Transposition of the Great Arteries

Results of Treatment with Early Palliation and Late Intracardiac Repair

By HOWARD P. GUTGESELL, M.D., AND DAN G. McNAMARA, M.D.

SUMMARY

A longitudinal study was made of 62 patients with dextro-transposition of the great arteries. The patients were managed with balloon atrial septostomy (BAS), palliative surgery if any operation was needed in the first year of life, and Mustard intra-atrial baffle repair after one year of age. Of the 47 patients with intact ventricular septum (IVS) treated in this manner, 41 (87%) were living at three months of age, 36 (77%) at one year, and 31 (66%) at two years of age. Survival rates were lower at each age for patients with associated ventricular septal defect (VSD). In relation to therapeutic interventions in the 62 patients, there were six medical deaths within one week of BAS, three deaths during palliative operations, and eight deaths associated with 31 Mustard procedures. Three patients with IVS and five with VSD developed pulmonary vascular obstructive disease, and two patients suffered strokes.

In view of the relatively low mortality rate after the neonatal period and our good results with palliative surgery, we continue to defer the Mustard procedure to the second year of life. This is in contrast to the policy of some centers which now advocate elective intracardiac repair in the first year of life.

Additional Indexing Words:
Rashkind balloon atrial septostomy  Blalock-Hanlon procedure  Mustard procedure
Pulmonary vascular obstructive disease  Cerebrovascular accident  Arrhythmias, postoperative

Prior to 1964, nearly all children with transposition of the great arteries (TGA) died in the first year of life, the majority of them before three months of age.1-3 Mustard’s intra-atrial baffle repair4 provides a physiologic, if not anatomic, correction of the defect and has become widely accepted as the definitive operation for TGA. Balloon atrial septostomy (BAS), introduced by Rashkind and Miller5 in 1966, has allowed many patients with TGA to survive the neonatal period and reach an age at which Mustard’s procedure can be performed. These two procedures, combined in some cases with an intermediate palliative operation, have become the important milestones in the management of the child with TGA. There is general agreement that BAS should be performed at the time of initial cardiac catheterization. There is disagreement, however, as to the optimal time for baffle repair. In some centers,6,7 including our own, a palliative operation is performed if surgical treatment becomes necessary in the first year of life and the Mustard procedure is deferred until the second or third year. More recently, others8-12 have advocated elective intracardiac operation in the first year of life without intermediate palliative operation.

The present report summarizes a longitudinal study of 62 patients with TGA managed with BAS, palliative surgery, and later Mustard repair. The results, which show significant improvement over the natural course of the disease, may be used as a baseline against which to judge alternate plans of management.

Material and Methods

The study group consisted of all patients with dextro-TGA who underwent BAS between September 1967 and December 1972, excluding those patients with single ventricle or atresia of any cardiac valve (although two patients with the Taussig-Bing form of TGA were included). Of the 62 patients so defined, 34 (55%) had simple TGA, 13 (21%) had a large ventricular septal defect (VSD), 13 (21%) had pulmonic stenosis (PS), and two (3%) had both VSD and PS. Six of the 62 patients also had a patent ductus arteriosus and one had coarctation of the aorta. This report includes patient evaluation through December 1973, providing a one to six year longitudinal study of this plan of treatment.

Pulmonic stenosis was considered to be present if the peak systolic pressure gradient between the left ventricle and the
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pulmonary artery was greater than 35 mm Hg\textsuperscript{13} or if there was angiographic evidence of left ventricular outflow tract obstruction. The ventricular septal defect was considered to be large on the basis of angiographic appearance or the presence of equal left and right ventricular pressure in the absence of PS. Two patients with small VSD are classified as simple TGA in this study.

The early results of BAS were classified according to our previous criteria.\textsuperscript{14}  

1) Good result: An increase in oxygen saturation of systemic arterial blood greater than 10 percentage points, a residual mean interatrial pressure gradient of less than 2 mm Hg, and definite clinical improvement.

2) Fair result: An increase in oxygen saturation of systemic blood less than 10%, a persistent atrial pressure gradient of 2-4 mm Hg, and only slight clinical improvement.

3) Unsatisfactory result: No increase in systemic oxygen saturation, a residual atrial mean pressure gradient of more than 4 mm Hg, lack of clinical improvement or BAS-related death.

Late results of BAS were classified according to the criteria of Vlad and Lambert:\textsuperscript{19} survival to six months of age with an arterial blood oxygen saturation greater than 60% and absence of stroke was considered "good;" anything less than this was "unsatisfactory."\textsuperscript{15} However, since patients with VSD frequently have a systemic arterial oxygen saturation of 60% or more even without BAS, only the patients with intact ventricular septum (IVS) were classified in this way.

By the end of the study, the survivors had been followed for an average of 37 months after BAS (range, 12 to 72 months). Final evaluation for this study was made of 32 of the 39 survivors at their latest visit during the last 12 months of the study. The status of six patients no longer under our care was determined by communication with their current physician. One patient has been lost to follow-up since April 1971, three years after BAS.

Plan of Management

All patients had BAS at the time of initial catheterization according to our previously described protocol.\textsuperscript{16} The age at the time of BAS ranged from one day to five months; 80% of the patients were under one month of age. Palliative surgery was performed if needed in the first year of life. Palliative surgery consisted of one or more of the following procedures: 1) Blalock-Hanlon atrial septectomy\textsuperscript{17} if the atrial septal defect proved to be inadequate, 2) systemic-to-pulmonary artery shunt for PS, 3) pulmonary artery banding for large VSD, or 4) ligation of a large ductus arteriosus.\textsuperscript{18}

The Mustard procedure was performed after 12 months of age with two exceptions (table 1). The majority of the patients weighed 16-25 pounds.

Aside from some technical changes in preoperative and postoperative care and in surgical technique, the plan of management remained basically unchanged over the 1967 to 1973 study period.

Results

Balloon Atrial Septostomy

The early and late results of BAS are shown in table

\begin{table}[h]
\centering
\caption{Operative Mortality of Mustard Procedure Related to Age and Weight}
\begin{tabular}{|c|c|c|c|}
\hline
\textbf{Age} & \textbf{Number of patients} & \textbf{Number of deaths} & \textbf{Mortality} \\
\textbf{(months)} & & & \\
\hline
6 or less & 1 & 1 & 100 \\
7 - 12 & 1 & 1 & 100 \\
13 - 18 & 5 & 1 & 20 \\
19 - 24 & 13 & 5 & 38 \\
25 - 30 & 5 & 0 & 0 \\
Over 30 & 6 & 0 & 0 \\
\hline
\textbf{Weight} & 31 & 8 & Over-all 26% \\
\textbf{(pounds)} & & & \\
\hline
15 or less & 1 & 1 & 100 \\
15 - 20 & 10 & 3 & 30 \\
21 - 25 & 14 & 3 & 21 \\
Over 25 & 6 & 1 & 17 \\
\hline
\end{tabular}
\end{table}

2. Fifty-five (89%) of the 62 patients had either a "good" or a "fair" early result. The four unsatisfactory results included one instance of perforation of the right atrium and fatal pericardial tamponade, one of a fatal arrhythmia produced by the BAS, one in which the patient died 12 hours after BAS of unrelieved congestive heart failure, and one in which the patient died during induction of anesthesia for emergency ligation of the ductus arteriosus and pulmonary artery banding immediately after BAS.

The result of BAS in three patients could not be classified by our previous criteria. Two were moribund

\begin{table}[h]
\centering
\caption{Results of Balloon Atrial Septostomy}
\begin{tabular}{|c|c|c|c|c|c|}
\hline
\textbf{Total number} & \textbf{TGA,} & \textbf{PS} & \textbf{VSD} & \textbf{PS + VSD} \\
\textbf{of patients} & \textbf{simple} & & & \\
\hline
\textbf{Early Results} & \textbf{Good} & 47 (76\%) & 25 & 13 & 7 & 2 \\
& Fair & 8 (13\%) & 3 & 0 & 3 & 0 \\
& Unsatisfactory & 4 (6\%) & 1 & 0 & 3 & 0 \\
& Not classified & 3 (5\%) & 3 & 0 & 0 & 0 \\
\hline
\textbf{Total} & \textbf{62} & \textbf{34} & \textbf{13} & \textbf{13} & \textbf{2} \\
\hline
\textbf{Late Results*} & \textbf{Good} & 25 (58\%) & 16 & 9 & - & - \\
& Unsatisfactory & 18 (42\%) & 14 & 4 & - & - \\
& Patients studied & 43 & 30 & 13 & - & - \\
\hline
\end{tabular}
\end{table}

\*Includes only patients with an early "good" or "fair" BAS result.

Abbreviations: TGA = transposition of the great arteries; PS = pulmonic stenosis; VSD = ventricular septal defect.

Patients with VSD or VSD and PS were not included under "late results" because the criteria were not applicable (see text).
and receiving assisted ventilation at the time of BAS. They had hemodynamic but not clinical improvement and died within four days. The third had hemodynamic and clinical improvement, but died of renal failure four days after BAS. Autopsy showed diffuse renal tubular necrosis, probably due to the hypoxemia present before BAS. Thus, seven of the 62 patients (11%) died within four days of BAS.

The initial improvement after BAS was transient in some cases. Among 43 patients with IVS who had an early "good" or "fair" result, only 25 had a good long-term result.

Palliative Surgery

Thirty-two patients underwent palliative operations (Table 3). Twenty patients had Blalock-Hanlon atrial septectomy, either alone or with other palliative procedures; one death occurred. The mortality of all palliative operations was 9% (three deaths in 32 operations), and these occurred in patients under two months of age.

Mustard Procedure

Thirty-one patients underwent Mustard repair during the time of this study. The operative mortality, eight (26%) of 31 patients, is shown related to age and weight in Table 1 and to associated lesion in Table 4. Among the 29 patients with IVS, there were six deaths (21% mortality), five in patients over 12 months of age. There were no deaths among the 11 patients operated upon during the last 18 months of this study.

The average length of postoperative follow-up was 15 months (median, 24 months; range, 2 to 53 months). Of the 23 patients surviving the Mustard procedure, two required subsequent operation for pulmonary vein obstruction. Another patient developed a leak around the patch resulting in a moderate left-to-right shunt and two had partial obstruction of the superior vena cava. None of the latter three patients has required another operation.

As reported previously,18-22 arrhythmias commonly occur following the Mustard procedure. Of the 23 patients in the study group who survived the operation, 14 (61%) had at least one recorded ectopic rhythm after the first postoperative week. Ten of these had well-tolerated junctional rhythm or intermittent atrioventricular dissociation; two had symptomatic bradycardia and two developed atrial flutter.

There was one late postoperative death. The patient underwent Mustard procedure at 24 months of age, but remained in chronic congestive heart failure and died 16 months after surgery. Postoperative cardiac catheterization had revealed satisfactory repair but poor contractility of both ventricles.

Over-all Mortality and Morbidity

At the end of the study, 39 of the original 62 patients were alive. The causes of death in 23 patients are shown in Table 5. The seven deaths at or within one week of BAS have been described above. The medical deaths later in the first year of life were due to

<table>
<thead>
<tr>
<th>Table 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Operative Mortality of Mustard Procedure Related to Associated Lesion</td>
</tr>
<tr>
<td>Associated lesion</td>
</tr>
<tr>
<td>-------------------</td>
</tr>
<tr>
<td>TGA, simple</td>
</tr>
<tr>
<td>TGA, complex</td>
</tr>
<tr>
<td>PS</td>
</tr>
<tr>
<td>VSD</td>
</tr>
<tr>
<td>VSD + PS</td>
</tr>
<tr>
<td><strong>Total</strong></td>
</tr>
</tbody>
</table>

Abbreviations: TGA = transposition of the great arteries; PS = pulmonic stenosis; VSD = ventricular septal defect.

<table>
<thead>
<tr>
<th>Table 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mortality of Palliative Operations in 30 Patients with TGA</td>
</tr>
<tr>
<td>Type of operation</td>
</tr>
<tr>
<td>-------------------</td>
</tr>
<tr>
<td>Blalock-Hanlon alone</td>
</tr>
<tr>
<td>Blalock-Hanlon plus:</td>
</tr>
<tr>
<td>Systemic-pulmonary shunt</td>
</tr>
<tr>
<td>Pulmonary artery banding</td>
</tr>
<tr>
<td>PDA ligation</td>
</tr>
<tr>
<td>Systemic-pulmonary shunt alone</td>
</tr>
<tr>
<td>Pulmonary artery banding:</td>
</tr>
<tr>
<td>Alone</td>
</tr>
<tr>
<td>With PDA ligation</td>
</tr>
<tr>
<td><strong>Total</strong></td>
</tr>
</tbody>
</table>

Abbreviations: PDA = patent ductus arteriosus; TGA = transposition of the great arteries.

<table>
<thead>
<tr>
<th>Table 5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cause of Death</td>
</tr>
<tr>
<td>Total number of patients</td>
</tr>
<tr>
<td>Survivors</td>
</tr>
<tr>
<td>Nonsurvivors</td>
</tr>
<tr>
<td>Medical deaths within 1 week of BAS</td>
</tr>
<tr>
<td>Medical deaths later in first year</td>
</tr>
<tr>
<td>Medical deaths in second year</td>
</tr>
<tr>
<td>Palliative operation</td>
</tr>
<tr>
<td>(includes 1 under 1 week)</td>
</tr>
<tr>
<td>Mustard procedure</td>
</tr>
<tr>
<td>early postoperative period</td>
</tr>
<tr>
<td>late postoperative period</td>
</tr>
</tbody>
</table>

Circulation, Volume 51, January 1975
pneumonia (1), sudden unexplained death at home (2), and congenital bowel obstruction (1). The one nonsurgical death in the second year of life was due to pneumonia.

The mortality for specific age intervals is shown in figure 1. The highest mortality (16%) occurred during the first three months of life, the next highest during the 19 to 24-month interval (15%). The high mortality during the latter period reflects the fact that a large number of Mustard procedures were performed at this age. No other age interval had a mortality higher than 6%.

A life-table was constructed for the study group using the method of Berkson and Gage for populations with varying length of follow-up. A graph of the survival rate is shown in figure 2 along with the data of Liebman et al. for the natural history of TGA.

Of the 47 patients with TGA and IVS, 41 (87%) survived to three months of age, 36 (77%) to one year, and 31 (66%) to two years. In relation to type of treatment five (11%) patients died within a week of BAS; one (2%) died at palliative surgery, four (8%) died waiting for corrective surgery, and six (13%) died at the time of the Mustard procedure. Twenty-three (49%) survived the Mustard procedure and eight (17%) await the operation.

Cerebrovascular accidents have been common in some reports but occurred in only two patients in our study group. Thirteen patients were in moderate or severe congestive heart failure at some point after BAS. Three of these had simple TGA, eight had a VSD, and two had a large surgical systemic-pulmonary artery shunt.

The tendency of patients with TGA to develop pulmonary vascular obstructive disease (PVOD) in early infancy has been well documented. Three of our patients with IVS developed PVOD before two years of age. Two of them died at the time of Mustard procedure and autopsies revealed pulmonary vascular obstructive disease (Heath-Edwards Grades I–IV). The third has not undergone operation. Two additional patients with VSD developed PVOD even though the VSD closed spontaneously in one and became small in the other. Both survived the Mustard procedure and were clinically well when last seen at three and four years after operation. Six other patients with VSD underwent pulmonary artery banding. Postoperative pulmonary artery pressure distal to the band was normal in four patients and moderately elevated in two. None of these patients have had corrective surgery.

Table 6 summarizes the status of the 62 patients at the end of the study. While 39 patients were alive, the long-term outlook for a number of them is uncertain, particularly for those with pulmonary hypertension and those with postoperative arrhythmias. Likewise, the surgical correction in those patients with VSD and pulmonary artery band may be associated with significant mortality.

Discussion

The results of this longitudinal study of 62 patients with TGA managed by early palliation and relatively late Mustard repair show considerable improvement over the natural course of the disease. Both Liebman et al. and Campbell described first-year mortalities.
Table 6
Status of Patients at Latest Evaluation

<table>
<thead>
<tr>
<th>Status</th>
<th>No.</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>I. Survivors</td>
<td>39</td>
<td>(63%)</td>
</tr>
<tr>
<td>Post-Mustard procedure</td>
<td></td>
<td></td>
</tr>
<tr>
<td>uncomplicated</td>
<td>16</td>
<td></td>
</tr>
<tr>
<td>arrhythmias</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>operation for pulmonary vein obstruction</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>total</td>
<td>22</td>
<td></td>
</tr>
<tr>
<td>No Mustard procedure to date</td>
<td>17</td>
<td></td>
</tr>
<tr>
<td>simple TGA</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>(one pulmonary hypertension)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>complex TGA</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>(includes five with VSD and banded PA)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>II. Nonsurvivors</td>
<td>23</td>
<td>(37%)</td>
</tr>
<tr>
<td>Operative deaths</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>palliative operation</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Mustard procedure</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>operative death</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>late death</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Medical deaths</td>
<td>11</td>
<td></td>
</tr>
<tr>
<td>first month of life</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>later in first year</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>second year</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>total</td>
<td>62</td>
<td>(100%)</td>
</tr>
</tbody>
</table>

approaching 90% before present methods of treatment were available. Kidd et al.\(^3\) found a mortality of 95% by age two years among patients with TGA seen prior to 1963. In comparison, 76% of the patients in our study group reached one year of age and 63% reached two years.

The first three months of life is the period of greatest risk for the patient with TGA. Our mortality of 11% within one week of BAS and 16% within the first three months of life is similar to the high neonatal mortality in other studies.\(^24\), \(^25\), \(^28\), \(^29\) Nonetheless, the three-month survival rate of 84% is an improvement over the survival rate of 25% before BAS was introduced.\(^1\)

In the past, palliative operations in patients with TGA have been associated with a formidable mortality. For the Blalock-Hanlon procedure alone, reported mortalities range from 22 to 57%.\(^30\)-\(^33\) At our institution, the mortality for palliative operation in patients with TGA was 30% before the introduction of BAS.\(^24\) In contrast, in the present series of 32 palliative operations, the mortality was 9% and there were no deaths among patients over two months of age. The improved survival rate is most likely because the patients in this series were older and in better condition at the time of operation; in the earlier study many of the patients were critically ill, and 80% of them were under three months of age. In our study group, BAS provided early palliation; only 31% of our patients were under three months at the time of surgical palliation.

The over-all results of the Mustard procedure are in general accord with other large series spanning the period involved in this study.\(^1\), \(^9\), \(^35\) Our recent experience, and that of other recent studies,\(^8\)-\(^11\) suggests that intra-atrial baffle repair can be performed in cases of TGA with IVS with a 10 to 15% mortality.

Despite the large number of reports of operative results in patients with TGA, there have been relatively few longitudinal studies. Tynan\(^24\) reviewed 80 cases of patients who had undergone BAS and found one- and two-year survival rates of 65% and 55%, respectively. The data of Kidd et al.\(^3\) show a two-year survival rate of 65% after BAS was available. Clarkson and colleagues\(^36\) reported one- and two-year survival rates of approximately 60% and 55% during a period when they employed surgical atrial septectomy. In each of the above studies, as in ours, patients were managed with early palliation and relatively late baffle repair.

The band in figure 3 represents a composite survival range obtained by combining our data with the above studies of Tynan,\(^24\) Kidd et al.\(^3\) and Clarkson et al.\(^36\) Although some differences in patient selection, surgical technique, and data collection have been disregarded, the graph shows the approximate range of survival rates one can expect when the plan of management described in this report is followed. Of note is the high mortality rate in the first three months of life followed by a more stable plateau over the next two years.

![Graph showing survival rates](https://csrc.ahajournals.org/doi/fig/10.1161/01.CIR.51.1.36)

**Figure 3**

Range of survival rates reported here and by Tynan\(^24\) and Kidd et al.\(^3\) for patients with TGA treated with BAS, palliative surgery, and late Mustard repair. Also included are those patients of Clarkson et al.\(^36\) treated with surgical atrial septectomy and late Mustard procedures.
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In the past few years, some investigators have advocated a major change in the management of TGA — namely, abandoning most palliative operations and performing elective Mustard procedures in the first year of life. Indeed, their early operative results have been very encouraging. Before adopting such a policy on a large scale, however, it is important to recognize that the consequences of TGA do not stop at the time of baffle repair. The problems of pulmonary vein and caval obstruction, arrhythmias, and posthypothermia central nervous system disturbances are well known, and there is at least some indication that such problems may be more common when the procedure is performed on very small infants.

Champsaur et al. reviewed the first 123 cases of baffle repair in Toronto and found a late death rate of 20% in the first five postoperative years. Breckenridge reported a late mortality of 8.5% for 200 cases of Mustard procedure. We have had one late death among the 23 survivors of the procedure. Thus, the survival rate after Mustard repair does not level off with time but continues to decrease at a variable rate. Whether this rate is greater if the procedure is performed in the first year of life cannot be determined from the data presently available.

In view of our good results with palliative surgery and the relatively low mortality rate between the neonatal period and one year of age, we still defer the Mustard procedure until at least 12 months of age in cases of simple TGA. However, the slow weight gain between 12 and 24 months of age plus the development of PVOD in some patients prior to 24 months of age suggest there is little advantage, and perhaps some risk, in waiting to even as much longer as 18 months.

Our management of patients with mild or moderate PS is similar to that of simple TGA; patients with severe PS are treated with systemic-to-pulmonary anastomosis and later correction. We still perform early banding and late correction for TGA with VSD.

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