Surgical Palliation of Tricuspid Atresia

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SUMMARY

In the 20-year period ending December 31, 1973 we operated on 105 patients for palliation of tricuspid atresia (TA) with reduced pulmonary blood flow. Potts’ anastomosis (85), Blalock-Taussig anastomosis (19), intrapericardial aorta (Ao)-to-right pulmonary artery (RPA) (18), Glenn procedure (3) and miscellaneous shunts (2) have been used. Of patients undergoing operation more than 15 years ago, 45% (9/20) are still alive. The over-all operative mortality was 9%. It was highest in patients less than one month of age (7/23, 30%). There were no operative deaths in patients older than 12 months. The median age at first operation for the entire series was five months. Our results indicate the Potts’ anastomosis and Ao-to-RPA anastomosis are superior to the Blalock-Taussig anastomosis (BT) for palliation of TA. Of patients surviving BT, 69% (11/16) required reoperation, compared to 22% (17/78) surviving Potts’ anastomosis and 13% (2/16) surviving Ao-to-RPA shunts. Operative mortality was 8% (7/85) for Potts’ anastomosis, 16% (3/19) for BT, and 11% (2/18) for Ao-to-RPA shunts. All three patients undergoing Glenn procedures (superior vena cava-to-RPA anastomosis) required reoperation or died without significant benefit.

Balloon atrial septostomy and Potts’ anastomosis appear to be indicated for infants less than six months of age, and Ao-to-RPA shunt with simultaneous atrial septectomy (if indicated) for older children.

Until recently palliative procedures were the only surgical therapy available for patients with tricuspid atresia. Fontan’s description1 of a “corrective” operation for this anomaly necessitates a thorough evaluation of the palliative treatment. Newer, more radical procedures for any condition must be measured against the yardstick of the prior methods of treatment. With this objective, we reviewed our experience with the surgical palliation of tricuspid atresia.

Materials and Methods

One hundred and five patients with tricuspid atresia and reduced pulmonary blood flow underwent surgical palliation at our institutions in the twenty-year period ending December 31, 1973. Patients with increased pulmonary blood flow (ten) or complex congenital heart disease (seven) of which tricuspid atresia was only one of multiple defects are excluded.

The 61 female and 44 male patients ranged from two days to 41 years at the time of their first operation. The median age of the patients at the time of their first procedure was five months. Twenty-three patients (22%) underwent operation before one month of age. One hundred and two patients underwent preoperative cardiac catheterization. Repeat cardiac catheterizations were performed when indicated by the clinical status of the patients.

For the purposes of comparison, the patients are divided into four groups. The operative techniques are described elsewhere.2

Group I

Eighty-five patients underwent descending aorta to left pulmonary artery shunts (Potts’ anastomosis). These patients ranged in age from two days to forty-one years. Excluding four patients over the age of 15 years, the average age at operation was 21 months. Twelve patients were less than one month of age and 47 were less than one year of age. The median age of the entire group was 4½ months.

Group II

Nineteen patients received subclavian artery-to-pulmonary artery anastomoses (Blalock-Taussig). Their average age was 32 months, ranging from less than one month to 12 years. The median age of this group was eight months. Ten patients were less than one year of age, and three of these were less than one month of age. Because the subclavian artery originating from the innominate artery is less likely to kink when anastomosed to the pulmonary artery than the subclavian artery originating from the aorta, these shunts were performed on the side opposite the aortic arch whenever possible. This approach has the additional benefit of leaving the pulmonary artery on the same side as the descending aorta available for a Potts’ anastomosis in the future.

Group III

All patients in this group underwent intrapericardial aorta-to-right pulmonary artery anastomosis. The 18 patients undergoing this procedure ranged from a few days of age to 18 years. The median age was 24 months. Five patients were less than one month of age.

Group IV

This group includes three patients who underwent

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superior vena cava (SVC) to-RPA-shunts (Glenn procedure). The anastomosis was end-to-end in each case. Also included in this group are two patients in whom individualized shunts were performed using Dacron grafts. One patient was a one-year-old male with a right aortic arch and a previous left Blalock-Taussig shunt. Because the bronchus separated the descending aorta from the right pulmonary artery, a short piece of 5 mm Dacron graft was used to join them. The other patient, a 29-year-old male, had undergone a Blalock-Taussig procedure at age ten, and an ascending aorta-to-RPA shunt at age 18. His third shunt was constructed with a short Dacron graft from the ascending aorta to the main pulmonary artery.

Atrial septectomy using a modified Blalock-Hanlon technique was performed in nine patients. Four patients had signs and symptoms of high right atrial pressure, consisting of hepatomegaly, abdominal discomfort, neck vein distention and peripheral edema. Two of these four patients required atrial septectomy because a systemic-pulmonary anastomosis increased return to the left atrium and partially obstructed a patent foramen ovale. Five other patients who needed a systemic-to-pulmonary anastomosis also had a gradient between the right and left atrium demonstrable at cardiac catheterization. These patients underwent simultaneous atrial septectomy even though they had no symptoms related to elevated right atrial pressure.

Results

Among the 105 surgically treated patients, 32 are known to be dead yielding an over-all mortality rate of 30% (table 1). One hundred and forty-four separate operative procedures were performed: 13 deaths occurred within the first 30 postoperative days resulting in an over-all operative mortality of 9%. Nineteen patients are known late deaths for a late mortality of 21% (19/92). This figure is probably low because eight patients are lost to follow-up and some of these may be presumed dead. If all eight patients are presumed dead, the late mortality rate would be 29% (27/92) and the over-all mortality would be 38% (40/105).

The early and late mortality are related to the patient's age at the time of the first shunt (fig. 1). The graph clearly demonstrates that the older the child was when the first shunt was required, the more likely it was that the child would survive the operation and that the palliation would be effective for a long time. Stated another way, the more deranged the child's anatomy and hemodynamics, the earlier the child will require some form of palliation, and the shorter his over-all life expectancy.

The majority (66%, 85/129) of the systemic-to-pulmonary anastomoses performed were group I (Potts' anastomosis). Despite having the lowest median age at operation, this group's operative mortality was 8% (7/85), less than either group II (3/19, 16%) or group III (2/18, 11%). The average age of group I at late death was 71 months. The median age at late death in this group was considerably less, 24 months. The mean and median age of seven operative deaths in this group was three weeks.

Of special note in the results of group I is that 15% (13/85) of the patients required reoperation because the shunt was too large (fig. 2). This is in marked contrast to groups II, III and IV in which no patient required reoperation for an excessively large shunt. The time interval between the Potts' anastomosis and the

Table 1

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Total operations</th>
<th>Thrombosed shunt</th>
<th>Outgrew shunt</th>
<th>Required shunt reduction</th>
<th>Operative deaths</th>
<th>Late deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group I</td>
<td>85</td>
<td>2 (3%)</td>
<td>2 (3%)</td>
<td>13* (17%)</td>
<td>7 (8%)</td>
<td>16 (21%)</td>
</tr>
<tr>
<td>Potts'</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Group II</td>
<td>19</td>
<td>6 (38%)</td>
<td>5 (31%)</td>
<td>—</td>
<td>3 (16%)</td>
<td>2 (13%)</td>
</tr>
<tr>
<td>Blalock-Taussig</td>
<td></td>
<td></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Group III</td>
<td>18</td>
<td>—</td>
<td>2 (13%)</td>
<td>—</td>
<td>2 (11%)</td>
<td>—</td>
</tr>
<tr>
<td>AO-RPA (intrapericardial)</td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Group IV</td>
<td>3</td>
<td>1 (33%)</td>
<td>1 (33%)</td>
<td>—</td>
<td>1 (33%)</td>
<td>1 (50%)</td>
</tr>
<tr>
<td>Glenn</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Group IV</td>
<td>2</td>
<td>1 (50%)</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Miscellaneous shunt</td>
<td></td>
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</table>

*One death.
Group III patients demonstrated a much lower incidence of late shunt failure, 13% (2/16), compared to the 69% (11/16) of group II and 22% (17/78) in group I. A partial explanation for this difference may be found in the higher median age of 24 months of the patients undergoing intrapericardial Ao-to-RPA anastomosis, considerably older than the 5 and 8 months median ages of groups I and II, respectively. Creation of a proper sized shunt is easier in an older child and survival is longer in the older children (fig. 1). The operative mortality of group III was 11% (2/18); one child was less than a week old and the other was undergoing reoperation at age seven months after the failure of a Potts' procedure.

Group IV contained three patients who received SVC-to-RPA anastomoses (Glenn procedure). Our results with this procedure were poor. One seven-month-old patient died ten days after a Glenn procedure and another 16-year-old patient died a few months after this procedure, without showing any improvement (table 1). The third patient underwent a Glenn procedure at five months of age and required a second shunt (Potts') at age eight.

Two patients in group IV received "individualized" shunts. A 5 mm Dacron graft placed between the descending aorta and RPA functioned satisfactorily, but the child eventually outgrew it and required an ascending Ao-to-RPA shunt four years later. The other patient in this group improved somewhat after an ascending aorta-to-main pulmonary artery shunt at age 29, 18 years after a Blalock-Taussig procedure and 11 years after a Potts' shunt.

The operative mortality for different shunt procedures varied only slightly in the high risk group, those children less than 1 month of age: mortality in group I was 33% (5/15); group II was also 33% (1/3); and group III was 20% (1/5). The over-all operative mortality for patients less than one month of age was 30% (7/23). Eleven of 58 patients younger than six months died within 90 days following operation for an operative mortality in this age group of 19%. Sixteen patients between 6 and 12 months of age underwent palliative shunt procedures with one death, an operative mortality of 6% in that age category. No operative deaths occurred in 31 patients undergoing their first shunt procedure when they were older than 12 months.

We considered the possibility that the improved results in group III (fig. 2) were due to the fact that the Ao-to-RPA shunt was not used until the latter part of the time period under review. Thus the apparent improved results in group III might be due to improvement acquired with experience rather than any superiority of the shunt itself. Analysis of our results in relationship to the year of operation revealed that this

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Results of different systemic to pulmonary shunts for patients with tricuspid atresia.

The necessity to reduce its size ranged from 4 months to 13 years. The average interval was 5.3 years. Operative mortality from constriction was 8% (1/13) and four late deaths (4/12, 33%) occurred, one each at 45 days, 5 years, 10 years and 13 years after constriction. Three patients required another shunt an average of 5 years after the Potts' was constricted. It is difficult to be certain whether the second shunt was necessary because of growth of the child or because the Potts' anastomosis was constricted excessively. Of the 12 late survivors after banding of the Potts', 25% required a further shunt. All the children undergoing constriction were severely symptomatic from refractory congestive heart failure, and erring on the side of too much rather than too little reduction in size appeared to be a safer course. In the children who did not develop congestive heart failure and required no banding only 3% (2/65) required further shunts.

The results of group II differ markedly from group I (fig. 2). The operative mortality is higher (16% vs 8%) despite a higher median age at operation for group II. Thirty-eight percent (6/16) of the survivors in group II experienced thrombosis of their shunt and 31% (5/16) outgrew their subclavian-to-pulmonary anastomosis and required another shunt. This resulted in a reoperation rate of 69% (11/16).
was not the case (fig. 3). It is apparent from the figure that the number of shunt failures and operative deaths has not changed noticeably over the period of time examined. As expected, fewer late deaths and outgrown shunts occurred in patients recently operated on but otherwise the distribution in the graph appears even across the twenty-year period. From this information one could infer that the Ao-to-RPA shunt must have some superior features, but this must be only a tentative conclusion because of the differences in the median age of the groups. Although we first used this shunt for tricuspid atresia in 1961, it was not used frequently until the last five years of this series and the passage of more time may reveal more patients who outgrow it.

Nine patients operated on before 1959 are still alive (fig. 3). The initial shunt performed was a Blalock-Taussig for six patients and a Potts’ for three. All patients with the Blalock-Taussig anastomosis required another shunt. The average interval between shunts was eight years. Four of these patients underwent a Potts’ anastomosis and two an Ao-to-RPA shunt as their second procedure. Three of these long-term survivors were less than 6 months old when their initial shunt was performed, and a fourth was 11 months of age at first operation. The remaining three patients surviving beyond 15 years had Potts’ anastomosis as their initial shunt. These patients were 4 months, 2 years and six years old at first operation. The two younger patients required constriction of their Potts’ anastomosis 12 and 13 years later and are doing well. Recent follow-up on these nine patients revealed that most are doing remarkably well. One patient does farm work, another runs a paint store and plays golf frequently, another attends college and writes that she almost keeps up with her friends. One 31-year-old patient is able to work full time at an office job. These nine patients represent a long-term survival of 45% (9/20) of the patients operated on before 1959. Three additional patients survived more than 15 years after their palliative surgery; one died of subacute bacterial endocarditis and two of unknown causes. The 15-year survival rate is thus 60% (12/20).

Nine patients required operative atrial septectomy, the Blalock-Hanlon procedure. There were two operative deaths (22%). One occurred in a patient undergoing a simultaneous Ao-to-RPA shunt and the other occurred in a patient undergoing atrial septectomy in whom the endotracheal tube accidentally dislodged. Eighty-six percent (6/7) of the survivors were significantly improved, one patient was only modestly improved. This latter patient had a Potts’ anastomosis performed at five weeks of age and subsequently developed congestive heart failure. He remained in congestive heart failure after the atrial septectomy and required banding of the Potts’ anastomosis one year later. He died suddenly seven days after the third procedure.

The average postoperative oxygen saturation of these patients was 84%, a considerable improvement over the average preoperative value of 65%. There was no significant difference between the shunts in immediate postoperative oxygen saturation values. The average preoperative saturation for those patients who died early or late was 62%, slightly less than the 66% average preoperative saturation for the survivors.

Comment

Tricuspid atresia, defined as absence of the tricuspid valve and inflow portion of the right ventricle,
is an uncommon malformation. Abbot found 21 instances in 1000 autopsies performed on congenital cardiac patients. Taussig reported 56 cases of tricuspid atresia in a group of 1037 patients undergoing the Blalock-Taussig operation. Campbell also reported that children with tricuspid atresia constitute approximately 5% of those with cyanotic congenital cardiac malformations.

The clinical course of patients with tricuspid atresia is highly variable, and depends on the amount of pulmonary blood flow. This may be decreased, normal or increased and results from the presence of normal or transposed great arteries (TGA) and the presence or absence of obstruction to pulmonary blood flow. The classification of Edwards and Burchell in this regard is generally accepted. The majority of patients have normally related great arteries (type I) and decreased pulmonary blood flow because of a restrictive VSD or pulmonary stenosis or atresia. Some patients with transposed great arteries (type II) may have low pulmonary blood flow if sufficient pulmonic stenosis is present, but patients with TGA constitute the great majority of tricuspid atresia patients with increased pulmonary blood flow. A few patients tricuspid atresia have anatomy resulting in almost normal pulmonary blood flow. These patients may survive into adult life without surgical intervention. This group is unfortunately small and the majority of patients with tricuspid atresia will be severely ill at a very young age because of insufficient pulmonary blood flow and will die without palliative surgery.

An adequate interatrial communication is important for these patients. The development of balloon atrial septostomy (BAS) by Rashkind is an important contribution to the care of these infants. When diagnostic catheterization is performed before six months of age, BAS should be performed if there is any pressure difference between the right and left atrium or if the presence of large "a" waves in the right atrium indicates that the foramen ovale is being forced open with each atrial contraction. If BAS is not performed before six months of age, surgical atrial septectomy may be necessary later.

Our over-all results follow the same trends reported by others, with the greatest operative mortality in the infants less than six months old. Deverall reported a hospital mortality of 57% in patients less than six months of age and a mortality of 12.5% in older patients. Mathur and Glenn reported a 12% operative mortality for the SVC-RPA shunt, but only 20% of their patients were less than one year of age. Their series is thus not strictly comparable to ours or that reported by Deverall.

Our long-term survival rate of 45% agrees closely with Taussig et al. who reported 18 of 43 patients (42%) alive 20 years after their initial operation. Taussig also found that longevity after palliative surgery is related to the age at which the initial shunt is performed. In her series only 6 of 21 patients (28%) less than four years old at the initial operation survived 20 years. This contrasts with 12 of 22 patients (54%) who survived for 20 years if their initial palliation was not necessary until after age four.

These observations are important in evaluating the success of the operations to "correct" tricuspid atresia. Most of these corrective procedures have been performed on older children or young adults and the mortality rate of these operations has been high compared to the negligible mortality rate for palliative procedures in this age group. The long-term effect of atrial arrhythmias on patients after corrective operation is yet unknown and may be serious. Miller reports an average saturation of 86% after corrective operation, which is not significantly better than the average postoperative saturation of 84% in our series of palliative operations. Miller also reported pleural effusion, hepatomegaly and ascites occurred in all patients, as well as transitory superior vena caval syndrome in three. His mortality was one of five (20%).

We have been interested in the Fontan procedure but have not encountered a case we considered suitable for the operation. The fact that we have had no mortality with the palliative procedure in children over 12 months of age influences our reluctance to apply a procedure which, in the hands of others, has resulted in significant morbidity and mortality.

Our current method of management for a small infant with tricuspid atresia is to perform a BAS and a Potts' anastomosis. We prefer the Potts' anastomosis because, technically, it is easier to perform. Also, this procedure does not require an opening of the pericardium and thus avoids formation of intrapericardial adhesions which might hinder any future corrective operation. A disadvantage of the Potts' anastomosis is the difficulty in surgically closing it at a future corrective procedure. At this time, the corrective procedures remain unproven in our opinion, and we favor the procedure that is most easily performed and likely to be successful in a tiny infant; and which probably offers palliation for the longest period of time. However, if the BAS is inadequate or impossible because the child is older than six months of age, we prefer the intrapericardial Ao-to-RPA shunt to an anterior thoracotomy in order that a Blalock-Hanlon atrial septectomy may be performed simultaneously. If the child outgrows the shunt at a later date, we would perform another shunt, usually in the opposite thoracic cavity.

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