Staged Surgical Management of Tetralogy of Fallot in Infants

LARRY W. STEPHENSON, M.D., SIDNEY FRIEDMAN, M.D., AND L. HENRY EDMUNDS, JR., M.D.

SUMMARY A cohort of 61 consecutive patients 24 months of age or younger had palliative shunts for symptoms of tetralogy of Fallot during a 12-year period. Thirty-six of these patients have been followed through definitive intracardiac repair or to death. For analysis palliative operations were separated into two six-year periods, 1965–1970 and 1971–1977. During the first period seven of 30 infants operated on died; all 31 infants operated on during the second period survived. The Waterston anastomosis was performed most frequently (67%) during the first period; the Blalock-Taussig anastomosis was performed in 68% of infants during the second period.

Of 54 hospital survivors, three died before definitive intracardiac repair. Two of the three interim deaths were related to heart disease. Twenty-six of the remaining 51 patients have had definitive intracardiac repair with two deaths (8%). Twenty-four in this group had intracardiac repair since 1973 with one hospital death (4%).

The cumulative mortality for the entire cohort is 25%, but more recent experience (1971–77) indicates a cumulative mortality near 5%. The recent mortality rate for staged management is less than the 14% rate reported by others for primary intracardiac repair of tetralogy of Fallot in 205 infants. We conclude that primary intracardiac repair has important advantages for infants with tetralogy of Fallot who have favorable anatomic features and no other associated cardiac lesions or medical problems. Staged management of tetralogy of Fallot is still recommended for infants with unfavorable anatomy, additional lesions or associated medical problems.

THE ROLE OF PALLIATIVE SHUNTS in the management of symptomatic infants with tetralogy of Fallot is controversial. Recent reports indicate that primary intracardiac repair can be carried out in infants with good results.1–18 However, advocates of palliative shunts claim that a shunt can be performed with a lower mortality risk in infancy than primary intracardiac repair and that intracardiac repair in older children is safer than in infancy. Infants who are first shunted are actually exposed to three periods of risk rather than one: the risk associated with the shunt procedure, the interval period between the shunt operation and intracardiac repair, and the risk of intracardiac repair. Are the cumulative risks of these three periods greater than for intracardiac repair in infancy?

In this study we review an experience with a cohort of symptomatic infants who had palliative shunts for tetralogy of Fallot. Over half of the patients within the cohort have been followed through intracardiac repair or to death. The data assess the cumulative risk of staged surgical repair, and are compared to published results of primary intracardiac repair of tetralogy of Fallot in infancy.

Materials and Methods

During a 12-year period from January 1965 through December 1977, 61 consecutive patients 24 months of age or younger had palliative shunts at Children's Hospital of Philadelphia for symptoms of tetralogy of Fallot. Thirty-three children were male and 28 female. Ages ranged from 2 days to 24 months, with a mean age of 9 months. Seven patients were less than 1 week old and 10 were 1 week to 3 months of age. The types of shunts performed included Blalock-Taussig (28), Waterston (29), Potts (one) and Glenn (three). Indications for surgery included hypoxic spells in 46, severe increasing cyanosis in 13 and respiratory insufficiency in two patients. Mean follow-up period was 52.7 months, and ranged from two to 150 months. Follow-up was conducted by review of hospital and clinic records and sometimes by telephone interview with referring physicians and/or patients.

Results

For purposes of analysis, the palliative operations were separated into two six-year periods — 1965–1970 and 1971–1977. Thirty operations were done during the first period, with seven deaths (23%) (table 1). Thirty-one were performed during the second period, with no deaths (table 2). Overall surgical mortality rate was 11% (seven of 61). Time of death ranged from the day of surgery to six weeks after operation. Operative mortality according to the type of shunt and age of patient is listed in table 1. During the first period, operative mortality was high in infants less than 3 months of age and in those who had Glenn anastomoses. Cardiac arrest during operation or shortly thereafter caused three deaths and respirator dependence led to four deaths. Three of the four patients who could not be weaned from the respirator ultimately died of sepsis.

The Waterston shunt was the most frequently performed shunt during the first six-year period (20

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Table 1. Palliative Shunts for Tetralogy of Fallot in Infants Under 2 Years of Age (1965-1970)

<table>
<thead>
<tr>
<th>Type of shunt</th>
<th>Under 1 month</th>
<th>1–3 months</th>
<th>4–6 months</th>
<th>7–12 months</th>
<th>13–24 months</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Waterston</td>
<td>5/3*</td>
<td>2/1</td>
<td>2</td>
<td>6</td>
<td>5</td>
<td>20/4</td>
</tr>
<tr>
<td>Rt. Blalock-Taussig</td>
<td></td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Glenn</td>
<td>1/1</td>
<td>1</td>
<td>1/1</td>
<td></td>
<td>3/2</td>
<td></td>
</tr>
<tr>
<td>Potts</td>
<td>1/1</td>
<td></td>
<td></td>
<td></td>
<td>1/1</td>
<td></td>
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<tr>
<td>Total</td>
<td>6/4</td>
<td>3/2</td>
<td>4</td>
<td>9/1</td>
<td>8</td>
<td>30/7</td>
</tr>
</tbody>
</table>

*Hospital deaths.

patients), and the Blalock-Taussig anastomosis was more commonly used after 1971 (22 patients) (table 2). There were no hospital deaths among the 28 patients having Blalock-Taussig shunts.

Five of the 61 patients (8%) were reoperated on for complications of the shunt. Two of these patients died and are included among the seven hospital deaths. A 7-month-old with absent pulmonary valve leaflets who had a Glenn shunt required reoperation on the first postoperative day. Although the Glenn shunt was patent, hypoxia was not relieved because the pulmonary vascular resistance was elevated. The right pulmonary arterial-caval anastomosis was taken down, and the right pulmonary artery was re-anastomosed to the ascending aorta. The child died shortly after leaving the operating room. A 2-month-old infant whose Waterston shunt thrombosed two weeks after initial operation died in the operating room during an attempted revision of the shunt. Thrombosis required revision of a Blalock-Taussig shunt four days after operation and of a Waterston shunt two months after operation. One narrow Blalock-Taussig shunt was revised two years following initial surgery.

One patient developed areas of ischemic necrosis of the right arm following a right Blalock-Taussig shunt. The arm required surgical debridement and skin grafting. No other serious complications occurred.

Interim Period

Of the 54 hospital survivors, three died before intracardiac repair was carried out. All had operations in the early years of this series. One infant with a Waterston shunt died suddenly at home one month after hospital discharge. A second patient who had heart failure following a Blalock-Taussig shunt was readmitted with a “flu-like” illness four months postoperatively and died suddenly during that hospitalization. The third patient, age 7 years, was symptomatically improved by a Waterston shunt made at age 15 months. Two months before death he was found to be slightly more cyanotic. Plans were made for restudy and definitive repair, but he died suddenly while playing in the snow. Two patients were successfully treated for at least one episode each of subacute bacterial endocarditis. No patients developed systemic thrombosis or brain abscess during the interim period.

Intracardiac Repair

Twenty-six of the surviving 51 patients have so far undergone intracardiac repair with two hospital deaths (8%). Twenty-four in this group had repair since 1973, with one hospital death (4%). Fifteen had previous Waterston anastomoses, 10 had Blalock-Taussig shunts and one had a Glenn anastomosis. One patient died during reoperation on the first postoperative day because of an additional anomaly involving the tricuspid valve. A second patient died of sepsis on the 11th postoperative day. Ages at the time of intracardiac repair ranged from 25 months to 10 years, 3 months (mean 6 years, 4 months). Mean duration from time of shunt to intracardiac repair was 5 years, 5 months (range 20 months to 9 years, 9 months). Intracardiac operations were elective in 18 patients and precipitated by increasing cyanosis or hypoxic spells in eight patients.

Height and weight were available for 23 of the 26 children at the time of complete surgical repair. These measurements were plotted against the patient’s age at the time of repair using a standard anthropometric chart (fig. 1). The median height for the eight boys fell in the third percentile and ranged from the second to the 97th percentile. The median weight also fell in the third percentile and ranged from the second to the 45th percentile. Likewise, the median weight for the 15 girls fell in the third percentile and ranged from the

Table 2. Palliative Shunts for Tetralogy of Fallot in Infants Under 2 Years of Age (1971-1977)

<table>
<thead>
<tr>
<th>Type of shunt</th>
<th>Under 1 month</th>
<th>1–3 months</th>
<th>4–6 months</th>
<th>7–12 months</th>
<th>13–24 months</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Waterston</td>
<td>4</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>Rt. Blalock-Taussig</td>
<td>1</td>
<td>1</td>
<td>7</td>
<td>6</td>
<td>6</td>
<td>15</td>
</tr>
<tr>
<td>Lt. Blalock-Taussig</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>Total</td>
<td>6</td>
<td>2</td>
<td>5</td>
<td>9</td>
<td>9</td>
<td>31</td>
</tr>
</tbody>
</table>
second to the 70th percentile. Their median height was in the 10th percentile and ranged from the third to the 75th percentile.

In 15 of the 26 patients (60%) a right ventricular outflow patch was inserted during definitive repair; one child required a valved external conduit.

One patient who survived a Glenn anastomosis at age 5 months for tetralogy of Fallot with absent pulmonary valve leaflets had definitive repair at age 11 years. The Glenn anastomosis was not taken down. Chronic heart failure developed and one year later the Glenn anastomosis was taken down, a heterograft pulmonary valve was inserted, a tricuspid annuloplasty was performed and a dacron graft was inserted between the right pulmonary artery and the main pulmonary artery. This child developed severe pulmonary hypertension and died 10 months later with necropsy evidence of severe bilateral pulmonary vascular disease. All of the other children who survived intracardiac repair are alive and well.

**Discussion**

Recommendations for intracardiac repair of tetralogy of Fallot in infancy were initially based upon the high mortality rate associated with shunt operations in this age group (table 3). In the years preceding widespread use of hypothermic extracorporeal techniques for open heart surgery in infants, palliative shunt operations had a mortality of 40% in infants less than 3 months of age and 25% in those less than 1 year.\(^{15-21}\) Furthermore, few infants less than 1 year of age had Blalock-Taussig anastomoses; most had Waterston anastomoses, which are difficult to control. With magnification and microsurgical techniques, Blalock-Taussig anastomoses can now be performed in most symptomatic infants with tetralogy of Fallot (table 2). This shunt is associated with fewer late complications and is easier to take down than the Waterston anastomosis.\(^ {24}\) The Potts and Glenn shunts are no longer recommended for these infants. As documented by the results of operations between 1971-1977, Blalock-Taussig and Waterston anastomoses can be performed in symptomatic infants with tetralogy of Fallot with low mortality risk.

Interim mortality is generally low; Puga reported two late deaths in a series of 56 children under 4 years of age who had Blalock-Taussig shunts.\(^ {15}\) Heart failure and anatomic distortion of the pulmonary artery at the site of the anastomosis are uncommon with the Blalock-Taussig shunt. The Waterston shunt is associated with more complications, specifically, heart failure, development of pulmonary vascular disease, and distortion or occlusion of the right pulmonary artery.\(^ {24, 25}\) Cerebrovascular occlusions or brain abscesses may also occur in these patients with persistent right-to-left shunts across the ventricular septum.\(^ {26}\)
Two of the three interim deaths in the present series were clearly related to problems of the shunt. Cardiac failure after a Waterston shunt caused the death of one infant one month after hospital discharge. The other death of the 7-year-old boy emphasizes the need for close follow-up and earlier rather than later intracardiac surgery. Elective intracardiac repair is preferred before the age of 5 years, but can be performed earlier if the shunt is inadequate or poorly tolerated. The third child who died in the interim period died of an infection which was probably unrelated to either the shunt or heart disease.

The current mortality rate for intracardiac repair of tetralogy of Fallot is reported to be 1–8% in children over 4 years of age. Two of the three interim deaths in the present series were clearly related to problems of the shunt. Cardiac failure after a Waterston shunt caused the death of one infant one month after hospital discharge. The other death of the 7-year-old boy emphasizes the need for close follow-up and earlier rather than later intracardiac surgery. Elective intracardiac repair is preferred before the age of 5 years, but can be performed earlier if the shunt is inadequate or poorly tolerated. The third child who died in the interim period died of an infection which was probably unrelated to either the shunt or heart disease.

The current mortality rate for intracardiac repair of tetralogy of Fallot is reported to be 1–8% in children over 4 years of age.2, 6, 11, 27, 28 Take down of Blalock-Taussig or Waterston anastomoses does not appear to increase mortality of the operation; however, take down of a Waterston shunt often requires pulmonary arterial angioplasty.5, 11, 24 The percentage of children requiring a right ventricular outflow patch should be less in older children than in those who undergo primary intracardiac repair in infancy, but the criteria for placing an outflow patch varies among surgeons. Occasionally, patients may require a valved conduit. In some patients, a ventriculotomy can be avoided entirely by the transatrial approach.28 All patients with tetralogy of Fallot, including those with anomalous left anterior descending coronary arteries, single pulmonary artery and multiple ventricular septal defects, can be treated initially by primary shunt operations; there are virtually no general exclusions. Late results of intracardiac repair are good to excellent in 85–90% of patients who have undergone staged repair.

The average mortality for the repair of tetralogy of Fallot in infancy is reported to be about 14% (table 4). Severe, permanent, neurologic complications associated with the extracorporeal, hypothermic circulatory arrest technique used in infancy has been reported.29 It is not known whether late sequelae of the early operation will be more serious or more common than after intracardiac repair at a later age.

There are significant social, economic and psychologic benefits of intracardiac repair in infancy, and experience may show that growth and development of the child are better than in patients who have had palliative shunts. This study, for example, shows that patients who undergo shunt first and the intracardiac repair later, have retardation of physical growth at least until the time that the total intracardiac repair is performed. Furthermore, in those patients who undergo primary intracardiac repair in infancy, anatomic complications of the shunt anastomosis are avoided and late hemodynamic measurements may show increased cardiac functional reserve. Closure of the ventricular septal defect prevents cerebrovascular insults from right-to-left shunting. At present the long-term benefits or complications of intracardiac repair of tetralogy of Fallot in infancy are not known.

<table>
<thead>
<tr>
<th>Author</th>
<th>Years spanned</th>
<th>Type of shunt</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Puga15</td>
<td>1955–1967</td>
<td>Blalock</td>
<td>9/3*</td>
</tr>
<tr>
<td>Mustard16</td>
<td>1964–1968</td>
<td>Mixed</td>
<td>30/12</td>
</tr>
<tr>
<td>Bernhard17</td>
<td>1964–1969</td>
<td>Waterston</td>
<td>12/6</td>
</tr>
<tr>
<td>Barrett-Boyce18</td>
<td>1960–1971</td>
<td>Waterston</td>
<td>12/6</td>
</tr>
<tr>
<td>Truecone19</td>
<td>1954–1972</td>
<td>Mixed</td>
<td>8/2</td>
</tr>
<tr>
<td>Lakes20</td>
<td>1961–1974</td>
<td>Blalock</td>
<td>12/3</td>
</tr>
<tr>
<td>Idriss41</td>
<td>1967–1974</td>
<td>Waterston</td>
<td>30/5</td>
</tr>
<tr>
<td>Chopra22</td>
<td>1972–1975</td>
<td>Blalock</td>
<td>7/0</td>
</tr>
<tr>
<td>Arciniegas23</td>
<td>1974–1977</td>
<td>Blalock</td>
<td>9/0</td>
</tr>
</tbody>
</table>

| Total           | 50/20         | 119/31        | 255/56         |

*Hospital deaths.
The overall experience at the Children’s Hospital in Philadelphia since 1965 indicates an 11.4% mortality for the shunt operation, 5.6% interim mortality, and a 7.7% mortality for intracardiac repair. This yields a 25% mortality rate for staged surgical management of symptomatic infants with tetralogy of Fallot. However, more recent experience (1971–1977) indicates that the mortality has been substantially reduced. In the recent period, the cumulative mortality rate of staged management is less than 5%. Almost all infants with tetralogy of Fallot are eligible for a Blalock-Taussig anastomosis; a few may require a Waterston shunt. If shunted patients are carefully followed and corrected before cyanosis and polycythemia become severe, interim mortality and morbidity risks are minimal. Thus, the cumulative mortality of staged surgical management of tetralogy of Fallot in infancy since 1971 is less than the 14% average mortality rate reported for primary intracardiac repair during a similar period.

Eventually, questions about long-term complications, hemodynamics and growth after primary repair of tetralogy of Fallot in infancy will be answered. If all of these considerations are equal, one operation is preferable to two. Operative mortality of primary intracardiac repair is decreasing as patients with unfavorable anatomic features are recognized and excluded. The present experience strongly justifies palliative shunt operations for infants with unfavorable anatomy, additional lesions, or unfavorable clinical conditions. Our data also show that the case for primary intracardiac repair of tetralogy of Fallot in infancy cannot rest upon mortality and morbidity data alone, as good or better results are obtained by staged management. Our recent experience and the experience of others (table 4) justifies primary intracardiac repair of tetralogy of Fallot in those infants who have favorable anatomy and have no additional lesions or medical problems. A first stage Blalock-Taussig anastomosis is preferred for those with associated lesions, unfavorable anatomy or additional medical problems.

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L W Stephenson, S Friedman and L H Edmunds, Jr

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