Systemic-Pulmonary Arterial Shunts in the First Year of Life

By Nestor J. Truccone, M.D., Frederick O. Bowman, Jr., M.D., James R. Malm, M.D., and Welton M. Gersony, M.D.

SUMMARY

Results of systemic to pulmonary arterial shunts during the first year of life are reported in 86 infants under one year of age. Survival rates were highest in older patients and among infants whose basic cardiac defects were considered to be eventually amenable to surgical repair. Persistent hypoxia accounted for the majority of deaths. Congestive heart failure secondary to too large anastomoses occurred in 32.5% of Waterston shunts, but could be successfully managed medically in the majority of patients. Surgical revision of the shunt was accomplished successfully in two infants with subsequent alleviation of symptoms. Congestive heart failure occurred in only one of 26 patients after a Blalock shunt. Ten of the twelve patients in this series survived open-heart repair for cardiac lesions. It will eventually be necessary to compare these data with the results of single stage corrective surgery in the infant group.

Additional Indexing Words:
- Blalock-Taussig
- Potts
- Waterston
- Cyanotic
- Congenital heart disease
- Tetralogy of Fallot
- Pulmonary atresia
- Tricuspid atresia

The value of systemic-pulmonary arterial shunt procedures as palliative treatment for cyanotic patients with diminished pulmonary blood flow secondary to congenital heart disease has been well established. While this type of surgical intervention can be carried out successfully with a relatively low risk in children, results have been less satisfactory in infants under one year of age.\(^1\,\,2\,\,3\)

Recently, however, the development of newer surgical techniques\(^4\) as well as improvements in diagnostic procedures, anesthesia, and infant perioperative care\(^5\) have contributed to a more favorable outlook for small infants requiring systemic-to-pulmonary arterial shunts. Furthermore, rapid progress in the techniques of open-heart surgery would be expected to allow total correction to become a reality for an increasing number of patients requiring early palliative shunting procedures.\(^6\) However, late results of attempts at open-heart repair for these patients have not been specifically reported.

The purpose of this paper is to present results of an extensive study of patients with systemic-pulmonary arterial anastomoses in the first year of life. The results attained by the use of various types of shunting procedures are evaluated in relation to the type of cardiac defect, the age at which surgery was carried out, and the complications encountered. Follow-up data regarding later corrective surgery are presented. It is expected that these data will serve as a baseline for comparison with results of primary total repair of similar cardiac malformations in infancy.

Materials and Methods

The records of 86 infants under one year of age, who underwent systemic-pulmonary arterial shunts at the Columbia-Presbyterian Medical Center between the years 1954 and 1972, were reviewed. Twenty-eight patients (32.5%) were less than 30 days of age at the time of surgery, 39 infants (45.3%) were between 30 days and six months, and 19 patients (22.0%) were operated upon after six months of age. The duration of the follow-up period ranged from three months to nine years (mean = 4 years). All of the patients had significant cyanosis, often associated with hypoxic episodes, precipitating surgical intervention during early infancy.

Potts\(^7\) and Waterston\(^8\,\,9\,\,10\) shunts were most often carried out in the infants under six months of age. The Waterston shunt replaced the Potts anastomosis after

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1964 as the procedure of choice for this age group. Throughout the study period, the Blalock-Taussig anastomosis was most often utilized after six months of age (table 1).

Diagnostic cardiac catheterization and angiography were carried out preoperatively in 78 patients (90.6%). The specific anatomic diagnosis was made by autopsy findings in four cases (4.7%), and by cardiac catheterization following the systemic-pulmonary shunt in three cases (3.5%). The diagnosis was based on clinical findings in one case (1.2%).

The patients were divided into two groups, depending upon feasibility of later total correction. Fifty-six infants (group A) had lesions potentially correctable by open-heart surgery (table 2). In this group the most common lesions were tetralogy of Fallot (31 patients), and pulmonary atresia with intact ventricular septum (13 patients). Thirty infants (group B) had complex cardiac anomalies, for which definitive corrective procedures are not currently available (table 3). The most common lesion in this group was tricuspid atresia (12 cases).

Results

Mortality data are shown in tables 2 and 3. Of the 56 infants with lesions potentially correctable by open-heart surgery (group A), 47 survived the initial systemic-pulmonary arterial shunt procedure (84%). Ten of these patients have since successfully undergone open-heart correction of their defects.

There have been 10 late deaths (>1 month after surgery) including two patients who died during attempted total correction. Of the 30 patients with complex cardiac anomalies not amenable to total corrective procedures (group B), only 11 patients are long-term survivors. There were 13 early deaths and six late deaths in this group.

Hypoxia, secondary to inadequately functioning systemic-pulmonary shunts (10 patients), and congestive heart failure due to excessively large anastomoses (two patients), accounted for the majority of the late deaths in both groups. The patients older than 30 days of age when surgery was carried out were better able to achieve long-term survival (table 4). The more mature infants were more likely to have a potentially correctable lesion (table 5), and were of sufficient size and weight to undergo a Blalock-Taussig operation rather than Potts or Waterston procedures. The latter operations were associated with a greater incidence of congestive heart failure in the postoperative period. However, surgical results for the youngest infants have improved recently. Seven of the last eight babies under 10 days of age who have had Waterston shunts have survived.

Twenty-one of the 86 patients in this series developed symptoms of congestive heart failure following the systemic-to-pulmonary arterial shunt. Fifteen were among the 46 patients who underwent Waterston shunts (32.6%). Two of these patients required revision of the anastomosis and both survived. Thirteen patients underwent vigorous medical therapy; eleven responded satisfactorily and two patients died. Five of 14 infants with Potts shunts developed congestive heart failure (35%), and there were three deaths. Only one of 26 patients with Blalock-Taussig anastomoses devel-

Table 1

<table>
<thead>
<tr>
<th>Age at surgery</th>
<th>Type of shunt</th>
<th>Waterston</th>
<th>Potts</th>
<th>Blalock</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt; 30 days</td>
<td></td>
<td>24</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>1 – 6 mos.</td>
<td></td>
<td>18</td>
<td>7</td>
<td>13</td>
</tr>
<tr>
<td>6 mos. – 1 yr.</td>
<td></td>
<td>4</td>
<td>3</td>
<td>13</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>46</td>
<td>14</td>
<td>26</td>
</tr>
</tbody>
</table>

Table 2

<table>
<thead>
<tr>
<th>Correctable Lesions</th>
<th>No. of patients</th>
<th>Long term survival</th>
<th>Early Deaths</th>
<th>Late Deaths</th>
<th>Successful repair</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot</td>
<td>31</td>
<td>23</td>
<td>5</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Pulmonary atresia with intact ventricular septum</td>
<td>13</td>
<td>6</td>
<td>3</td>
<td>4(1)</td>
<td>2</td>
</tr>
<tr>
<td>VSD with pulmonary atresia</td>
<td>6</td>
<td>4</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>T.G.V., V.S.D. and pulmonary atresia</td>
<td>3</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Double outlet R.V. and P.S.</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Tricuspid stenosis with small V.S.D.</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1(1)</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>56</td>
<td>37(66%)</td>
<td>9</td>
<td>10(2)</td>
<td>10</td>
</tr>
</tbody>
</table>

Early Deaths: < than 30 days postoperative or before hospital discharge.
Late Deaths: > than 30 days postoperative.
( ) Died as result of attempted total repair.
Abbreviations: TGV = transposition of the great vessels; VSD = ventricular septal defect; PS = pulmonic stenosis; RV = right ventricle.
Tricuspid atresia
Corrected T.G.V., single ventricle, and pulmonary atresia
Single ventricle with pulmonary atresia
Ebstein's anomaly
T.G.V., hypoplastic left ventricle and P.S.
Tricuspid stenosis with hypoplastic right ventricle
T.G.V., with intact ventricular septum, pulmonary atresia
and P.A.P.V.R.
Total

Abbreviations: TGV = transposition of the great vessels; PAPVR = partial anomalous pulmonary venous return; FS = pulmonic stenosis.

ophed cardiac decompensation in the postoperative period (3.8%), and this child responded well to medical management.

In the over-all series, a second systemic-to-pulmonary shunt was required in ten patients because of inadequate function of the initial systemic-pulmonary arterial shunts. Repeat surgery accounted for one of the early deaths and four patients succumbed later.

One of 86 infants in this series developed bacterial endocarditis during the follow-up period. The child was managed successfully with antibiotic therapy, and total correction was carried out 1½ years later with an excellent outcome.

Discussion

Systemic-to-pulmonary arterial shunts have been associated with a higher mortality rate in the first six months of life than later in childhood.1 2 3 The increased risk in early infancy is due to two primary factors. First, there is more likelihood that the cardiac lesion will be of greater complexity. Our study demonstrates that babies with cyanotic heart disease severe enough to virtually preclude the possibility of future repair have higher early and late mortality rates after shunts than infants with potentially correctable defects. In this situation, pulmonary outflow tract obstruction may be only one of numerous hemodynamic abnormalities. Thus, systemic-to-pulmonary arterial shunting may not improve the patient's status to the degree which may be achieved in infants with less complicated anatomic lesions. Bernhard and his associates1 also have reported additional cardiac anomalies as a major cause of mortality in a series of 80 infants who underwent Waterston shunts.

The second factor accounting for higher rates of mortality in the younger patients is related to the type of surgical procedure which is carried out. With few exceptions, small infants require a Waterston or Potts anastomosis since the Blalock-Taussig operation has achieved lillt success in this age group.10, 11 Although technically more feasible, the side-to-side pulmonary artery-aortic anastomosis allows little margin for error between the extremes of an anastomosis being too small and the risk of clotting, and an anastomosis being too large resulting in postoperative congestive heart failure. The Potts anastomosis was the procedure utilized for small infants in the years prior to 1964. In the past decade, however, the Waterston shunt has been utilized because this anastomosis may be closed more easily at the time of total correction.

Experience at this institution indicates that congestive heart failure secondary to excessive pulmonary blood flow is the prominent complication of Waterston and Potts shunts carried out.

Table 3

Non-correctable Lesions

<table>
<thead>
<tr>
<th>Defect</th>
<th>Total no. of patients</th>
<th>Long term survivors</th>
<th>Early deaths</th>
<th>Late deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tricuspid atresia</td>
<td>12</td>
<td>5</td>
<td>5</td>
<td>2</td>
</tr>
<tr>
<td>Corrected T.G.V., single ventricle, and pulmonary atresia</td>
<td>9</td>
<td>2</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>Single ventricle with pulmonary atresia</td>
<td>4</td>
<td>0</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Ebstein's anomaly</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>T.G.V., hypoplastic left ventricle and P.S.</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Tricuspid stenosis with hypoplastic right ventricle</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>T.G.V., with intact ventricular septum, pulmonary atresia and P.A.P.V.R.</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>30</td>
<td>11 (37%)</td>
<td>13 (43%)</td>
<td>6 (20%)</td>
</tr>
</tbody>
</table>

Table 4

Surgical Results According to Age Group

<table>
<thead>
<tr>
<th>Age</th>
<th>No. of pts.</th>
<th>Early deaths</th>
<th>Late deaths</th>
<th>Long term survivors</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 – 30 days</td>
<td>28</td>
<td>8 (29%)</td>
<td>8</td>
<td>12</td>
</tr>
<tr>
<td>1 – 6 mo.</td>
<td>38</td>
<td>10 (26%)</td>
<td>7</td>
<td>21</td>
</tr>
<tr>
<td>6 mos. – 1 yr.</td>
<td>20</td>
<td>4 (20%)</td>
<td>1</td>
<td>15</td>
</tr>
<tr>
<td>Total</td>
<td>86</td>
<td>22 (26%)</td>
<td>16 (19%)</td>
<td>48 (56%)</td>
</tr>
</tbody>
</table>

Table 5

Distribution of Correctable and Non-correctable Lesions According to Age

<table>
<thead>
<tr>
<th></th>
<th>0-30 days</th>
<th>30 days-6 mos.</th>
<th>6 mos.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Correctable</td>
<td>12</td>
<td>25</td>
<td>19</td>
</tr>
<tr>
<td>Non-correctable</td>
<td>16</td>
<td>13</td>
<td>1</td>
</tr>
</tbody>
</table>

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during infancy. In most instances, the patient can be managed successfully with digitalis and diuretics. However, when the clinical condition continues to deteriorate, despite an optimal medical regimen, a revision of the anastomosis to reduce the size of the shunt may be attempted with reasonable hope of success. This was accomplished in two patients with Waterston shunts in this series. If the size of the anastomosis is too small, resulting in initial inadequate pulmonary blood flow and persistent hypoxemia, further surgery to enlarge the size of the anastomosis may be indicated. If the initial palliative operation allows the patient to reach the age of six months or more, it is preferred to carry out a corrective procedure rather than a second palliative operation. Indeed, with increasing success in open-heart surgery for infants, it is now possible to carry out corrective surgery in lieu of initial palliative shunts at any age in patients with a suitable anatomy.

There was not a high incidence of bacterial endocarditis in this series of patients followed during the first decade of life. One child developed bacterial endocarditis five years after a Waterston shunt for tetralogy of Fallot. The incidence of bacterial endocarditis in this group of infants and young children is similar to that reported by Cole and his associates in children of all ages with systemic-to-pulmonary anastomoses for tetralogy of Fallot.

On the basis of this follow-up study, it may be concluded that a vigorous surgical approach is indicated for hypoxic infants with pulmonary outflow obstruction, since the possibilities for long-term survival are good. If corrective operations are not considered advisable, Waterston shunts are advocated for the patient less than six months of age, and Blalock-Taussig anastomoses are carried out after the age of six months.

References

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