The Need for Systemic–Pulmonary Artery Shunts in the Surgical Management of Tetralogy of Fallot

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In 1944, Blalock and Taussig demonstrated that surgical establishment of shunts between the aorta and pulmonary artery brought partial, but dramatic, relief for patients suffering from the tetralogy of Fallot. Literally thousands of humans have received palliation from this important operation. In 1955, Lillicrith and his associates were the first to develop methods for the total, intracardiac correction of the deformity. With this later and brilliant step, the feeling developed in the minds of many cardiologists and surgeons that shunt operations were of little or no further use. This attitude came about because of the belief that total correction could probably be offered to all patients, could be completed in a single operative step, and the mortality rate of such treatment could be brought down to an acceptable level. However, in the last few years, surgical experience in many cardiac centers throughout this country clearly shows that while total correction in one step can often be achieved and a highly satisfactory result obtained, a very large number of deaths occur when complete repair of the cardiac malformation is attempted in a single stage. This has led us to believe that there is probably still a very important place for the use of systemic–pulmonary arterial shunts in preparing some patients for the more drastic procedure of total correction. We have therefore restudied, and summarized herewith, our past experience with operative establishment of shunts, proposing to formulate some policies which will govern us in the management of tetralogy cases in the future.

Clinical Data

Series Studied

Systemic–pulmonary arterial anastomoses (Potts, 223; Blalock-Taussig, 164) were created in 370 patients with tetralogy of Fallot during the years 1947 to 1960 at the Children’s Hospital Medical Center. Seventy-six were infants aged 2 years or under at the time of operation, while 294 ranged from 2 to 26 years of age. A second shunt was required in 17 patients (of both age groups) because of shunt closure or inadequate pulmonary blood flow.

Mortality Statistics

The operative mortality (death occurring within 30 days after shunt operation) in the infant group was 10 per cent (8 of 80 cases), compared with 6.5 per cent (21 of 307 cases) for patients older than 2 years. Early deaths were attributed primarily to shunt thrombosis or intractable congenital heart failure. Follow-up data ranging from 2 to 14 years revealed that an additional 10 per cent of the infants who survived operation had since expired. In older patients, there was an additional 6.5 per cent mortality during the same period, with death caused by congestive heart failure, thrombosis of shunts, thrombosis of branches of the circle of Willis, and brain abscess (table 1).

Clinical Appraisal of Survivors

An evaluation of all living patients is presented in table 2.

In the infant group, it was found that there was a higher percentage of good and excellent results with the Potts anastomoses than with the Blalock shunts.

Of 17 patients who underwent a second systemic–pulmonary arterial shunt because of shunt

Table 1

<p>| Mortality in Systemic–Pulmonary Arterial Shunts |</p>
<table>
<thead>
<tr>
<th>Infants</th>
<th>Patients over two years old</th>
</tr>
</thead>
<tbody>
<tr>
<td>Operative mortality</td>
<td>10</td>
</tr>
<tr>
<td>Late deaths</td>
<td>10</td>
</tr>
<tr>
<td>Overall mortality rates</td>
<td>20</td>
</tr>
</tbody>
</table>

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thrombosis or inadequate pulmonary blood flow, nine had their initial shunt at less than two years of age, and all these shunts had been subclavian-pulmonary arterial anastomoses.

Postoperative Catheterization Studies
Cardiac catheterization was carried out in 50 of the shunted patients (table 3). This was a highly selected group of cases. Some were chosen for study because of suspected pulmonary arterial hypertension on the basis of the patient’s clinical evaluation, serial electrocardiograms, and x-rays. Others were catheterized for study prior to total corrective operation. The interval between shunt operation and catheterization ranged from 1½ to 13 years, the average being 9 years. A patient was considered to have pulmonary arterial hypertension if the pulmonary arterial systolic pressure was greater than 30 mm. Hg. Resistances greater than 3.0 units/M.² were considered to be abnormally elevated.

Ten of the 50 patients studied by catheterization showed pulmonary hypertension. Seven of these had increased pulmonary vascular resistance. All of the 10 had undergone aortic–pulmonary arterial (Potts) anastomoses. Since most of these catheterizations were done many years after establishment of the shunts, we do not have any information concerning the time when the pulmonary arterial hypertension developed.

Discussion
The recognition of the great difference in surgical outlook between the “pink” tetralogy and the severely cyanotic patient is important in consideration of mortality rates of one-stage, total correction of tetralogies. While there is not a sharp differentiation between the two groups, in general they can be roughly separated. In the pink type, there is a minimal infundibular obstruction and a predominant left-to-right shunt; the resting arterial saturation may be normal or depressed to as low as 85 per cent. In contrast, the “blue” tetralogy has a more marked infundibular (or pulmonic valve) block, a predominant right-to-left shunt, and a resting arterial oxygen saturation below 85 per cent. Published reports, and data from our clinic, clearly show that one-stage, total correction is highly satisfactory for the pink variety, but that it involves a very high risk for the cyanotic type.

In all new fields of surgery, increasing experience and development of technical skill on the part of the surgeon will gradually reduce mortality rates of an operative procedure. Hopefully, such a trend will be noticed in the future handling of correction of the tetralogy of Fallot as surgeons achieve greater mastery of the reparative techniques. However, it is obvious that the presently high mortality rates cannot be blamed entirely on the surgeons’ inexperience. The anomaly itself has certain inherent factors which can bring on fatality, even though an excellent anatomical repair has been made in the heart. The bronchial blood flow may be very high, giving a congested “pump lung” postoperatively. The pulmonary vessels may be small and may not permit enough blood to get through to the left heart. The left ventricle may be underdeveloped and unable to handle a full work load. The blood may have clotting deficiencies leading to postoperative bleeding. All of these various factors can militate against a successful operative result.

Table 2
Clinical Appraisal of 259 Survivors with Systemic–Pulmonary Arterial Shunts

<table>
<thead>
<tr>
<th>Patients over two years old</th>
<th>Infants</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excellent</td>
<td>8</td>
<td>12 (6%)</td>
</tr>
<tr>
<td>Good</td>
<td>79</td>
<td>122 (59%)</td>
</tr>
<tr>
<td>Fair</td>
<td>25</td>
<td>47 (22%)</td>
</tr>
<tr>
<td>Poor</td>
<td>14</td>
<td>27 (13%)</td>
</tr>
</tbody>
</table>

Excellent = asymptomatic with full range of activities and no cyanosis.
Good = normal activities with moderate cyanosis and dyspnea.
Fair = improvement since surgery, but physical activity still limited.
Poor = no improvement with shunt.
When faced with a severely cyanotic infant or young child with symptoms (such as syncope) serious enough to demand operation, it is widely agreed that creation of a palliative systemic–pulmonary arterial anastomosis is the procedure of choice. Total correction can be contemplated some years later. Such a two-stage approach has been widely used and successfully managed in many clinics. The problem demanding our attention is how to handle the patient over seven to eight years old who has had no previous shunt, but who now seeks surgical relief. If he has the pink variety of clinical picture, total correction can be offered with relative safety. However, if he is a cyanotic type, with resting arterial saturations below 85 per cent, severe incapacitation, marked polycythemia, and small pulmonary arteries (by angiogram), open correction in one stage has very little hope of successful result, regardless of the fact that the patient may be of rather large size and aged even above 10 to 12 years. Therefore, we have come to believe that all patients of this type should (regardless of an older age or a large body size) be handled first by creating a shunt between the aorta and pulmonary artery (preferably of the Blalock type). Such a shunt subsequently enlarges the pulmonary arteries, increases the capacity of the left ventricle, diminishes the polycythemia, and decreases the abnormal bronchial circulation. Admittedly, this policy subjects an occasional individual to two operations when he might have been cured by one; however, when the principle is applied to a series of patients, almost certainly more of them will survive.

Summary

When managing infants and small children with severe enough symptoms to require operative intervention, there can be no doubt that the establishment of a shunt is highly efficacious. Total repair can be contemplated some years later. If technically feasible, a shunt of the Blalock-Taussig type is the desired form of procedure. When dealing with very small infants, a Potts anastomosis has a higher probability of remaining patent and is therefore more desirable, even though it might be more difficult to obliterate at the second operation.

In older or larger children with the pink tetralogy, total correction in one stage can be advised with an acceptably low mortality rate. However, in older subjects with severe cyanosis, it is now our policy in nearly all cases to employ a two-stage attack in the form of a preliminary systemic–pulmonary arterial shunt, proceeding with complete correction some years later.

References


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