Recent Experiences with the Mustard Procedure for the Complete Transposition of the Great Arteries by Means of “Bypass Hypothermia”

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ISHIZAWA, E., HORIUCHI, T., ITO, T., YAMAKI, S., SATO, K., SATO, N., TANAKA, S., SUZUKI, Y. and TADOKORO, M. Recent Experiences with the Mustard Procedure for the Complete Transposition of the Great Arteries by Means of “Bypass Hypothermia”. Tohoku J. Exp. Med., 1979, 127 (2), 189–195 — This paper presents our recent results of the Mustard procedure (intra-atrial baffle operation) for the complete transposition of the great arteries performed in 24 infants and children during the past 6 years at the Tohoku University Hospital. All intracardiac repairs were performed using “bypass hypothermia” (surface-induced deep hypothermia, circulatory arrest, and limited cardiopulmonary bypass). The hospital mortality rate was 8% in patients with simple transposition without pulmonary hypertension (Group I, one death in 12 patients), 50% in patients with ventricular septal defect (VSD) and pulmonary hypertension (Group II, 3 deaths in 6 patients), and 20% in patients with VSD and pulmonary stenosis (Group III, one death in 5 patients). Various types of longstanding dysrhythmia were found in 6 out of 18 long-term survivors, and subsequent pace-maker implantation was necessary in one infant. Postoperative pulmonary venous obstruction occurred in 3 infants, and surgical relief of the obstruction was successfully undertaken in 2 of them. Prevention of postoperative dysrhythmia, pulmonary venous obstruction, and possible brain damage are also discussed. — complete transposition of the great arteries; Mustard procedure; bypass hypothermia

Since May 1963 when Mustard first succeeded in the hemodynamic correction of the complete transposition of the great arteries (TGA), the prognosis of this anomaly has been considered to have changed completely. Especially, in most cases of the simple transposition, it has become possible to save lives by applying the two-stage operation of Rashkind’s balloon atrioseptostomy (Rashkind and Miller 1966) and the Mustard procedure (Mustard 1964). In recent years, however, various problems of this procedure, such as late occurrence of pulmonary venous obstruction and conduction disturbances, have become evident, and many efforts to reduce such late complications have been made in many institutions.

Since 1970 when we achieved our first success in the hemodynamic correction of TGA, we have striven to improve early and late results of this procedure (Horiuchi

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et al. 1971; Ishizawa et al. 1971, 1973, 1974). This paper presents our experiences in the past 6 years.

**Patients and Methods**

From April 1970 to July 1975, 24 cases of TGA were operated upon at the Tohoku University Hospital. Their ages at the time of surgery ranged from 5 months to 5 years; one case was just less than 6 months of age; 8 cases, 7–12 months; 9 cases, 13–18 months; and 6 cases, 19 months – 5 years.

Associated lesions and anomalies classified into Group I (simple TGA) included 13 cases involving small ventricular septal defect (VSD) in 2 and patent ductus arteriosus (PDA) in other 2 cases. Among 6 cases which belonged to Group II with VSD and pulmonary hypertension (PH), 2 cases had pulmonary artery banding (PA banding). Finally, there were 5 cases in Group III with VSD and pulmonary stenosis (PS) (Table 1).

**Table 1. Results of the Mustard procedure by the ages of the patients.**

<table>
<thead>
<tr>
<th>Age of the Patients</th>
<th>Group I</th>
<th>Group II (with VSD &amp; PH)</th>
<th>Group III (with VSD &amp; PS)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>4–6 months</td>
<td>3</td>
<td>1(1)</td>
<td>1</td>
<td>13(2)</td>
</tr>
<tr>
<td>7–12 months</td>
<td>6(1)</td>
<td>2(1)</td>
<td>4(1)</td>
<td>13(2)</td>
</tr>
<tr>
<td>13–18 months</td>
<td>9(1)</td>
<td>1</td>
<td>1</td>
<td>5(1)</td>
</tr>
<tr>
<td>19 months or more</td>
<td>12(1)</td>
<td>2</td>
<td>6(5)</td>
<td>13(2)</td>
</tr>
</tbody>
</table>

( ) Hospital death. PFO, patent foramen ovale; ASD, atrial septal defect; VSD, ventricular septal defect. PDA, patent ductus arteriosus; PH, pulmonary hypertension; PS, pulmonary stenosis.

The average ages of the patients at the time of surgery were 12 months, 15 months, and 41 months in Group I, Group II, and Group III, respectively. Among these 24 cases, 13 in Group I and 5 in Group II underwent balloon atrioseptostomy between the 2nd and 60th days after birth. Prior to the Mustard procedure, one case in Group I underwent ligation of PDA, while one case in Group II received the Blalock-Hanlon operation and PA-banding, and another case had ligation of PDA and PA-banding.

To prevent the hemorrhagic tendency (reduction of platelet count and adhesiveness, and depletion of anti-hemophilic factors V and VIII) which was observed primarily in patients with PS and VSD, we had been administering 4 mg/kg/day of heparin for 3 days or 1 mg/kg/day of prednisolone for 2 days for preoperative treatment.

For the ill infants, “bypass hypothermia” (Tanaka et al. 1974) developed from simple hypothermia (Horiuchi et al. 1963) has usually been used with the aims of (1) shortening the bypass time as much as possible to prevent the tendency toward postoperative bleeding and pulmonary complications, and (2) performing a bloodless operation under circulatory arrest.

With regard to the anesthesia, after administration of 0.4 mg/kg of Vesprin, GOF slow induction was done, which was followed by deep ether anesthesia. The patient was then moved to an ice water bath where surface cooling began. In order to regulate the cooling
speed of the patient, it was necessary to record the pharyngeal and rectal temperature every 5 min and to prevent the difference between the two from becoming too wide. The administration of a dose of 10 ml/kg of low molecular Dextran before the start of cooling was considered effective to improve the peripheral circulation and to maintain the systemic blood pressure.

In the present cases, usually in about 40 min of surface cooling, the pharyngeal temperature reached 27–28°C, allowing the ultimate pharyngeal temperature to be 24°C. The after-drop of temperature was around 3–4°C. In the process of core cooling, the patient was then moved to the operating table, and the heart was exposed through mid-sternal splitting. Using a heart-lung machine with a Temptrol Q-130 added to a Brown-Harrison heat exchanger (primed with 1000 ml of heparinized fresh blood diluted to 20% with lactated Ringer’s solution), a flow rate of 120–140 ml/kg min was maintained. Rapid cooling was avoided by controlling the temperature drop to about 2°C/5 min, the pharyngeal temperature being about 17°C. In this procedure, it was also important to minimize the difference of temperature between the pharyngeal and rectal levels. During heart-lung bypass, 25 mg/kg of prednisolone was administered to the patient, and in order to prevent the development of metabolic acidosis in the core cooling, 7% sodium bicarbonate was given every 30 min during heart-lung bypass. This step was taken to keep the change of base deficit around 10 mEq/liter. The time of circulatory arrest necessary for intracardiac repair averaged 49 min for Group I, 60 min for Group II, and 55 min for Group III. Serial circulatory arrest was tried for the more complicated cases in Groups II and III.

It has been our routine to remove blood from the cardiac cavity as well as from the pericardial sac in order to prevent the destruction of blood components within the heart-lung machine (Ito et al. 1974). This measure forestalled the development of postoperative hemorrhage, pulmonary complications and renal failure. For rewarming, the core temperature of the heart-lung perfusate was raised above 27°C, and the warming was discontinued when the pharyngeal temperature reached 34°C. At this point, the rectal temperature was usually 27–28°C. An electric blanket or warm water bath was required for further rewarming.

After completion of the heart-lung bypass, heparin was neutralized with protamine sulfate, and platelet-rich plasma, fresh frozen plasma, and anti-hemophilic factor were administered to the patient. Up to the present we have been using autologous pericardium as much as possible to construct baffles. The pericardium in use has been a relatively large trapezoidal piece, approximately 5 by 6 cm. With the exception of one case, the right atriotomy has been performed with a longitudinal incision into the center of the right atrium, with every precaution paid against the incision and sutures approaching the area of the S-A node.

It is desirable that the area of the atrial septum to be resected will be as large as possible not to interfere with venous return. Especially, it is necessary to remove a large section of the muscular atrial septum at the upper part of the limbus fossae ovalis to avoid obstruction on the superior vena cava. The septum in the area of the coronary sinus should not be cut deeply to avoid the damage to the A-V node. Without performing a coronary cutback, a Hegar dilator was inserted retrograde from the coronary sinus, and a new orifice was constructed in the left atrium. The orifice was closed with 5–0 Nylon shallow stitches. Just before completion of suturing the baffle, the occlusion of the venous return was released, removing the air from behind the baffle. And, blood leakage was confirmed. Excessive expansion of the baffle was controlled by plication with additional 5–0 Nylon stitches.

In the presence of PS or previous PA-banding, release of PS or pulmonary artery debanding was done at first, and then an intracardiac repair was carried out.

In coping with VSD, a transatrial approach was the first choice to accomplish its closure. Yet, due to the size and location of the defect, there were cases where a transventricular approach was thought to be more an appropriate method than a transatrial approach.
Results

Employing the steps for open-heart surgery described above, we have undertaken the Mustard procedure for correction of TGA in 24 cases, among which 6 succumbed shortly after surgery (Table 1). The causes of death in these 6 cases were as follows: low cardiac output syndrome due to advanced pulmonary vascular obstruction (2 cases), hemorrhagic diathesis due to prolonged extracorporeal circulation (1 case), hyperkalemia originating in the heart-lung machine perfusate (1 case), and unrelated accidents (2 cases).

In general, the postoperative course was good in the cases which were not complicated with PH. This made removal of the endotracheal tube possible after 24 hr postoperatively in most cases. In those with PH, however, reinsertion of the endotracheal tube was often necessary during the postoperative period because of recurrent pulmonary atelectasis.

We have encountered 4 cases of dysrhythmia that suggested various types of S-A node dysfunction such as atrioventricular nodal rhythm, sinus bradycardia and left atrial rhythm, etc. (Fig. 1). Recently, however, we have found an increasing number of patients showing signs of normal sinus rhythm, and early recovery from the dysrhythmia postoperatively. It was also noticed in the follow-up period (Figs. 2 and 3).

Fig. 1. Dysrhythmia after the Mustard procedure for TGA in 4 cases.
A: Atrioventricular nodal rhythm (Y.K., 34 months after operation).
B: Left atrial rhythm (Mirowski type I) (M.S., 22 months after operation).
C: Alternating sinus rhythm and atrioventricular nodal rhythm (G.T., 27 months after operation).
D: Sinus bradycardia with junctional escaped beats (K.N., 18 months after operation).
We have encountered 3 cases of pulmonary venous obstruction 4 to 24 months after the operation, 2 of which were successfully treated with patch enlargement of the pulmonary venous vessels to a functional left atrial channel.

**DISCUSSION**

Since 6 out of 24 patients terminated shortly after surgery, our results of operations are still unsatisfactory. Four of these 6 fatal cases were those that potentially could have been dealt successfully with appropriate technical steps and postoperative management. On the other hand, only one immediate death occurred in 12 cases in Group I which were unaccompanied by PH. It is thought that one of the most serious complications associated with this type of operation is the subsequent dysrhythmias. Especially its late occurrence is a major concern. With regard to the Mustard procedure, it demands a complex intraatrial operation. Some doubts have been raised whether or not complete prevention of dysrhythmias can be achieved with this method, and various trends of counter measures to prevent this complication have been reported from many institutions (El-Said et al. 1972; Isaacson et al. 1973; Angelini and Sandiford. 1973). As described above, we have
been operating to avoid surgical intervention in the vicinity of the S-A and A-V nodes, and have found an earlier recovery of the sinus rhythm postoperatively. Yet, since the number of our cases is still small and the follow-up study is not long enough, it is necessary to follow up these patients with great care for definite evaluation of this operation.

Pulmonary venous obstruction is an extremely serious complication, and recovery without surgical treatment cannot be expected. As mentioned previously, we encountered 3 cases of the postoperative pulmonary venous obstruction which implied that the incidence of the complication was higher than we had previously believed. The course of development of pulmonary venous obstruction is not yet known. We conjecture that the pericardium has a tendency to become thicker and shorter within the blood stream, which is accompanied by the adhesion of the baffle to the atrial wall and causes it to constrict. Further improvement in our procedure for the atriotomy and tailoring of the pericardial baffle is now under investigation.

As mentioned previously, heparin and prednisolone were given as a routine preoperative treatment, especially in patients with PS and VSD. Blood from the cardiac cavity as well as from the pericardial sac was discarded during intracardiac repair, and then platelet-rich plasma, fresh frozen plasma and antihemophilic factors were administered to the patients after completion of the heart-lung bypass. Use of this kind of methods dramatically reduced the bleeding in the postoperative period, and led to neither hemolysis nor hemoglobinuria. The incidence of lung complication and operative mortality were both drastically reduced.

The most serious potential problem in profound hypothermia is brain damage. The problem of brain damage due to bypass hypothermia in infants was studied by Ito et al. (1976) from our Department. They reported on the results of electroencephalography and neurological examinations. A developmental quotient (D. Q.) applicable to infants was recommended for the operative candidates prior to surgery and from 1 to 3 months after surgery. So far in one case which had circulatory arrest for 73 min with a lowest temperature of 16°C, temporary postoperative epileptic foci were detected. But in the other cases, no one showed either organic brain damage or intelligence disturbances. From this observation, a maximum of 60 min circulatory arrest at a minimum temperature of 17°C is completely safe, if a favorable tissue perfusion is achieved during the cooling process.

It is concluded that ‘bypass hypothermia’ provides a satisfactory condition for complex intracardiac surgery in infants and is a reasonably safe method.

References

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