioning systemic-pulmonary arterial shunts and underwent total correction with eight surviving the period of hospitalization. One previously untreated patient, 24 months of age, received a right subclavian-pulmonary artery anastomosis because he had a thrombosed inferior vena cava and was considered an unacceptable candidate for complete repair. Twenty-one patients underwent primary complete intracardiac correction of tetralogy of Fallot and are the subject of this review. These 21 patients represent an unselected consecutive series of patients whose ages ranged from 3½ to 48 months, having a mean age of 25.5 months. During this period other infants with tetralogy of Fallot and mild or no symptoms were seen and are being followed.

A technique of profound hypothermia with total circulatory arrest was used during operation in nine patients. Each patient was initially cooled to a nasopharyngeal temperature of about 30°C, using ice bags applied to the body after induction of anesthesia. A median sternotomy was made and cardiopulmonary bypass with a bubble oxygenator (Bentley Laboratories) used to further reduce body temperature. A three-minute period of hypercarbosis (10% CO₂) was employed when the nasopharyngeal temperature was below 25°C to augment cerebral blood flow and appropriately shift the oxyhemoglobin dissociation curve. Total circulatory arrest was established when nasopharyngeal tempera-
ture reached 20° C. The patent foramen ovale was closed through a right atriotomy. Relief of infundibular and valvular stenosis and closure of the ventricular septal defect was accomplished through a transverse right ventriculotomy. Cardiopulmonary bypass was reestablished, and rewarming begun. When the temperature reached 30° C, bypass was transiently discontinued to allow measurement of right ventricular/left ventricular (RV/LV) systolic pressure ratios. If these were satisfactory, cardiopulmonary bypass was reestablished and rewarming continued until the temperature reached 35° C. Just before closing the sternotomy incision, RV/LV systolic pressure ratios were remeasured and these are the ones presented in this paper.

Cardiopulmonary bypass with profound cardiac cooling was used in 12 patients. Cooling proceeded as described above, but two venous cannulae were used. Body temperature was reduced to 22°-24° C, and during this cooling period, the foramen ovale was closed, and a left atrial or ventricular vent inserted. When the desired temperature was reached, flow was reduced to 0.5 L/min/m² for 10 to 30 min, and then was increased to 1.6 until rewarming was started. The aorta was crossclamped for about 40 min, while the intraventricular repair was accomplished.

Postoperatively, all patients except one were managed for at least 24 hr by the technique of spontaneous breathing with a continuous positive airway pressure system. One patient, the child who died from pulmonary edema with low left atrial pressure, was ventilated with a volume respirator. Each patient was attached to an automated care system, and left and right atrial and arterial pressure, electrocardiogram and rectal temperature continuously monitored. The measurement of cardiac output was made in nine patients from indicator dilution curves with injection of indocyanine green dye into right atrium and sampling from radial artery.

### Results

The age and weight distribution of the 21 patients and the hospital mortality is shown in Table 1. The youngest patient was 3½ months and the oldest 48 months of age. Twelve patients were less than two years of age and the single death occurred in a 24-month-old girl. She succumbed on the first postoperative day from hemorrhagic pulmonary edema in the presence of low left atrial pressure. There have been no late deaths and no instances of permanent heart block.

Figure 1 shows the distribution of the two techniques of perfusion, according to patient age. Early in this experience, the technique of surface cooling and limited cardiopulmonary bypass with profound hypothermia and total circulatory arrest was used.

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**Table 1**

<table>
<thead>
<tr>
<th>Age (Months)</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>9</td>
</tr>
<tr>
<td>9</td>
<td>15</td>
</tr>
<tr>
<td>15</td>
<td>27</td>
</tr>
<tr>
<td>21</td>
<td>33</td>
</tr>
<tr>
<td>27</td>
<td>39</td>
</tr>
<tr>
<td>33</td>
<td>45</td>
</tr>
</tbody>
</table>

- **Cardiopulmonary Bypass with Profound Cardiac Cooling**
- **Profound Hypothermia Total Circulatory Arrest**
- **Outflow Patch**
- **Death**

*Mean age 25.5 Months.*

**Figure 1**

The distribution according to age of the two techniques of patient support are shown. The single fatality and the patient receiving outflow tract reconstruction are indicated.
CORRECTING TF IN CHILDREN UNDER FOUR

Table 1
Primary Repair Tetralogy of Fallot < 4 yrs of age*

<table>
<thead>
<tr>
<th>Age (mos.)</th>
<th>Patients (no.)</th>
<th>Weights (kg.)</th>
<th>Mortality (no.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>3 - 6</td>
<td>2</td>
<td>5.1 - 7.0</td>
<td>0</td>
</tr>
<tr>
<td>7 - 12</td>
<td>3</td>
<td>6.8 - 7.2</td>
<td>0</td>
</tr>
<tr>
<td>13 - 18</td>
<td>2</td>
<td>10.0 - 11.0</td>
<td>0</td>
</tr>
<tr>
<td>19 - 24</td>
<td>5</td>
<td>9.0 - 13.2</td>
<td>1†</td>
</tr>
<tr>
<td>25 - 36</td>
<td>6</td>
<td>9.5 - 13.7</td>
<td>0</td>
</tr>
<tr>
<td>37 - 48</td>
<td>3</td>
<td>14.0 - 17.0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>21</td>
<td></td>
<td>1 (4.8%)</td>
</tr>
</tbody>
</table>

*Mean age 25.5 months.
†Hemorrhagic pulmonary edema.

for all patients less than 2 years of age or 13 kg in weight. Thus, of 21 patients nine were operated upon using this method. There were no deaths in this group. Later in our experience, because of its greater surgical flexibility, cardiopulmonary bypass with profound hypothermia was used more frequently for infant cardiac surgery, and 12 patients were operated upon by this method (age 12 to 48 months). One death occurred in this group and one patient required a Dacron outflow patch. No neurologic problems were observed in the patients in either group.

The ratio of RV/LV systolic pressure measured in the operating room after completion of repair and just before closure of the sternum is shown according to patient age and the perfusion technique in figure 2. Ratios ranged from 0.28 to 1.22 and the mean value for the group was 0.55. A Dacron outflow patch was used in one patient, reducing the right ventricular/left ventricular pressure ratio from 1.33 to 0.40. All ratios were less than 0.81, excepting one which was 1.22. In this patient, the hemodynamic state was good, and left atrial pressure was higher than right atrial pressure. The patient convalesced satisfactorily. This child was restudied 12 months postoperatively and found to have persistent severe right ventricular hypertension. Reoperation will be required. There was no relation between patient age or the technique of perfusion and the ability to relieve infundibular obstruction in this group of patients.

The duration of cardiopulmonary bypass and of total circulatory arrest for the two groups of patients is shown in figure 3. The nine patients
operated upon using the technique of limited cardiopulmonary bypass, profound hypothermia and total circulatory arrest had perfusion times (cooling + rewarming) varying between 22 and 38 (mean 31) minutes. The duration of total circulatory arrest, established at a nasopharyngeal temperature of 20°C, varied between 49 and 59 (mean 55.3) minutes. Twelve patients were operated upon using cardiopulmonary bypass with profound hypothermia and the duration of bypass varied between 49 and 76 (mean 61.9) minutes. This included one patient who had placement of a Dacron outflow patch.

Cardiac indices were measured in nine patients between 1 and 4, 8 and 10, and at 24 hours after operation.* These data are shown in figure 4. Cardiac index varied between 1.11 and 3.50 L/min/m² and mean values for each period were 2.59, 2.21 and 2.56 L/min/m² respectively. The two patients whose cardiac indices were less than 2 L/min/m² were four and 12 months of age, the youngest patients in whom this measurement was made.

All patients except one were extubated 24 hours postoperatively. One patient was maintained on the continuous positive airway system for 48 hours because of right upper lobe atelectasis and a large alveolar-arterial oxygen gradient.

All patients were well at the time of dismissal, which varied from eight to 18 days after operation. There have been no late deaths.

Discussion

Controversy exists regarding the proper patient management program for infants and small children who are symptomatic from the tetralogy of Fallot. Lillehei and colleagues reported successful intracardiac repair in several small children in 1955, using...
controlled cross-circulation rather than a pump-oxygenator.\textsuperscript{6} Infants 9, 18, and 22 months of age survived and were reported to have obtained excellent results. In our early experience with the tetralogy of Fallot, we did successfully perform primary repair in infants and small children.\textsuperscript{7} By 1960, however, we believed that the risk of open operation for this malformation was greater in children less than about four years of age than in older children,\textsuperscript{8} and by 1965 had adopted a policy of performing intracardiac repair at age five years and an anastomotic operation if symptoms became severe before that age.\textsuperscript{9} Most groups have followed such a policy in recent years. Puga, DuShane and McGoon at the Mayo Clinic have recently published their justification for continuing such a policy.\textsuperscript{10} An important part of this is their high hospital mortality rate for complete repair in infants and small children, but the data come from operations done prior to 1966.

Dobell, Charrette and Chughtai\textsuperscript{11} and Starr and associates\textsuperscript{12, 13} have advocated intracardiac repair in at least some infants and small children for some time. Starr has recently reported excellent results from intracardiac repair in selected infants and small children.\textsuperscript{14} Barratt-Boyes, in about 1970, began performing primary intracardiac repair for all symptomatic infants with the tetralogy of Fallot. He recently reported 17 such cases, with one death (6\%).\textsuperscript{2} Four patients were less than two months of age, and the one death was in one of these. Eight were between two and 12 months old, and five were between one and two years of age. Three of the four infants less than two months of age required an outflow patch, which is not surprising since probably most infants severely symptomatic in the first few months of life have a small pulmonary valve ring. He employed more outflow patches in the older infants than we have, perhaps in part because of different indications for a patch and in part because of differences in technique.

The risks inherent in the two-stage procedure were recently reviewed by Kerr and Barratt-Boyes.\textsuperscript{2} In their group of patients palliated by systemic-pulmonary artery anastomosis in the first year of
life, only 72% were alive at late follow-up and some of these had still to undergo the intracardiac repair. In contrast, over 90% of patients are alive and well following intracardiac repair.\textsuperscript{1, 2, 15}

Our recent experience, reported in this paper, and that of Barratt-Boyes, indicates to us that in centers properly prepared for infant intracardiac surgery, primary intracardiac repair should be done whenever patients with the tetralogy of Fallot become symptomatic, and without waiting for the symptoms to become severe. In any event, the malformation should be repaired by the age of about five years. Possible criticisms of this position are several. We have no experience, by chance, with primary repair in infants three months of age or less. Six such infants have been operated upon by Barratt-Boyes, and five survived.\textsuperscript{2} The published hospital mortality rates for palliative operations for the tetralogy of Fallot in the first 12 months of life are higher than this.\textsuperscript{2, 16–20} On the basis of our review of angiocardiograms made in the past in such infants severely symptomatic in the first three months of life, we believe they are correctable by primary definitive operation. Another possible criticism, particularly with reference to Barratt-Boyes' and Starr's experience, is that a higher incidence of outflow patches is required when the intracardiac operation is done in infancy than when it is done later in life. We do not believe this to be the case. We have found the need for outflow patches to be about the same incidence in infants and young children as in older children. We do believe that as we operate upon infants in trouble before the age of three months we will experience a higher incidence of outflow patches (Barratt-Boyes found such a patch needed in five of six patients repaired at age three months or less), since the pulmonary valve ring is often small in patients severely symptomatic in the first few months of life. We believe the ring does not enlarge after a palliative operation, and that an outflow patch would be required at whatever age the operation was done. Heart block has not occurred in Barratt-Boyes' experience, nor in Starr's nor ours. Secure repair of the ventricular septal defect has been obtained. Good relief of the pulmonary stenosis has been obtained (except in the one patient in whom we should have placed an outflow patch).

The actual surgical technique may be important. We have routinely opened the right atrium and closed the foramen ovale if it is seen to be patent. A transverse incision is made in the right ventricle unless the pulmonary valve ring is deemed so narrow as to require enlargement with an outflow patch. The pulmonary stenosis is relieved by the usual techniques.\textsuperscript{1} The ventricular septal defect is repaired by suturing in a patch with a continuous 5-0 or 4-0 prolene suture swedged on a special small needle, taking care to avoid the area believed to contain the bundle of His.\textsuperscript{2} The surgical technique inside these small hearts must be absolutely precise, and we sometimes use optical magnification as an aid. At present, after closing the ventriculotomy the patient is rewarmed by the perfusion until the temperature (in the nasopharynx) is 30°C. Cardiopulmonary bypass is then temporarily discontinued and pressures measured and the bypass reestablished for rewarmin to 37°C. If the RV/LV systolic pressure is more than about 0.75, an outflow patch is placed across the pulmonary valve ring while rewarmin proceeds.

Profound hypothermi, limited cardiopulmonary bypass, and total circulatory arrest (the Kyoto-Barratt-Boyes technique) was used in some of the infants and small children in this series. We now usually prefer initial surface cooling to 30°C, and cardiopulmonary bypass with two venous cannulae for cooling to 22°-24°C.\textsuperscript{4} The foramen ovale can be closed and a small left atrial vent inserted during cooling. The intraventricular repair is performed during total circulatory arrest or during cardiopulmonary bypass at a low perfusion flow rate (0.5 to 1.6 L/min/m²). The aorta is crossclamped one time, usually for about 40 minutes. The ventriculotomy is closed during rewarmin.

The postoperative care includes measurement of cardiac index, left and right atrial and arterial pressures, mixed venous oxygen tension, and calculation of oxygen consumption. Currently, a nasotracheal tube is left in place, and the infant's respirations are controlled with a ventilator until he is active and his hemodynamic state is good. Then he breathes spontaneously with continuous positive airway pressure. We usually extubate these children 24 to 48 hours postoperatively.

References


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