Transposition of the Great Arteries
Clinical and Physiological Observations on 74 Patients
Treated by Palliative Surgery

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SUMMARY
Experience with 74 consecutive patients with transposition of the great arteries treated by palliative methods at the Children's Hospital Medical Center in Boston from June 1950 to September 1966 forms the basis for this report. Four palliative techniques were considered in detail. The specific palliative approach suggested depended on the anatomy and physiology in the individual case. Atrial septal defect creation was used largely for patients with an intact ventricular septum to improve arterial saturation, vent the left atrium, and prevent development of pulmonary vascular obstructive changes. Pulmonary artery banding was the procedure used mainly for infants with large ventricular septal defect, adequate mixing, and pulmonary arterial hypertension. The combined operation was performed on infants with ventricular defect in whom arterial-venous mixing was less than optimal through a relatively small ventricular septal defect. Arterial shunt operations were used for cyanotic infants, not in congestive failure, with ventricular septal defect and severe pulmonic stenosis.

The authors recommend palliative surgery in the neonatal period for all patients with transposition of the great arteries, except for the few with type IIA lesions who have just the optimal amount of arterial saturation and absence of congestive heart failure. At present at least an 84% survival rate can be expected from the palliative surgical procedures even in the critically ill infants in the first days of life.

Additional Indexing Words:
Creation of atrial septal defect
Arterial shunt operations
Pulmonary hypertension
Oxygen saturation
Hyperbaric condition
Pulmonary artery banding
Ventricular septal defect

A previous publication from this institution has reviewed the variable clinical profile of transposition of the great arteries (TGA) and suggested surgical approaches toward palliation. At that time no corrective surgery, except the Senning procedure, designed principally for older children, was available for these critically ill patients. Within recent years the Mustard operation, in effect a modification of the Senning idea, has been proposed as an effective corrective procedure applicable to young children as well as to older individuals. The availability of this technique, for patients as young as 2 to 3 years of age, has made it all the more important to employ palliative surgical measures in early infancy so that these babies may survive to an age when they will be candidates for complete correction.

In view of these considerations it seemed appropriate to review our experience with the palliative surgical treatment of infants with transposition of the great arteries. We
Table 1

Palliative Operations in Patients with Transposition of Great Arteries (TGA)

<table>
<thead>
<tr>
<th>Classification of TGA</th>
<th>Total</th>
<th>ASD</th>
<th>PAB &amp; ASD Shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>40</td>
<td>38</td>
<td>1 1 0</td>
</tr>
<tr>
<td>Type IIA</td>
<td>12</td>
<td>2</td>
<td>0 0 10</td>
</tr>
<tr>
<td>Type IIB</td>
<td>1</td>
<td>1</td>
<td>0 0</td>
</tr>
<tr>
<td>Type IIC</td>
<td>18</td>
<td>1</td>
<td>8 9 0</td>
</tr>
<tr>
<td>Type NK</td>
<td>3</td>
<td>3</td>
<td>0 0</td>
</tr>
<tr>
<td>Total</td>
<td>74</td>
<td>45</td>
<td>9 10 10</td>
</tr>
</tbody>
</table>

Classification of TGA:
Type I = TGA with intact or virtually intact ventricular septum.
Type IIA = TGA with ventricular septal defect (VSD) and severe pulmonary stenosis (PS).
Type IIB = TGA with VSD and pulmonary obstructive disease (PVO).
Type IIC = TGA with VSD and large pulmonary blood flow.
Type NK = patients in whom the size and function of the VSD could not be adequately evaluated.

Abbreviations: ASD = atrial septal defect creation; PAB = pulmonary artery banding.

will emphasize the physiological considerations leading to the choice of operation and will present not only the immediate surgical results but also the long-term follow-up. We will not discuss the details of the clinical profile of these patients but rather will stress only deviations from the picture presented in the earlier publication.1

Methods

Seventy-four (74) consecutive patients with TGA, treated by palliative methods at the Children's Hospital Medical Center between June 1950 and September 1966, form the basis of this report. The palliative techniques to be discussed include (1) creation of an atrial septal defect, (2) banding of the pulmonary artery, (3) creation of an atrial septal defect with banding of the pulmonary artery, and (4) establishment of a shunt from a systemic to a pulmonary artery.

We will not discuss the results of complete correction (Senning, Mustard) nor will we present the patients dealt with by means of Baffes' procedure or a Glenn shunt, neither of which has been part of our surgical approach during the past 4 years.

The preoperative diagnosis of TGA was based on cardiac catheterization with selective angiographic studies in 61 instances. In five critically ill patients the characteristic clinical picture led to surgical confirmation of the diagnosis. The diagnosis of TGA was not established preoperatively in eight infants with ischemic lung fields, in whom a systemic to pulmonary artery shunt was performed; the diagnosis of transposition was made only at postoperative catheterization in six and at surgery in two. Autopsy examination was available in 20 patients.

All but one of our patients had classical d-transposition4; one patient had l-transposition.

Table 2

Shunting Procedures in Type IIA TGA

<table>
<thead>
<tr>
<th>No.</th>
<th>Age (yr)</th>
<th>Operation Type</th>
<th>Pressure</th>
<th>P.O. follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3 days</td>
<td>Asc. ao. to RPA &amp; ASD</td>
<td>HB</td>
<td>3/12 A</td>
</tr>
<tr>
<td>2</td>
<td>70 days</td>
<td>Asc. ao. to RPA &amp; ASD</td>
<td>HB</td>
<td>11/12 A</td>
</tr>
<tr>
<td>3*</td>
<td>1 9/12</td>
<td>R-Blalock-Taussig</td>
<td>NB</td>
<td>15 11/12 A</td>
</tr>
<tr>
<td>4</td>
<td>1 10/12</td>
<td>Potts</td>
<td>NB</td>
<td>6 3/12 D†</td>
</tr>
<tr>
<td>5</td>
<td>1 11/12</td>
<td>Potts</td>
<td>NB</td>
<td>16 6/12 A</td>
</tr>
<tr>
<td>6*</td>
<td>2 6/12*</td>
<td>L-Blalock-Taussig</td>
<td>HB</td>
<td>5 4/12 A</td>
</tr>
<tr>
<td>7</td>
<td>2 10/12</td>
<td>Potts</td>
<td>NB</td>
<td>10 9/12 A</td>
</tr>
<tr>
<td>8</td>
<td>3 9/12</td>
<td>Potts</td>
<td>NB</td>
<td>15 11/12 A</td>
</tr>
<tr>
<td>9</td>
<td>4 7/12</td>
<td>Potts</td>
<td>NB</td>
<td>15 2/12 A</td>
</tr>
<tr>
<td>10</td>
<td>7 8/12</td>
<td>Asc. ao. to MPA (Teflon prosthesis) &amp; ASD</td>
<td>HB</td>
<td>8/12 A</td>
</tr>
</tbody>
</table>

*These patients had subsequent Blalock-Taussig operations on the opposite sides at 14 2/12 and 6 3/12 years, respectively.
†Died 6 1/4 years after operation in congestive heart failure.

Abbreviations: Asc. ao. = ascending aorta; RPA = right pulmonary artery; MPA = main pulmonary artery; ASD = atrial septal defect creation; HB = hyperbaric; NB = normobaric; A = alive; D = dead.
Infants with dextrocardia, "corrected transposition of the great arteries," tricuspid or mitral atresia, and coarctation of the aorta in association with transposition were not included in this study. The anatomic classification proposed earlier of types I and IIA, B, and C will be adhered to (table 1) with the exception that we have included in type I some patients whose ventricular septum was almost but not completely intact. These are patients who show no large left-to-right shunt at the ventricular level, though possibly a small shunt may be proven by selective angiography or suggested by a slight increase in oxygen saturation at the right ventricular or systemic arterial level.

All patients were examined by at least one of the authors. Twelve-lead electrocardiograms and 7-foot radiograms were obtained in all. Routine right heart catheterization, exclusively by the femoral approach, was performed preoperatively in 61 infants. The types of surgery performed in various transposition groups are presented in table 1. The specific techniques employed for shunts are summarized in table 2.

Results

Atrial Septal Defect Created

As seen in table 1, 45 patients with TGA were treated by creating, or enlarging, an interatrial communication. It is clear from this table that the great majority of these patients (38 out of 45) belonged to type I (intact ventricular septum), and thus the clinical description to follow refers largely to this group. The age distribution at operation is presented in figure 1, together with the levels of preoperative oxygen saturation. All these infants were severely cyanotic. Tachypnea, a consequence of anoxia and left-sided congestive heart failure, was almost as striking in these babies as the cyanosis. Right-sided failure was shown by hepatomegaly, while peripheral edema was less common. These infants seemed anxious, fretful, sweaty, and sick. Among the auscultatory findings, the presence of a split second sound ($S_2$) with $P_2$ of average intensity should be stressed as an important differential diagnostic clue in a severely cyanotic baby, excluding critical pulmonic stenosis with or without a ventricular septal defect. The respiratory

Figure 1

Age and systemic arterial oxygen saturation at time of palliative surgery.
Transposition of the Great Arteries

Cardiac enlargement related to age in type I TGA.

Variations in splitting may be hard to appreciate with the rapid heart and respiratory rates of these critically ill infants. The murmurs, as described earlier, were unimpressive and not particularly helpful in differential diagnosis. Of eight patients operated on within the first week of life, none had murmurs louder than grade II, four had no significant murmurs at all, and only one had a grade II murmur. Gallaher and associates emphasized, and we certainly agree, that in the very young newborn the size and shape of the cardiac silhouette in the radiogram is usually normal or only slightly enlarged (fig. 2). Similarly, the pulmonary vasculature is not unusual in most instances, although one may note that it is not decreased. The commonly described “egg on side” shape is seen only in babies who survive beyond the first weeks of life. The electrocardiogram is also of relatively little help in the newborn, since it frequently shows only slightly more right ventricular

Figure 2
Cardiac enlargement related to age in type I TGA.

Figure 3
Preoperative and postoperative pulmonary artery pressures (A) (see text) and oxygen saturation (B) in patients undergoing ASD creation. Data from patients with type I TGA are presented in chart form. Data from patients with other types undergoing ASD creation are presented in inserts. (→ = mean value.)
hypertrophy than expected for age. Of seven patients under 1 week of age, three had entirely normal tracings and the remaining four exhibited only questionable right ventricular hypertrophy (frontal plane axis of over 150° or a deep S in V6). Most patients beyond the first week of life showed variable, but seldom maximal, degrees of right ventricular hypertrophy. The physiological data on these patients, most of whom fitted into type I of our classification, are presented in figures 3 and 4. All patients had low arterial oxygen saturations before operation, due to inadequate left-to-right shunting (fig. 3B; see also fig. 6). The estimation of pulmonary arterial pressure is difficult in these patients since the pulmonary artery cannot be easily entered by conventional methods. As a matter of fact we have entered the pulmonary artery in only two of the patients in this group. In each, pulmonary artery pressure was approximately 60% of systemic arterial pressure. Figure 3A presents these two pulmonary arterial pressures, as well as left ventricular pressures, in terms of percentage of systemic arterial pressures, in those patients in whom we assume that no significant degree of pulmonic stenosis was present. Significant pulmonic stenosis was felt to be excluded by means of one of the following criteria: (1) autopsy (13 patients); (2) left ventricular pressure, in the absence of a significant murmur, at 50%, or less, of the simultaneous systemic arterial pressure (10 patients); (3) left ventricular pressure, in the absence of significant murmur, at more than 50% of simultaneous systemic arterial pressure, but with a pulmonary venous wedge pressure over 50% of left ventricular pressure (four patients); and (4) postoperative low (under 50% of systemic arterial pressure) left ventricular pressure (five patients).

It may be seen then from figure 3A that in 12 out of 28 patients the estimated preop-
operative pulmonary arterial pressure was 75% or more of systemic arterial pressure, and in only six out of 28 was the pulmonary pressure half or less that of the systemic arterial pressure. No significant differences with age were noted in infants under the age of 2 months. None of the three infants who were over 2 months of age, however, had pressures less than 50% of systemic arterial pressure. Figure 4A and B present the preoperative and postoperative left atrial pressures as well as left atrial to right atrial pressure gradients. It may be seen that LA pressures postoperatively averaged half of the preoperative values and the gradients were abolished in all but one instance.

In summary then, this operation was performed on very young infants with severe cyanosis, dyspnea, and inadequate mixing at the atrial level and no mixing, or only poor exchange, at the ventricular level. A comparison of operative mortality by age in the groups operated on under hyperbaric and normobaric conditions is presented in figure 5. The atrial septal defects in all the infants operated upon under hyperbaric conditions were created under direct vision by means of inflow occlusion. The techniques used in the normobaric group consisted of various modifications of the Blalock-Hanlon technique. Within the past few months, atrial septal defects have been created in seven patients by means of the Rashkind balloon technique. Only two of these babies are presented in this series; the others were admitted to the hospital after September 1966.
Nevertheless, in view of the recent great interest in this approach, it may be worth stating that we have had one fatality among the seven infants so treated. Among the remaining six patients we have had two late deaths, one due to bronchial obstruction, the other to overwhelming pneumonia.

Clearly the results of operation under hyperbaric conditions offer a definite advantage over our previous surgical methods (fig. 5). The possible explanation for this improved survival is discussed on page 329. Twenty-seven of 32 patients (84%) operated upon at increased environmental pressure and eight out of 13 (62%) operated upon under normobaric conditions survived, resulting in a total of 35 surgical survivors. Among these who left the hospital postoperatively, we have had five late deaths. One died with a brain abscess at 5 years of age, two died in congestive heart failure within 6 months after palliative surgery, and the remaining two succumbed during second operations performed within 5 months to improve the mixing at the atrial level. Both operations in these latter patients were carried out under hyperbaric conditions.

We thus have 30 long-term survivors among the 45 patients with transposition of the great arteries in whom creation of an atrial septal defect was the only palliative procedure performed. The mere fact that these patients are still alive, at the writing of this paper, is the most important fact of the follow-up, since as has been repeatedly demonstrated, approximately half of the infants with type I TGA die before the age of 2 weeks, and almost 90% are dead by the age of 4 months. The average length of postoperative follow-up is 31 months, with a range of 3 to 64 months. All patients, as indicated earlier, had tachypnea preoperatively. The postoperative respiratory rate was uniformly lower. In spite of significant improvement in oxygen saturation (fig. 3B), 14 patients were still maintained on digitalis, although none required diuretics postoperatively. Only four still had significant cardiac enlargement when last seen. No satisfactory assessment of a change in growth and development is possible since most infants were operated upon at a very young age, but 22 of the 30 patients with long-term follow-up are still below the third percentile in weight.

It may be said then that, although these infants are alive and doing reasonably well clinically, with oxygen saturation at reasonable levels, most of them show evidence of failure to thrive.

Repeated catheterization was performed postoperatively in 14 of the survivors of surgery. The results are presented in figures 3 and 4.

### Table 3

<table>
<thead>
<tr>
<th>Operation</th>
<th>Pressure</th>
<th>Total</th>
<th>Survivors (%)</th>
<th>Deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD</td>
<td>NB</td>
<td>13</td>
<td>8 (62%)</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td>HB</td>
<td>32</td>
<td>27 (84%)</td>
<td>5</td>
</tr>
<tr>
<td>PAB</td>
<td>NB</td>
<td>4</td>
<td>1 (25%)</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>HB</td>
<td>5</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>PAB &amp; ASD</td>
<td>NB</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>HB</td>
<td>9</td>
<td>8</td>
<td>1</td>
</tr>
<tr>
<td>Shunts</td>
<td>NB</td>
<td>6</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>HB</td>
<td>4</td>
<td>4</td>
<td>0</td>
</tr>
</tbody>
</table>

**Figure 6**

*Preoperative systemic arterial oxygen saturation, type of TGA, and surgical treatment.*

*Oxygen saturation from patients with other than type I TGA undergoing ASD creation has been omitted.*

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Pulmonary Artery Banding

Nine patients were subjected to pulmonary artery banding (table 1). It may be seen that all but one had TGA of type IIC (VSD and large pulmonary flow). The remaining patient was classified under type I since the VSD was proven to be small at autopsy. The age distribution of these infants together with the arterial oxygen saturation are presented in figure 1. In contrast to the babies in whom only an atrial septal defect was created, cyanosis was not a very prominent feature of this group (fig. 6). Left-sided congestive heart failure, on the other hand, was invariably present and severe. The presence of congestive heart failure in a baby without striking cyanosis, appearing in most instances beyond the first 2 weeks of life, suggested the diagnosis of a large ventricular septal defect, initially, in many of these patients. The cardiac findings at physical examination were somewhat more striking than in the group discussed earlier in that the cardiac impulse was more hyperkinetic, the left chest deformity was more marked, and the systolic murmur along the lower left sternal border was louder. Diastolic rumbles were heard in five. The second heart sound was usually split (seven out of nine) with more or less increase in the intensity of the pulmonic component. The cardiac silhouette, by the time the baby entered our hospital, was usually large, with impressive active and passive pulmonary vascular engorgement. The electrocardiogram was not markedly abnormal in these patients either. One baby had an entirely normal electrocardiogram while three showed evidence of right atrial hypertrophy as the only abnormality. Of the remaining five, the one with pure right ventricular hypertrophy had type I transposition, the other four had left ventricular hypertrophy alone or in combination with right ventricular hypertrophy.

Cardiac catheterization indicated an arterial oxygen saturation of 80% or more in seven patients (fig. 6). Since the pulmonary artery of these patients was banded, they by definition had to have a ventricular septal defect. The presence of such was suggested by near equilibration of the pressures between the two ventricles and the great arteries, and confirmed by catheter passage

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**Figure 7**

LA pressure, type of TGA, and surgical treatment. *LA pressures from patients with other than type I TGA undergoing ASD creation have been omitted.

**Figure 8**

LA-RA pressure gradient, type of TGA, and surgical treatment. *LA-RA gradients from patients with other than type I TGA undergoing ASD creation have been omitted.
or contrast studies of both or by oxygen saturation data. A large pulmonary blood flow was suggested by the relatively high systemic arterial oxygen saturation (figs. 1 and 6) and proven in six instances by calculated pulmonary to systemic flow ratios averaging 3.2. The left atrial pressures and the LA-RA gradients presented in figures 7 and 8 indicate that the majority of these patients have high LA pressures presumably due to the large pulmonary blood flow.

The patients, then, who were subjected to banding of the pulmonary artery as the only palliative surgical intervention were babies beyond the first 2 weeks of life without marked cyanosis but with a significant degree of congestive heart failure due to very large pulmonary blood flow, high pulmonary arterial pressure, and a large ventricular septal defect. The operative mortality related to age is presented in figure 9. All patients operated on in the hyperbaric chamber survived surgery and left the hospital. By contrast only one of four banded under normobaric conditions survived surgery. There were two late deaths among the six survivors; one died abroad with evidence of increasing hypoxia, the other under our care in intractable cardiac failure.

Although the prognosis for patients with type IIC lesions, who make up all but one of the patients subjected to pulmonary artery banding may not be quite as poor as that for patients with type I, Keith and associates\textsuperscript{11} found that only three of 34 patients with this anatomic situation survived beyond 1 year. Thus, the mere fact that four of nine patients operated upon are still alive at an average of 35 months after operation attests to the helpfulness of surgical intervention. The overall heart size, the pulmonary plethora, and the severity of congestive heart failure...
failure have all diminished in the four survivors. Although they are growing at a more rapid rate than they did prior to surgery, only one of the four has reached the third percentile for weight and only two, the third percentile for height.

In addition to the clinical and physiological improvement accomplished by the banding procedure, the fact that the pulmonary vasculature in these patients is no longer exposed to high pressure is a further obvious advantage of the operation.

Atrial Septal Defect Creation and Pulmonary Artery Banding

Pulmonary artery banding in combination with creation of an atrial septal defect was the procedure of choice in 10 patients (table 1). The ages at operation and the arterial oxygen saturations are presented in figure 1. The symptoms, as well as the findings at physical examination, were closely similar to those found in patients in whom pulmonary artery banding alone was performed, except that the patients were in general somewhat more cyanotic (fig. 6). The radiological and electrocardiographic features were also almost identical in the two groups. The infants subjected to the combined palliative approach were characterized clinically by cyanosis and severe congestive heart failure. The physiological data indicated the presence of a ventricular septal defect, an inadequate atrial septal defect, with LA hypertension (figs. 7 and 8), arterial unsaturation (fig. 6), pulmonary artery hypertension, and varying degrees of pulmonary vascular obstruction.

The operative mortality is presented in figure 10. It may be seen that eight of nine patients operated upon in the hyperbaric chamber survived. The one infant treated under normobaric conditions succumbed. There were no late deaths in this group, and
the average postoperative follow-up is 19 months. The advantages of surgery are those discussed in the section on pulmonary artery banding alone. Seven of the eight survivors have less cardiac enlargement and less pulmonary plethora than before operation. All have less clinical evidence of heart failure than before. Six are still markedly underweight and small but two, surprisingly, have reached the twenty-fifth percentile in height and weight despite being at the third percentile at the time of surgery.

Arterial Shunts

In 10 patients a systemic to pulmonary artery shunt was performed. Since most of these patients were operated upon some time ago, preoperative arterial saturations are not available in five of them. The others had arterial oxygen saturations of 26, 61, 73, 74, and 87%, respectively (fig. 1).

The clinical picture was characterized by marked cyanosis and dyspnea on exertion but without clear-cut anoxic spells. Congestive heart failure was absent. On physical examination the striking cyanosis, with an ejection systolic murmur and a single second sound were the dominant features. Cardiac enlargement was moderate at best, and pulmonary vascularity was diminished, though not as strikingly as in patients with severe tetralogy of Fallot. The electrocardiogram showed right axis deviation with severe right ventricular hypertrophy in all. At cardiac catheterization, performed postoperatively in eight and preoperatively in two, the presence of a ventricular septal defect and pulmonary stenosis was occasionally proven by traversing the left ventricular outflow tract but more often demonstrated by selective angiography from the left ventricle. Briefly then, the patients in whom a systemic-to-pulmonary
artery shunt was performed were almost indistinguishable, clinically as well as physiologically, from patients with tetralogy of Fallot, though differentiation from other types of transposition was relatively easy. The only clinical clue was sometimes furnished by the absence of maximal pulmonary ischemia in the x-rays.

All patients survived surgery (table 2) and improved symptomatically. As an expression of their relatively adequate pulmonary blood flow, one may cite the average postoperative arterial oxygen saturation of 87% (76 to 95%).

The advantage of palliative surgery in these patients, in addition to the obvious immediate and gratifying relief from symptoms of anoxia, may be preservation of the pulmonary vascular bed, without intimal changes and thrombotic processes, thus leaving the door open for eventual complete repair.

Discussion

The significant increase in the number of babies with transposition operated upon in this institution is worth comment (fig. 11). We doubt that this increase is based on an increased incidence of transposition of the great arteries which has been estimated to represent 5% of the cases of congenital heart disease.11 We believe that increasingly aggressive diagnostic and surgical approaches to these babies have resulted in increased referral to the hospital at an earlier age. It should be stressed here that suspicion of the diagnosis of type I TGA should be raised concerning any sick newborn with persistent cyanosis, no matter how mild, irrespective of heart size, electrocardiographic abnormalities, or even the absence of heart murmur. In addition, the diagnosis of type IIc transposition should be suspected at a very young age for all infants presenting as patients with ventricular septal defect and very severe failure. Consideration of these simple crib-side hints may lead to the early diagnosis of TGA by the pediatrician.

The difficulty in entering the pulmonary artery in patients with transposition in a laboratory which has an extensive teaching program and thus a large number of different individuals performing catheterizations is obvious. A new approach to this problem has been suggested by Carr and Wells who use a coaxial flow-guided polyethylene tube to enter the pulmonary artery.12

The primary purpose of creating an atrial septal defect in these babies is to increase arteriovenous mixing and thus improve the systemic arterial oxygen saturation. This indeed has been accomplished with reasonable success in the majority of patients (fig. 3B). The other, not quite so obvious, but equally important result of the operation is decompression of left atrial hypertension, thereby eliminating or significantly decreasing pulmonary venous congestion. This alone may result in a decreasing respiratory rate. A third favorable effect of atrial septal defect creation may be the result of a combination of the first two factors, in other words, protection from pulmonary vascular obstructive changes. The integrity of the pulmonary vasculature may be preserved both by an absence of pulmonary and left atrial hypertension, as well as by the relatively normal arterial oxygen saturation. Atrial septal defect creation then, we believe, improves the hypoxia of these babies, relieves their dyspnea, and may protect them from the development of pulmonary vascular obstructive disease. On the other hand, we are not certain of the effects of this palliative approach on the relief of congestive heart failure. First, some cardiac enlargement tends to persist, and digitalis is not commonly discontinued. As further evidence of the persistent, chronic, though often low-grade, heart failure, one may cite the poor growth of most of these infants in spite of relatively adequate systemic arterial saturations. Certainly this phenomenon serves as a stimulus for as early performance of the Mustard operation as is practical.

One word about the Rashkind ballooning procedure, first proposed in 19669 and performed in more than 100 cases in various centers of North America and Europe with
an 80 to 85% success, may be appropriate. At the present time, in our institution, this is the first choice of attack on the atrial septum; it can be performed, if indicated, with safety in expert hands as part of the initial catheterization procedure. This should result in an immediate increase in arterial oxygen saturation and decrease in left atrial pressure. If the post-ballooning arterial saturation is over 75% and the left atrial pressure approximates right atrial pressure, this may be all that has to be done for palliation, and the Mustard operation can be performed later in a patient with an intact pericardium. If, on the other hand, the immediate improvement following ballooning is not satisfactory or the initial success is not maintained because of resealing of the defect, then open creation of an ASD during inflow occlusion can still be performed days or weeks later, as an elective procedure on a baby in a somewhat better state of oxygenation. There is a real place for the balloon maneuver even in an institution like ours, where surgical creation of an atrial septal defect is a safe and effective procedure.

The prime purpose of pulmonary artery banding is the relief of congestive heart failure by decreasing the pulmonary blood flow. The relief of hypoxia that may result from this procedure is minimal but, as indicated, hypoxia is not a major problem in the patients subjected to pulmonary artery banding. Pulmonary artery banding alone was performed in patients in congestive heart failure, with a VSD, and a systemic arterial O₂ saturation of 80% or higher. The operation itself may not be any more difficult than it is in patients with simple ventricular septal defect although the posterior position of the pulmonary artery presents some problems. The principal technical difficulty is to determine how far to constrict the pulmonary artery without interfering with the pulmonary blood flow to an extent that mixing, a vital factor for survival of the patient with transposition of the great arteries, is not critically curtailed. As a rough guide we aim at halving the pulmonary artery mean pressure as measured in the operating room.

The combination of pulmonary artery banding with creation of an atrial septal defect should be considered in two groups of patients. In type I, with pulmonary arterial hypertension and a small ventricular septal defect, banding may be considered, after atrial septal defect creation, to relieve congestive heart failure and to protect the pulmonary vasculature. Our impression, however, is that in most instances, if the VSD is truly small, this may not be necessary. The favorable consequences of atrial septal defect creation enumerated earlier would all tend to diminish pulmonary arterial pressure and retard the development of pulmonary vascular obstructive changes. Serial catheterization may be necessary to arrive at an accurate conclusion but our present policy is not to band patients with type I TGV routinely, even if a small ventricular septal defect is present. The second group of patients to be considered are those with a type IIIC lesion. Here banding, as discussed, is mandatory, and probably helpful enough in lowering left atrial pressure, by diminishing pulmonary blood flow, so that a simultaneous creation of an atrial septal defect may not be necessary in most instances in which preoperative catheterization has demonstrated an appreciable inter-atrial communication. If, on the other hand, no significant ASD was found at preoperative catheterizations or at surgery after banding, and if the pressure in the left atrium still seems elevated, or if the arterial saturation drops significantly, an atrial septal defect should be created simultaneously. Furthermore, infants successfully banded but not doing well postoperatively, in terms of low arterial oxygen saturations, should be recatheterized and creation of an atrial septal defect by ballooning or surgery should be considered. In other words, in patients with type IIIC lesions, a combination of banding and atrial septal defect creation is recommended only if the hemodynamics after banding indicate that the mixing now afforded by the ventricular septal defect is not satis-
factory enough to result in an arterial oxygen saturation of over 75%, or the decrease in pulmonary blood flow has not lowered left atrial pressure to acceptable levels.

The indications for arterial shunts in patients having type IIA lesions are essentially the same as those commonly recognized for infants with tetralogy of Fallot, that is, hypoxia of severe degree resulting in "spells" or severe limitations of effort. In our institution, at the present time, the approach is to create a shunt between the ascending aorta and right pulmonary artery in infants under 1 year and to carry out a Blalock procedure in most patients over this age limit. The results are on the whole satisfactory and comparable to those cited for patients with tetralogy of Fallot.

The creation of an atrial septal defect, simultaneously with a systemic arterial shunt should be considered, and as a matter of fact has been performed in our two youngest infants in this group, in those situations where adequate mixing does not take place through the ventricular septal defect despite an adequate pulmonary blood flow via the shunt. These infants may be identified, rarely, preoperatively by angiographic demonstration of a small VSD with pulmonary stenosis. At the operating table, if the systemic arterial saturation does not rise adequately after establishment of an atrial shunt, or if in similar circumstances the left atrial pressure rises to excessive levels, an atrial septal defect should be created.

It may be seen from the foregoing discussion that the authors believe that opening of the atrial septum, as suggested originally by Blalock and Hanlon is an integral part of the palliative surgical treatment of transposition of the great arteries. This is the only procedure that is recommended for transposition with an intact ventricular septum and should be considered seriously as a supplementary maneuver for type II patients as well. It should be emphasized that a large left atrial-to-right atrial pressure gradient obviously indicates only a small inter-atrial communication, but the absence of such a gradient may also be compatible with a relatively intact atrial septum. It follows from this that creation of an atrial septal defect may be necessary, and beneficial, even if left atrial and right atrial pressures are identical (fig. 12). We believe that identity of atrial pressures, with only a small foramen ovale present, may be based on the similar compliance-flow ratios in the two ventricles.

Finally, a word of explanation is indicated regarding the improved results obtained under hyperbaric conditions. We do not believe that any single factor is responsible for the excellent survival rate in these critically ill infants. Several factors contribute. First of all, due to the increased optimism created by the Mustard procedure, more infants are referred to the hospital at a very young age and in relatively good condition. Secondly, a team of surgeons, cardiologists, nurses, and technicians with a special interest in neonatal cardiology handle these delicate infants. Thirdly, the metabolic needs of the babies before, during, and after operation are analyzed and met with appreciably more skill than in the past. Finally, an unknown, but we believe a significant contribution to the survival of these infants is the increased availability of oxygen provided in an hyperbaric environment. The in-
crease in arterial $P_O_2$ as well as in pH at 3 atmospheres of pressure,\textsuperscript{7} improves the metabolic state of the cells and thus allows survival through anesthesia, surgery, and necessary periods of inflow occlusion.

References


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